CASE REPORT

Idiopathic Inflammatory Neuroretinitis Simulating Optic Nerve Sheath Dural Ectasia

Süleyman Sami Ilker¹, Göktuğ Seymenoğlu², Serdar Tarhan³, Özgür Uzun⁴, and Seçil Şencan⁴

¹Department of Neuro-Ophthalmology, ²Department of Retinal Diseases, ³Department of Radiology, and ⁴Department of Ophthalmology, Celal Bayar University, Faculty of Medicine, Manisa, Turkey

ABSTRACT

We report a case of a 32-year-old female patient who presented with decreased vision on both eyes and headache. In fundus examination, both eyes had elevation of the optic disc and star shaped hard exudates in the macula. Magnetic resonance imaging was completely normal except the saccular dilatation of bilateral optic nerve sheath. The patient was treated with oral steroids following high-dose intravenous methylprednisolone. She displayed good anatomical and functional results during the follow-up. This case raises the possibility that optic nerve sheath enlargement, probably induced by an idiopathic inflammatory optic neuritis, may simulate dural ectasia of the optic nerve sheath.

Keywords: Magnetic resonance imaging, optic nerve sheath dural ectasia, optic neuritis

INTRODUCTION

Neuroretinitis is an inflammatory optic neuropathy characterised by optic disc oedema associated with a macular star. There is ipsilateral visual acuity and field loss, dyschromatopsia, and a relative afferent pupillary defect. Prior to macular star development, neuroretinitis is virtually indistinguishable from papillitis.¹

Optic nerve sheath dural ectasia is saccular dilatation of the optic nerve sheath which is characterised by expansion of the cerebrospinal fluid (CSF) around the optic nerve without orbital and intracranial neoplasm and inflammation. Imaging of the orbit using magnetic resonance imaging (MRI) in patients with dural ectasia reveals tube-like enlargement of the optic nerve/sheath complex.^{2,3}

There are few reports of neuroimaging studies in patients with neuroretinitis.^{4–6} We report a case of optic neuritis which not only had a clinical course typical for neuroretinitis, but also had neuroradiologic findings most commonly seen with optic nerve sheath dural ectasia.

CASE REPORT

A 32-year-old woman was referred to our clinic with complaints of decreased vision in both eyes and headache with a duration of two weeks. The best corrected visual acuity was 20/32 and 20/50 in the right and left eyes, respectively. Extraocular movements, anterior segment examination and intraocular pressures were normal bilaterally. There were no inflammatory features on slit lamp examination, e.g. vitreous cells on both eyes. She was able to perceive all Ishihara colour plates and no afferent pupillary defect was present. Humphrey central 30° visual field analysis revealed serious concentric constriction up to 5–10° and a generalised depression. On fundus examination, both eyes had elevation of the optic disc, hard exudates originating from the fovea and extending to the optic disc in a radial fashion, and splinter haemorrhages around the disc (Figure 1A and B).

Laboratory studies for Bartonella, angiotensin-converting enzyme, Lyme, Toxoplasma, syphilis, and chest radiograph were all negative. A brain and orbital MRI scan was performed. On orbital axial

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Correspondance: Göktuğ Seymenoğlu, Cengiz Topel cad. No:38/4, Karşıyaka, PO Box 35540, Izmir, Turkey. Tel: 00905323557196. Fax: 00902323362959. E-mail: gseymeno@gmail.com

FIGURE 1. Fundus photographs of right (A) and left (B) eyes: both eyes had an elevation of the optic disc, hard exudates originating from the fovea and extending to the optic disc in a radial fashion, and splinter haemorrhages around the disc.

FIGURE 2. MRI. (A) The brain MRI was completely normal except the saccular dilatation of both optic nerve sheaths. (B) On orbital axial images, the diameter of the optic nerve/sheath complex was 8.2 mm in both eyes, the optic discs were swollen.

images, a dilated optic nerve sheath was identified as containing a CSF-intensity lesion that was hyperintense on T2-weighted images. The optic nerves were normal. The diameter of the optic nerve/sheath complex was 8.2 mm in both eyes. There was no evidence of tumour in the brain or orbit (Figure 2A and B).

Complete blood count and routine blood biochemistry were normal. Erythrocyte sedimentation rate was 38 mm/h, serum C-reactive protein level was 5.15 mg/L (normal range: 0–3), serum rheumatoid factor level was <10 IU/ml (normal range: 0–15). Systemic blood pressure was 120/80 mm Hg. A lumbar puncture was performed. The CSF opening pressure was 150 mm H₂O (normal range: 60–200 mm H₂O). Chemical analysis of CSF showed that glucose, chloride, and protein levels were within the normal limits. There was no leukocytosis and protein

electrophoresis. It was negative for oligoclonal bands. Repeated lumbar punctures were performed and the CSF pressure was again found to be less than or equal to 150 mm H₂O. Therefore dynamic spinal MRI was performed and it revealed no signs of CSF transport obstruction in the subarachnoid space between the brain and the spinal cord. A fundus fluorescein angiography was performed and it revealed intense leakage from both of the optic discs in the early stages.

The patient was treated with 1000 mg of intravenous methylprednisolone for three days followed by 1 mg/kg/day oral steroids as a maintenance therapy. The visual acuity was 20/20 in both eyes four weeks after the therapy and she remained stable in all follow-up examinations. Humphrey visual field analysis that was performed with 8–11 day intervals

FIGURE 3. Humphrey's visual field analysis that is performed with 8–11 days intervals: a series of visual fields showed significant reduction in visual field defects.

showed significant reduction in visual field defects (Figure 3). At day 50 of the treatment; optic disc oedema was significantly reduced on both eyes, haemorrhages were partially absorbed but hard exudates were still present. On orbital axial MRI, the diameter of the optic nerve/sheath complex was decreased on both eyes (Figure 4). At day 100 of the treatment; both fundi showed optic disc pallor and a resolving macular star (Figure 5A and B).

DISCUSSION

Our patient had neuroretinitis of unknown cause, and neuroimaging demonstrated involvement of the optic nerve sheaths. Neuroretinitis is an acute inflammatory disorder in which the specific aetiology is rarely found. When an aetiology is found, it is most often due to cat scratch disease, syphilis, Lyme disease, toxoplasmosis, or leptospirosis,⁷ but laboratory studies were not able to identify an aetiological factor in this patient.

A dilatation of the optic nerve sheath could be secondary to neoplasms and malformations of the orbit and also inflammatory processes. Neuroimaging abnormalities in patients with neuroretinitis have been infrequently reported. Zhang *et al.*⁴ reported a case of idiopathic neuroretinitis. Computed

FIGURE 4. At day 50 of the treatment, orbital axial MRI: the diameter of the optic nerve/sheath complex was decreased.

tomography scan, which does not usually provide enough resolution to distinguish perineural from neural enhancement, revealed "tubular enlargement of the optic nerve and postcontrast enhancement." Histopathologic examination was consistent with optic perineuritis. Wals *et al.*⁵ reported a similar case

FIGURE 5. At day 100 of the treatment, fundus photographs of right (A) and left (B) eyes: Both fundi showed optic disc pallor and a resolving macular star.

of neuroretinitis of unknown cause associated with optic perineuritis. The MRI demonstrated enhancement and thickening of the involved optic nerve sheath with sparing of the nerve. Fat suppression of the orbital soft tissue further supported the diagnosis of optic perineuritis. Vaphiades *et al.*⁸ reported two patients with neuroretinitis in which a contrast enhanced fat-suppressed orbital MRI revealed diffuse enhancement of the optic nerve and sheath on both patients.

Headache and temporary or permanent vision loss can be observed in patients with optic nerve sheath ectasia.² Our patient experienced acute onset of headache and progressive visual loss. MRI scan revealed a dilated optic nerve sheath. Although this combination of symptoms and MRI findings are highly suggestive of dural ectasia, the patient's star configuration of lipid exudates in the macula is atypical.

Neuroimaging findings on our patient further support that orbital optic nerve enhancement may be seen in neuroretinitis and a macular star does not exclude retrobulbar pathology. Perhaps, in some cases of neuroretinitis, the inflammation is so severe and widespread that it extends posteriorly from the optic disc to include the retrobulbar optic nerve. Blood-brain barrier breakdown, similar to what occurs at the level of the optic disc, may produce retrobulbar optic nerve enhancement on orbital MRI.

Schmalfuss *et al.*⁶ evaluated the MRI characteristics of patients with optic neuropathy. In five of the nine patients, who were diagnosed with cat scratch disease, optic nerve enhancement was present on contrasted orbital MRI, but, only one of these five patients had the macular findings of neuroretinitis at the time of MRI scanning. Hence the patients might not have macular involvement and MRI findings simultaneously.

Optic nerve or sheath enlargement, probably induced by an idiopathic inflammatory perioptic neuritis, may simulate dural ectasia of the optic nerve sheath. These kinds of cases still pose a problem in respect of differential diagnosis. A prompt diagnosis has to be made due to the fact that dural ectasia of the optic nerve sheath may sometimes be associated with rapidly progressive and permanent visual loss if not treated at the right time.⁹ Our patient had a clinical course typical for neuroretinitis but also had neuroradiologic findings most commonly seen with optic nerve sheath dural ectasia, which initially might have led to an incorrect diagnosis. In the case illustrated here, clinical presentation of the disease process was correctly applied to the interpretation of the neuroradiologic findings which prevented an unnecessary surgical intervention in this particular patient. In conclusion, clinicians must be aware of the fact that neuroretinitis is an important condition for the differential diagnosis of visual loss when macular involvement is not present but MRI demonstrates enlargement of the optic nerve sheath.

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