Grand Rounds

A 24-year-old woman with blurred vision and eye pain

Aristeidis Konstantinidis,^a Ioannis Athanasiadis,^b Nikolaos Kozeis,^c Claire Workmann,^d and Yajati Ghosh

Author affiliations: ^aRoyal United Hospital Bath, UK;

^bMilton Keynes General Hospital, UK;

^cHippokrateio General Hospital, Greece;

^dCoventry & Warwickshire University Hospital, UK

History

A 24-year-old Caucasian woman presented at the eye casualty complaining of blurred vision and pain with ocular movements in the left eye for one week. Her past medical history was unremarkable and the patient was not on any medication.

Examination

The patient was a high myope with best corrected visual acuity (BCVA) of 6/9 (20/60) in the right eye and 6/60 (20/200) in the left eye. Visual acuity did not improve with pinhole in either eye. On examination there was no relative afferent pupillary defect (RAPD) and both optic discs were normal. However, color vision (tested with Ishihara's pseudo-isochromatic plates) was impaired in the left eye and normal in the right. Red desaturation was present in the left eye.

The neurological examination was normal. Visual field testing showed a diffuse reduction in sensitivity of the left eye with a dense centrocecal scotoma and a superior temporal quadrantanopia in the right eye (Figure 1), which was confirmed with a second visual field test on the following day (Figure 2). This pattern of defects is a junctional scotoma.

Ancillary Testing

An intravenous fluorescein angiogram was normal.

Blood work revealed a normal B12 level, negative anti-nuclear antibody (ANA) level, negative anti-double stranded DNA antibody level, and normal angiotensin

Figure 1. Dense cecocentral scotoma with diffuse reduction of the sensitivity of the left eye associated with a superior temporal quadrantanopia in the right eye (junctional scotoma).

Figure 2. Similar junctional scotoma confirmed on the following day.

converting enzyme level. Chest x-ray showed no evidence of interstitial lung disease or hilar adenopathy.

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Correspondence: Aristeidis Konstantinidis (email: aristeidiskon@hotmail.com).

Figure 3. T1-weighted, gadolinium-enhanced MRI of brain and orbits. (a) Lesions consistent with demyelination plaques on the left side of the optic chiasm (arrow). (b) Lesions consistent with demyelinating areas in the periventricular region.

The patient also underwent magnetic resonance imaging (MRI) of the brain and orbits, which revealed lesions consistent with demyelinating disease at the junction of the left optic nerve and optic chiasm. This is shown on T1-weighted, gadolinium-enhanced scans (Figure 3a). On T1-weighted scans, there were also multiple periventricular demyelinating lesions consistent with demyelinating disease (Figure 3b).

Treatment

Because there was bilateral involvement, the patient received a three day course of intravenous steroids (1000 mg prednisolone), which rapidly improved the BCVA in the left eye to 6/9 (20/60).

Differential Diagnosis

- Multiple sclerosis (MS)
- Space occupying lesion (e.g. tumours, aneurysms, arachnoid cysts or hydrocephalus)
- Vasculitis (e.g. systemic lupus erythematosus)
- Inflammation (e.g. sarcoidosis)
- Trauma
- Vitamin B12 deficiency
- Empty sella syndrome
- Pathological myopia

The diagnoses of space occupying lesion and empty sella syndrome were excluded after the initial scans. Our patient did not give a history of trauma and the diagnosis of vitamin B12 deficiency was also excluded as she did not have any symptoms consistent with vitamin B12 deficiency and its serum levels were normal.

Sarcoidosis was not likely to be the cause of the scotoma as chest X-ray did not reveal any relevant signs and angiotensin converting enzyme levels were normal. Pathological myopia was an unlikely diagnosis according to the fundoscopic and intravenous fluorescein angiogram findings.

Vasculitis was a possible diagnosis along with multiple sclerosis. Systemic lupus erythematosus (SLE), albeit rare, is a well established cause of chiasmatis in the international literature.^{1,2,3} It can affect the visual pathway by either thrombosis of the visual pathway vasculature or by immunological mechanisms such as vasculitis.³ Both mechanisms cause ischemia of the affected structures. The differential diagnosis between MS and SLE becomes more confusing by the fact that involvement of the central nervous system by SLE can cause the development of multiple areas of increased signal intensity in the subcortical white matter on T2 weighted images.¹ In addition to that, there is clinical overlap between neurologic manifestations of MS and SLE. Furthermore 81% of patients with MS are positive for anti-nuclear antibodies (ANA).²

In the case of our patient, ANA and anti-double strand DNA antibodies were negative and medical history and systematic examination did not reveal symptoms or signs of SLE. The course of the disease confirms the diagnosis of MS.

Diagnosis

The most common process affecting the optic chiasm is a compressive lesion such as tumor, aneurysm, arachnoid cyst, or hydrocephalus. More rare etiologies include demyelination, vasculitis (e.g. systemic lupus erythematosus), inflammation (e.g. sarcoidosis), trauma, vitamin B12 deficiency and empty sella syndrome.^{4,5,6} Other authors have described patients with chiasmal visual field defects in whom MS was either the definitive or most likely diagnosis.^{4,7} None of the patients described a junctional scotoma associated with typical multiple sclerosis findings of periventricular white matter and chiasmal lesions showing enhancement with gadolinium, without the presence of RAPD. Our patient was eventually diagnosed with MS based on a new attack in her lower extremities, which occurred several months after her initial ocular complaints. In addition, she had CSF studies, which demonstrated oligoclonal bands and lymphocytosis.

The knee of Wilbrand which was initially described by Hermann Wilbrand^{8,9} is believed to be made up of crossing fibers that originate from the inferonasal retina

(superotemporal visual field), which travel for a short course in the posterior segment of the contralateral optic nerve before they loop back in the contralateral optic tract. As the presence of a RAPD implies an asymmetry of light input between the two eyes, it is likely that the difference in the number of optic nerve fibers affected in the two eyes of our patient was not as great as to clinically produce a RAPD.

The existence of Wilbrand's knee has been disputed¹⁰ and recent sophisticated studies have not confirmed its presence either in humans or other primates.¹¹ Despite these findings the junctional scotoma remains a classic example of damage to the optic chiasm.^{12,13,14} Neurological and radiological investigations are warranted in a case presenting with junctional scotoma in order to identify the nature of the lesion.

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