Neuro-ophtalmology: Disorders of the Afferent Visual Pathway

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The visual system is a fundamental nervous system component. Neuro-ophthalmologic problems present special challenges in regard to the physician’s knowledge of neuroanatomy and clinical examination skills. Viewing the fundus offers real-time inspection of the nervous system and its pathology. This view reveals a vast arrangement of neural structures dedicated to capturing, processing, and scrutinizing light. Given the evolutionary importance of vision, visual pathways occupy a great extent of the neurologic landscape. Thus, a keen understanding of vision is crucial for appreciating many, if not most, neurologic illnesses.

This manual, the first part of a 2-part review of neuro-ophthalmology, focuses on disorders of the afferent visual pathway and uses clinical cases to illustrate essential concepts guiding the physician to the location of pathology as well as the underlying process of disease. The afferent visual system includes the retina, optic nerve, optic tract, optic chiasm, and retrochiasmal pathways, including the optic radiations and the cortical/higher cognitive areas of visual representation. The afferent visual system, which will be the focus of the next manual, encompasses the pupil as well as the mechanism of ocular motility and cranial nerves involved in eyelid function.

DISEASE OF THE RETINA

CASE 1 PRESENTATION

A 66-year-old woman with a history of hypertension and lower extremity deep vein thrombosis (DVT) presents to the emergency department (ED) after a 5-minute episode of vision loss in the right eye. The patient had been well until earlier that day, when she developed sudden-onset loss of vision from the right eye beginning in the upper visual field. She covered her right eye and observed that vision from her left eye was normal. The patient had no eye pain, change in cognition or language, weakness, or other difficulties. Spontaneous and rapid recovery of vision ensued, after which she noticed no visual or neurologic deficits.

- What is the most likely cause of this patient’s symptoms? What disorders are in the differential diagnosis?

DIFFERENTIAL DIAGNOSIS OF TRANSIENT MONOCULAR VISUAL LOSS

In order to establish diagnostic possibilities in this patient, the evaluation begins with localizing the site of pathology. The patient’s visual symptoms are consistent with transient monocular visual loss (TMVL), which may reflect pathology in various sites including the retina, optic nerve, and even the more posterior visual structures including the cortex. Given the brevity and acuity of her symptoms and the altitudinal pattern of visual loss, the most likely site of disease is the retinal arterial vessels. In this setting, the probable source of arterial disruption is atheroembolism from a stenotic ipsilateral internal carotid artery (ICA).

Vascular Causes

TMVL is most often considered a disease of the retina, and more specifically the retinal vasculature (eg, the ipsilateral ICA).1,2 A severe stenosis of the ipsilateral ICA is made more likely when the patient reports rapid onset of symptoms and TMVL lasting 1 to 10 minutes.3 Visual loss respecting either the horizontal or vertical meridian commonly occurs in this setting and also points to disease of the ipsilateral ICA,4 although alternative sources of embolization are possible. Patients may perceive a curtain being pulled over the eye. Despite a lack of persistent symptoms, retinal emboli may be seen on fundoscopic examination (Figure 1; see page 8).

Additional important causes of TMVL via arterial disease of the retina or optic nerve include giant cell arteritis (GCA),5,6 other ischemic optic neuropathies,7 retinal