CHAPTER 12

Topical Diagnosis of Chiasmal and Retrochiasmal Disorders

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TOPICAL DIAGNOSIS OF OPTIC CHIASMAL LESIONS

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TOPICAL DIAGNOSIS OF OPTIC CHIASMAL LESIONS

The optic chiasm is one of the most important structures in neuro-ophthalmologic diagnosis. The arrangement of visual fibers in the chiasm accounts for the characteristic visual field defects caused by such diverse lesions as tumor, inflammation, demyelination, ischemia, and infiltration. In addition, damage to neurologic and vascular structures adjacent to the chiasm produces typical additional symptoms. Thus, accurate diagnosis depends on having knowledge of both the neuro-ophthalmologic and non–neuro-ophthalmologic manifestations of chiasmal lesions. Two early papers are of fundamental importance: Adler et al. (1) described early field changes in chiasmal lesions, and Gartner (2) outlined ocular pathology in the chiasmal syndrome. A paper of historical interest describes a case of chiasmal compression from the 16th century (3).

VISUAL FIELD DEFECTS

Although there are many variations in the visual field defects caused by damage to the optic chiasm, the essential feature is some type of bitemporal defect, the hallmark of damage to fibers that cross within the chiasm. The bitemporal defects may be superior, inferior, or complete, as well as peripheral, central, or both. Bitemporal field defects are also called heteronymous field defects, a term that distinguishes them from homonymous field defects.

Visual field defects caused by lesions of the optic chiasm are often classified according to the general site of the damage. In many cases this is an oversimplification, since many lesions that arise in the region of the chiasm affect not only the entire chiasm but the intracranial optic nerves as well. Nevertheless, it is reasonable to use this approach because it helps determine the precise management of the lesion and predict the visual outcome following treatment. This chapter classifies visual field defects produced by lesions that damage the optic chiasm as a result of damage at one of three locations: (a) the anterior angle of the chiasm, (b) the body of the chiasm, or (c) the posterior angle of the chiasm. A small number of lesions may damage nerve fibers at the lateral aspects of the chiasm.

Lesions that Damage the Distal Portion of One Optic Nerve at the Anterior Angle of the Optic Chiasm

Traquair (4) pointed out that it is at the anterior angle of the optic chiasm that the “junction” scotoma occurs because of the separation of nasal crossed and temporal uncrossed fibers. When a small lesion damages only the crossing fibers of the ipsilateral eye, the field defect is monocular and tem-
poral and has a midline hemianopic character that extends to the periphery of the field. When only the macular crossed fibers from one eye are damaged, the resultant field defect is still monocular and temporal but is scotomatous and located in the paracentral region. If there is extensive damage to the visual fibers in an optic nerve, an extensive field defect or total blindness develops in the ipsilateral eye. In such cases, but also in less severe cases, the crossed ventral fibers that originate from ganglion cells inferior and nasal to the fovea of the contralateral eye may also be damaged, producing a defect in the superior temporal field of the contralateral eye (Fig. 12.1).

This contralateral field defect, which occurs in an eye without other evidence of visual dysfunction, may be overlooked when kinetic perimetry is performed unless the superior temporal region of the “normal” eye is carefully tested by the examiner, but it is almost always detected when automated static perimetry is used (Fig. 12.1). Bird (5) detailed the findings in eight patients with the anterior chiasmal syndrome. In each case, a central scotoma was present in the visual field of the eye on the side of the lesion, and there was temporal field loss in the contralateral eye. In five cases, the contralateral field loss was in the superior temporal field only, and in two of these five cases, the peripheral field was normal by kinetic perimetry, with the defect being scotomatous and detectable only in the paracentral upper temporal field with small test objects. In the remaining three patients, the contralateral field loss was in the paracentral temporal region, without preferential loss above or below the horizontal meridian.

The mechanism by which the fibers from the contralateral eye are damaged was ascribed to their anterior extension into the affected ipsilateral optic nerve to form the structure called Wilbrand’s knee (6,7) (see Chapters 1 and 4). Horton (8) suggested that this eponymous structure is more likely an artifact that develops during atrophy of the ipsilateral optic nerve. Against this are findings of superior temporal visual field defects in patients with acute avulsion injuries of the anterior chiasm (9,10). Whatever the mechanism, the importance of identifying the usually asymptomatic field defect cannot be sufficiently stressed, because it is at this point that the examiner can make an absolute diagnosis of an anterior optic chiasmal (distal optic nerve) syndrome, at a stage at which treatment of the underlying lesion is most likely to result in improvement in visual function.

Lesions that Damage the Body of the Optic Chiasm

Lesions that damage the body of the optic chiasm characteristically produce a bitemporal defect that may be quadrantic or hemianopic and that may be peripheral, central, or a combination of both, with or without so-called splitting of the macula (Fig. 12.2). In most cases, visual acuity is normal. In some patients, however, visual acuity is diminished, even though no field defect other than a bitemporal hemianopia is present (11). When the lesion compresses the chiasm from below, such as occurs with a pituitary adenoma, the field defects follow a stereotyped pattern (12). When the peripheral fibers are principally affected, the field defects usually begin in the outer upper quadrants of both eyes (Fig. 12.3). In the field of the right eye, the defect usually progresses in a clockwise direction and in the left eye in a counterclockwise direction (13).

The field defects may be unequal in the two eyes: one
Figure 12.2. Optic chiasmal syndrome. A, Kinetic perimetry in a patient with a large pituitary adenoma reveals a complete bitemporal hemianopia. B, Static perimetry, using a Humphrey 24-2 Threshold Test, in another patient with a pituitary adenoma, reveals an incomplete bitemporal hemianopia.

Figure 12.3. Bilateral superior temporal defects in patients with a pituitary adenoma. A, Kinetic perimetry in one patient demonstrates defects that are restricted to the superior temporal quadrants of the visual fields of both eyes (a bitemporal superior quadrantanopia). B, Static perimetry, using a Humphrey 24-2 Threshold Test, in another patient with a pituitary adenoma demonstrates bitemporal hemianopic defects that are much denser superiorly.
eye may become almost or completely blind, whereas the
defect in the field of the other eye remains mild. In the charting
of visual field defects resulting from pituitary adenomas and
other compressive lesions, scotomas in the peripheral parts
of the visual fields are usually dense and are not likely to be
overlooked, although small relative paracentral scotomas are
frequently missed when kinetic perimetry alone is per-
formed. Automated perimetry can help in detecting some of
the earliest signs of chiasmal compression, such as a subtle
depression respecting the vertical meridian, detected by
comparing pairs of thresholds across the meridian (14).

Pituitary adenomas are not, of course, the only lesions
that can produce bitemporal field defects that are denser
below. Suprasellar but infrachiasmal lesions, such as tuber-
culum sellae and medial sphenoid ridge meningiomas, cra-
iopharyngiomas, and aneurysms, can also produce such
defects. The field defects caused by such lesions are indistin-
guishable from those caused by pituitary adenomas.

Various suprasellar and suprachiasmal compressive le-
ssions, such as tuberculum sellae meningiomas (15), cranio-
pharyngiomas (16,17), aneurysms (18–21), and dolichoec-
tatic anterior cerebral arteries, may damage the superior
fibers of the optic chiasm, as may infiltrating lesions, such
as germinomas (22–24), benign and malignant gliomas (25),
and cavernous angiomas (26). The defects in the visual fields
in such cases are still bitemporal but are located in the infe-
rior rather than the superior fields of both eyes (Fig. 12.4).
In addition, papilledema, which is quite unusual in patients

![Figure 12.4](image-url)

Figure 12.4. Bilateral inferior temporal field defects in a 32-year-old man with a suprasellar mass. Kinetic perimetry reveals
inferior temporal quadrant defects in the visual fields of the left (A) and right (B) eyes. The defects are scotomatous. Static
perimetry, using a Humphrey 24-2 Threshold Test, in the same patient confirms the inferior temporal quadrant nature of the
defects. C, Visual field of left eye. D, Visual field of right eye. The patient underwent a craniotomy and was found to have
a suprasellar suprachiasmatic germinoma.

CLINICAL NEURO-OPHTHALMOLOGY
with suprasellar, infrachiasmal lesions, is somewhat more common in suprachiasmal lesions because such lesions can extend into and occlude the third ventricle. Infiltrating tumors, such as gliomas and germinomas, as well as inflammatory and demyelinating lesions that affect the optic chiasm, may produce bitemporal field defects (Fig. 12.5), but such lesions may also produce other types of field defects, such as arcuate defects and nonspecific reduction in sensitivity, that do not necessarily correlate with the location, size, or extent of the lesion.

Figure 12.5. Optic chiasmal syndrome in multiple sclerosis. The patient was a 50-year-old woman with a previous history of transient lower extremity weakness who developed progressive loss of vision in both eyes. The patient reported that her vision would worsen considerably whenever she took a steam bath. Visual acuity was counting fingers at 3 feet OD and 20/100 OS. Color vision was diminished in both eyes, and there was a right relative afferent pupillary defect. A, Kinetic perimetry at presentation shows a complete temporal hemianopia in the field of vision of the left eye and only a small nasal island in the field of vision of the right eye. B, Unenhanced T1-weighted coronal magnetic resonance image shows an apparently normal optic chiasm. C and D, T1-weighted coronal magnetic resonance images after intravenous injection of contrast show enhancement of the intracranial portions of the optic nerves (C) and the optic chiasm (D). (Figure continues.)
Although most compressive and infiltrative or inflammatory lesions that damage the body of the chiasm produce defects that are incomplete and that usually have a relative component, it is not uncommon for a tumor to produce a complete bitemporal hemianopia (Fig. 12.6). Successful decompression of the chiasm in such cases not infrequently results in improvement in the visual field, an outcome that might not have been expected from the severity of the visual field defect, and which can occur within days of surgery (27).
Lesions that Damage the Posterior Angle of the Optic Chiasm

Lesions that damage the posterior aspect of the optic chiasm produce characteristic defects in the visual fields, typically bitemporal hemianopic scotomas (28) (Fig. 12.7). Such defects may be mistaken for cecocentral scotomas and attributed to a toxic, metabolic, or even hereditary process rather than to a tumor; however, true bitemporal hemianopic scotomas are almost always associated with normal visual acuity and color perception, whereas cecocentral scotomas are invariably associated with reduced visual acuity and dyschromatopsia.

Traquair (4) wrote that a lesion that affects the posterior portion of the optic chiasm may be sufficiently focal that it damages only the crossed nasal fibers from the opposite retina, producing a monocular temporal field defect, or only the uncrossed temporal fibers from the ipsilateral eye, producing a monocular nasal field defect. In fact, this is uncommon, and most organic monocular nasal or temporal hemianopic field defects are caused by lesions affecting the optic nerve rather than the optic chiasm.

Lesions located toward the posterior aspect of the optic chiasm may damage one of the optic tracts, producing a homonymous field defect that is combined with whatever field defect has occurred from damage to the optic chiasm.
Bitemporal homonymous scotomas are particularly important in localizing a lesion, or at least the effects of such a lesion, to the posterior aspect of the optic chiasm. Lesions that produce such defects may be more difficult to treat successfully and are more likely to cause permanent residual field defects after surgical therapy or radiotherapy.

Lesions that Damage the Lateral Aspects of the Optic Chiasm

The lateral aspect of the optic chiasm occasionally is damaged by various tumors and, according to some authors, by pressure from the supraclinoid portion of sclerotic internal carotid arteries. When such damage occurs, both the uncrossed temporal fibers from the ipsilateral eye and crossed nasal fibers from the contralateral eye are affected, producing a contralateral homonymous hemianopic defect that cannot be differentiated from that produced by damage to the ipsilateral optic tract (29,30). Although a binalar hemianopia may rarely originate from pressure or other influences affecting the lateral aspects of the optic nerves, it probably never originates from damage to the lateral aspects of the optic chiasm.

Visual Field Defects Caused by Lesions that Damage the Optic Chiasm After Initially Damaging the Optic Nerve or Optic Tract

If there is extension of a lesion from the optic nerve or optic tract to the optic chiasm, the blind eye is usually on the side of the lesion. For example, if a patient with a blind right eye exhibits a defect in the temporal field of the left eye, the lesion obviously is on the right. Similarly, if there has been a left homonymous hemianopia from a right optic tract lesion and if there is extension of the lesion to affect the optic chiasm, blindness (or an extensive field defect) develops in the right eye. Conversely, if a lesion of the optic chiasm that has produced a bitemporal hemianopia extends to the right optic nerve, it will eventually produce loss of vision of the right eye. Similarly, if a chiasmal lesion extends into the right optic tract, there is again loss of vision of the right eye. In other words, if there is extension from an optic nerve or optic tract to the optic chiasm, the blind eye is on the side of the lesion, whereas when there is extension of a lesion from the optic chiasm to the optic nerve or to the optic tract, the blind eye is on the side of extension of the lesion. In such cases, it may be impossible to determine the nature and extent of the pathologic process from an examination of visual sensory function, and particularly visual fields, alone. Rather, it is the history of visual loss and its progression that provides an explanation of the visual findings.

Etiologies of the Optic Chiasmal Syndrome

Damage to the optic chiasm can occur from the direct or indirect effects of a variety of lesions. Bitemporal field defects are usually caused by damage to the optic chiasm from a cerebral mass lesion (31,32). In Rosen’s (33) series, for example, tumors accounted for 80% of the cases of bitemporal field defects. The most common causes of an optic chiasmal syndrome are pituitary adenomas, suprasellar meningiomas, craniopharyngiomas, gliomas, and aneurysms originating from the internal carotid artery (34–38). Chiasmitis (39), particularly related to multiple sclerosis, sarcoi-