Decoding Visual Pathway Lesions

Because visual information from the eye traverses the axial length of the brain to the occipital cortex, much ophthalmic and neurologic pathology may affect the visual pathway. By taking a systematic approach, clinicians can identify unique clinical patterns that can lead to accurate localization and diagnosis of visual pathway lesions.

Anatomy of the Visual Pathway

The afferent visual pathway begins in the retina and, as with other somatosensory pathways, has a three-order neuronal pathway, which ultimately relays information to the occipital cortex. First-order neurons are formed by the bipolar cells of the retina, which synapse with retinal ganglion cells. These second-order neurons extend their axons along the innermost portion of the retina, coalescing at the optic disc. These axons exit the globe and continue posteriorly via the optic nerve, and both optic nerves join in the suprasellar region to form the optic chiasm. The nasal retinal fibers (temporal visual field) decussate contralaterally, while the temporal retinal fibers (nasal visual field) remain ipsilateral. The conjoined optic nerve fibers now form the optic tracts. The optic tract axons synapse in the lateral geniculate body (LGB) of the thalamus. These, now third-order neurons, relay information from the LGB via the optic radiations (either the dorsal bundle or Meyer loop) to the visual cortex in the occipital lobe.

Conceptualizing the visual pathway in three major sections—optic nerve, chiasm, and retrochiasm—can be helpful in identifying patterns of disease and localization because pathology will vary based on which section of the visual pathway is affected.

The Major Sections

In patients with visual pathway lesions, the presenting signs and symptoms may depend on the site affected, degree of axonal loss, and amount of visual function lost. In some cases, systemic features may be associated with some of these lesions.

Optic nerve lesions. Optic nerve pathology tends to cause symptoms of dimming or graying of vision and color desaturation, as opposed to blurring or positive visual phenomena seen in other ocular conditions. In addition, a relative afferent pupillary defect (RAPD) is typically present in unilateral or asymmetric bilateral optic nerve pathology.

The most anterior portion, the intraorbital segment of the optic disc, has a separate vascular supply, making it particularly susceptible to ischemic damage, which may translate to defects on exam, visual fields, and OCT findings that respect the horizontal meridian.

Intracanalicular and intracranial portions of the optic nerve may have other associated focal neurologic deficits/cranial neuropathies (e.g., orbital apex syndrome) to help with further localization. Examples of common conditions are listed with associated clinical and imaging features in Table 1. A wide array of pathology may affect the optic nerve, including ischemic, compressive, inflammatory, or infiltrative processes. Common conditions include optic neuritis, ischemic optic neuropathy (arteritic and nonarteritic), infiltrative optic neuropathies (leukemia, lymphoma, sarcoid), infectious optic neuropathy (syphilis, tuberculosis, Lyme disease), traumatic optic neuropathy, compressive optic neuropathy, and hereditary and toxic optic neuropathies.

Papilledema is bilateral, passive, disc swelling as a result of raised intracranial pressure and not a lesion of the visual pathway. But, since it presents as disc swelling and can mimic optic nerve lesions, it is being discussed here.

Chiasmal lesions. Chiasmal syndrome classically presents with bilateral heteronymous field defects.[1] The optic chiasm is prone to compression given its suprasellar location. Inferior compression of the chiasm is most typical, but it also can be compressed anteriorly, centrally, laterally, posteriorly, or superiorly. Most commonly, pituitary adenomas are the cause of chiasmal syndrome, but craniopharyngioma and meningiomas are also somewhat common. Intrinsic infectious, inflammatory, and infiltrative lesions should

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also be considered (e.g., chiasmitis as a presenting sign of neuromyelitis optica spectrum disorder).[2] Table 2 outlines identifying characteristics of various types of compression.

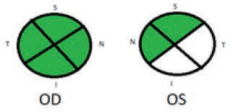
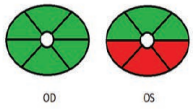
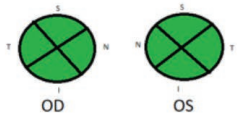
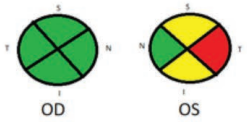
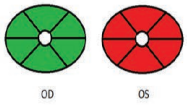
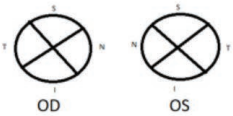
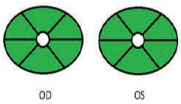
Retrochiasmal lesions. All lesions that affect the visual pathway posterior to the chiasm are characterized by contralateral homonymous field defects. Generally, the congruity of field defects increases with a more posterior location of the lesion.

Retrochiasmal lesions may or may not be accompanied by an RAPD. Lesions of the optic tract can have an RAPD, as there is an asymmetric crossing of fibers at the optic chiasm. However, fibers involved in the pupillary reflex exit the optic tract just prior to the LGB; therefore lesions at, or distal, to the LGB should not have an RAPD.

Common causes of retrochiasmal lesions are metastatic tumors, primary central nervous system tumors, trauma,

TABLE 1: Optic Nerve Lesions

The findings are described assuming a left-sided lesion.

CONDITION	RAPD	Optic Disc Features	Neurological Features	Visual Fields	OCT-ppRNFL	OCT-GCIPL
NAION	+	Sectoral edema, fellow eye crowded disc, "disc at risk"	None	Altitudinal and/or arcuate scotoma	 RNFL sectoral edema	 Corresponding sectoral ganglion cell layer (GCL) loss precedes RNFL thinning. Visual loss correlates with central macular thickness.[4]
Optic neuritis (Multiple sclerosis–related)	+	Normal in retrobulbar neuritis (2/3 of all cases); subsequently develop temporal pallor	MRI with demyelinating lesions; may have other focal neurologic symptoms (e.g., internuclear ophthalmoplegia, focal numbness or weakness)	Central and/or centrocecal scotoma	 Acute presentation  Presentation with time 1/3 of patients present with RNFL edema initially. Subsequent thinning is seen more in the temporal quadrant, even in fellow eyes (around 90 µm vs. 110 µm in normal eyes).[5,6]	 Thinning seen at as early as two weeks of disease onset. Microcystic macular edema is common and a sign of worse visual outcome.
Papilledema	-	Variable degree of bilateral disc edema based on severity, hyperemia, obliteration of the cup	Headaches, pulsatile tinnitus, transient visual obscurations, diplopia	Enlarged blind spot progresses to inferonasal constriction, which progresses to diffuse constriction	 RNFL edema bilaterally, upward deflection of Bruch membrane	 Normal at onset; concordant reduction in GCIPL may indicate treatment failure.

demyelinating disease, vascular injury (ischemic/hemorrhagic infarct).[3] Table 3 covers lesions of the optic tract, LGB, and optic radiations.

Testing and Management

Testing. Although visual field analysis usually is sufficient for localizing a visual pathway lesion, ancillary testing or imaging can be helpful. OCT provides an added advantage in the three follow-

Continued on page 41.

TABLE 2: Chiasmal Lesions

Chiasmal lesions of any type may be accompanied by an RAPD. In addition, nasal or band atrophy (bow-tie pattern) may be associated with bitemporal field defect. (Please note visual fields are shown via standard convention with OS being on the left side and OD on the right side.)

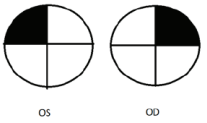
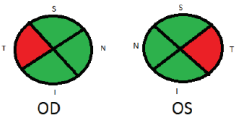
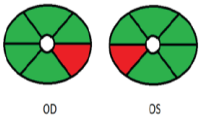
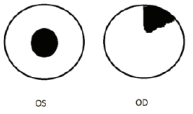
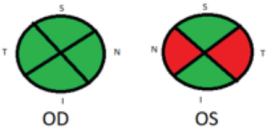
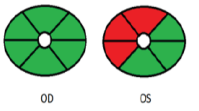
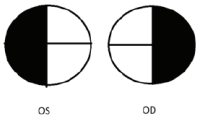
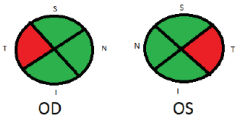
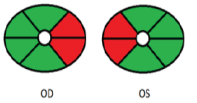
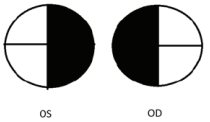
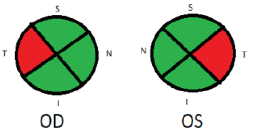
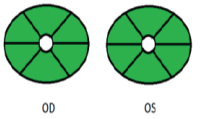
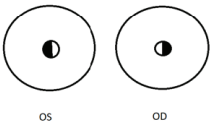
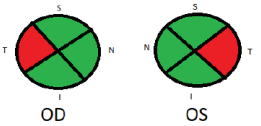
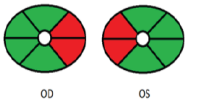
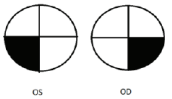
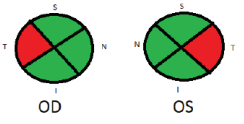
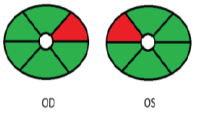
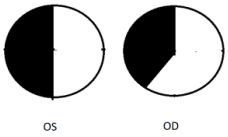
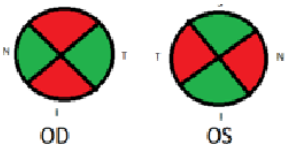
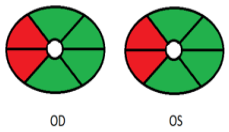
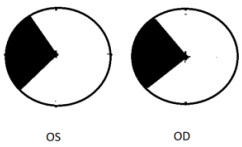
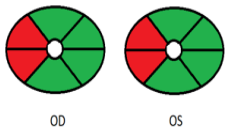
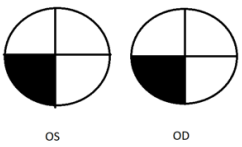
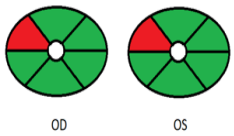
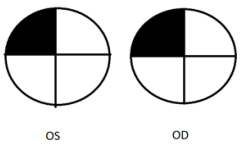
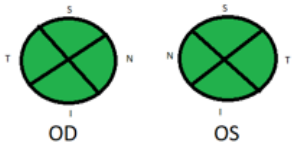
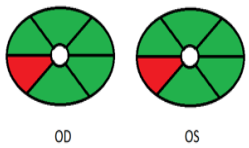
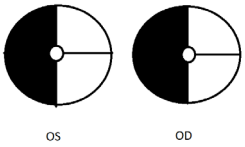
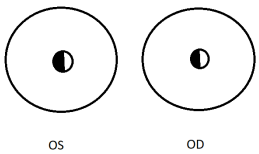
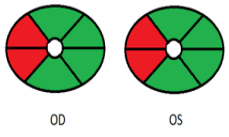
Location of Compression	Visual Fields	OCT-ppRNFL	OCT-GCIPL
Inferior (most common)	 Superotemporal quadrantanopia	 Bitemporal RNFL loss of varying degrees	 Inferonasal thinning
Anterior (affecting the left>right chiasmal fibers)	 Junctional scotoma	 Thinning predominant in the nasal and temporal quadrant of the ipsilateral RNFL with normal contralateral RNFL thickness	 Ipsilateral nasal GCL thinning with normal GCL of the contralateral eye
Central	 Bitemporal hemianopia	 Bitemporal RNFL loss of varying degrees	 Binasal thinning
Lateral	 Binasal hemianopia	 Superotemporal thinning on both sides	 Normal (because the macular fibers are usually central)
Posterior	 Paracentral bitemporal hemianopic scotoma	 Temporal thinning	 Binasal thinning
Superior	 Inferotemporal quadrantanopia	 Bitemporal RNFL loss of varying degrees	 Superonasal thinning

TABLE 3: Retrochiasmal Lesions

Retrochiasmal lesions of any type may be accompanied by demyelination, tumors, and/or infarcts. RAPD is seen in lesions proximal to the LGB. Lesions distal to the LGB do not have an RAPD. (This table is constructed for a right-sided lesion. The terminology ipsilateral/contralateral is in reference to the site of the lesion and not the visual field defect. Please note visual fields are shown via standard convention with OS being on the left side and OD on the right side.)

Site of Lesion	Optic Disc Features	Visual Fields	OCT-ppRNFL	OCT-GCIPL
Optic tract	Ipsilateral temporal pallor with contra-lateral bow-tie atrophy	 Contralateral homonymous hemianopias	 Ipsilateral superior and inferior thinning with contralateral nasal and temporal thinning (bow-tie or band atrophy)[7]	 Homonymous thinning of the ipsilateral side
Lateral geniculate body	Normal-appearing disc	 Contralateral homonymous sectoranopia	Normal ppRNFL thick ness at presentation (May develop temporal thinning with time due to retrograde degenration of retinal ganglion cells[8])	 Homonymous thinning of the ipsilateral side
Optic radiations-dorsal bundle		 Contralateral homon-ymous inferotemporal quadrantanopia		 Homonymous superior quadrant GCL thinning
Optic radiations-Meyer loop		 Contralateral homonymous superotemporal quadrantanopia		 Homonymous inferior quadrant GCL thinning
Occipital cortex		 Contralateral congruous homonymous hemianopia with macular sparing		
Tip of the occipital cortex		 Contralateral congruous homonymous hemianopic scotoma		 Ipsilateral temporal hemimacular thinning and contralateral nasal hemimacular thinning[8]

ing situations: 1) When the patient is unable to perform a visual field analysis due to poor vision or limited understanding or comprehension; 2) when objective measurement of axonal loss is desired; 3) when prognostication of recovery is desired. OCT analysis of peripapillary retinal nerve fiber layer (ppRNFL) thickness and macular ganglion cell–inner plexiform layer (GCIPL) thickness provides clinicians with objective data that can aid in diagnosis and monitoring of neuro-ophthalmic conditions. Additionally, GCIPL has shown increased sensitivity in the detection of early compressive lesions.

Management. The underlying cause for a visual pathway lesion may be established through examination, imaging, or lab testing. Treatment depends on the etiology and may require a multidisciplinary approach.

Conclusion

Lesions can affect any portion of the visual pathway, and they may have a characteristic presentation. A complete ophthalmic exam, with emphasis on disc appearance, visual field defects, and pattern of nerve fiber loss on OCT helps in localizing the lesion. When visual field analysis cannot be performed, the pattern of ppRNFL and GCIPL thinning on OCT can serve as an adjunct.

1 Astorga-Carballo A et al. *Saudi J Ophthalmol.* 2017;31(4):229-233.
2 Rui W et al. *J Neuroophthal.* 2019;39(4):515-515.
3 Fadzli F et al. *Clin Radiol.* 2013;68(10):e538-e551.
4 Papchenko T et al. *Acta Ophthalmol.* 2012;90(6):e463-e469.
5 Noval S et al. *Mult Scler Int.* 2011;2011:472790.
6 Bennett JL et al. *Mult Scler.* 2015;21(6):678-688.
7 Lloyd-Smith AJ et al. *Neurology.* 2016;87(19):2063-2064.
8 Yamashita T et al. *J Ophthalmol.* 2016;2016:2394957.

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