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Case Report

Idiopathic optic perineuritis: Disguised as recurrent optic neuritis

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1. Initial presentation

A 53-year-old woman with history of mitral valve prolapse and gastroesophageal reflux, presented to the Emergency Department with right eye pain and blurry vision for 2 days. She had acute onset of retro-orbital pain and headache, eye pain with eye movement but no nausea or vomiting. She also reported seeing gray spots and had 'blurry vision' in the right eye. She reported that the temporal visual field was entirely gray in the right eye. She had no prior visual symptoms and no family history of eye disease. She had no other neurological complaints.

Her neurological examination showed extra ocular movements to be intact. She had a relative afferent pupillary defect in the right eye. On Humphrey 24-2 visual field, she had an enlarged blind spot and constriction peripherally. On ophthalmoscopic examination the patient had disk swelling in the right eye (Fig. 1). There were no hemorrhages or exudates. There was pain on eye movement. The remainder of her neurological and general physical examination was normal.

Patient was admitted for evaluation of what was thought to be optic neuritis. MRI brain with and without contrast did not reveal any lesions suggestive of multiple sclerosis or other demyelinating disease. Lumbar puncture was normal. She was started on high dose intravenous corticosteroids with rapid resolution of the pain and her vision improved remarkably. The right temporal visual field defect improved.

2. Relapse

Four months after her initial attack of suspected optic neuritis she had an analogous episode of visual loss in the left eye and a repeat evaluation looking for an etiology. MRI of the brain and spine did not reveal any lesions suggestive of multiple sclerosis (MS). CSF and serum studies including neuromyelitis optica (NMO) antibody, were all negative.

MRI of orbits revealed hyperintensity on FLAIR of the meninges surrounding the affected optic nerve which responded to high dose corticosteroids. She later on had two subsequent relapses affecting the left eye at 4 and 8 month's interval following the first initial episode which affected the right eye.

With each attack she had severe eye pain, worse with eye movement and blurring of vision, but no other neurologic symptoms. She had no diplopia, injection of sclera or conjunctiva during any attack.

She became asymptomatic after treatment of each flare up with intravenous corticosteroids and the exam after her last exacerbation revealed visual acuity of 20/20 in both eyes, and no residual relative afferent pupillary defect (Fig. 2).

MR imaging of the orbits revealed slight enhancement of the left intraorbital optic nerve sheath which led to the diagnosis of perioptic neuritis with the inflammation surrounding the optic nerve rather than causing the inflammation of the nerve itself [Image 1].

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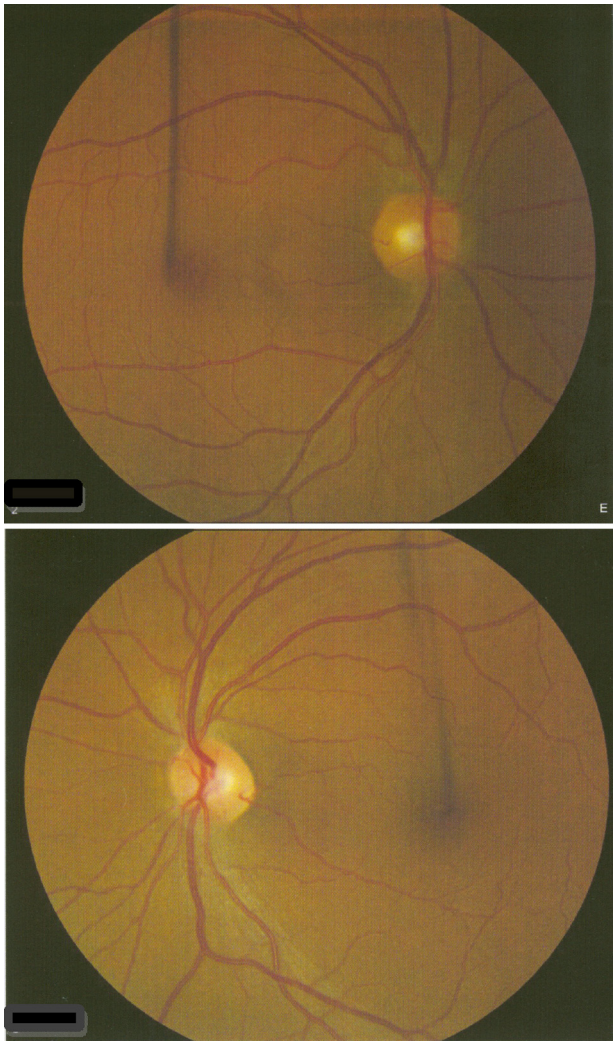

Fig. 1. The right eye has a .3 cup to disk ratio and shows a pink neuroretinal rim. The left eye has .25 cup to disk ratio with nerve fiber layer drop out from the 1 o' clock to 5 o' clock position. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

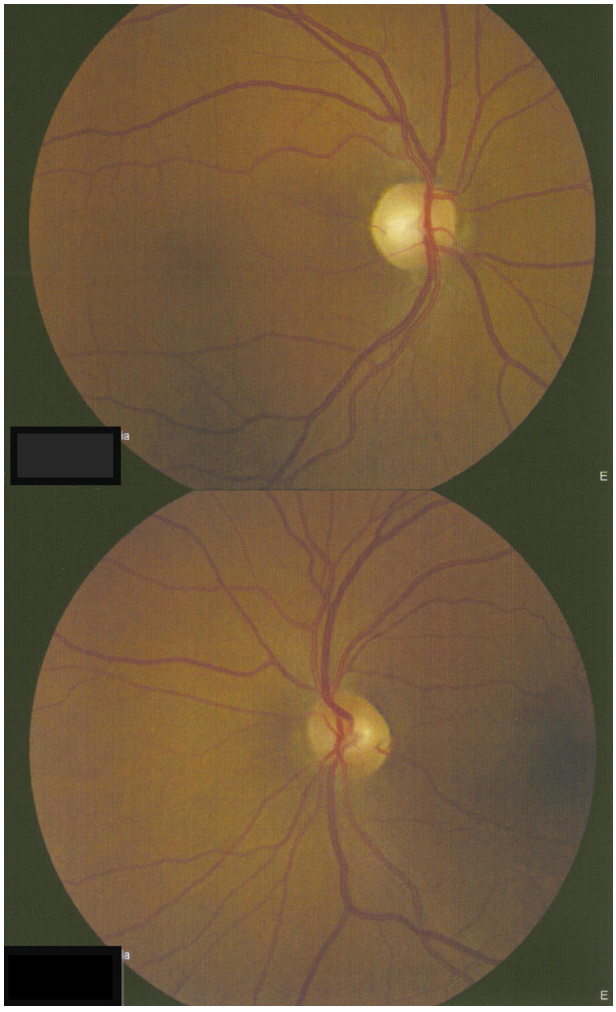

Fig. 2. The right eye has a cup to disk ratio of .5 with neuroretinal rim pallor. The left eye has .25 cup to disk ratio with pink neuro retinal rim. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

3. Discussion

Optic perineuritis (OPN) also known as perioptic neuritis, is a rare orbital inflammatory disorder of the optic nerve dural sheath and is distinct from demyelinating optic neuritis. It has been mostly described as an isolated condition of unknown cause; however, it has also been reported to have occurred with other infectious and inflammatory conditions. OPN usually does not have a recurrence.

In the past two decades, reports with MRI have begun to clearly demarcate this disease from optic neuritis (ON). This differentiation stems primarily from the differences in the clinical course, the MRI appearance and the response to treatment of these two diseases.

OPN, more often than not, presents as an isolated idiopathic disorder. Cases have been reported where it has presented as a manifestation of an underlying pathology such as Wegener's granulomatosis, giant cell arteritis [1], Crohn's disease [2], during the acute phase of (secondary) syphilis [3] as well as a manifestation of neurosyphilis [4], and with pre-B-cell lymphocytic leukemia [5].

ON, meanwhile, is associated with multiple sclerosis (MS) and in most cases is a predictor of MS in at least 50% of the cases. It can occur secondary to autoimmune disorders such as systemic lupus erythematosus (SLE), sarcoidosis, or, due to infections such as syphilis, Lyme disease or tuberculosis.

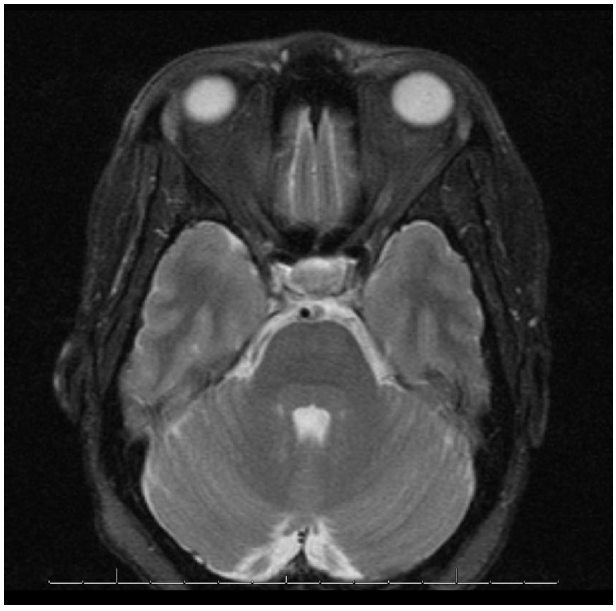

Image 1. MRI of the orbits, T2 axial image of the left eye in the posterior orbit preoptic canal sheath shows a tram track sign consistent with diagnosis of optic perineuritis.