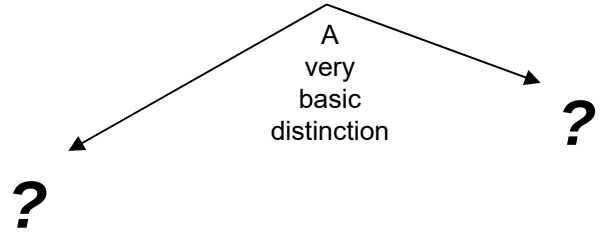
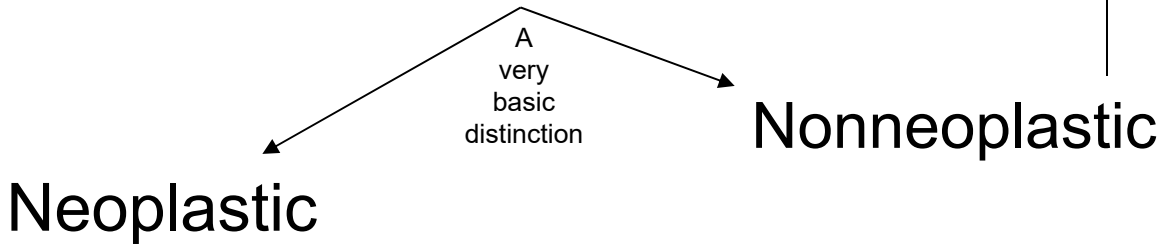
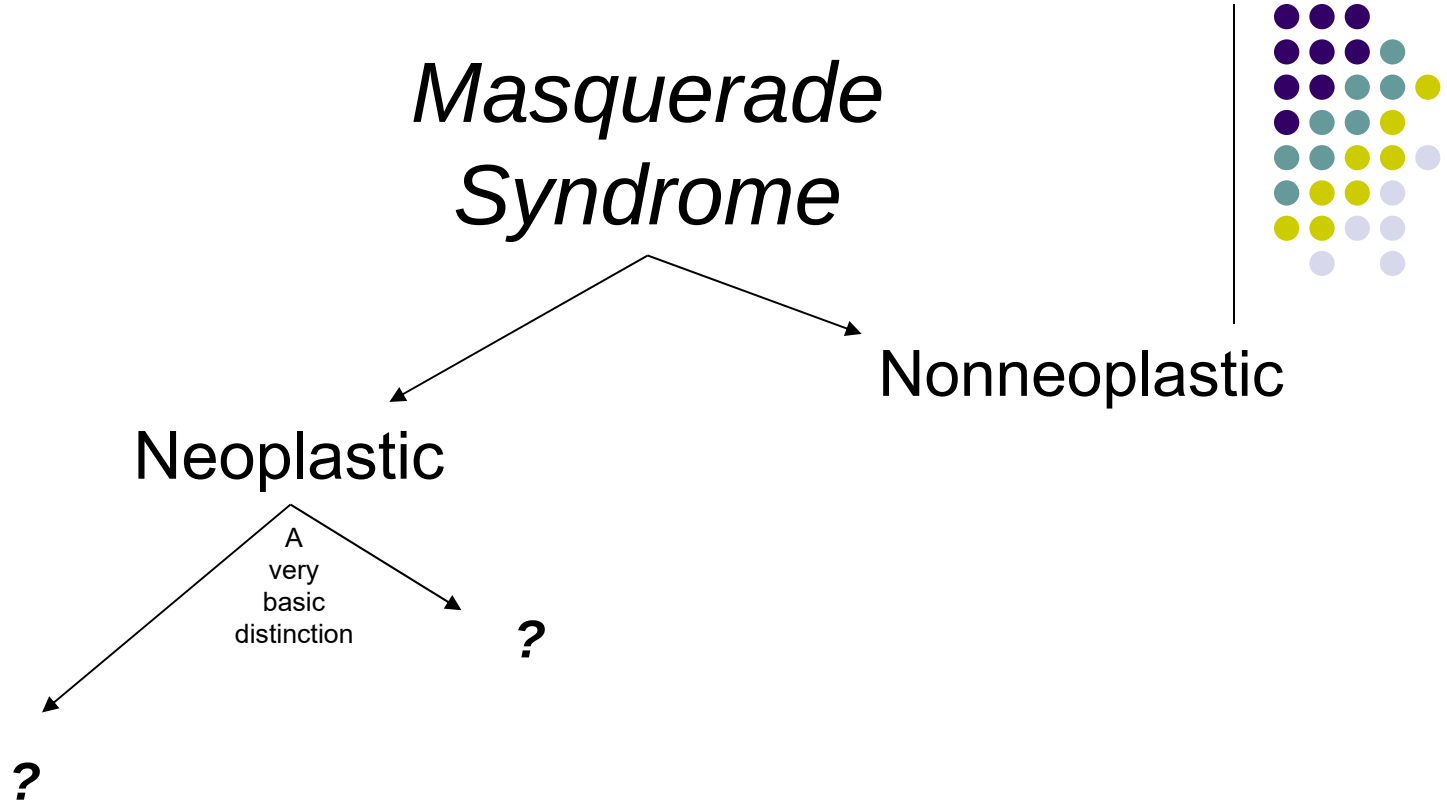
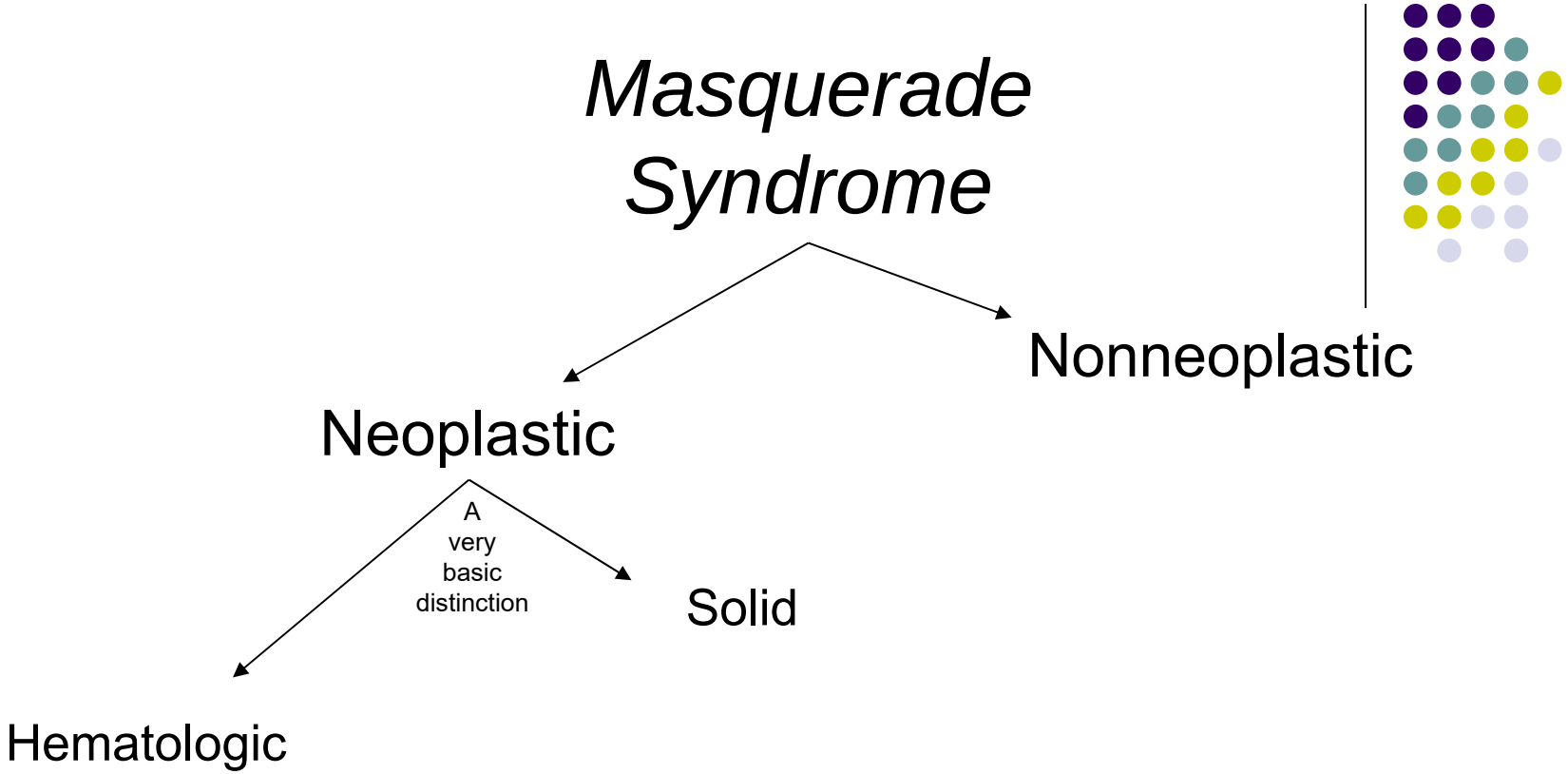
Masquerade Syndrome

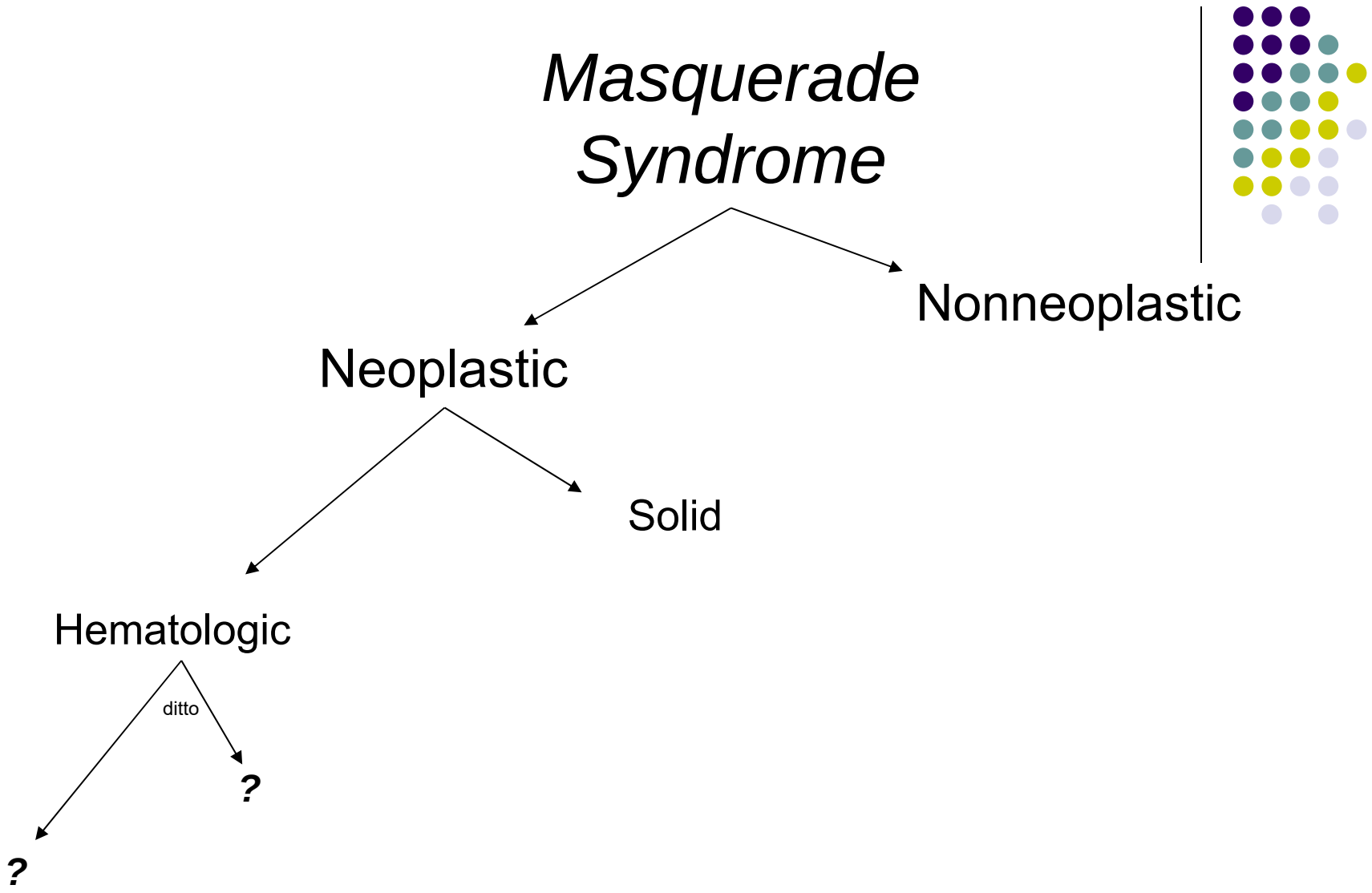


Masquerade Syndrome

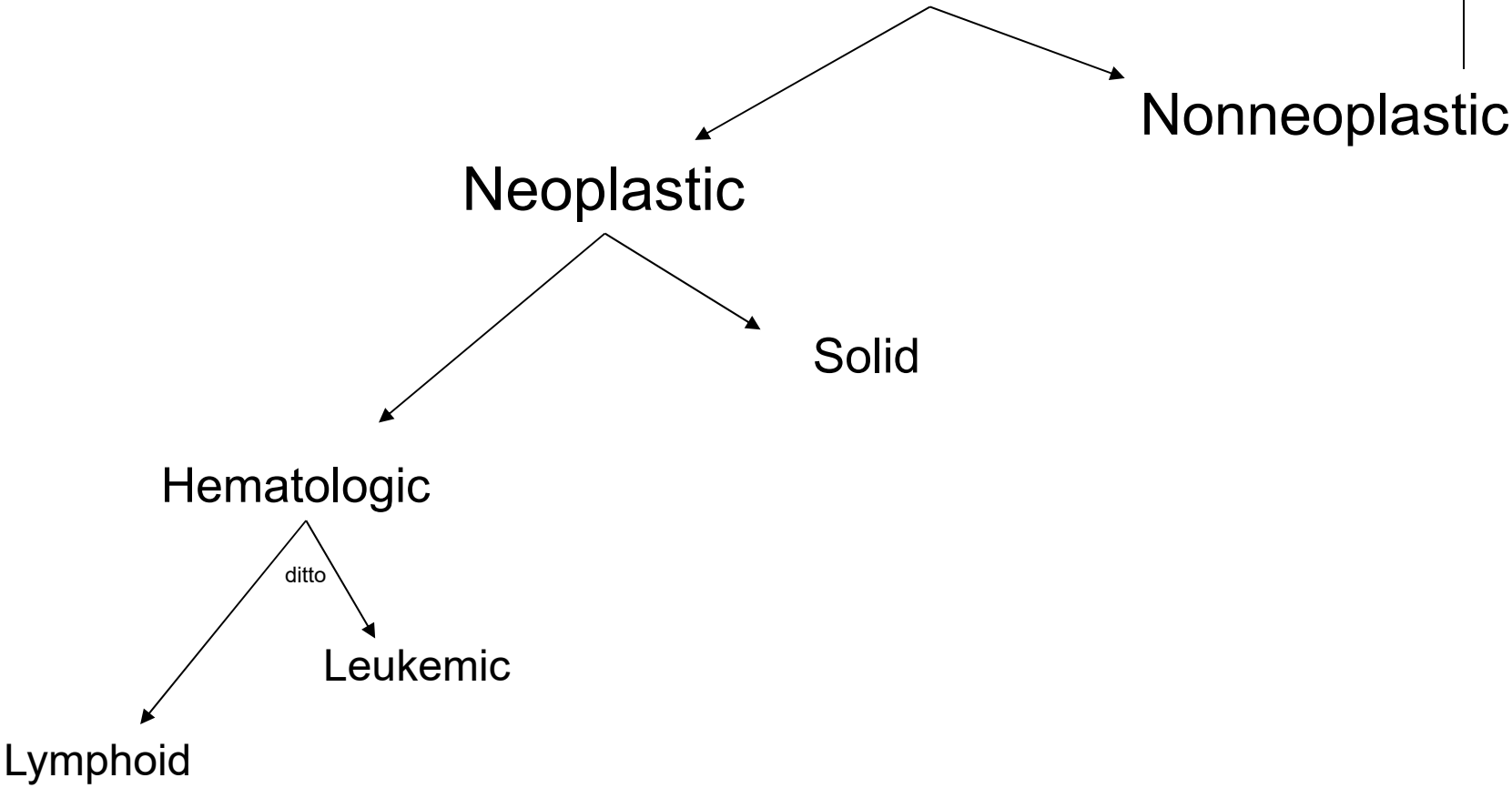


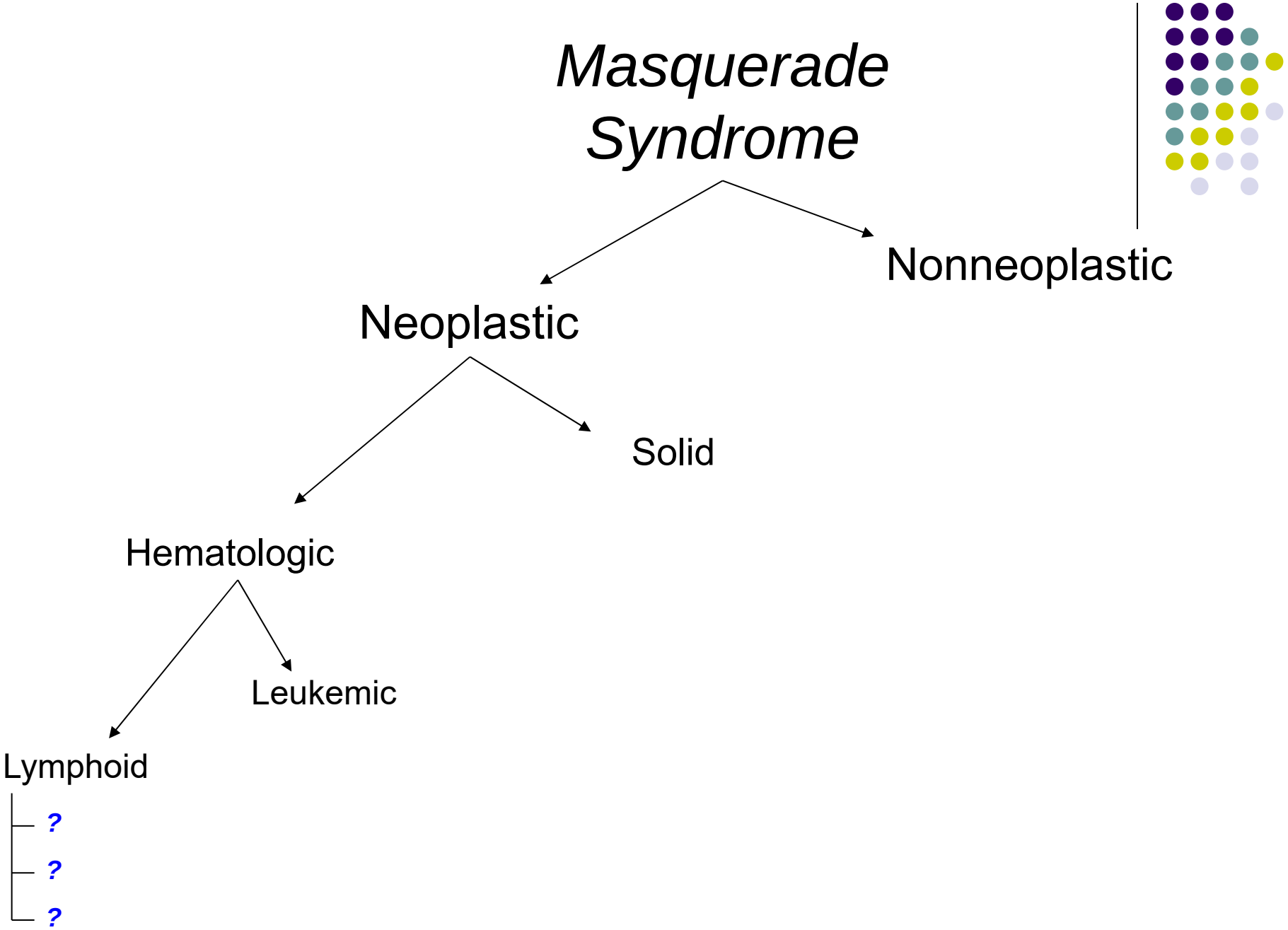

Masquerade Syndrome
Nonneoplastic
Neoplastic
A very basic distinction
?
?


Masquerade Syndrome
Nonneoplastic
Neoplastic
A very basic distinction
Solid
Hematologic

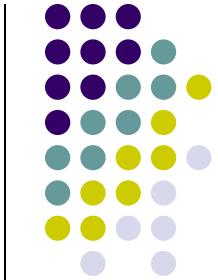

Masquerade Syndrome
Nonneoplastic
Neoplastic
Solid
Hematologic
ditto
?
?

Masquerade Syndrome



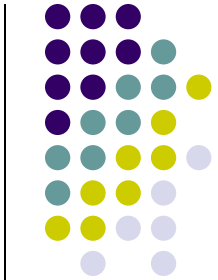

Masquerade Syndrome
Nonneoplastic
Neoplastic
Solid
Hematologic
Leukemic
Lymphoid
?
?
?

Masquerade Syndrome



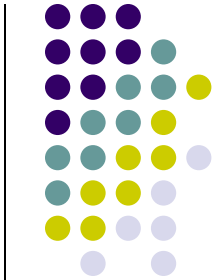
- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

Masquerade Syndrome

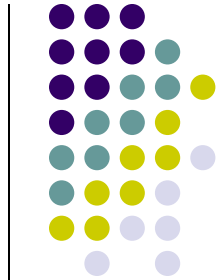


- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
      - *?*
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

Masquerade Syndrome



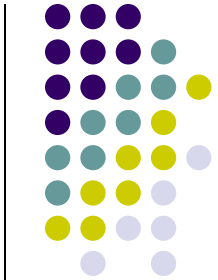
- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz



Masquerade Syndrome

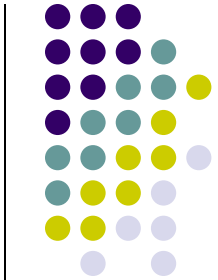
- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - Solid
    - ?
    - ditto
      - ?

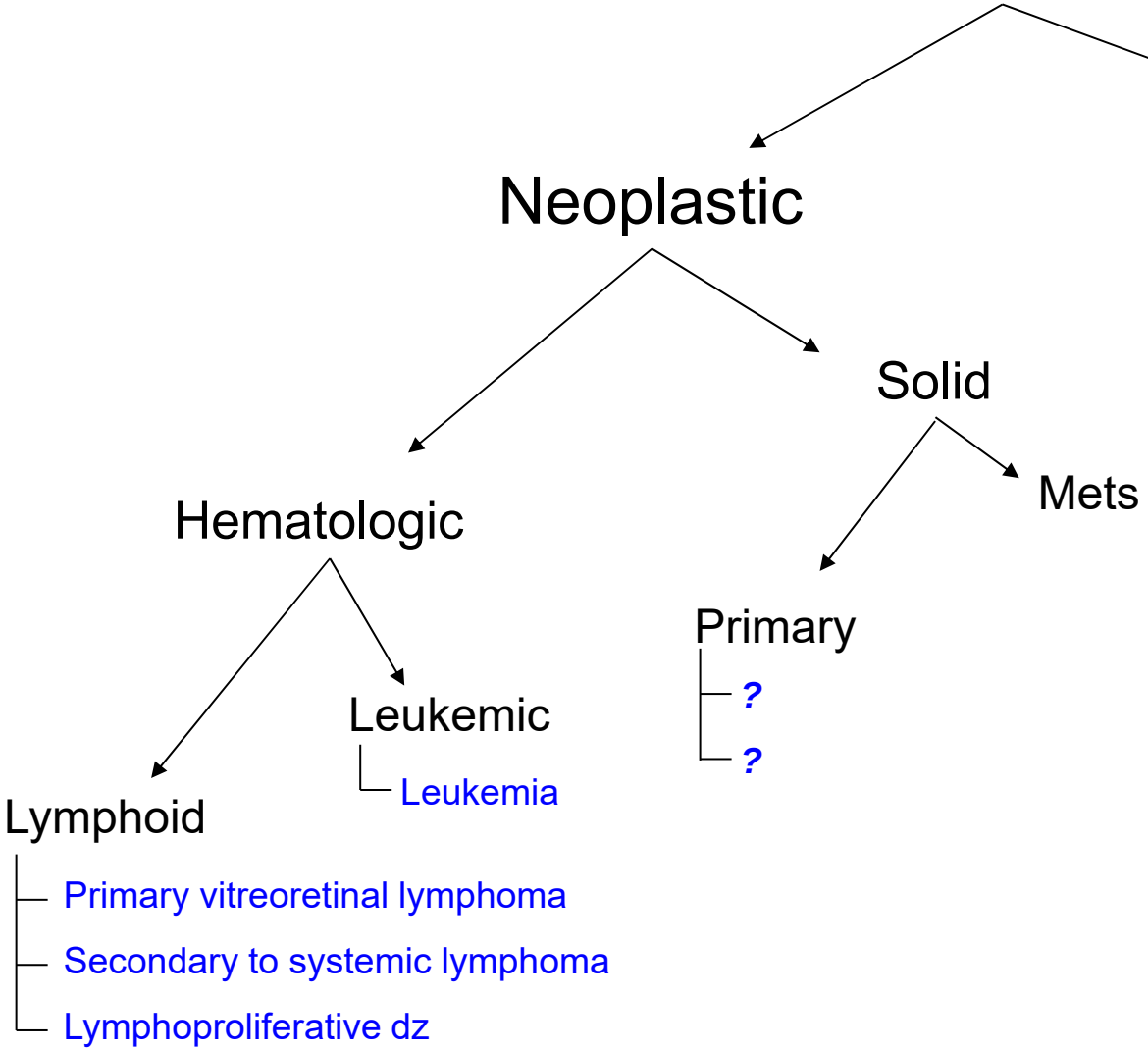
Masquerade Syndrome



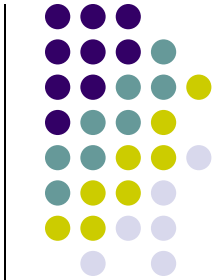
- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - Solid
    - Primary
    - ditto
    - Mets

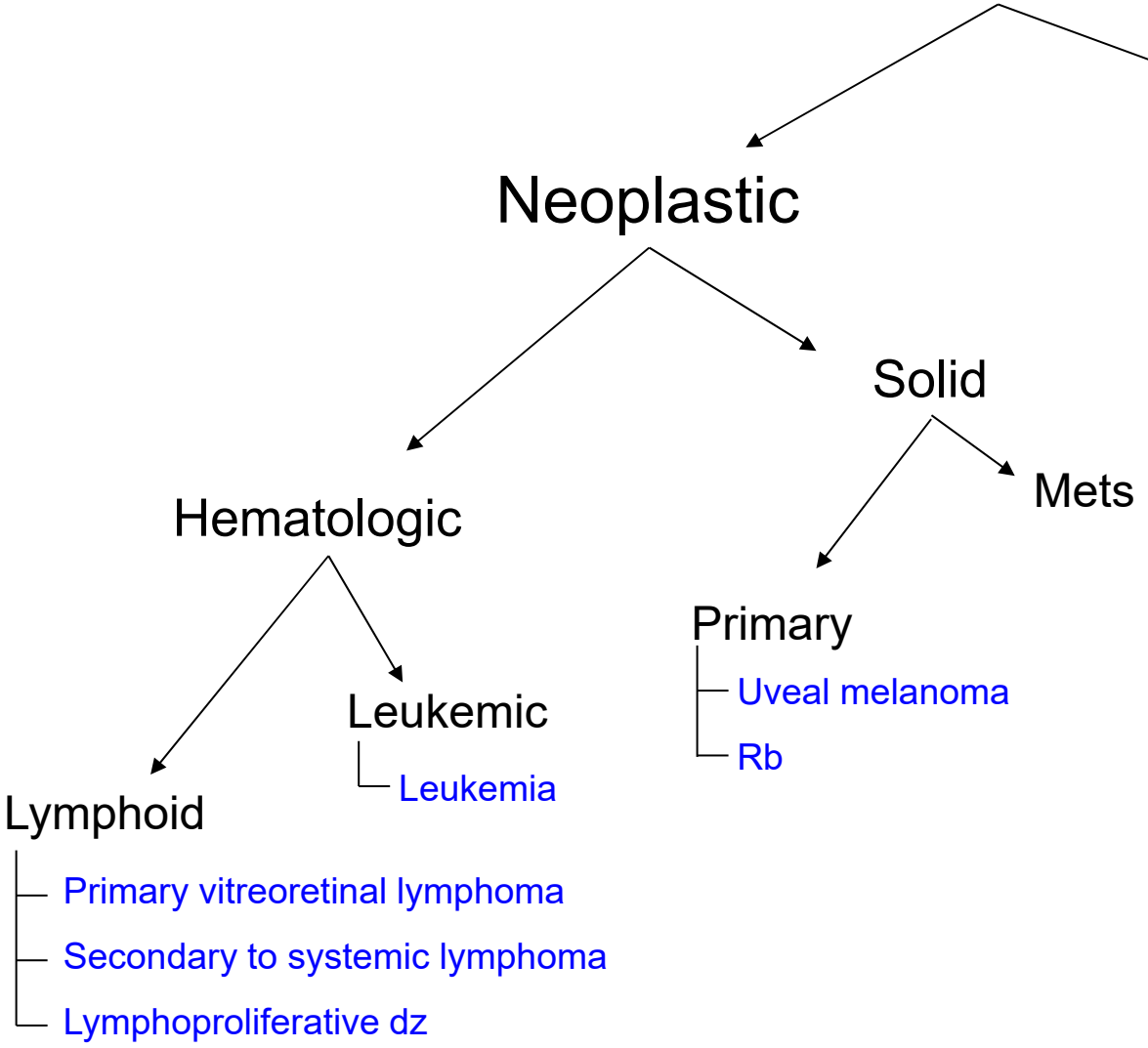
Masquerade Syndrome



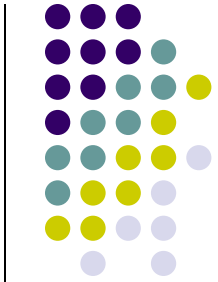


Masquerade Syndrome





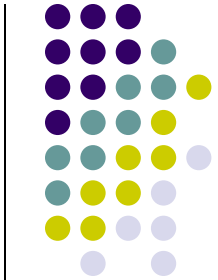
Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - *Primary vitreoretinal lymphoma?*
      - *Secondary to systemic lymphoma?*
      - *Lymphoproliferative dz?*
    - Leukemic
      - *Leukemia?*
  - Solid
    - Mets
    - Primary
      - *Uveal melanoma?*
      - *Rb?*

Of these, which is the most common etiology of <u>neoplastic</u> *masquerade syndrome?*

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
      - Rb

Of these, which is the most common etiology of <u>neoplastic</u> *masquerade syndrome?*
Primary vitreoretinal lymphoma (PVRL), by a mile

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
      - Rb
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

PVRL is a subtype of what malignancy?

gy of

neoplastic masquerade syndrome?

Primary vitreoretinal lymphoma (PVRL), by a mile

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
      - Rb
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

PVRL is a subtype of what malignancy?
Primary CNS lymphoma (PCNSL)

gy of
neoplastic *masquerade syndrome?*
Primary vitreoretinal lymphoma (PVRL), by a mile

Masquerade Syndrome

Nonneoplastic

N

What type of lymphoma is most common in PVRL?

Hematologic

Leukemic

Leukemi

Lymphoid

Primary vitreoretinal lymph

Secondary to systemic lymph

Lymphoproliferative dz

Masquerade Syndrome

Nonneoplastic

N

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Hematologic

Leukemic

Leukemi

Lymphoid

Primary vitreoretinal lymph

Secondary to systemic lymph

Lymphoproliferative dz

Masquerade Syndrome

Nonneoplastic

N

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

There are three main populations whose immunocompromised status puts them at risk for PVRL—what are they?
--?
--?
--?

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

There are three main populations whose immunocompromised status puts them at risk for PVRL—what are they?
--HIV/AIDS pts
--Organ transplantation on chronic immunosuppressives
--Cancer pts on chemo

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

What eye complaint(s) do these pts present with?

Masquerade Syndrome

- Nonneoplastic
- N
- Hematologic
  - Leukemic
    - Leukemi
  - Lymphoid
    - **Primary vitreoretinal lymph**
    - Secondary to systemic lymph
    - Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

What eye complaint(s) do these pts present with?
Decreased vision and/or floaters

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

Primary vitreoretinal lymph

Secondary to systemic lymph

Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

What eye complaint(s) do these pts present with?
Decreased vision and/or floaters

In addition to visual complaints, what other signs/symptoms may clue the astute clinician to the possibility of PVRL?

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

What eye complaint(s) do these pts present with?
Decreased vision and/or floaters

In addition to visual complaints, what other signs/symptoms may clue the astute clinician to the possibility of PVRL?
Those due to CNS involvement (eg, confusion, weakness, memory loss)

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

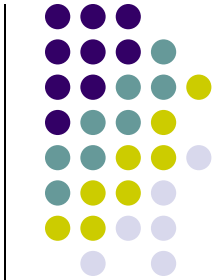
What systemic status increases the risk of developing PVRL?
Being immunocompromised

What eye complaint(s) do these pts present with?
Decreased vision and/or floaters

In addition to visual complaints, what other signs/symptoms may clue the astute clinician to the possibility of PVRL?
Those due to CNS involvement (eg, confusion, weakness, memory loss)

What other CNS signs/symptoms may be present?

Masquerade Syndrome



Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

What eye complaint(s) do these pts present with?
Decreased vision and/or floaters

In addition to visual complaints, what other signs/symptoms may clue the astute clinician to the possibility of PVRL?
Those due to CNS involvement (eg, confusion, weakness, memory loss)

What other CNS signs/symptoms may be present?
The most common is changes in behavior/personality. Other, more obvious S/S include seizures, cerebellar signs, hemiparesis and cranial nerve palsies.

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

Le

Leukemi

What proportion of PVRL pts will end up with CNS involvement, ie, with PCNSL?

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

In addition to visual complaints, what other signs/symptoms may clue the astute clinician to the possibility of PVRL?
Those due to **CNS involvement** (eg, confusion, weakness, memory loss)

What other CNS signs/symptoms may be present?
The most common is changes in behavior/personality. Other, more obvious S/S include seizures, cerebellar signs, hemiparesis and cranial nerve palsies.

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Le

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

What proportion of PVRL pts will end up with CNS involvement, ie, with PCNSL?
Most—over 2/3

In addition to visual complaints, what other signs/symptoms may clue the astute clinician to the possibility of PVRL?
Those due to **CNS involvement** (eg, confusion, weakness, memory loss)

What other CNS signs/symptoms may be present?
The most common is changes in behavior/personality. Other, more obvious S/S include seizures, cerebellar signs, hemiparesis and cranial nerve palsies.

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

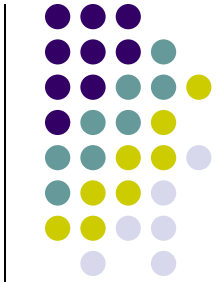
Given its CNS manifestations, are there findings on brain imaging suggestive of PCNSL?

ay clue the

mory loss)

more obvious S/S include seizures, cerebellar signs, hemiparesis and cranial nerve palsies.

Masquerade Syndrome



Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being immunocompromised

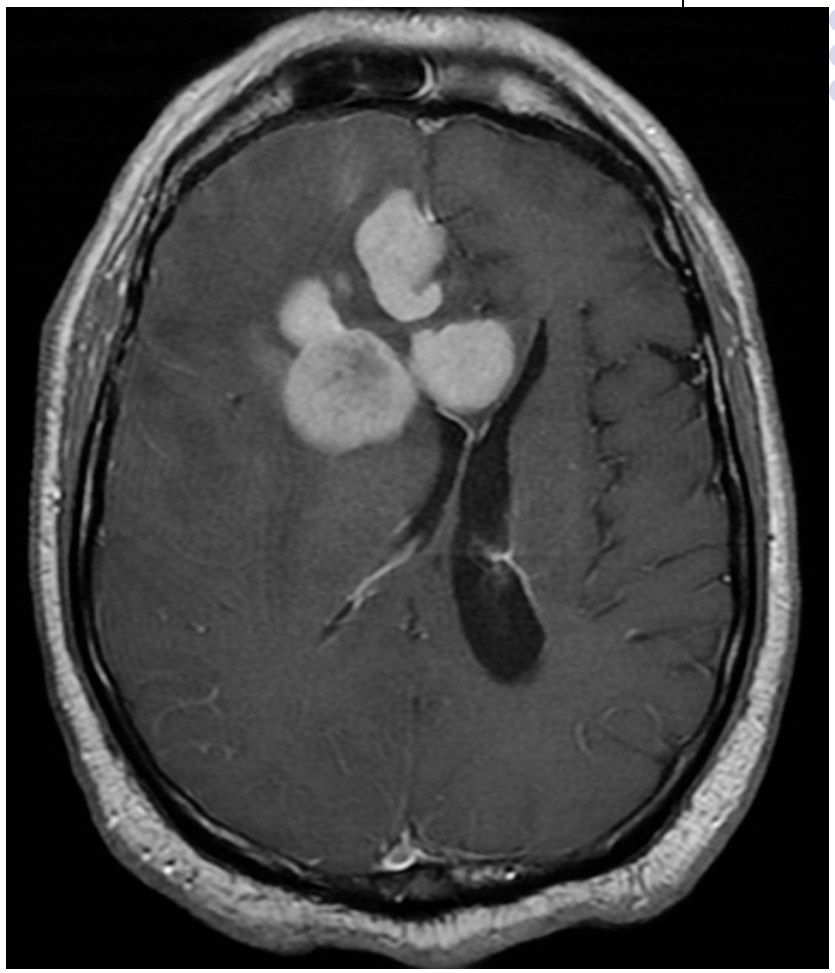
Given its CNS manifestations, are there findings on brain imaging suggestive of PCNSL?
Yes--periventricular white-matter lesions

ay clue the

mory loss)

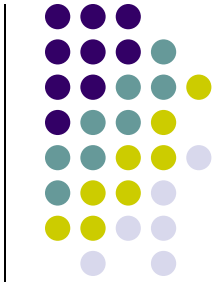
more

obvious S/S include seizures, cerebellar signs, hemiparesis and cranial nerve palsies.



PCNSL imaging

Masquerade Syndrome



Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being **immunocompromised**

Given its CNS manifestations, are there findings on brain imaging suggestive of PCNSL?
Yes--periventricular white-matter lesions

Is there a difference in the appearance of the lesions found in immunocompetent vs immunocompromised pts?

ay clue the

mory loss)

more

obvious S/S include seizures, cerebellar signs, hemiparesis and cranial nerve palsies.

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being **immunocompromised**

Given its CNS manifestations, are there findings on brain imaging suggestive of PCNSL?
Yes--periventricular white-matter lesions

Is there a difference in the appearance of the lesions found in immunocompetent vs immunocompromised pts?
There can be. The lesions in immunocompetent pts tend to be uniformly bright, whereas those in immunocompromised pts tend to be something-like

ay clue the

mory loss)

more

obvious S/S include seizures, cerebellar signs, hemiparesis and cranial nerve palsies.

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Leukemi

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being **immunocompromised**

Given its CNS manifestations, are there findings on brain imaging suggestive of PCNSL?
Yes--periventricular white-matter lesions

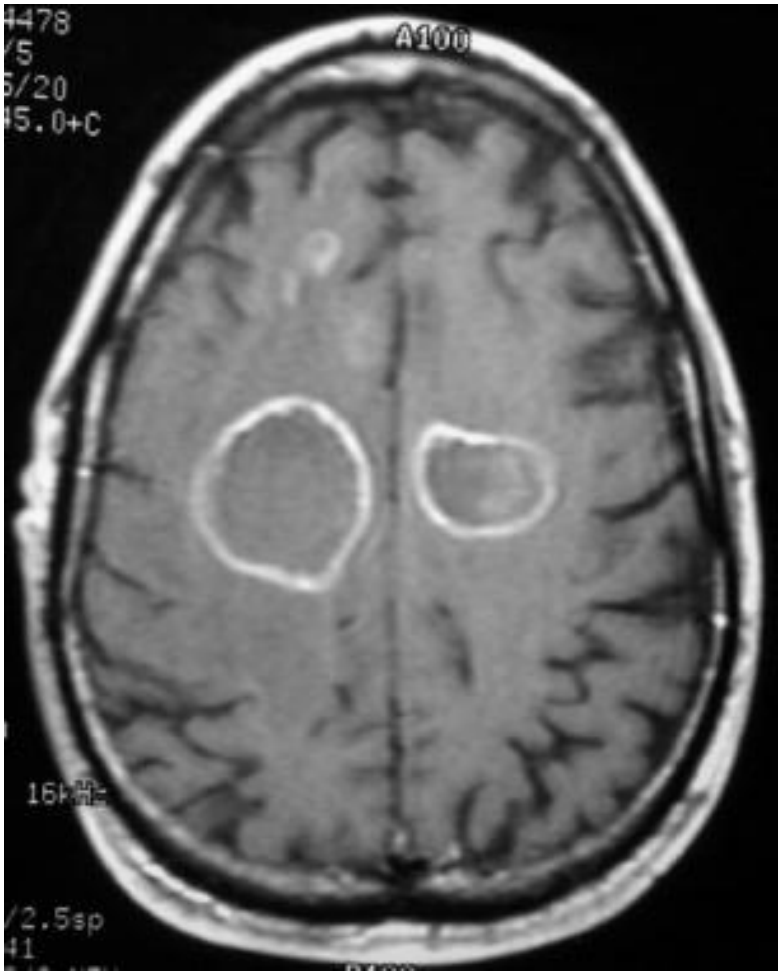
Is there a difference in the appearance of the lesions found in immunocompetent vs immunocompromised pts?
There can be. The lesions in immunocompetent pts tend to be uniformly bright, whereas those in immunocompromised pts tend to be ring-like.

ay clue the

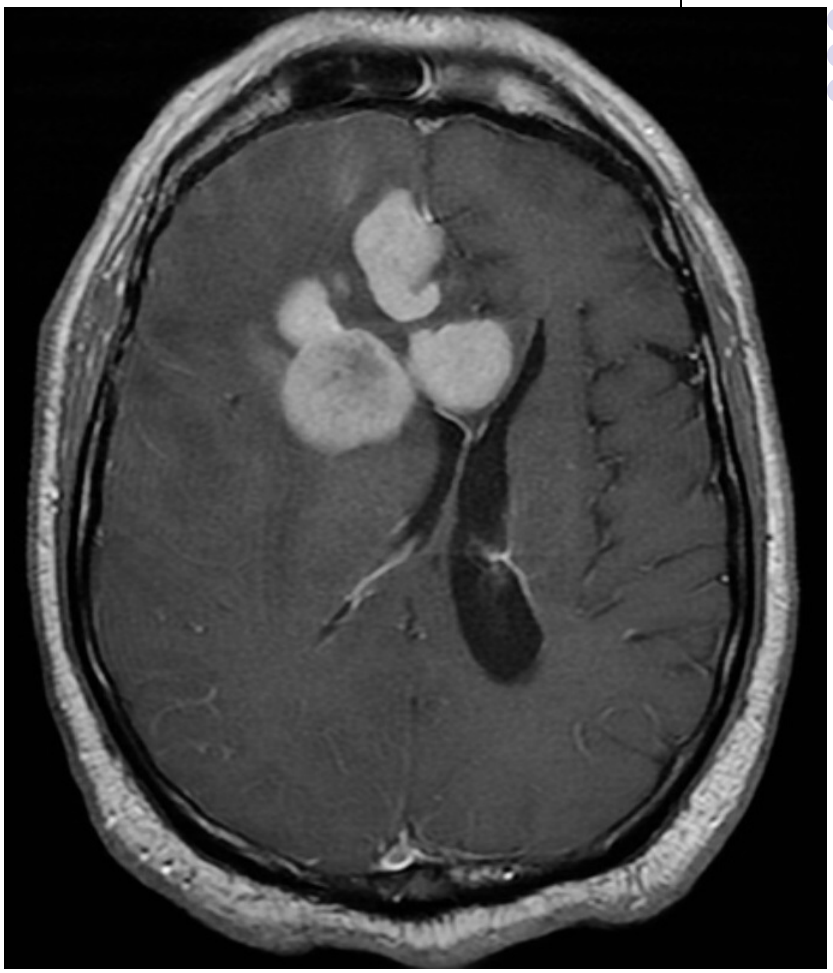
mory loss)

more

obvious S/S include seizures, cerebellar signs, hemiparesis and cranial nerve palsies.



PCNSL in immunocompromised pt



PCNSL in immunocompetent pt

PCNSL imaging

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being **immunocompromised**

Given its CNS manifestations, are there findings on brain imaging suggestive of PVRL?
Yes--periventricular white-matter lesions

There is another opportunistic condition in immunocompromised individuals that can present with ring-shaped brain lesions on imaging--what is it?

the

ss)

There can be. The lesions in immunocompetent pts tend to be uniformly bright, whereas **those in immunocompromised pts tend to be ring-like.**

re

obvious S/S include seizures, cerebellar signs, hemiparesis and cranial nerve palsies.

Masquerade Syndrome

Nonneoplastic

N

Hematologic

Leukemic

Lymphoid

- **Primary vitreoretinal lymph**
- Secondary to systemic lymph
- Lymphoproliferative dz

What type of lymphoma is most common in PVRL?
Virtually all PVRLs are non-Hodgkin B-cell lymphomas

Who is the typical PVRL pt?
An adult in their 50s-60s

What systemic status increases the risk of developing PVRL?
Being **immunocompromised**

Given its CNS manifestations, are there findings on brain imaging suggestive of PVRL?
Yes--periventricular white-matter lesions

There is another opportunistic condition in immunocompromised individuals that can present with ring-shaped brain lesions on imaging--what is it?
CNS toxoplasmosis in AIDS

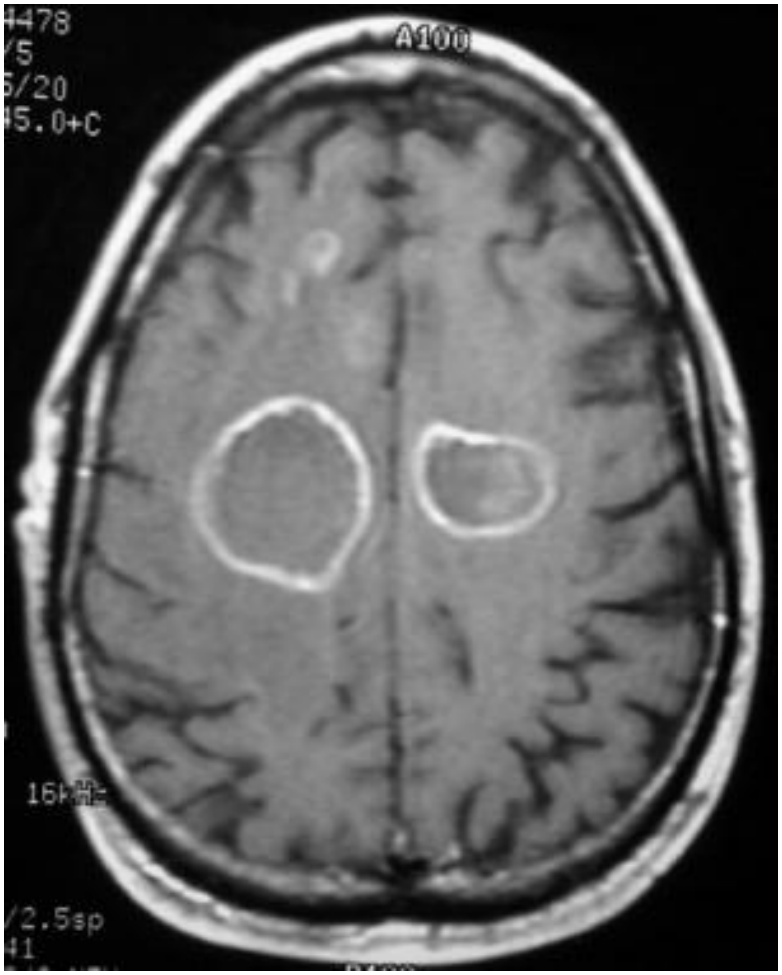
the

ss)

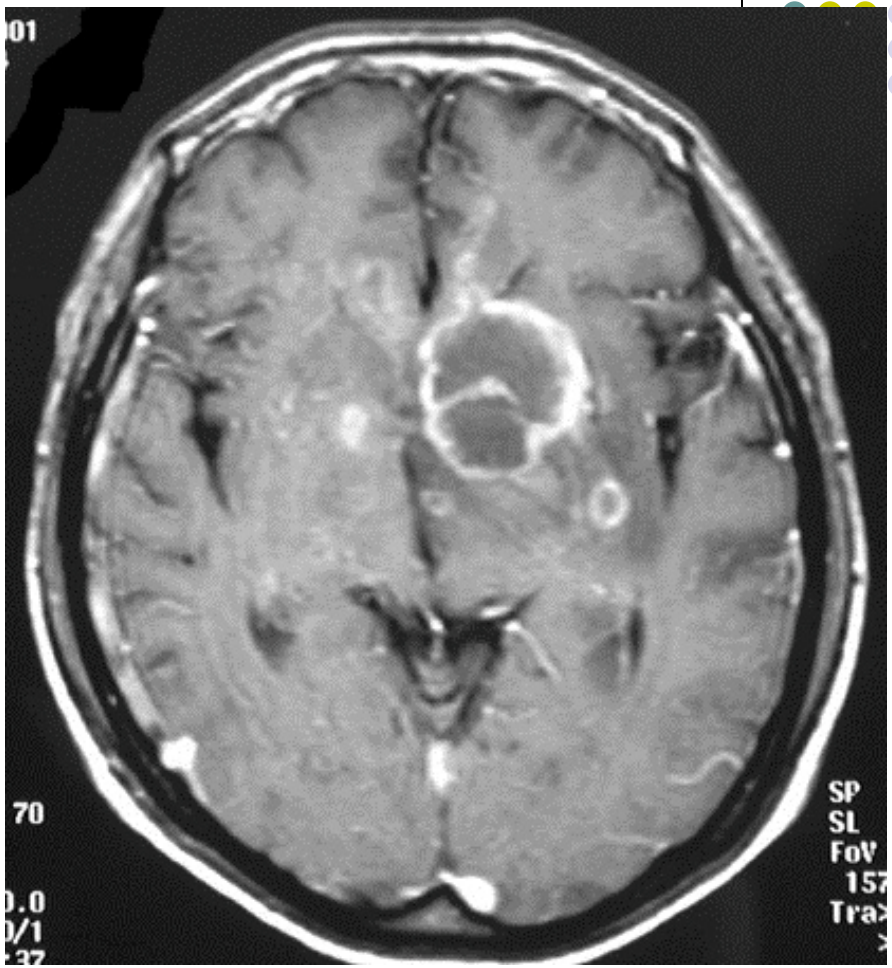
There can be. The lesions in immunocompetent pts tend to be uniformly bright, whereas **those in immunocompromised pts tend to be ring-like.**

re

obvious S/S include seizures, cerebellar signs, hemiparesis and cranial nerve palsies.



PCNSL in immunocompromised pt



CNS toxo in immunocompromised pt

PCNSL imaging?

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal lymp**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

What does DFE typically reveal in PVRL?

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal lymp**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

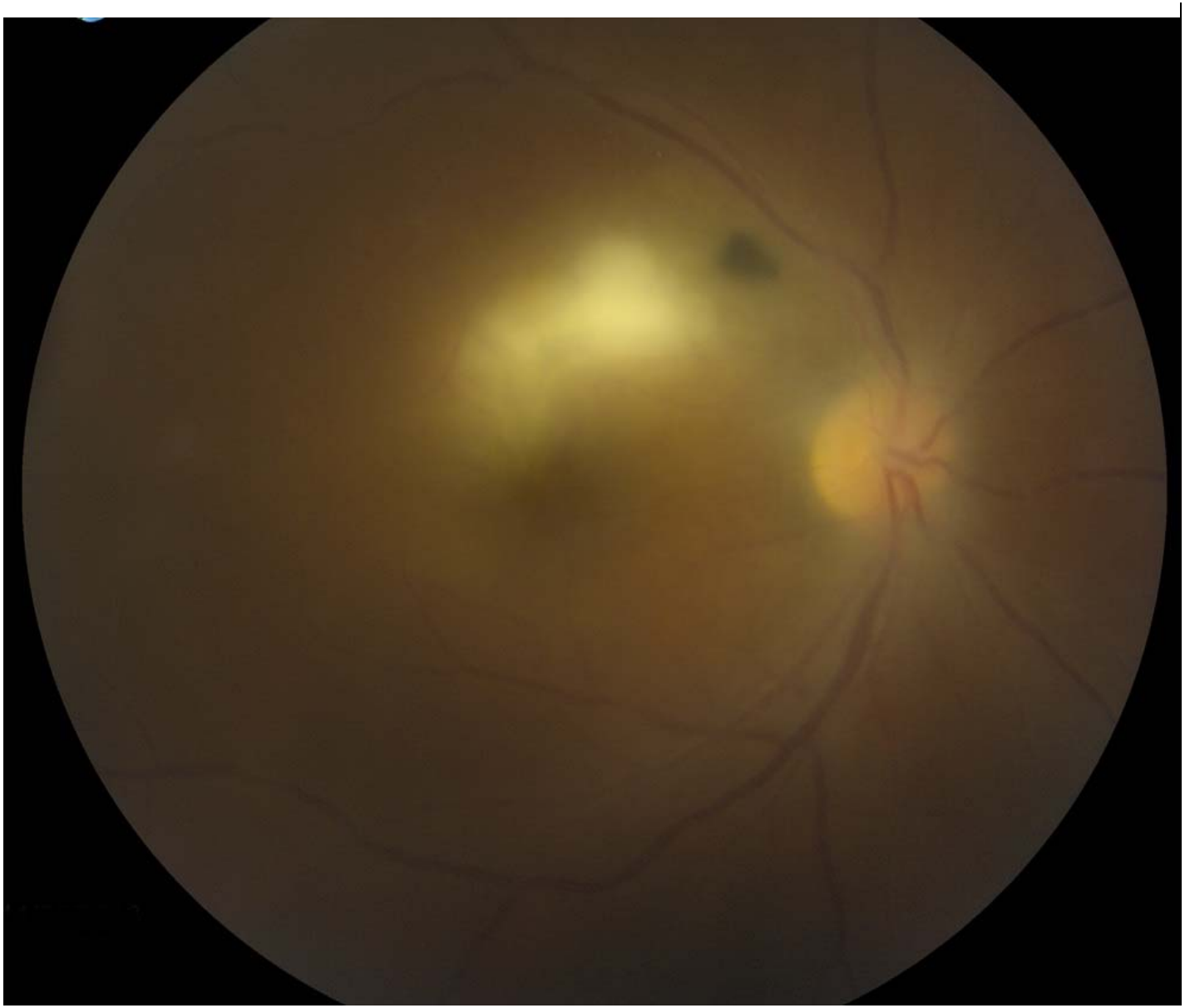
What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as "**creamy yellow**" in color.

PVRL: Typical white-yellow subretinal infiltrates

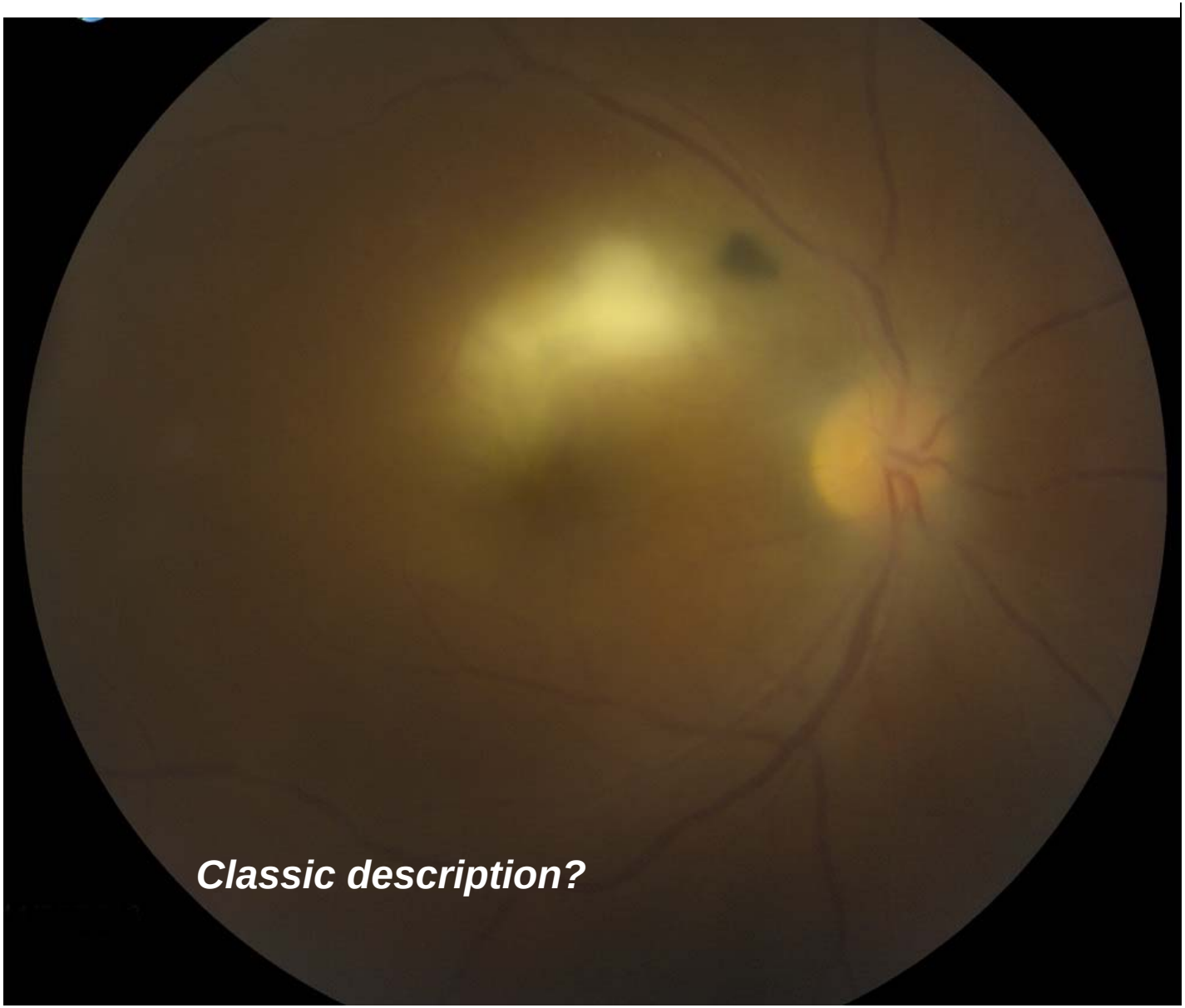
Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal lymp**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as "**creamy yellow**" in color. The infiltrates can mimic the findings of other, more common conditions (eg, toxoplasmosis).



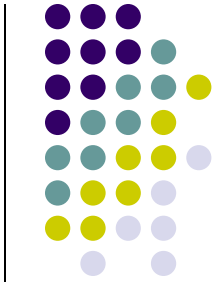
Ocular toxoplasmosis



Ocular toxoplasmosis

Ocular toxoplasmosis

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal**
      - Secondary to systemic
      - Lymphoproliferative dz

What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as "**creamy yellow**" in color. **The infiltrates can mimic the findings of other, more common conditions (eg, toxoplasmosis).**

There is another opportunistic condition in immunocompromised individuals that can present with ring-shaped brain lesions on imaging--what is it?
CNS toxoplasmosis in AIDS

Given that both the DFE and imaging presentations of PVRL are similar to those of toxo, make sure you keep PVRL in your DDx for toxo (and vice versa)!

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal lymp**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as "**creamy yellow**" in color. The infiltrates can mimic the findings of other, more common conditions (eg, toxoplasmosis).

What about vitritis and/or AC cell?

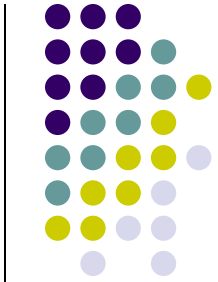
Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal lymp**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as "**creamy yellow**" in color. The infiltrates can mimic the findings of other, more common conditions (eg, toxoplasmosis).

What about vitritis and/or AC cell?
One or both may be present

Masquerade Syndrome



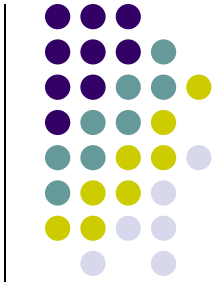
- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - L
    - Lymphoid
      - **Primary vitreoretinal**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as "**creamy yellow**" in color. The infiltrates can mimic the findings of other, more common conditions (eg, toxoplasmosis).

*What about **vitritis** and/or AC cell?*
One or both may be present

Can the vitritis be severe enough to give a 'headlight in the fog' impression a la toxoplasmosis?

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - L
    - Lymphoid
      - **Primary vitreoretinal**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as "**creamy yellow**" in color. The infiltrates can mimic the findings of other, more common conditions (eg, toxoplasmosis).

*What about **vitritis** and/or AC cell?*
One or both may be present

Can the vitritis be severe enough to give a 'headlight in the fog' impression a la toxoplasmosis?
Indeed it can

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal lymp**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as **"creamy yellow"** in color. The infiltrates can mimic the findings of other, more common conditions (eg, toxoplasmosis).

What about vitritis and/or AC cell?
One or both may be present

What is the classic (mis)diagnosis of PVRL?

Masquerade Syndrome

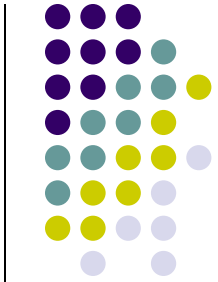
- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal lymp**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as "**creamy yellow**" in color. The infiltrates can mimic the findings of other, more common conditions (eg, toxoplasmosis).

What about vitritis and/or AC cell?
One or both may be present

What is the classic (mis)diagnosis of PVRL?
As being an autoimmune uveitis

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal lymp**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as "**creamy yellow**" in color. The infiltrates can mimic the findings of other, more common condi

Another characteristic retinal finding in PVRL can easily be mistaken as evidence of autoimmune uveitis. What is it?

What
One

What is the classic (mis)diagnosis of PVRL?
As being a**n autoimmune uveitis**

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal lymp**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

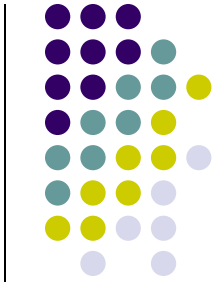
What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as "**creamy yellow**" in color. The infiltrates can mimic the findings of other, more common condi

Another characteristic retinal finding in PVRL can easily be mistaken as evidence of autoimmune uveitis. What is it?
Retinal vasculitis. 'Frosted-branch' changes and/or arteriolar obstruction may occur.

What
One

What is the classic (mis)diagnosis of PVRL?
As being a**n autoimmune uveitis**

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal lymp**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

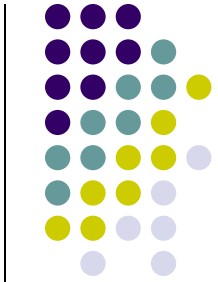
What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as **"creamy yellow"** in color. The infiltrates can mimic the findings of other, more common conditions (eg, toxoplasmosis).

What about vitritis and/or AC cell?
One or both may be present

What is the classic (mis)diagnosis of PVRL?
As being an autoimmune uveitis

Does PVRL respond to systemic steroids?

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukem
      - Leuken
    - Lymphoid
      - **Primary vitreoretinal lymp**
      - Secondary to systemic lymp
      - Lymphoproliferative dz

What does DFE typically reveal in PVRL?
The classic finding is of subretinal infiltrates described as "**creamy yellow**" in color. The infiltrates can mimic the findings of other, more common conditions (eg, toxoplasmosis).

What about vitritis and/or AC cell?
One or both may be present

What is the classic (mis)diagnosis of PVRL?
As being an autoimmune uveitis

Does PVRL respond to systemic steroids?
Yes, but only briefly (which is a telltale clue that one is not dealing with an autoimmune uveitis)

Masquerade Syndrome

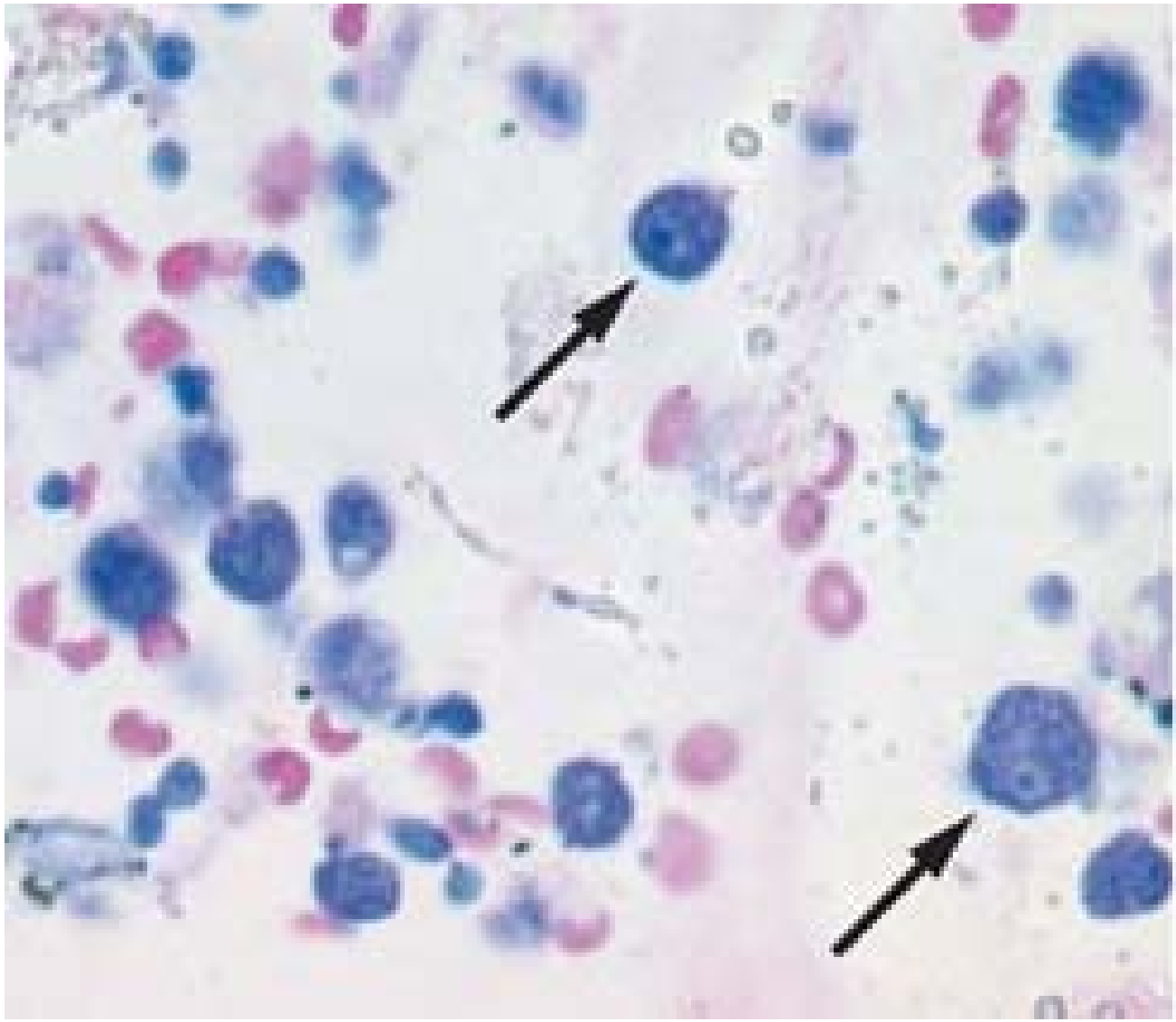
- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL diagnosed?

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL diagnosed?
Via finding 'big blue cells' on biopsy (Remember: If cancer's the answer, tissue's the issue)



PVRL: Big blue cells

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL diagnosed?
Via finding 'big blue cells' on biopsy (Remember: If cancer's the answer, tissue's the issue)

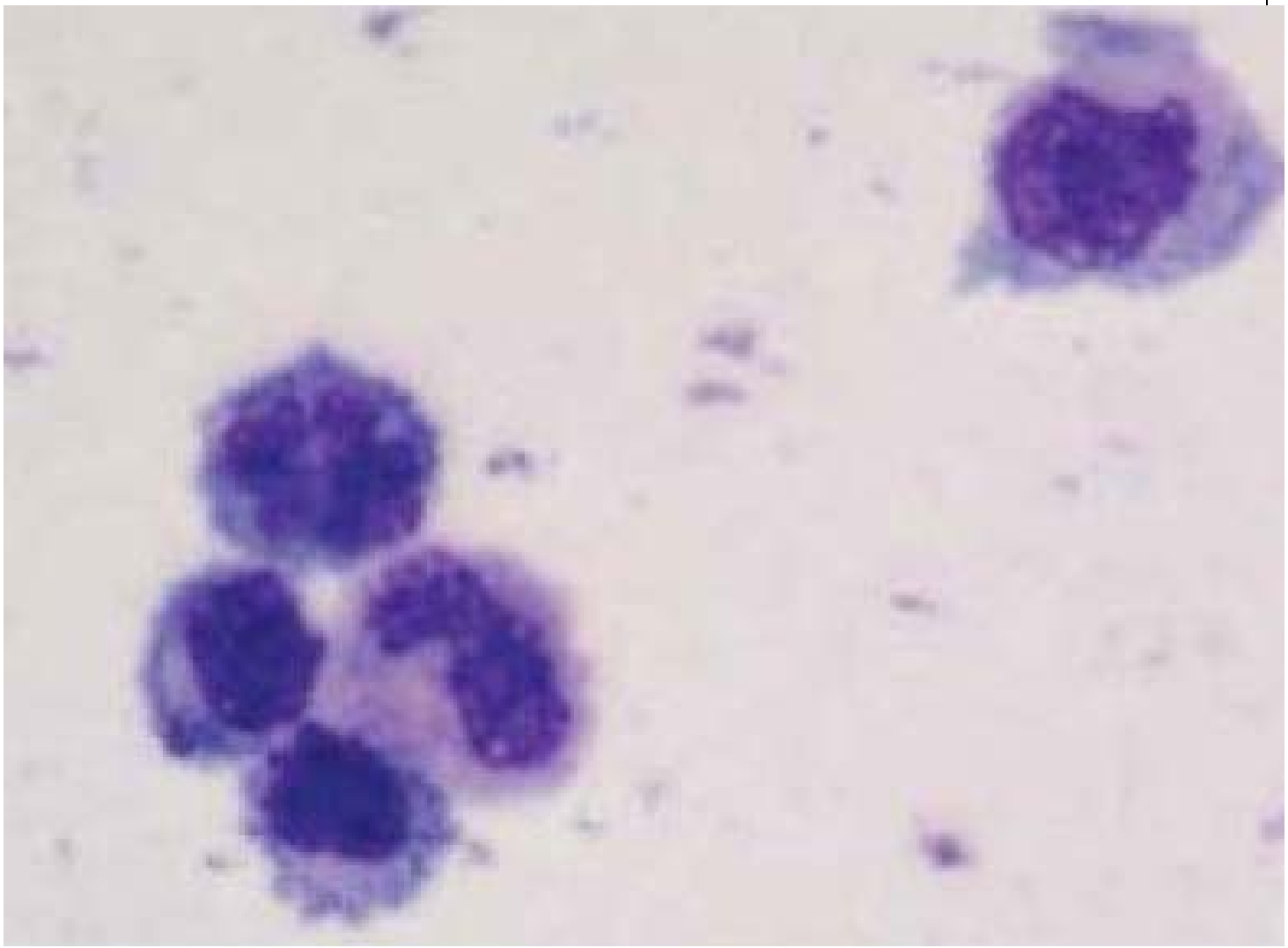
Where should one look for these cells?

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

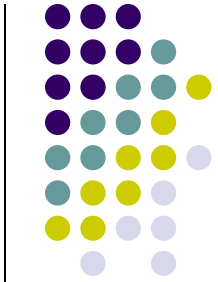
How is PVRL diagnosed?
Via finding 'big blue cells' on biopsy (Remember: If cancer's the answer, tissue's the issue)

Where should one look for these cells?
On a vitreous biopsy



Typical cytology of PVRL cells from the vitreous showing several atypical lymphoid cells with basophilic cytoplasm and large prominent irregular nuclei

Masquerade Syndrome



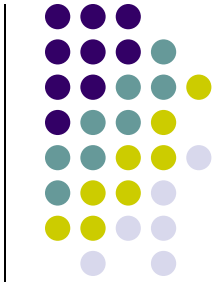
- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL diagnosed?
Via finding 'big blue cells' on biopsy (Remember: If cancer's the answer, tissue's the issue)

Where should one look for these cells?
On a vitreous biopsy

If the biopsy result is negative, what should be the next step?

Masquerade Syndrome



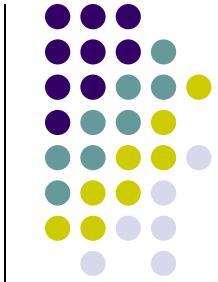
- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL diagnosed?
Via finding 'big blue cells' on biopsy (Remember: If cancer's the answer, tissue's the issue)

Where should one look for these cells?
On a vitreous biopsy

If the biopsy result is negative, what should be the next step?
Do it again (as many as a third of vitreous biopsies yield false-negative results)

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL diagnosed?
Via finding 'big blue cells' on biopsy (Remember: If cancer's the answer, tissue's the issue)

Where should one look for these cells?
On a vitreous biopsy

If the biopsy result is negative, what should be the next step?
Do it again (as many as a third of vitreous biopsies yield false-negative results)

If that *biopsy is negative as well, what next?*

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

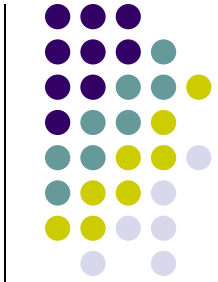
How is PVRL diagnosed?
Via finding 'big blue cells' on biopsy (Remember: If cancer's the answer, tissue's the issue)

Where should one look for these cells?
On a vitreous biopsy

If the biopsy result is negative, what should be the next step?
Do it again (as many as a third of vitreous biopsies yield false-negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL diagnosed?
Via finding 'big blue cells' on biopsy (Remember: If cancer's the answer, tissue's the issue)

In addition to cytology, another analysis can be performed on vitreous aspirate that is very helpful in distinguishing PVRL from an inflammatory process. What is it?

negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

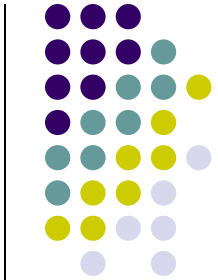
How is PVRL diagnosed?
Via finding 'big blue cells' on biopsy (Remember: If cancer's the answer, tissue's the issue)

In addition to cytology, another analysis can be performed on vitreous aspirate that is very helpful in distinguishing PVRL from an inflammatory process. What is it?
Cytokine analysis. In PVRL, one expects elevated levels of IL- # . In contrast, IL- # levels are elevated in inflammatory processes.

negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL diagnosed?
Via finding 'big blue cells' on biopsy (Remember: If cancer's the answer, tissue's the issue)

negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

In addition to cytology, another analysis can be performed on vitreous aspirate that is very helpful in distinguishing PVRL from an inflammatory process. What is it?
Cytokine analysis. In PVRL, one expects elevated levels of IL- 10 . In contrast, IL- 6 levels are elevated in inflammatory processes.

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL diagnosed?
Via finding 'big blue cells' on biopsy (Remember: If cancer's the answer, tissue's the issue)

In addition to cytology, another analysis can be performed on vitreous aspirate that is very helpful in distinguishing PVRL from an inflammatory process. What is it?
Cytokine analysis. In PVRL, one expects elevated levels of IL- 10 . In contrast, IL- 6 levels are elevated in inflammatory processes. Thus, an **elevated IL-10/IL-6 ratio suggests a diagnosis of PVRL.**

negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Hem
  - Lymphoid
    - **Primary vitreoretinal lymphoma**
    - Secondary to systemic lymphoma
    - Lymphoproliferative dz

In addition to biopsy-ing the vitreous, what other diagnostic steps must be taken?

s

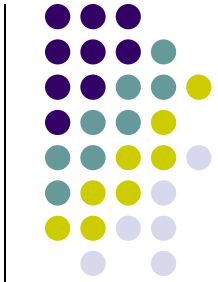
te that it?

Cytokine analysis: In PVRL, one expects elevated levels of IL- 10 . In contrast, IL- 6 levels are elevated in inflammatory processes. Thus, an **elevated IL-10/IL-6 ratio suggests a diagnosis of PVRL.**

false-negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

Masquerade Syndrome



Nonneoplastic

Neoplastic

In addition to biopsy-ing the vitreous, what other diagnostic steps must be taken?
The pt must be checked for the presence of CNS involvement, ie, PCNSL

Hem

Lymphoid

- **Primary vitreoretinal lymphoma**
- Secondary to systemic lymphoma
- Lymphoproliferative dz

levels are elevated in inflammatory processes. Thus, an **elevated IL-10/IL-6 ratio suggests a diagnosis of PVRL.**

false-negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

Masquerade Syndrome

Nonneoplastic

Neoplastic

Hem

Lymphoid

- **Primary vitreoretinal lymphoma**
- Secondary to systemic lymphoma
- Lymphoproliferative dz

In addition to biopsy-ing the vitreous, what other diagnostic steps must be taken?
The pt must be checked for the presence of CNS involvement, ie, PCNSL

All, pts, or only those exhibiting signs of CNS involvement?

s

te that it?

Cytokine analysis: In PVRL, one expects elevated levels of IL- 10 . In contrast, IL- 6 levels are elevated in inflammatory processes. Thus, an **elevated IL-10/IL-6 ratio suggests a diagnosis of PVRL.**

false-negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Hem
  - Lymphoid
    - **Primary vitreoretinal lymphoma**
    - Secondary to systemic lymphoma
    - Lymphoproliferative dz

In addition to biopsy-ing the vitreous, what other diagnostic steps must be taken?
The pt must be checked for the presence of CNS involvement, ie, PCNSL

All, pts, or only those exhibiting signs of CNS involvement?
All pts

s

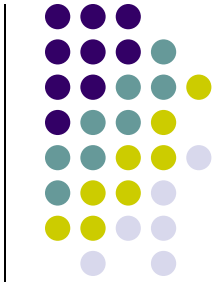
te that it?

Cytokine analysis: In PVRL, one expects elevated levels of IL- 10 . In contrast, IL- 6 levels are elevated in inflammatory processes. Thus, an **elevated IL-10/IL-6 ratio suggests a diagnosis of PVRL.**

false-negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

Masquerade Syndrome



Nonneoplastic

Neoplastic

Hem

In addition to biopsy-ing the vitreous, what other diagnostic steps must be taken?
The pt must be checked for the presence of CNS involvement, ie, PCNSL

All, pts, or only those exhibiting signs of CNS involvement?
All pts

What tests should be performed?

s

te that it?

Cytokine analysis. In PVRL, one expects elevated levels of IL- 10 . In contrast, IL- 6 levels are elevated in inflammatory processes. Thus, an **elevated IL-10/IL-6 ratio suggests a diagnosis of PVRL.**

false-negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

Lymphoid

- **Primary vitreoretinal lymphoma**
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome

Nonneoplastic

Neoplastic

Hem

In addition to biopsy-ing the vitreous, what other diagnostic steps must be taken?
The pt must be checked for the presence of CNS involvement, ie, PCNSL

All, pts, or only those exhibiting signs of CNS involvement?
All pts

What tests should be performed?
Imaging as previously discussed, along with a two words to check for

two diff words

Lymphoid

- **Primary vitreoretinal lymphoma**
- Secondary to systemic lymphoma
- Lymphoproliferative dz

levels are elevated in inflammatory processes. Thus, an **elevated IL-10/IL-6 ratio suggests a diagnosis of PVRL.**

false-negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

Masquerade Syndrome

Nonneoplastic

Neoplastic

Hem

Lymphoid

- **Primary vitreoretinal lymphoma**
- Secondary to systemic lymphoma
- Lymphoproliferative dz

In addition to biopsy-ing the vitreous, what other diagnostic steps must be taken?
The pt must be checked for the presence of CNS involvement, ie, PCNSL

All, pts, or only those exhibiting signs of CNS involvement?
All pts

What tests should be performed?
Imaging as previously discussed, along with a lumbar puncture to check for lymphoma cells

s

te that it?

Cytokine analysis. In PVRL, one expects elevated levels of IL- 10 . In contrast, IL- 6 levels are elevated in inflammatory processes. Thus, an **elevated IL-10/IL-6 ratio suggests a diagnosis of PVRL.**

false-negative results)

If that *biopsy is negative as well, what next?*
Gotta go with a retinal biopsy, or biopsy of a sub-retinal lesion

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL treated?

Masquerade Syndrome

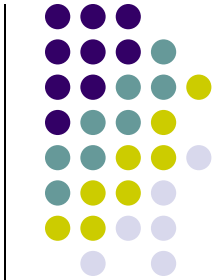
- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL treated?

A two-pronged approach is standard—intravitreal chemo (abb. +/-) + XBRT.*

*External-beam radiation therapy

Masquerade Syndrome



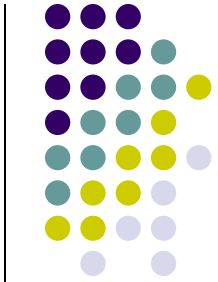
- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL treated?

A two-pronged approach is standard—intravitreal chemo (MTX +/- rituximab) + XBRT.*

*External-beam radiation therapy

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL treated?
A two-pronged approach is standard—intravitreal chemo (MTX +/- rituximab) + XBRT.*

What if CNS involvement is present?

*External-beam radiation therapy

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How is PVRL treated?
A two-pronged approach is standard—intravitreal chemo (MTX +/- rituximab) + XBRT.*

What if CNS involvement is present?
In such cases, systemic and/or intrathecal chemo is deployed as well

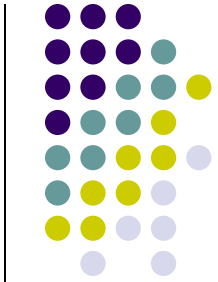
*External-beam radiation therapy

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
  - Hematologic
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

What is the prognosis for PVRL?

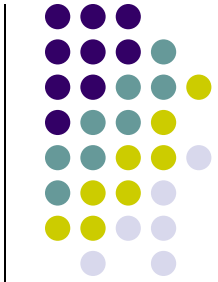
Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
  - Hematologic
    - Lymphoid
      - **Primary vitreoretinal lymphoma**
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

What is the prognosis for PVRL?
Untreated, it is dismal—on the order of a couple of months.
With treatment the 5-year survival rate is 60%.

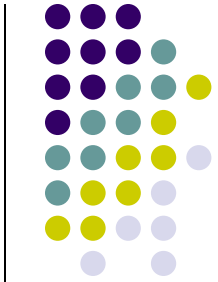
Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - **Secondary to systemic lymphoma**
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
      - Rb

Is it common for systemic lymphoma to 'masquerade' as intraocular lymphoma?

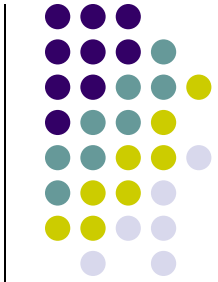
Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - **Secondary to systemic lymphoma**
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
      - Rb

Is it common for systemic lymphoma to 'masquerade' as intraocular lymphoma?
No, it is very rare

Masquerade Syndrome

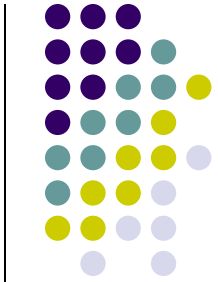


- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
      - Rb
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - **Secondary to systemic lymphoma**
      - Lymphoproliferative dz

Is it common for systemic lymphoma to 'masquerade' as intraocular lymphoma?
No, it is very rare

How does systemic lymphoma masquerade syndrome present?

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
      - Uveal melanoma
      - Rb
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - **Secondary to systemic lymphoma**
      - Lymphoproliferative dz

Is it common for systemic lymphoma to 'masquerade' as intraocular lymphoma?
No, it is very rare

How does systemic lymphoma masquerade syndrome present?
In a manner essentially identical to PVRL, but **without the CNS manifestations**

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemi
      - Leukem
    - Lymphoid
      - Primary vitreoretinal lymphon
      - Secondary to systemic lymph
      - **Lymphoproliferative dz**

How does uveal lymphoid proliferation masquerade syndrome present?

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemi
      - Leukem
    - Lymphoid
      - Primary vitreoretinal lymphon
      - Secondary to systemic lymph
      - **Lymphoproliferative dz**

How does uveal lymphoid proliferation masquerade syndrome present?
The posterior findings are similar to those of PVRL.

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemi
      - Leukem
    - Lymphoid
      - Primary vitreoretinal lymphon
      - Secondary to systemic lymph
      - **Lymphoproliferative dz**

How does uveal lymphoid proliferation masquerade syndrome present?
The posterior findings are similar to those of PVRL. However, lymphoproliferative disease is more likely to have an acute anterior-uveitis response—a red angry eye with pain/photophobia and increased IOP (from cells clogging the TM).

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemi
      - Leukem
    - Lymphoid
      - Primary vitreoretinal lymphon
      - Secondary to systemic lymph
      - **Lymphoproliferative dz**

How does uveal lymphoid proliferation masquerade syndrome present?
The posterior findings are similar to those of PVRL. However, lymphoproliferative disease is more likely to have an acute anterior-uveitis response—a red angry eye with pain/photophobia and increased IOP (from cells clogging the TM).

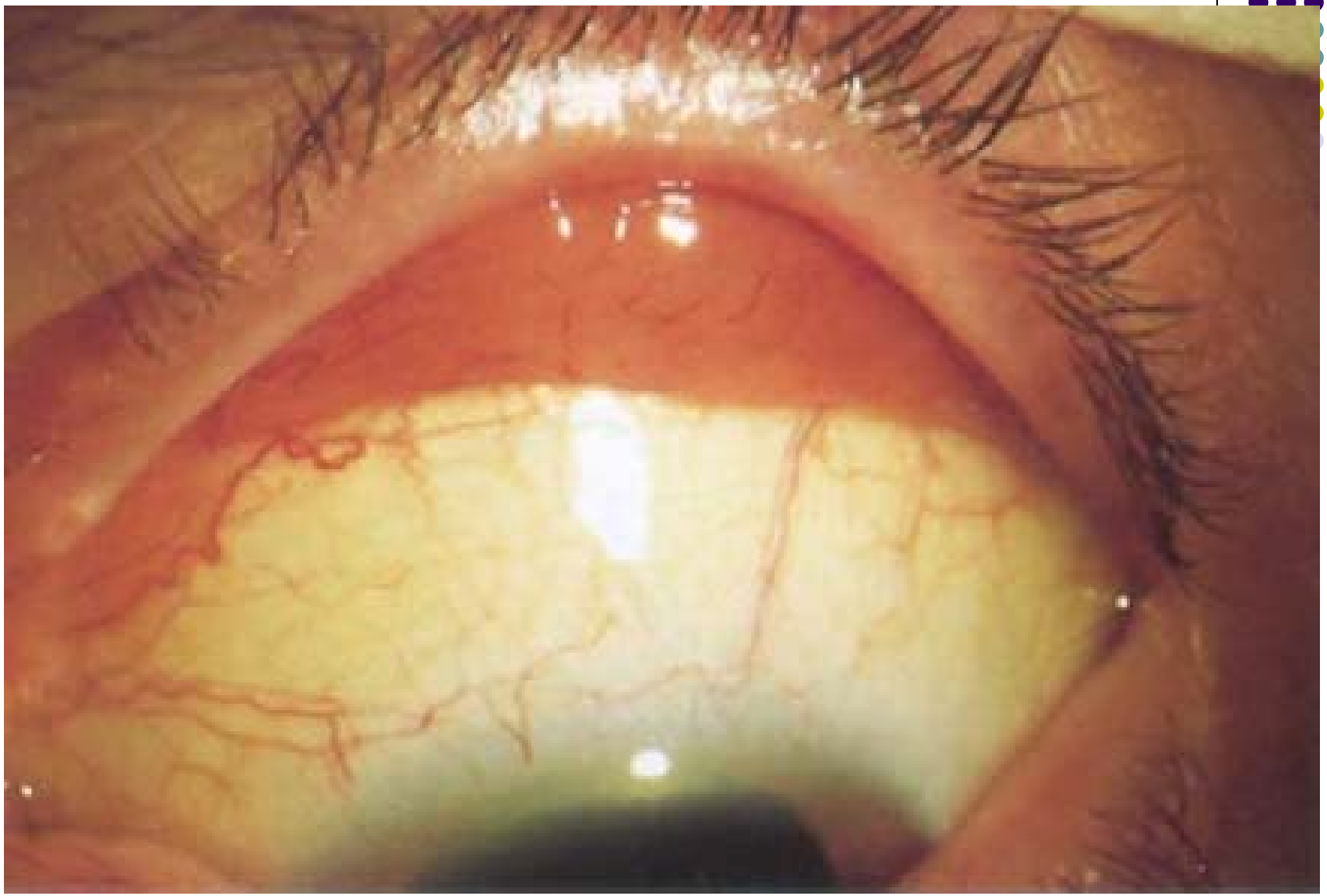
What additional anterior-segment finding may occur?

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemi
      - Leukem
    - Lymphoid
      - Primary vitreoretinal lymphon
      - Secondary to systemic lymph
      - **Lymphoproliferative dz**

How does uveal lymphoid proliferation masquerade syndrome present?
The posterior findings are similar to those of PVRL. However, lymphoproliferative disease is more likely to have an acute anterior-uveitis response—a red angry eye with pain/photophobia and increased IOP (from cells clogging the TM).

What additional anterior-segment finding may occur?
The presence of **salmon-pink nodules** under the conj



Lymphoproliferative disease: Sub-conj salmon patch

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemi
      - Leukem
    - Lymphoid
      - Primary vitreoretinal lymphon
      - Secondary to systemic lymph
      - **Lymphoproliferative dz**

How does uveal lymphoid proliferation masquerade syndrome present?
The posterior findings are similar to those of PVRL. However, lymphoproliferative disease is more likely to have an acute anterior-uveitis response—a red angry eye with pain/photophobia and increased IOP (from cells clogging the TM).

What additional anterior-segment finding may occur?
The presence of **salmon-pink nodules** under the conj

But such nodules can occur in lymphoma as well. How do these differ?

Masquerade Syndrome

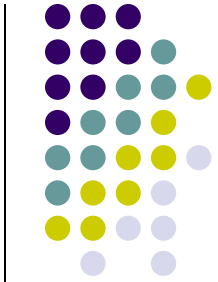
- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemi
      - Leukem
    - Lymphoid
      - Primary vitreoretinal lymphon
      - Secondary to systemic lymph
      - **Lymphoproliferative dz**

How does uveal lymphoid proliferation masquerade syndrome present?
The posterior findings are similar to those of PVRL. However, lymphoproliferative disease is more likely to have an acute anterior-uveitis response—a red angry eye with pain/photophobia and increased IOP (from cells clogging the TM).

What additional anterior-segment finding may occur?
The presence of **salmon-pink nodules** under the conj

But such nodules can occur in lymphoma as well. How do these differ?
Unlike the mobile conj nodules seen in lymphoma, the nodules in lymphoproliferative dz are **firmly fixed** to the underlying sclera

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemi
      - Leukem
    - Lymphoid
      - Primary vitreoretinal lymphon
      - Secondary to systemic lymph
      - **Lymphoproliferative dz**

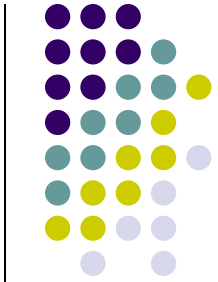
How does uveal lymphoid proliferation masquerade syndrome present?
The posterior findings are similar to those of PVRL. However, lymphoproliferative disease is more likely to have an acute anterior-uveitis response—a red angry eye with pain/photophobia and increased IOP (from cells clogging the TM).

What additional anterior-segment finding may occur?
The presence of **salmon-pink nodules** under the conj

But such nodules can occur in lymphoma as well. How do these differ?
Unlike the mobile conj nodules seen in lymphoma, the nodules in lymphoproliferative dz are **firmly fixed** to the underlying sclera

How is the diagnosis made?

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
  - Hematologic
    - Leukemi
      - Leukem
    - Lymphoid
      - Primary vitreoretinal lymphon
      - Secondary to systemic lymph
      - **Lymphoproliferative dz**

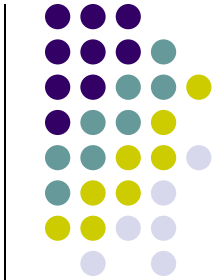
How does uveal lymphoid proliferation masquerade syndrome present?
The posterior findings are similar to those of PVRL. However, lymphoproliferative disease is more likely to have an acute anterior-uveitis response—a red angry eye with pain/photophobia and increased IOP (from cells clogging the TM).

What additional anterior-segment finding may occur?
The presence of **salmon-pink nodules** under the conj

But such nodules can occur in lymphoma as well. How do these differ?
Unlike the mobile conj nodules seen in lymphoma, the nodules in lymphoproliferative dz are **firmly fixed** to the underlying sclera

How is the diagnosis made?
Only via biopsy, which will reveal well-differentiated, mature cells

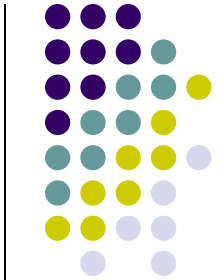
Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
  - Hematologic
    - Leukemic
      - **Leukemia**
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

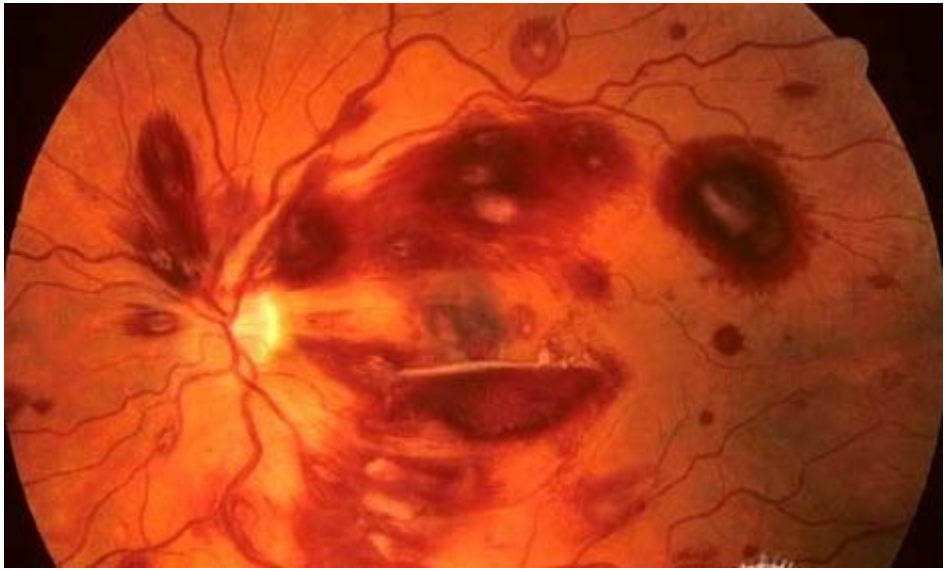
How does leukemic masquerade syndrome present?

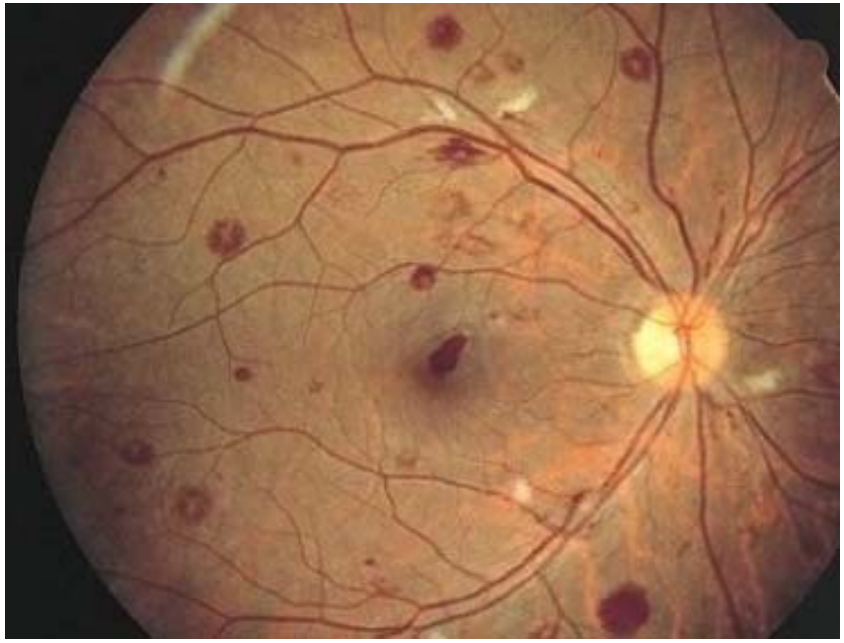
Masquerade Syndrome

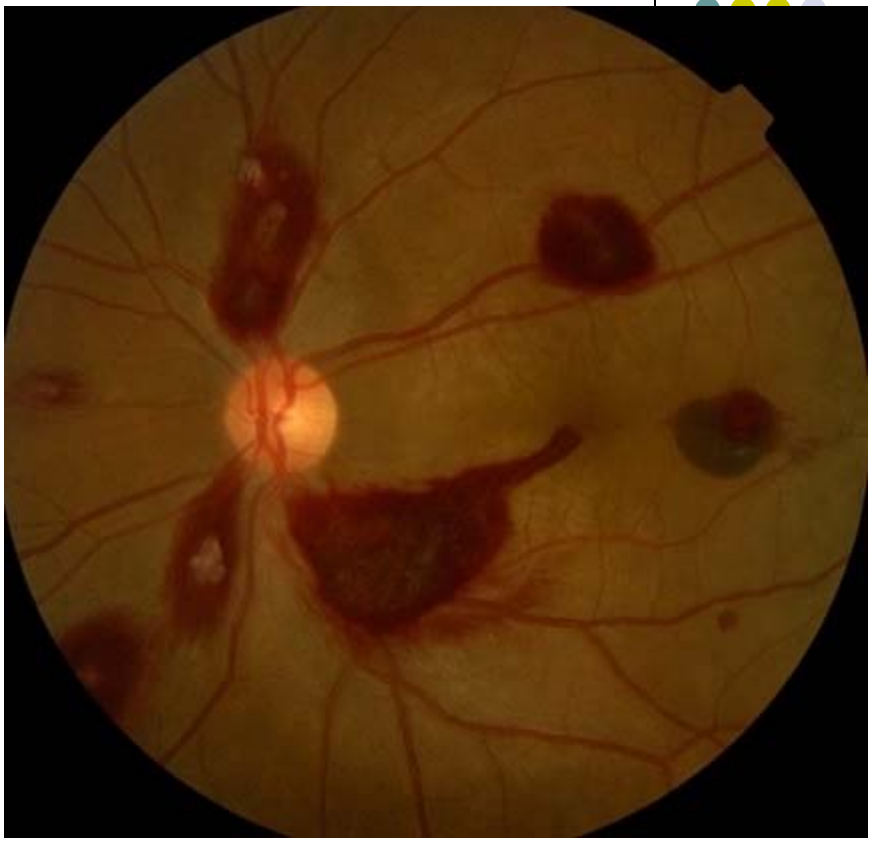


- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
  - Hematologic
    - Leukemic
      - **Leukemia**
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How does leukemic masquerade syndrome present?
The posterior findings are primarily **retinal**, and include hemorrhages (which may be white-centered), cotton-wool spots and peripheral neo

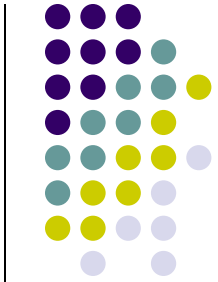






Leukemia: CWS and white-centered hemorrhages

Masquerade Syndrome

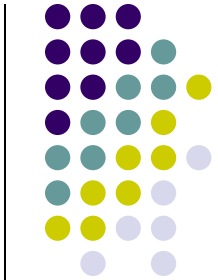


- Nonneoplastic
- Neoplastic
  - *What is the eponymous name for white-centered hemorrhages?*
  - Hematologic
    - Leukemic
      - **Leukemia**
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How does leukemic masquerade syndrome present?

The posterior findings are primarily **retinal**, and include hemorrhages (which may be white-centered), cotton-wool spots and peripheral neo

Masquerade Syndrome

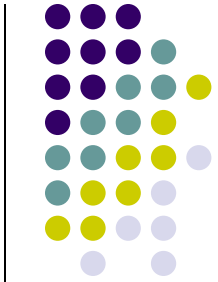


- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - **Leukemia**

What is the eponymous name for white-centered hemorrhages?
Roth spots

How does leukemic masquerade syndrome present?
The posterior findings are primarily **retinal**, and include hemorrhages (which may be white-centered), cotton-wool spots and peripheral neo

Masquerade Syndrome



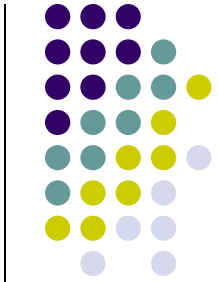
- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - **Leukemia**

What is the eponymous name for white-centered hemorrhages?
Roth spots

What is the DDx for Roth spots?
--Leukemia
--
--
--
--(There are many others)

How does leukemic masquerade syndrome present?
The posterior findings are primarily **retinal**, and include hemorrhages (which may be white-centered), cotton-wool spots and peripheral neo

Masquerade Syndrome



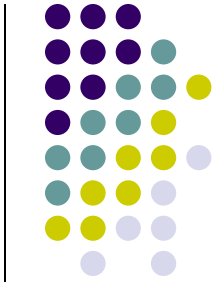
- Nonneoplastic
- Neoplastic
  - Hematologic
    - Leukemic
      - **Leukemia**
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

What is the eponymous name for white-centered hemorrhages?
Roth spots

What is the DDx for Roth spots?
--Leukemia
--Subacute bacterial endocarditis
--Anemia
--Endophthalmitis
--(There are many others)

How does leukemic masquerade syndrome present?
The posterior findings are primarily **retinal**, and include hemorrhages (which may be white-centered), cotton-wool spots and peripheral neo

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Hematologic
    - Leukemic
      - **Leukemia**
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

What is the eponymous name for white-centered hemorrhages?

Roth spots

Of these, which is most commonly associated with Roth spots?

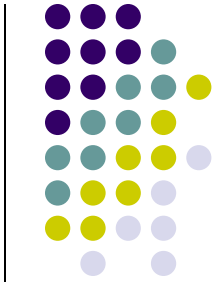
What is the DDx

--Leukemia?
--Subacute bacterial endocarditis?
--Anemia?
--Endophthalmitis?
--(There are many others)

How does leukemic masquerade syndrome present?

The posterior findings are primarily **retinal**, and include hemorrhages (which may be white-centered), cotton-wool spots and peripheral neo

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - **Leukemia**

What is the eponymous name for white-centered hemorrhages?
Roth spots

Of these, which is most commonly associated with Roth spots?
SABE

What is the DDx
--Leukemia
--Subacute bacterial endocarditis
--Anemia
--Endophthalmitis
--(There are many others)

How does leukemic masquerade syndrome present?
The posterior findings are primarily **retinal**, and include hemorrhages (which may be white-centered), cotton-wool spots and peripheral neo

Masquerade Syndrome

Nonneoplastic

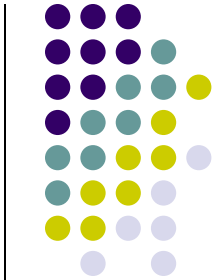
Neoplastic

What is the eponymous name for white-centered hemorrhages?

Roth spots

Note: There is inconsistency across Academy sources with regard to the term *Roth spots:*
--Early in the BCSC *Path* book, *Roth spot* and *white-centered hemorrhage* are used interchangeably and are said to occur "in a number of conditions;" later, the term ***pseudo-****Roth spot* is used to refer to white-centered hemorrhages secondary to leukemia. (Per the *Master Index*, *pseudo-Roth spot* appears nowhere else in the *BCSC*.)
--The *Uveitis* book uses *Roth spots* when referring to white-centered hemorrhages secondary to bacterial endophthalmitis, but not when referring to those secondary to leukemia (these are termed 'white-centered hemorrhages').
--The *Peds* book simply says retinal hemorrhages in leukemia "may have white centers."
--The online source *EyeWiki* uses *Roth spot* to refer to white-centered hemorrhages of *any* cause.
--Puzzlingly, neither *Roth spot* nor *white-centered hemorrhage* appear in the index of the *Retina* book.
What's the correct usage? I dunno. Caveat emptor.

Masquerade Syndrome

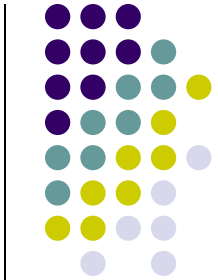


- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
  - Hematologic
    - Leukemic
      - **Leukemia**
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How does leukemic masquerade syndrome present?
The posterior findings are primarily **retinal**, and include hemorrhages (which may be white-centered), cotton-wool spots and peripheral neo

What about anterior findings?

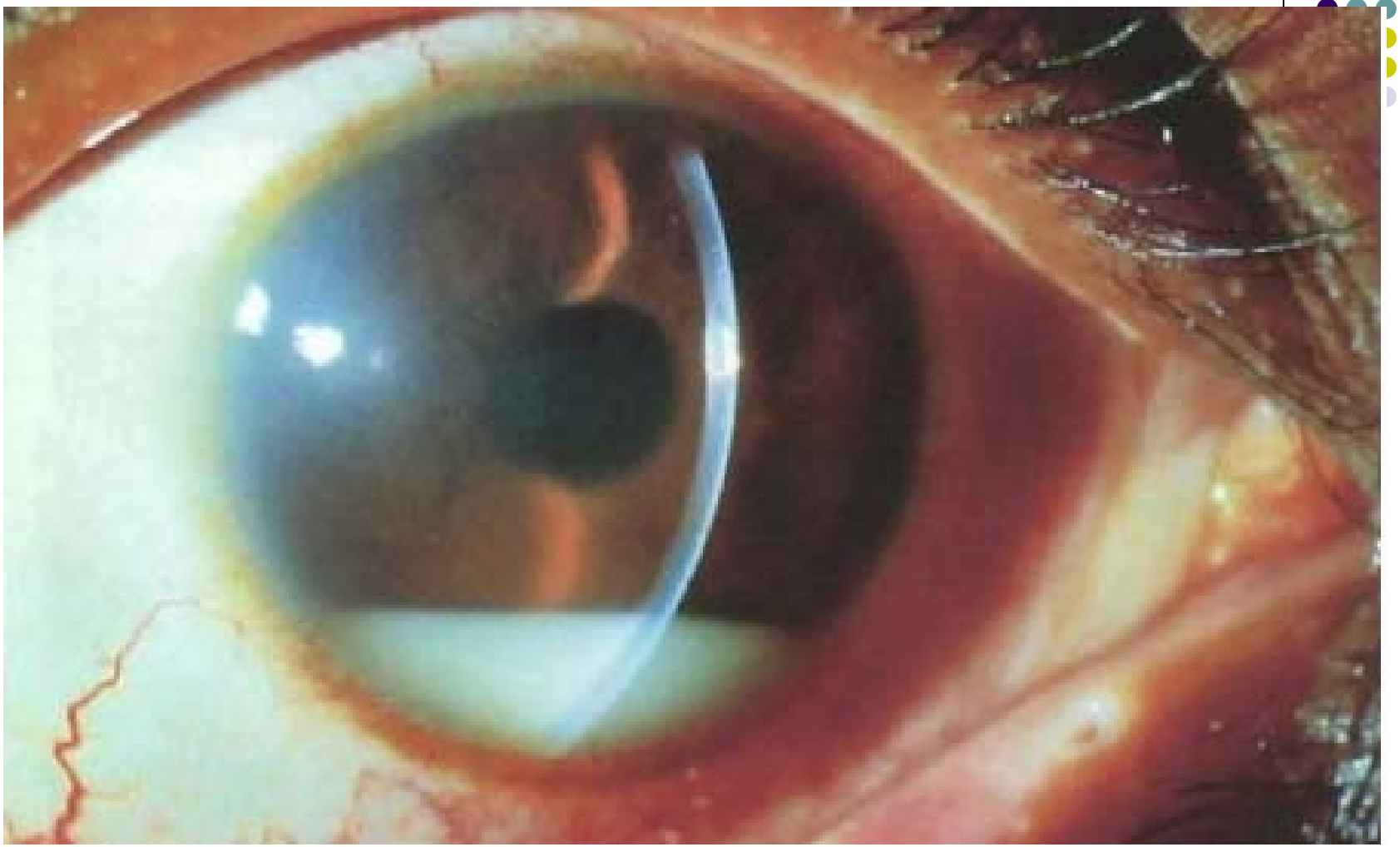
Masquerade Syndrome



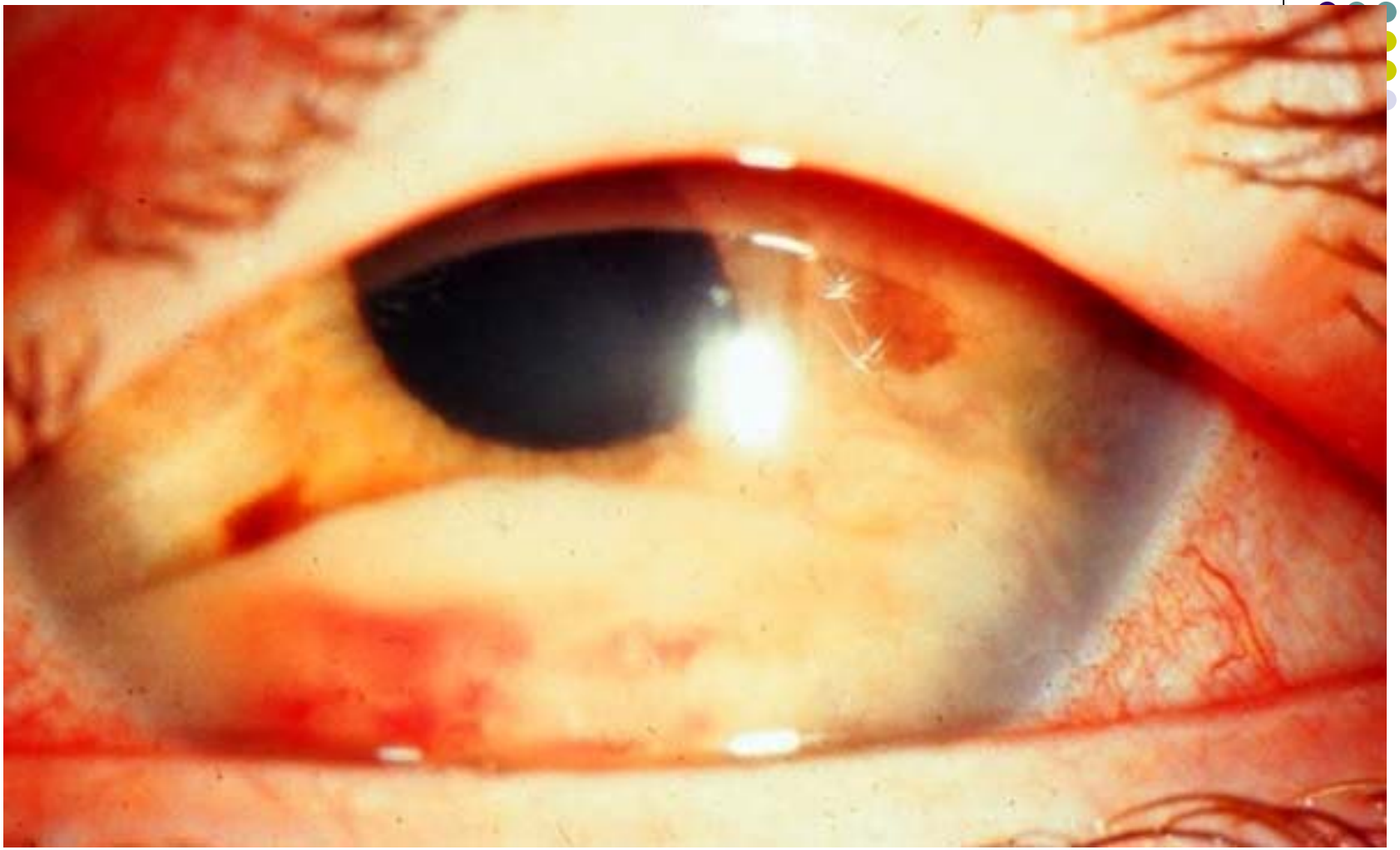
- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
    - Primary
  - Hematologic
    - Leukemic
      - **Leukemia**
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

How does leukemic masquerade syndrome present?
The posterior findings are primarily **retinal**, and include hemorrhages (which may be white-centered), cotton-wool spots and peripheral neo

What about anterior findings?
These are common, and can include hyphema, iris heterochromia, and/or pseudohypopyon

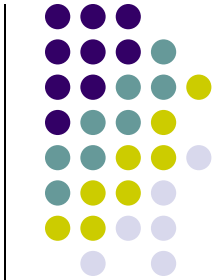


Leukemia: Pseudohypopyon in ALL



Leukemia: Pseudohypopyon with hyphema in ALL

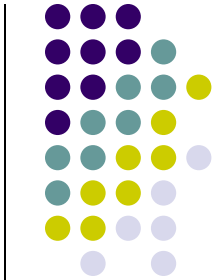
Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - Solid
    - Mets
    - Primary
      - **Uveal melanoma (multiple slide-sets)**

A small minority of uveal melanomas present in an inflammatory fashion (ie, with some form of uveitis, up to and including an endophthalmitis/panuveitis picture). Uveal melanomas are discussed in detail in multiple silde-sets.

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - S
    - Primary
      - Uveal me
      - **Rb**

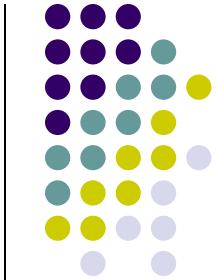
What are the three variants/forms of retinoblastoma?

--

--

--

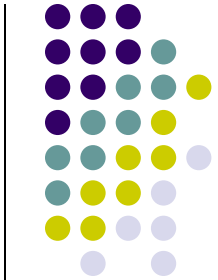
Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - S
    - Primary
      - Uveal me
      - **Rb**

What are the three variants/forms of retinoblastoma?
--Exophytic
--Endophytic
--Diffuse infiltrating

Masquerade Syndrome

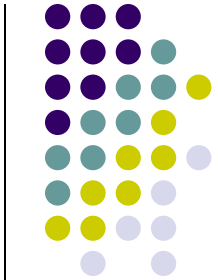


- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - S
    - Primary
      - Uveal me
      - **Rb**

What are the three variants/forms of retinoblastoma?
--Exophytic
--Endophytic
--Diffuse infiltrating

Which form can masquerade as uveitis?

Masquerade Syndrome

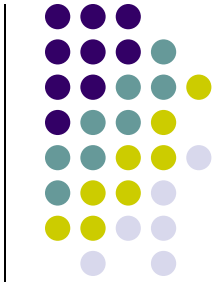


- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - S
    - Primary
      - Uveal me
      - **Rb**

What are the three variants/forms of retinoblastoma?
--Exophytic
--Endophytic
--Diffuse infiltrating

Which form can masquerade as uveitis?
Diffuse infiltrating

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - S
    - Primary
      - Uveal me
      - **Rb**

What are the three variants/forms of retinoblastoma?
--Exophytic
--Endophytic
--Diffuse infiltrating

Which form can masquerade as uveitis?
Diffuse infiltrating

Diffuse infiltrating Rb often presents with a (pseudo)hypopyon. What two attributes of the hypopyon should alert you to the possibility it stems from diffuse infiltrating Rb?

Masquerade Syndrome

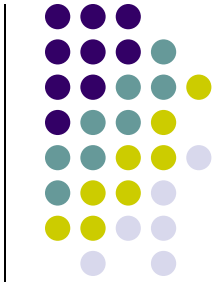
- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - S
    - Primary
      - Uveal me
      - **Rb**

What are the three variants/forms of retinoblastoma?
--Exophytic
--Endophytic
--Diffuse infiltrating

Which form can masquerade as uveitis?
Diffuse infiltrating

Diffuse infiltrating Rb often presents with a (pseudo)hypopyon. What two attributes of the hypopyon should alert you to the possibility it stems from diffuse infiltrating Rb?
--If it a structural tendency of the hypopyon
--If it is in color

Masquerade Syndrome



- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - S
    - Primary
      - Uveal me
      - **Rb**

What are the three variants/forms of retinoblastoma?
--Exophytic
--Endophytic
--Diffuse infiltrating

Which form can masquerade as uveitis?
Diffuse infiltrating

Diffuse infiltrating Rb often presents with a (pseudo)hypopyon. What two attributes of the hypopyon should alert you to the possibility it stems from diffuse infiltrating Rb?
--If it shifts easily with head movements
--If it is **snow-white** in color

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
  - S
    - Primary
      - Uveal me
      - Leukemia

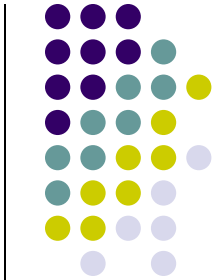
What are the three variants/forms of retinoblastoma?
--Exophytic
--Endophytic
--Diffuse infiltrating

Which form can masquerade as uveitis?
Diffuse infiltrating

A hypopyon that shifts with head movements is characteristic of what 'genuine' uveitis?

hypopyon should alert you to the possibility it stems from diffuse infiltrating Rb?
--If it shifts easily with head movements
--If it is **snow-white** in color

Masquerade Syndrome



Nonneoplastic

Neoplastic

Hematologic

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

S

Primary

What are the three variants/forms of retinoblastoma?
--Exophytic
--Endophytic
--Diffuse infiltrating

Which form can masquerade as uveitis?
Diffuse infiltrating

A hypopyon that shifts with head movements is characteristic of what 'genuine' uveitis?
Behçet

hypopyon should alert you to the possibility it stems from diffuse infiltrating Rb?
--If it shifts easily with head movements
--If it is **snow-white** in color

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
  - S
    - Primary
      - Uveal me
      - **Rb (R2)**

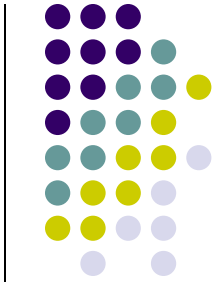
What are the three variants/forms of retinoblastoma?
--Exophytic
--Endophytic
--Diffuse infiltrating

Which form can masquerade as uveitis?
Diffuse infiltrating

Diffuse infiltrating Rb often presents with a (pseudo)hypopyon. What two attributes of the hypopyon should alert you to the possibility it stems from diffuse infiltrating Rb?
--If it shifts easily with head movements
--If it is **snow-white** in color

For more on diffuse-infiltrating Rb (and other forms), see slide-set R2.

Masquerade Syndrome



Nonneoplastic

Neoplastic

Which two primary malignancies are most likely to be implicated in masquerade syndrome?

Solid

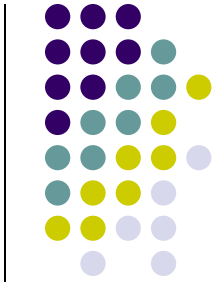
Mets

- *?*
- *?*

elanoma (multiple slide-sets)

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



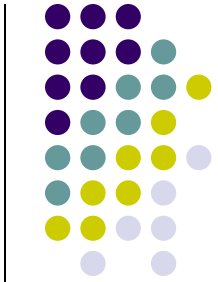
- Nonneoplastic
- Neoplastic
  - *Which two primary malignancies are most likely to be implicated in masquerade syndrome?*
    Lung and breast
  - Solid
    - Mets
      - **Lung**
      - **Breast**
    - elanoma (multiple slide-sets)
  - Primary vitreoretinal lymphoma
  - Secondary to systemic lymphoma
  - Lymphoproliferative dz

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - *Which two primary malignancies are most likely to be implicated in masquerade syndrome?*
    Lung and breast

    What is the most common manifestation in the eye?
  - Solid
    - Mets
      - **Lung**
      - **Breast**
    - elanoma (multiple slide-sets)
  - Primary vitreoretinal lymphoma
  - Secondary to systemic lymphoma
  - Lymphoproliferative dz

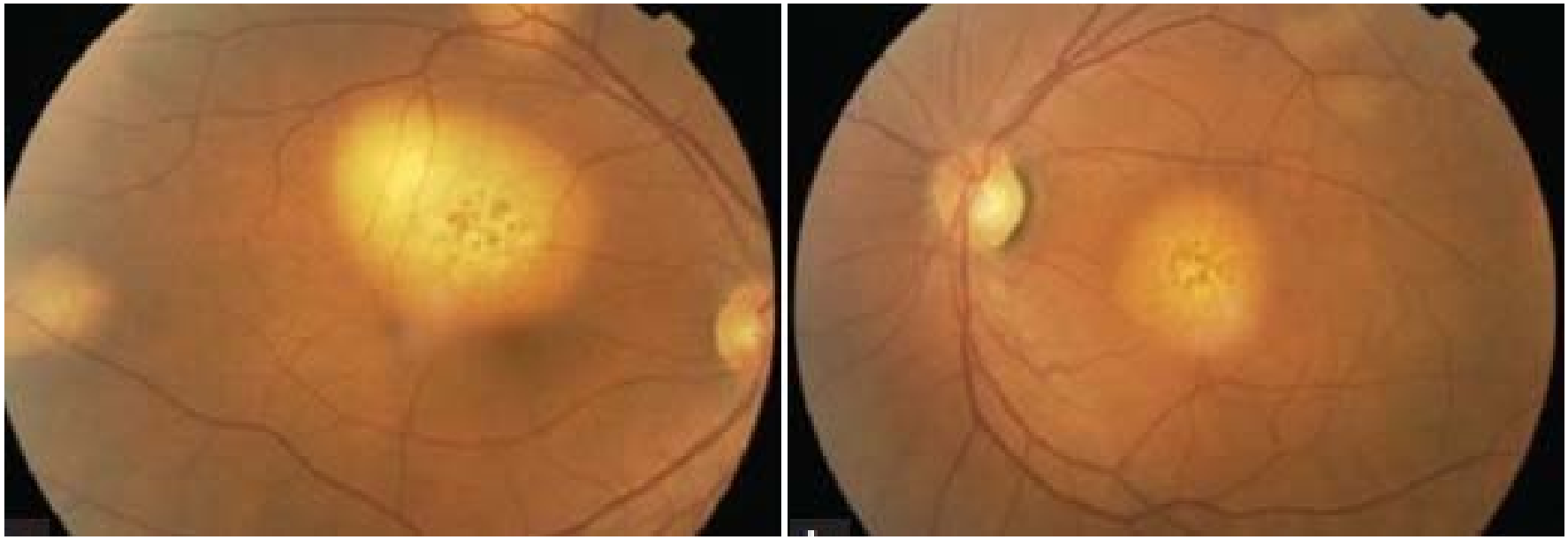
Masquerade Syndrome



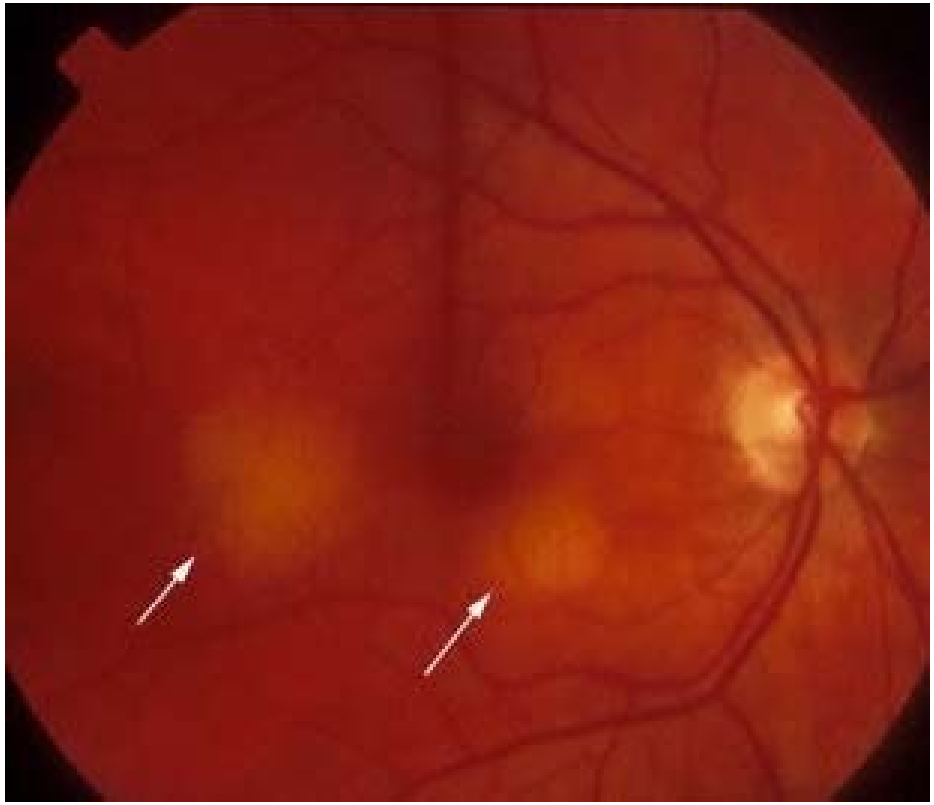
- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
      - **Lung**
      - **Breast**
    - elanoma (multiple slide-sets)
  - Primary vitreoretinal lymphoma
  - Secondary to systemic lymphoma
  - Lymphoproliferative dz

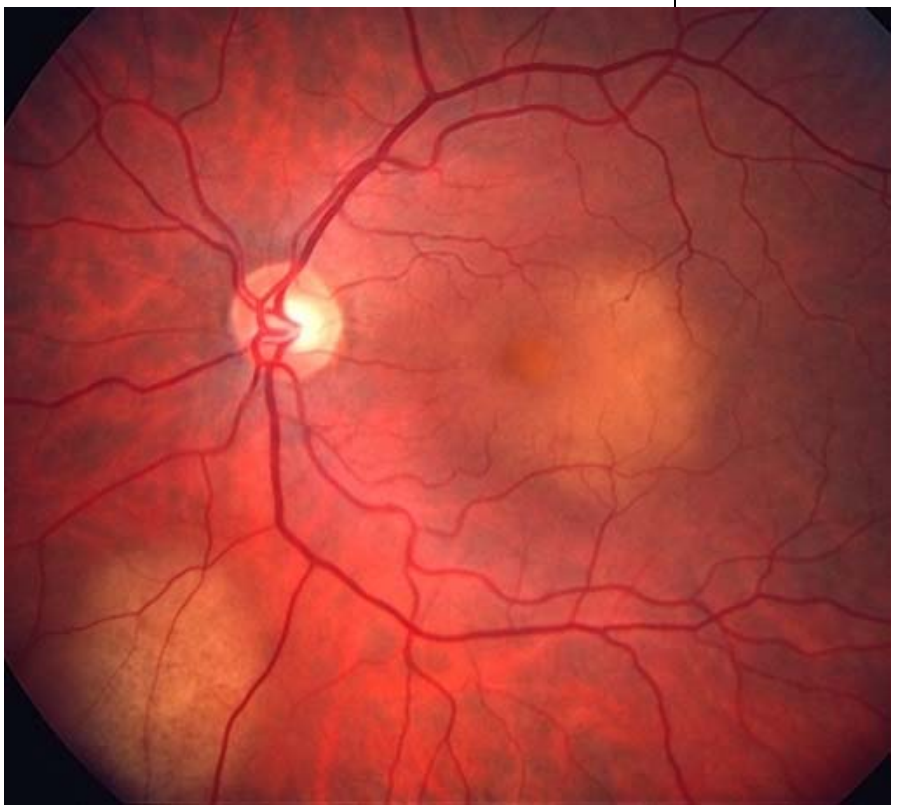
Which two primary malignancies are most likely to be implicated in masquerade syndrome?
Lung and breast

What is the most common manifestation in the eye?
Bilateral multifocal choroidal lesions mimicking choroiditis



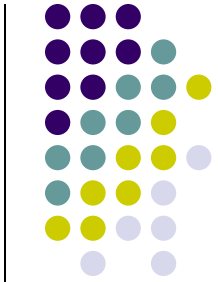
Bilateral metastatic lung cancer





Metastatic breast cancer

Masquerade Syndrome



Nonneoplastic

Neoplastic

Which two primary malignancies are most likely to be implicated in masquerade syndrome?
Lung and breast

What is the most common manifestation in the eye?
Bilateral multifocal choroidal lesions mimicking choroiditis

Is vitritis present?

Solid

Mets
- **Lung**
- **Breast**

elanoma (multiple slide-sets)

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome

Nonneoplastic

Neoplastic

Which two primary malignancies are most likely to be implicated in masquerade syndrome?
Lung and breast

What is the most common manifestation in the eye?
Bilateral multifocal choroidal lesions mimicking choroiditis

Is vitritis present?
Usually, but not always

Solid

Mets

- **Lung**
- **Breast**

elanoma (multiple slide-sets)

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
      - **Lung**
      - **Breast**
    - elanoma (multiple slide-sets)
  - Primary vitreoretinal lymphoma
  - Secondary to systemic lymphoma
  - Lymphoproliferative dz

Which two primary malignancies are most likely to be implicated in masquerade syndrome?
Lung and breast

What is the most common manifestation in the eye?
Bilateral multifocal choroidal lesions mimicking choroiditis

Is vitritis present?
Usually, but not always

Can the mets mimic an acute **anterior** *uveitis?*

Masquerade Syndrome

- Nonneoplastic
- Neoplastic
  - Solid
    - Mets
      - **Lung**
      - **Breast**
    - elanoma (multiple slide-sets)
  - Primary vitreoretinal lymphoma
  - Secondary to systemic lymphoma
  - Lymphoproliferative dz

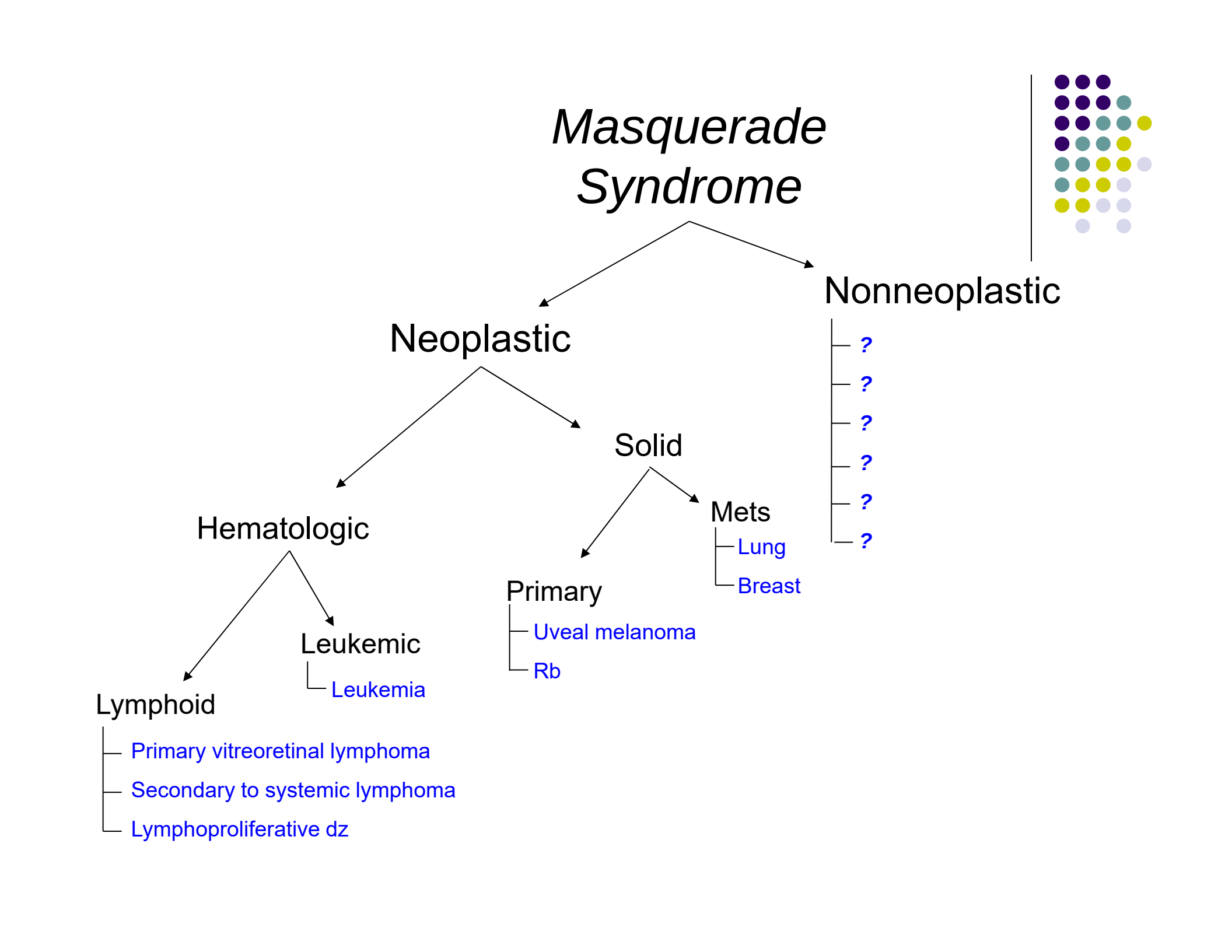
Which two primary malignancies are most likely to be implicated in masquerade syndrome?
Lung and breast

What is the most common manifestation in the eye?
Bilateral multifocal choroidal lesions mimicking choroiditis

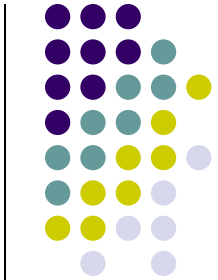
Is vitritis present?
Usually, but not always

Can the mets mimic an acute **anterior** *uveitis?*
Yes, although this is a less common presentation

Metastatic lung cancer: Pseudohypopyon

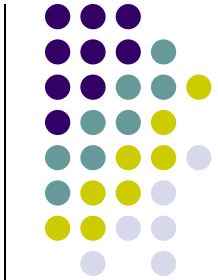

Masquerade Syndrome
Nonneoplastic
?
?
?
?
?
?
Neoplastic
Solid
Mets
Lung
Breast
Primary
Uveal melanoma
Rb
Hematologic
Leukemic
Leukemia
Lymphoid
Primary vitreoretinal lymphoma
Secondary to systemic lymphoma
Lymphoproliferative dz

Masquerade Syndrome



- Nonneoplastic
  - Retinitis pigmentosa
  - Ocular ischemic syndrome
  - Chronic rhegmatogenous RD
  - Intraocular foreign body
  - Pigment dispersion syndrome
  - Juvenile xanthogranuloma
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - Solid
    - Primary
      - Uveal melanoma
      - Rb
    - Mets
      - Lung
      - Breast

Masquerade Syndrome



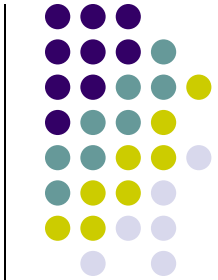
What two common findings in RP might (mis)lead one to conclude the pt had a uveitic condition?

--

--

- Nonneoplastic
  - **Retinitis pigmentosa**
  - Ocular ischemic syndrome
  - Chronic rhegmatogenous RD
  - Intraocular foreign body
  - Pigment dispersion syndrome
  - Juvenile xanthogranuloma
- Solid
  - Mets
    - Lung
    - Breast
  - Primary
    - Uveal melanoma
    - Rb
- Hematologic
  - Leukemic
    - Leukemia
  - Lymphoid
    - Primary vitreoretinal lymphoma
    - Secondary to systemic lymphoma
    - Lymphoproliferative dz

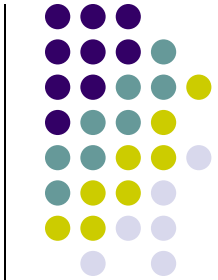
Masquerade Syndrome



What two common findings in RP might (mis)lead one to conclude the pt had a uveitic condition?
--Vitreous cell
--Cystoid macular changes

- Nonneoplastic
  - **Retinitis pigmentosa**
  - Ocular ischemic syndrome
  - Chronic rhegmatogenous RD
  - Intraocular foreign body
  - Pigment dispersion syndrome
  - Juvenile xanthogranuloma
- Solid
  - Mets
    - Lung
    - Breast
  - Primary
    - Uveal melanoma
    - Rb
- Hematologic
  - Leukemic
    - Leukemia
  - Lymphoid
    - Primary vitreoretinal lymphoma
    - Secondary to systemic lymphoma
    - Lymphoproliferative dz

Masquerade Syndrome



What two common findings in RP might (mis)lead one to conclude the pt had a uveitic cor

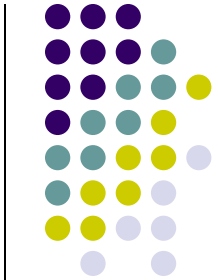
--Vitreous cell

--Cystoid macula

But if they have vitreous cell, doesn't that mean they have vitritis, and therefore **do** *have a uveitic condition?*

- Nonneoplastic
  - **Retinitis pigmentosa**
  - Ocular ischemic syndrome
  - Chronic rhegmatogenous RD
  - Intraocular foreign body
  - Pigment dispersion syndrome
  - Juvenile xanthogranuloma
- Mets
  - Lung
  - Breast
- Primary
  - Uveal melanoma
  - Rb
- Hematologic
  - Leukemic
    - Leukemia
  - Lymphoid
    - Primary vitreoretinal lymphoma
    - Secondary to systemic lymphoma
    - Lymphoproliferative dz

Masquerade Syndrome

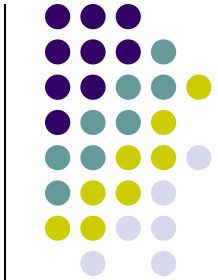


What two common findings in RP might (mis)lead one to conclude the pt had a uveitic cor
--Vitreous cell
--Cystoid macula

But if they have vitreous cell, doesn't that mean they have vitritis, and therefore **do** *have a uveitic condition?*
No, because the vitreous cells in RP are not inflammatory in origin; rather, they are mainly degenerated RPE cells liberated from Bruch's membrane

- Nonneoplastic
  - **Retinitis pigmentosa**
  - Ocular ischemic syndrome
  - Chronic rhegmatogenous RD
  - Intraocular foreign body
  - Pigment dispersion syndrome
  - Juvenile xanthogranuloma
- Mets
  - Lung
  - Breast
- Primary
  - Uveal melanoma
  - Rb
- Hematologic
  - Leukemic
    - Leukemia
  - Lymphoid
    - Primary vitreoretinal lymphoma
    - Secondary to systemic lymphoma
    - Lymphoproliferative dz

Masquerade Syndrome



Nonneoplastic

- **Retinitis pigmentosa**
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

What two common findings in RP might (mis)lead one to conclude the pt had a uveitic condition?
--Vitreous cell
--Cystoid macular changes

What specific sorts of cystoid macular changes occur in RP?

Hematologic

Leukemic

- Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

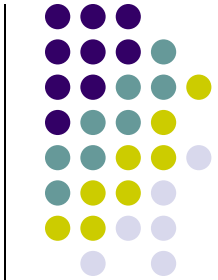
Primary

- Uveal melanoma
- Rb

Mets

- Lung
- Breast

Masquerade Syndrome



Nonneoplastic

- **Retinitis pigmentosa**
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

What two common findings in RP might (mis)lead one to conclude the pt had a uveitic condition?
--Vitreous cell
--Cystoid macular changes

What specific sorts of cystoid macular changes occur in RP?
Cystoid macular edema (CME), and cystoid macular degeneration (CMD)

Hematologic

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Leukemic

- Leukemia

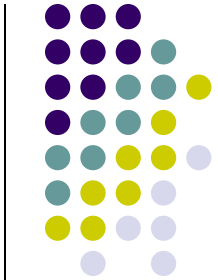
Primary

- Uveal melanoma
- Rb

Mets

- Lung
- Breast

Masquerade Syndrome



Nonneoplastic

- **Retinitis pigmentosa**
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

What two common findings in RP might (mis)lead one to conclude the pt had a uveitic condition?
--Vitreous cell
--Cystoid macular changes

What specific sorts of cystoid macular changes occur in RP?
Cystoid macular edema (CME), and cystoid macular degeneration (CMD)

What's the difference?

Hematologic

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Leukemic

- Leukemia

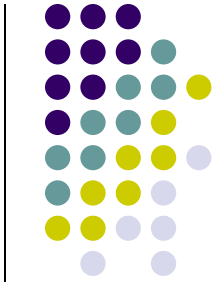
Primary

- Uveal melanoma
- Rb

Mets

- Lung
- Breast

Masquerade Syndrome



Nonneoplastic

- **Retinitis pigmentosa**
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

What two common findings in RP might (mis)lead one to conclude the pt had a uveitic condition?
--Vitreous cell
--Cystoid macular changes

What specific sorts of cystoid macular changes occur in RP?
Cystoid macular edema (CME), and cystoid macular degeneration (CMD)

What's the difference?
CME leaks on FA; CMD doesn't

Hematologic

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Leukemic

- Leukemia

Primary

- Uveal melanoma
- Rb

Mets

- Lung
- Breast

Masquerade Syndrome

What is ocular ischemic syndrome *(OIS)?*

Nonneoplastic

- RP
- **OIS**
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

H

de-sets)

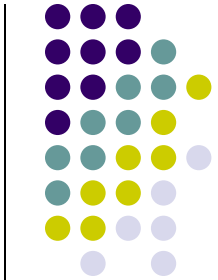
Rb (R2)

Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



What is ocular ischemic syndrome *(OIS)?*
A constellation of ocular abnormalities owing to prolonged hypoperfusion of the globe

Nonneoplastic

- RP
- **OIS**
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

H

de-sets)

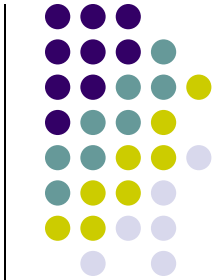
Rb (R2)

Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



What is ocular ischemic syndrome *(OIS)?*
A constellation of ocular abnormalities owing to prolonged hypoperfusion of the globe

What is the classic cause?

Nonneoplastic

- RP
- **OIS**
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

H

de-sets)

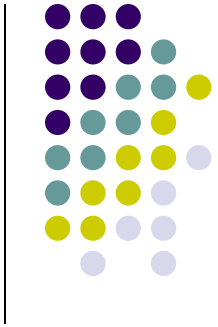
Rb (R2)

Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



What is ocular ischemic syndrome *(OIS)?*
A constellation of ocular abnormalities owing to prolonged hypoperfusion of the globe

What is the classic cause?
Ipsilateral carotid stenosis

Nonneoplastic

- RP
- **OIS**
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

H

de-sets)

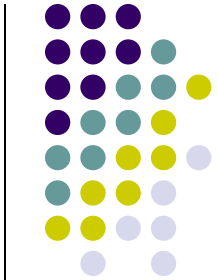
Rb (R2)

Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



What is ocular ischemic syndrome *(OIS)?*
A constellation of ocular abnormalities owing to prolonged hypoperfusion of the globe

What is the classic cause?
Ipsilateral carotid stenosis

Who is the typical pt?

Nonneoplastic
- RP
- **OIS**
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

H

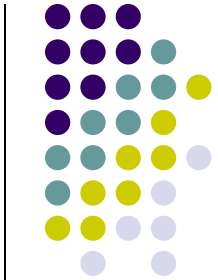
de-sets)

Rb (R2)

Leukemia

Lymphoid
- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



What is ocular ischemic syndrome *(OIS)?*
A constellation of ocular abnormalities owing to prolonged hypoperfusion of the globe

What is the classic cause?
Ipsilateral carotid stenosis

Who is the typical pt?
An elderly (65+) vasculopathic male

Nonneoplastic
- RP
- **OIS**
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

H

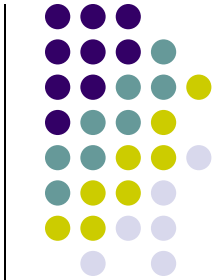
de-sets)

Rb (R2)

Leukemia

Lymphoid
- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



What is ocular ischemic syndrome *(OIS)?*
A constellation of ocular abnormalities owing to prolonged hypoperfusion of the globe

What is the classic cause?
Ipsilateral carotid stenosis

Who is the typical pt?
An elderly (65+) vasculopathic male

What four findings, common in OIS, might (mis)lead one to conclude the pt had a uveitic condition?
--
--
--
--

H

Nonneoplastic
- RP
- **OIS**
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

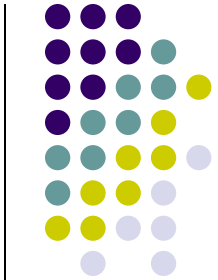
de-sets)

Rb (R2)

Leukemia

Lymphoid
- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



What is ocular ischemic syndrome *(OIS)?*
A constellation of ocular abnormalities owing to prolonged hypoperfusion of the globe

What is the classic cause?
Ipsilateral carotid stenosis

Who is the typical pt?
An elderly (65+) vasculopathic male

What four findings, common in OIS, might (mis)lead one to conclude the pt had a uveitic condition?
--AC cell and flare
--Low IOP
--NVI/NVA
--Cataract more advanced on that side

Nonneoplastic
- RP
- **OIS**
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

H

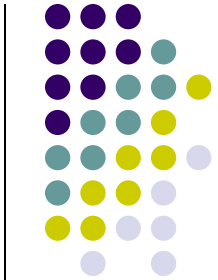
de-sets)

Rb (R2)

Leukemia

Lymphoid
- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



Nonneoplastic

- RP
- OIS
- **Chronic rhegmatogenous RD**
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Neoplastic

How *might a chronic rhegmatogenous retinal detachment mimic uveitis?*

Hematologic

Leukemic

- Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

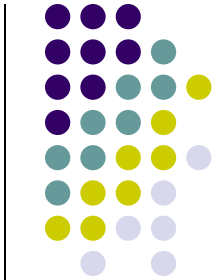
Primary

- Uveal melanoma
- Rb

Mets

- Lung
- Breast

Masquerade Syndrome



Nonneoplastic

- RP
- OIS
- **Chronic rhegmatogenous RD**
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Neoplastic

How *might a chronic rhegmatogenous retinal detachment mimic uveitis?* Liberated photoreceptor outer segments can find their way into the vitreous and/or anterior chamber, and give the erroneous impression of inflammation

Mets

- Lung
- Breast

Primary

- Uveal melanoma
- Rb

Hematologic

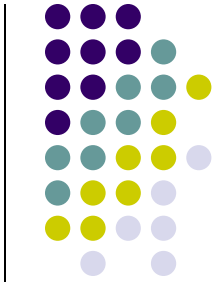
Leukemic

- Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- **Intraocular foreign body**
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

As masquerade syndromes go, how does intraocular foreign body (IOFB) differ from the others?

Primary

- Uveal melanoma
- Rb
- Breast

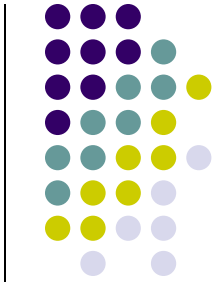
Leukemic

- Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- **Intraocular foreign body**
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

As masquerade syndromes go, how does intraocular foreign body (IOFB) differ from the others?

Unlike the others, what appears to be an inflammatory response in IOFB is in fact an inflammatory response--it's just not autoimmunologic in its origin

Primary

- Breast
- Uveal melanoma
- Rb

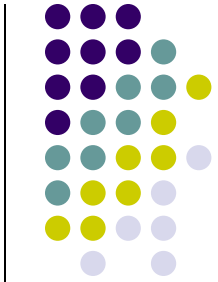
Leukemic

- Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- **Intraocular foreign body**
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

As masquerade syndromes go, how does intraocular foreign body (IOFB) differ from the others?
Unlike the others, what appears to be an inflammatory response in IOFB is in fact an inflammatory response--it's just not autoimmunologic in its origin

Why is it important to maintain an index of suspicion for the possibility of an IOFB?

Primary

- Breast
- Uveal melanoma
- Rb

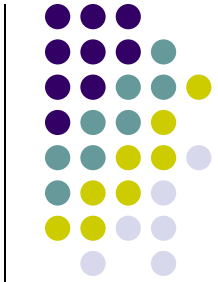
Leukemic

- Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

Masquerade Syndrome



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- **Intraocular foreign body**
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

As masquerade syndromes go, how does intraocular foreign body (IOFB) differ from the others?
Unlike the others, what appears to be an inflammatory response in IOFB is in fact an inflammatory response--it's just not autoimmunologic in its origin

Why is it important to maintain an index of suspicion for the possibility of an IOFB?
Because timely removal is usually curative, whereas delayed removal may result in permanent loss of visual function (or even the eye)

Primary

- Breast
- Uveal melanoma
- Rb

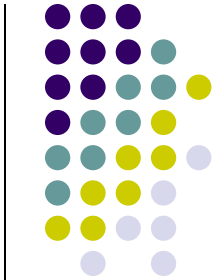
Leukemic

- Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

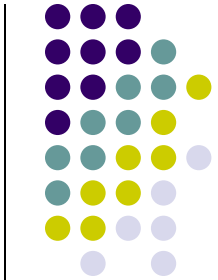
Masquerade Syndrome



- Nonneoplastic
  - RP
  - OIS
  - Chronic rhegmatogenous RD
  - IOFB
  - **Pigment dispersion syndrome**
  - Juvenile xanthogranuloma
- Neoplastic
  - Solid
    - Breast
    - Primary
      - Uveal melanoma
      - Rb
  - Leukemic
    - Leukemia
  - Lymphoid
    - Primary vitreoretinal lymphoma
    - Secondary to systemic lymphoma
    - Lymphoproliferative dz

How does pigment dispersion syndrome (PDS) mimic uveitis?

Masquerade Syndrome



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- **Pigment dispersion syndrome**
- Juvenile xanthogranuloma

Neoplastic

How does pigment dispersion syndrome (PDS) mimic uveitis?
The hallmark of PDS is the liberation of posterior iris pigment, which subsequently migrates into the anterior chamber. These pigment granules in the AC can be misconstrued as inflammatory cells.

Solid

- Breast

Primary

- Uveal melanoma
- Rb

Leukemic

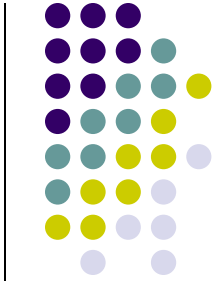
- Leukemia

Lymphoid

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?

e

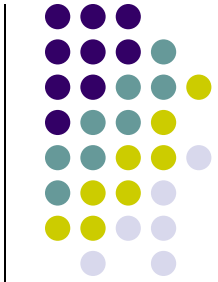


Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**



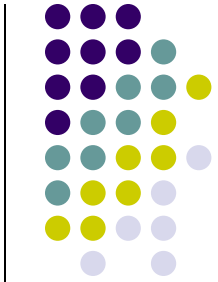
Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

Note: In its chapter entitled Masquerade Syndromes, the BCSC *Uveitis* book identifies JXG as a 'nonlymphoid malignancy.' This is clearly an error--JXG is **not** neoplastic, much less a malignancy. (See, eg, the *Peds, Pathology* and *Cornea* books.)



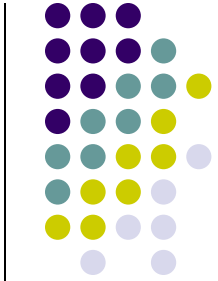
Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

lipid-laden macrophages
foamy histiocytes

Don't be fooled if they're referred to as 'lipid-laden' instead of 'foamy,' and/or if the term 'histiocytes' is used instead of 'macrophages'!

Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

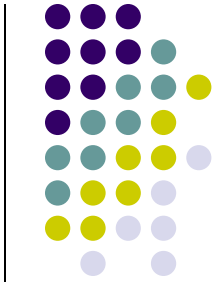
- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?

e



Nonneoplastic

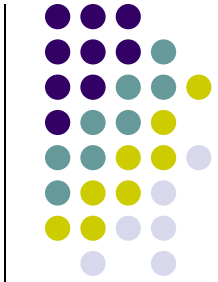
- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

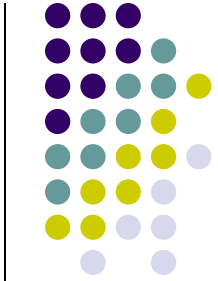
In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)

e



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

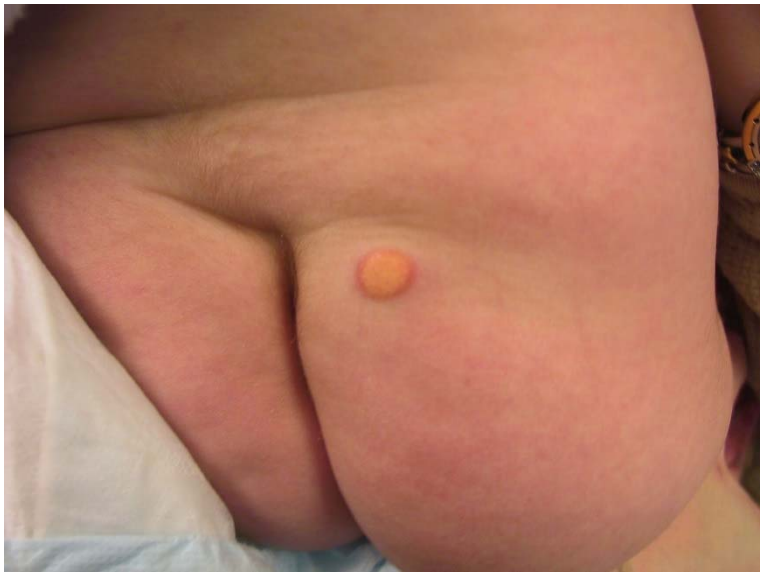
At what age does JXG present?
The majority before age 1 year, and almost all by age 2

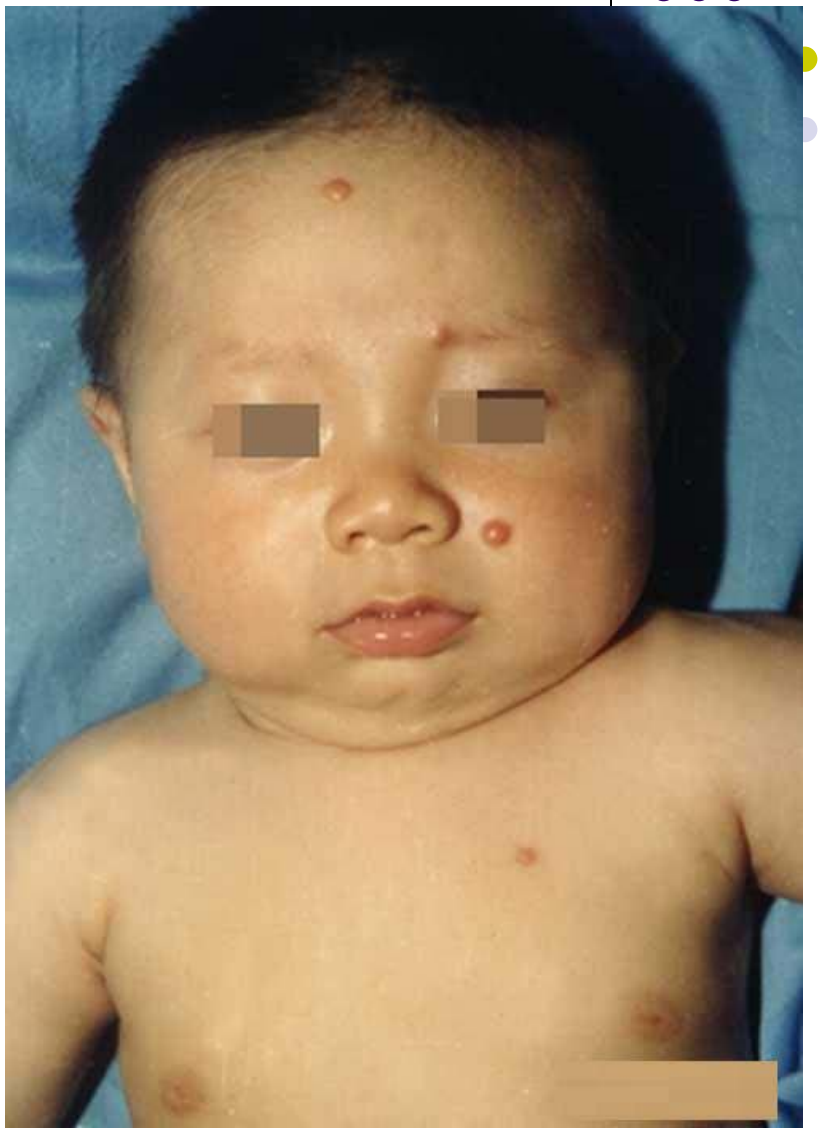
How does JXG usually present? (Hint: It's not ophthalmic)
As **orange skin papules**

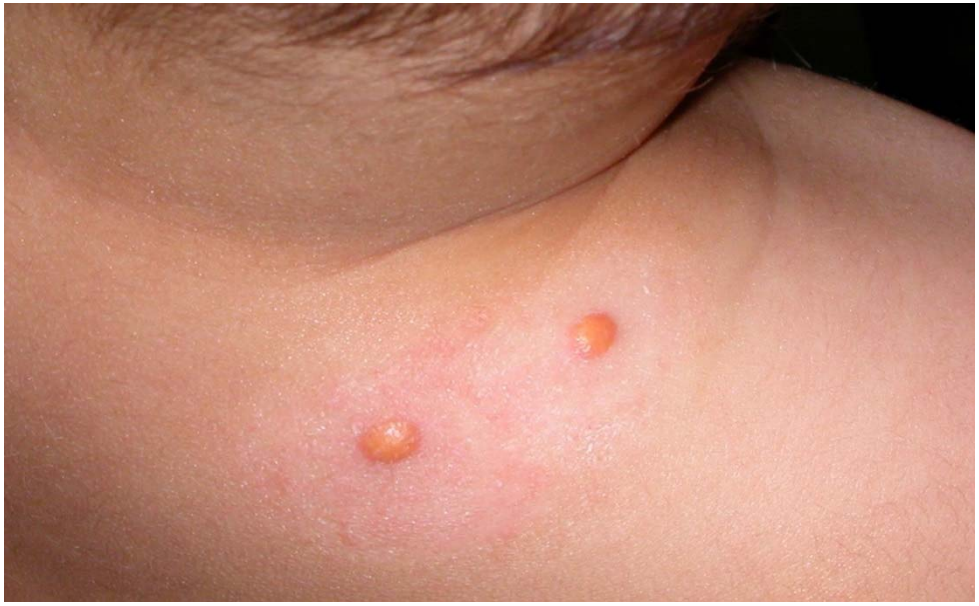
Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma







JXG: Skin papules

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

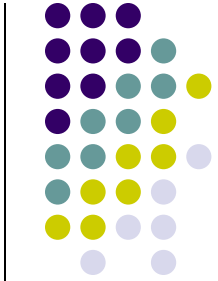
What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As **orange skin papules**

How do they present with respect to 'masquerading'?

e



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

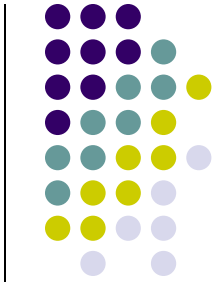
What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As **orange skin papules**

How do they present with respect to 'masquerading'?
As fleshy, tan-brown **iris nodules**

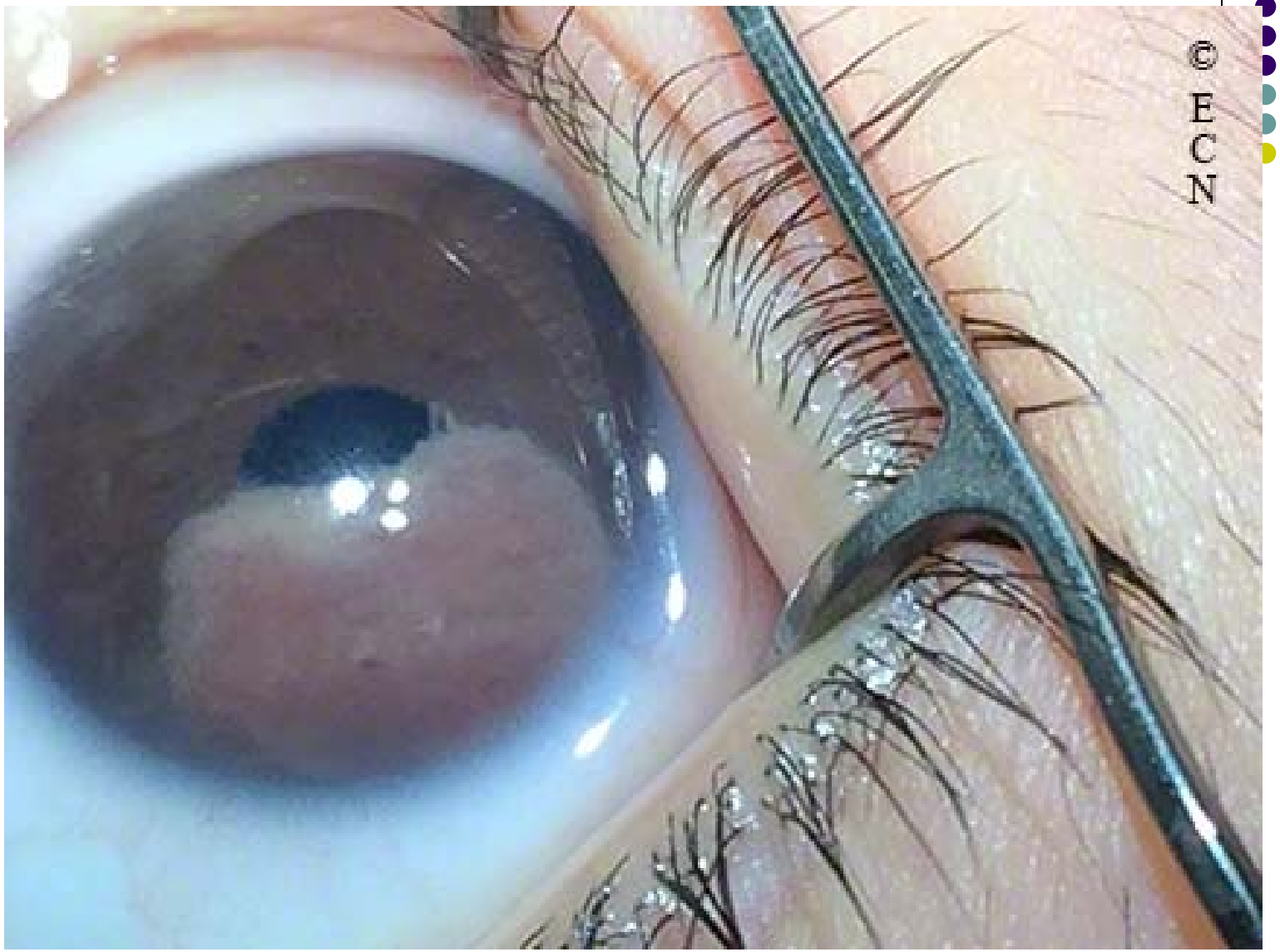
e



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

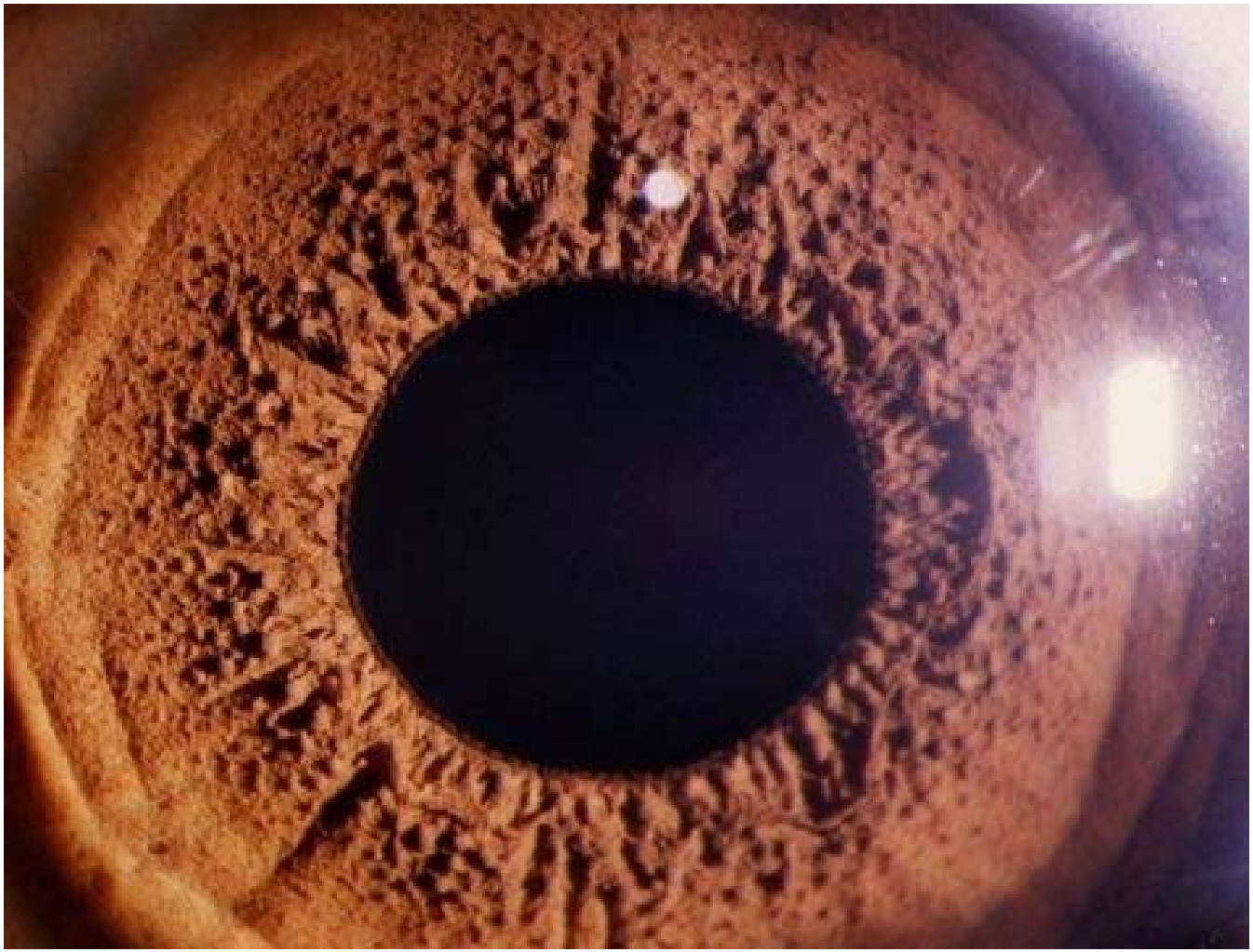
- Secondary to systemic lymphoma



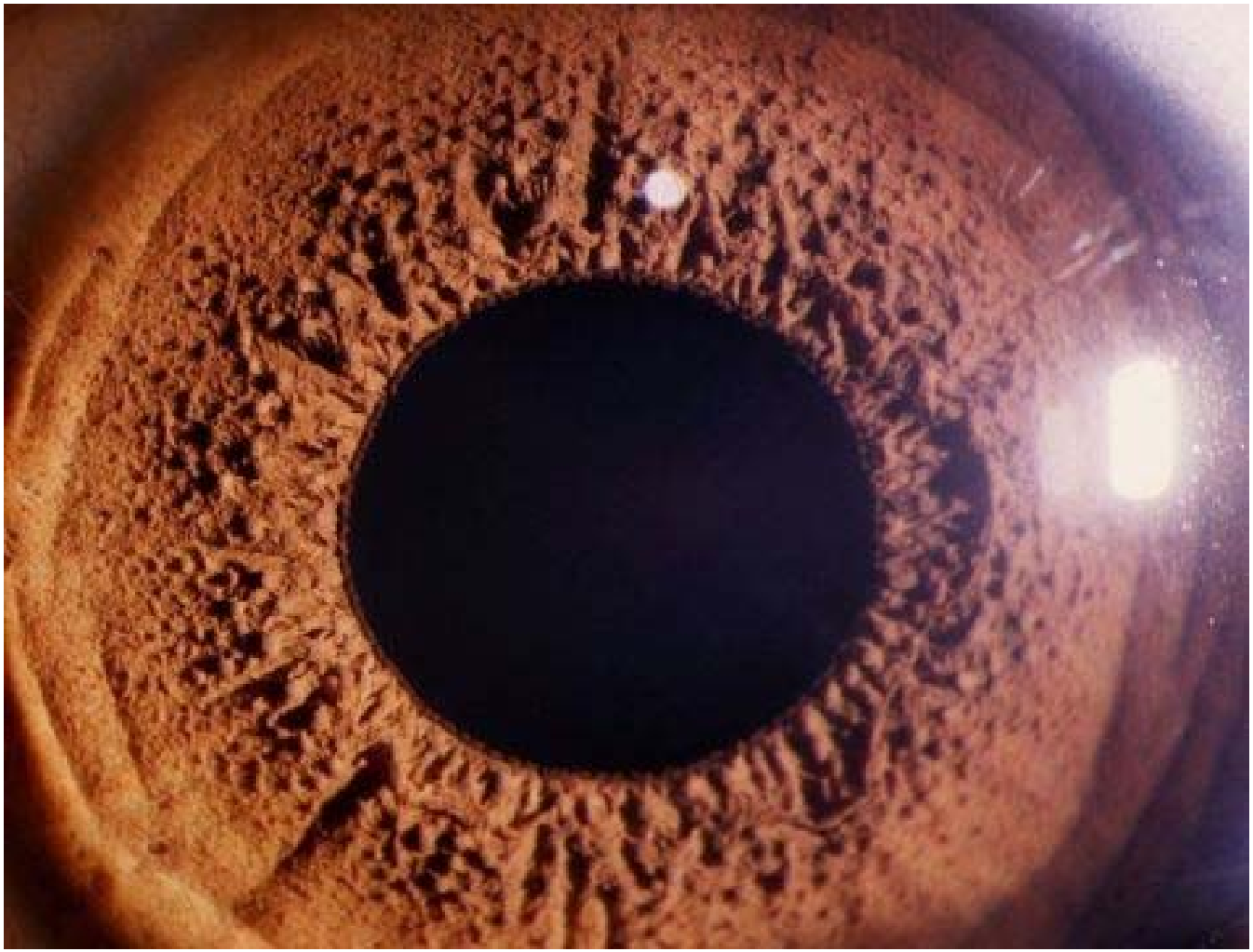
JXG: Iris nodule

JXG: Iris nodules?

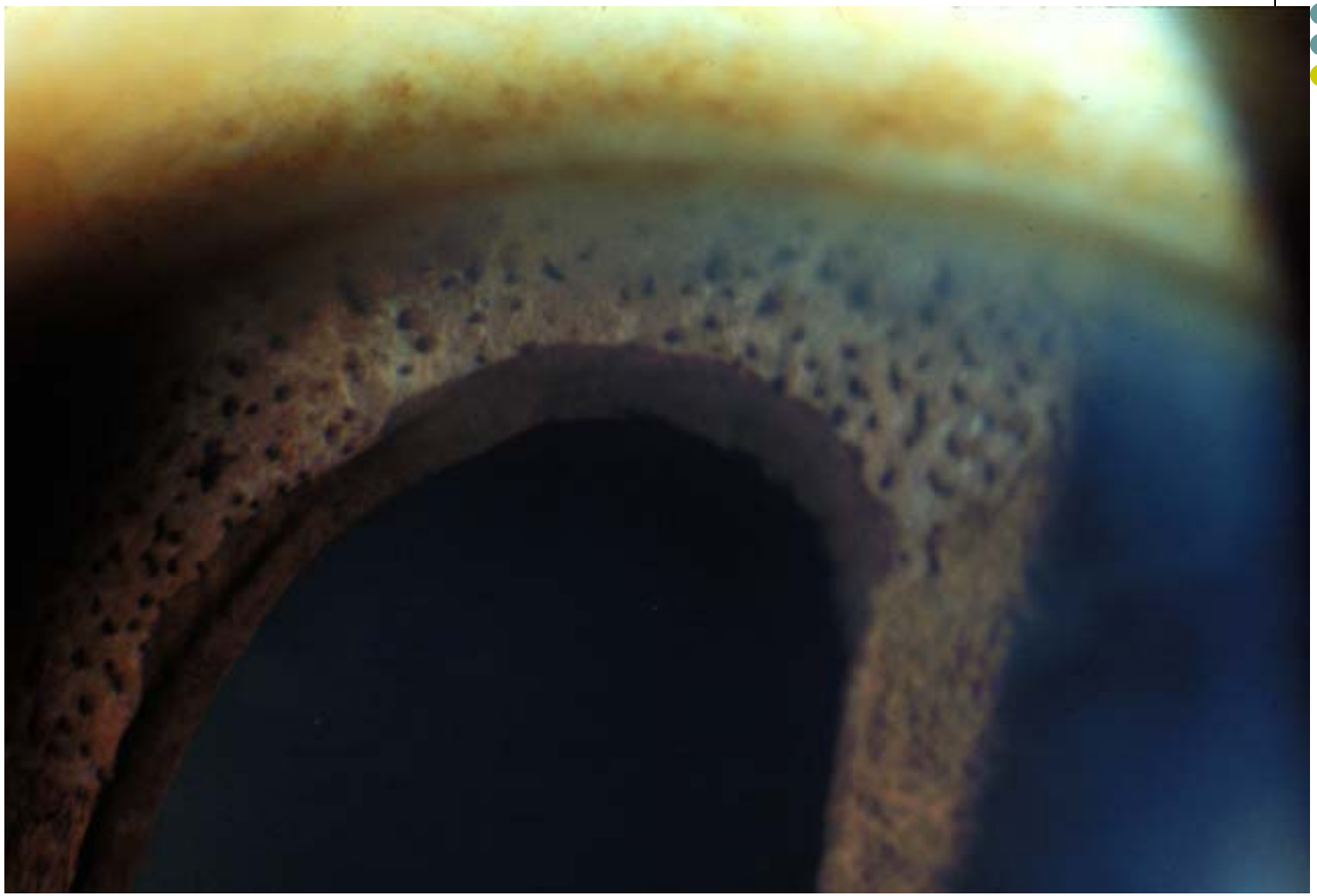
Lisch nodules



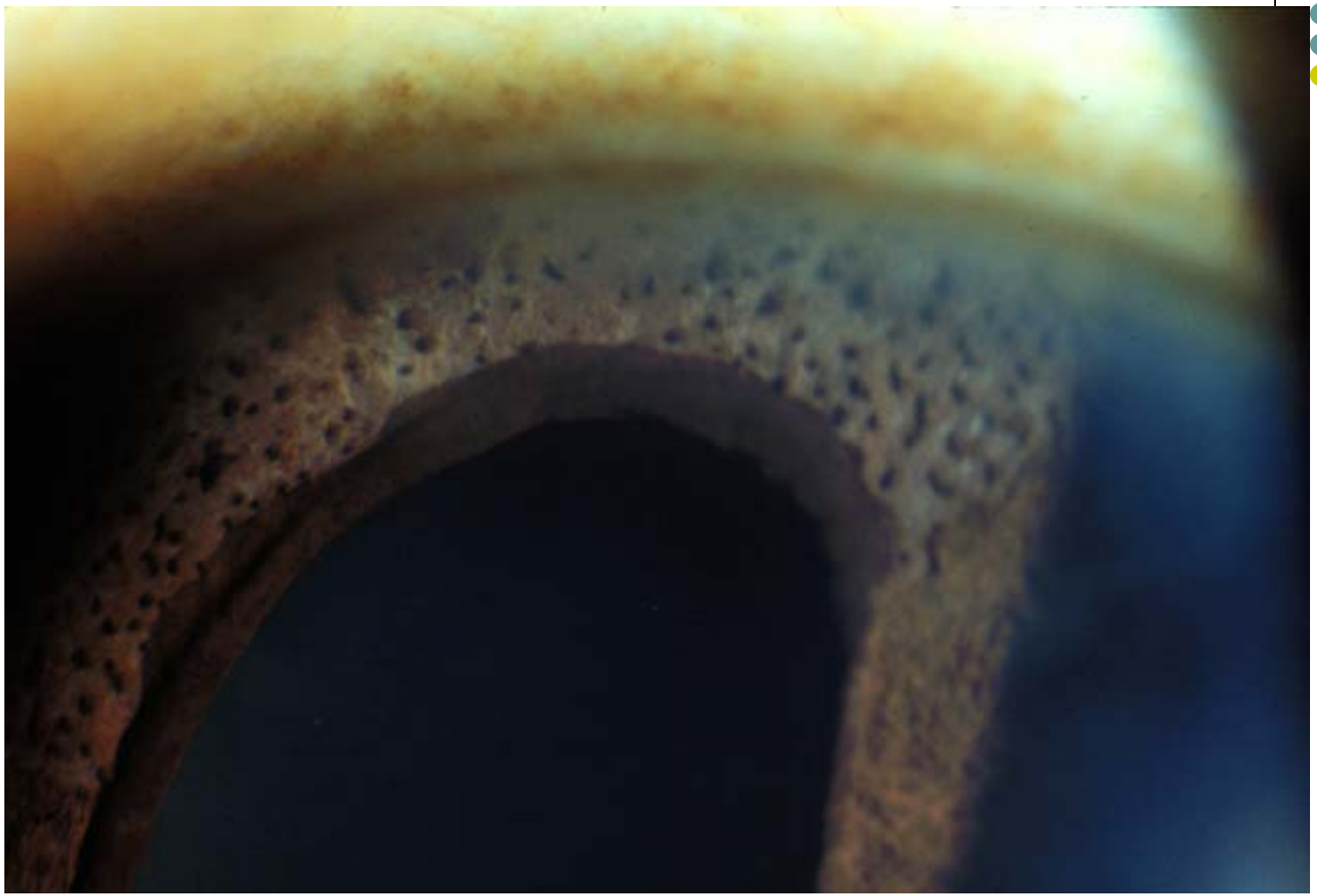
JXG: Iris nodules?



Iris mammillations



JXG: Iris nodules?



Iris nevus (aka Cogan-Reese) syndrome

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As **orange skin papules**

How do they present with respect to 'masquerading'?
As fleshy, tan-brown **iris nodules**

When iris JXG nodules are present, is it uni-, or bilaterally?

e

Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

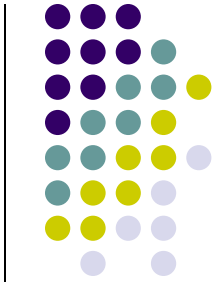
At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As **orange skin papules**

How do they present with respect to 'masquerading'?
As fleshy, tan-brown **iris nodules**

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

e



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

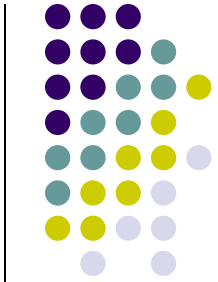
How does JXG usually present? (Hint: It's not ophthalmic)
As **orange skin papules**

How do they present with respect to 'masquerading'?
As fleshy, tan-brown **iris nodules**

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In addition to 'masquerading,' how are the iris nodules clinically significant?
--
--

e



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As **orange skin papules**

How do they present with respect to 'masquerading'?
As fleshy, tan-brown **iris nodules**

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In addition to 'masquerading,' how are the iris nodules clinically significant?
--They may bleed, producing hyphema and secondary glaucoma
--If numerous enough, they will produce heterochromia iridis

Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As **orange skin papules**

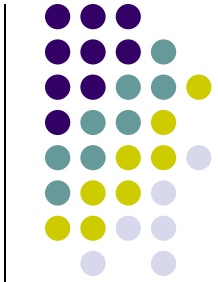
How do they present with respect to 'masquerading'?
As fleshy, tan-brown **iris nodules**

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In addition to 'masquerading,' how are the iris nodules clinically significant?
--They may bleed, producing hyphema and secondary glaucoma
--If numerous enough, they will produce heterochromia iridis

Given this, should they be removed surgically?

e



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

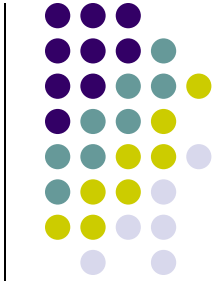
How does JXG usually present? (Hint: It's not ophthalmic)
As **orange skin papules**

How do they present with respect to 'masquerading'?
As fleshy, tan-brown **iris nodules**

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In addition to 'masquerading,' how are the iris nodules clinically significant?
--They may bleed, producing hyphema and secondary glaucoma
--If numerous enough, they will produce heterochromia iridis

Given this, should they be removed surgically?
Only if the glaucoma is uncontrollable



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As **orange skin papules**

How do they present with respect to 'masquerading'?
As fleshy, tan-brown **iris nodules**

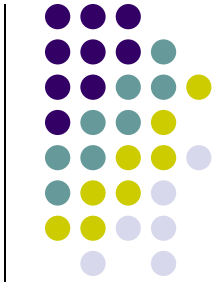
When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In addition to 'masquerading,' how are the iris nodules clinically significant?
--They may bleed, producing hyphema and secondary glaucoma
--If numerous enough, they will produce heterochromia iridis

Given this, should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?

e



Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

In three words, what sort of condition is juvenile xanthogranuloma (JXG)?
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
The presence of…**Touton giant cells** and **'foamy macrophages**'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As **orange skin papules**

How do they present with respect to 'masquerading'?
As fleshy, tan-brown **iris nodules**

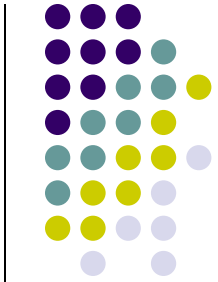
When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In addition to 'masquerading,' how are the iris nodules clinically significant?
--They may bleed, producing hyphema and secondary glaucoma
--If numerous enough, they will produce heterochromia iridis

Given this, should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years

e

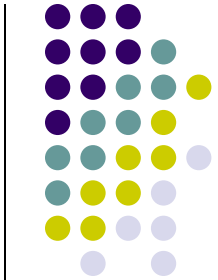


Nonneoplastic

- RP
- OIS
- Chronic rhegmatogenous RD
- IOFB
- PDS
- **Juvenile xanthogranuloma**

- Secondary to systemic lymphoma

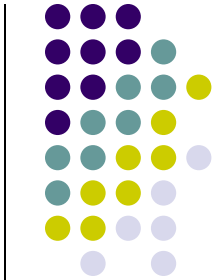
Masquerade Syndrome



- Nonneoplastic
  - Retinitis pigmentosa
  - Ocular ischemic syndrome
  - Chronic rhegmatogenous RD
  - Intraocular foreign body
  - Pigment dispersion syndrome
  - Juvenile xanthogranuloma
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - Solid
    - Primary
      - Uveal melanoma
      - Rb
    - Mets
      - Lung
      - Breast
      - ?

Finally: There's one more category of masquerade syndrome. It's neither neoplastic nor nonneoplastic. What is it?

Masquerade Syndrome

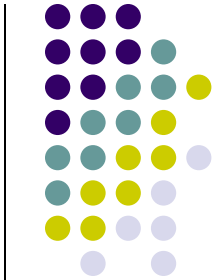


- Nonneoplastic
  - Retinitis pigmentosa
  - Ocular ischemic syndrome
  - Chronic rhegmatogenous RD
  - Intraocular foreign body
  - Pigment dispersion syndrome
  - Juvenile xanthogranuloma
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - Solid
    - Primary
      - Uveal melanoma
      - Rb
    - Mets
      - Lung
      - Breast
- Paraneoplastic

Finally: There's one more category of masquerade syndrome. It's neither neoplastic nor nonneoplastic. What is it?

Paraneoplastic

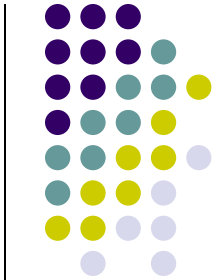
Masquerade Syndrome



- Nonneoplastic
  - Retinitis pigmentosa
  - Ocular ischemic syndrome
  - Chronic rhegmatogenous RD
  - Intraocular foreign body
  - Pigment dispersion syndrome
  - Juvenile xanthogranuloma
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - Solid
    - Primary
      - Uveal melanoma
      - Rb
    - Mets
      - Lung
      - Breast
- Paraneoplastic
  - ***?***

The Masquerade Syndromes *chapter of the* Uveitis *book discusses one paraneoplastic syndrome. What is it?*

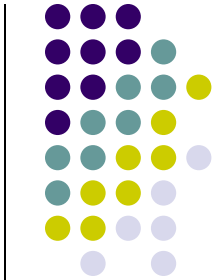
Masquerade Syndrome



- Nonneoplastic
  - Retinitis pigmentosa
  - Ocular ischemic syndrome
  - Chronic rhegmatogenous RD
  - Intraocular foreign body
  - Pigment dispersion syndrome
  - Juvenile xanthogranuloma
- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz
    - Leukemic
      - Leukemia
  - Solid
    - Primary
      - Uveal melanoma
      - Rb
    - Mets
      - Lung
      - Breast
- Paraneoplastic
  - BDUMP

The Masquerade Syndromes *chapter of the* Uveitis *book discusses one paraneoplastic syndrome. What is it?*
Bilateral diffuse uveal melanocytic proliferation (BDUMP)

Masquerade Syndrome



- Neoplastic
  - Hematologic
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - S
      - L
    - Leukemic
      - Leukemia
  - Solid
    - Primary
      - Uveal melanoma
      - Rb
    - Mets
      - Lung
      - Breast
- Nonneoplastic
  - Retinitis pigmentosa
  - Ocular ischemic syndrome
  - Chronic rhegmatogenous RD
  - Intraocular foreign body
  - Pigment dispersion syndrome
  - Juvenile xanthogranuloma
- Paraneoplastic
  - BDUMP

Point of clarification: The *Masquerades* chapter doesn't use the term 'paraneoplastic syndrome' when discussing BDUMP; however, the *Retina* book does, as do *EyeWiki* and *EyeRounds.org*.

erade Syndromes *chapter of the* Uveitis *book*
ne ***paraneoplastic syndrome****. What is it?*
fuse uveal melanocytic proliferation (BDUMP)

Masquerade Syndrome

Nonneoplastic

- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

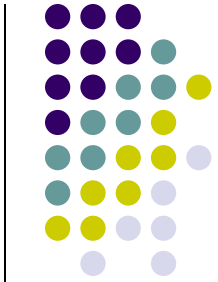
Mets

- Lung
- Breast

Paraneoplastic

- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

lid

Mets
- Lung
- Breast

oma

Lyr

Paraneoplastic
- **BDUMP**

Masquerade Syndrome

Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

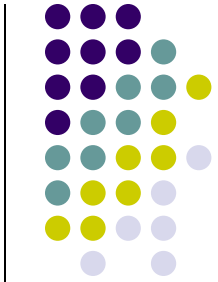
Who is the typical BDUMP pt?

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Mets
- Lung
- Breast

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

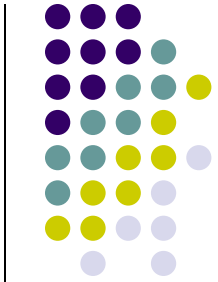
Who is the typical BDUMP pt?
An adult 50+

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Mets
- Lung
- Breast

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

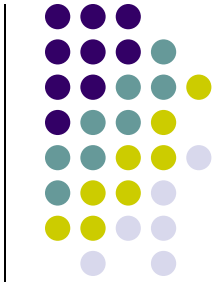
Is there a gender predilection?

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Mets
- Lung
- Breast

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

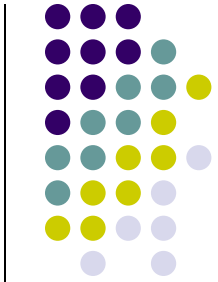
Is there a gender predilection?
No

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Mets
- Lung
- Breast

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associated?

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

lid

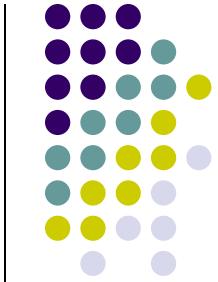
Mets
- Lung
- Breast

oma

Lyr

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associated?
Gynecologic in women; lung and pancreatic in men

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

lid

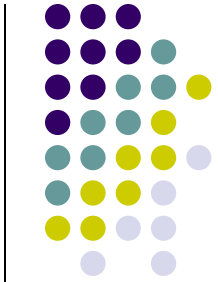
Mets
- Lung
- Breast

oma

Lyr

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associated?
Gynecologic in women; lung and pancreatic in men

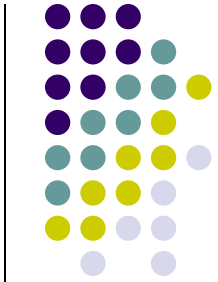
What is the classic presenting ocular complaint?

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Mets
- Lung
- Breast

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associated?
Gynecologic in women; lung and pancreatic in men

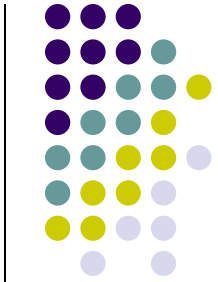
What is the classic presenting ocular complaint?
Bilateral rapid vision loss

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Mets
- Lung
- Breast

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associated?
Gynecologic in women; lung and pancreatic in men

What is the classic presenting ocular complaint?
Bilateral rapid vision loss

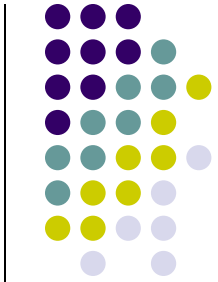
What are the three classic findings on exam?
Bilateral…
--
--
--

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Mets
- Lung
- Breast

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associated?
Gynecologic in women; lung and pancreatic in men

What is the classic presenting ocular complaint?
Bilateral rapid vision loss

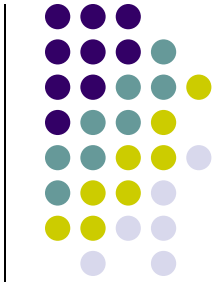
What are the three classic findings on exam?
Bilateral…
--rapid cataract progression
--serous/exudative RD
--multiple large ‘nevi’ of the posterior choroid

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Mets
- Lung
- Breast

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associated?
Gynecologic in women; lung and pancreatic in men

What is the classic presenting ocular complaint?
Bilateral rapid vision loss

What are the three classic findings on exam?
Bilateral…
--rapid cataract progression
--serous/exudative RD
--multiple large 'nevi' of the posterior choroid

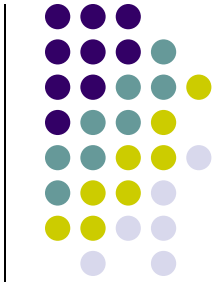
What is the prognosis?

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Mets
- Lung
- Breast

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associated?
Gynecologic in women; lung and pancreatic in men

What is the classic presenting ocular complaint?
Bilateral rapid vision loss

What are the three classic findings on exam?
Bilateral…
--rapid cataract progression
--serous/exudative RD
--multiple large 'nevi' of the posterior choroid

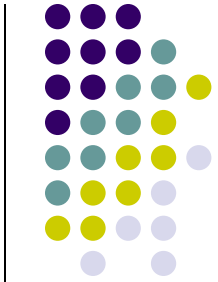
What is the prognosis?
Poor, in terms of both vision and life expectancy

Nonneoplastic
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

Mets
- Lung
- Breast

Paraneoplastic
- **BDUMP**

Masquerade Syndrome



Nonneoplastic

- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD

Lyr

Paraneoplastic

- **BDUMP**

Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associate
Gynecologic in women; lung and panc

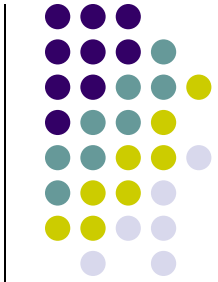
What is the classic presenting ocular c
Bilateral rapid vision loss

What are the three classic findings on
Bilateral…
--rapid cataract progression
--serous/exudative RD
--multiple large 'nevi' of the posterior choroid

What is the prognosis?
Poor, in terms of both vision and life expectancy

I'm confused. BDUMP doesn't sound particularly inflammatory—no cell; no creamy-yellow choroidal infiltrates; etc. Why is it being covered in the masquerade syndrome slide-set?

Masquerade Syndrome



Nonneoplastic

- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD

Paraneoplastic

- **BDUMP**

Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associate
Gynecologic in women; lung and panc

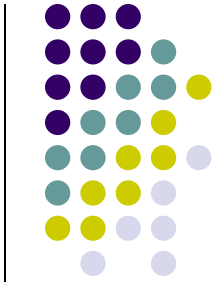
What is the classic presenting ocular c
Bilateral rapid vision loss

What are the three classic findings on
Bilateral…
--rapid cataract progression
--serous/exudative RD
--multiple large 'nevi' of the posterior choroid

What is the prognosis?
Poor, in terms of both vision and life expectancy

I'm confused. BDUMP doesn't sound particularly inflammatory—no cell; no creamy-yellow choroidal infiltrates; etc. Why is it being covered in the masquerade syndrome slide-set?
Because of its association with

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associate
Gynecologic in women; lung and panc

What is the classic presenting ocular c
Bilateral rapid vision loss

What are the three classic findings on
Bilateral…
--rapid cataract progression
--**serous/exudative RD**
--multiple large 'nevi' of the posterior choroid

What is the prognosis?
Poor, in terms of both vision and life expectancy

Nonneoplastic

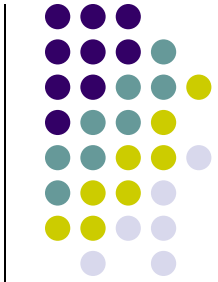
- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD

I'm confused. BDUMP doesn't sound particularly inflammatory—no cell; no creamy-yellow choroidal infiltrates; etc. Why is it being covered in the masquerade syndrome slide-set?
Because of its association with bilateral serous/exudative RDs

Paraneoplastic

- **BDUMP**

Masquerade Syndrome



Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associate
Gynecologic in women; lung and panc

What is the classic presenting ocular c
Bilateral rapid vision loss

What are the three classic findings on
Bilateral…
--rapid cataract progression
--**serous/exudative RD**
--multiple large 'nevi' of the posterior choroid

What is the prognosis?
Poor, in terms of both vision and life expectancy

Nonneoplastic

- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD

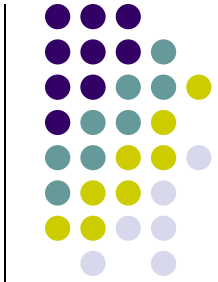
I'm confused. BDUMP doesn't sound particularly inflammatory—no cell; no creamy-yellow choroidal infiltrates; etc. Why is it being covered in the masquerade syndrome slide-set?
Because of its association with bilateral serous/exudative RDs

When you hear 'bilateral serous RDs,' what inflammatory condition should leap to mind?

Paraneoplastic

- **BDUMP**

Masquerade Syndrome



Nonneoplastic

- Retinitis pigmentosa
- Ocular ischemic syndrome
- Chronic rhegmatogenous RD

Lyr

Paraneoplastic

- **BDUMP**

Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associate
Gynecologic in women; lung and panc

What is the classic presenting ocular c
Bilateral rapid vision loss

What are the three classic findings on
Bilateral…
--rapid cataract progression
--**serous/exudative RD**
--multiple large 'nevi' of the posterior choroid

What is the prognosis?
Poor, in terms of both vision and life expectancy

I'm confused. BDUMP doesn't sound particularly inflammatory—no cell; no creamy-yellow choroidal infiltrates; etc. Why is it being covered in the masquerade syndrome slide-set?
Because of its association with bilateral serous/exudative RDs

When you hear 'bilateral serous RDs,' what inflammatory condition should leap to mind?
Vogt-Koyanagi-Harada (VKH) syndrome