Right pupil was reacting sluggishly to light with a relative afferent papillary defect (RAPD). Fundus examination revealed bilateral pale discs. Remaining neurological examination was unremarkable. Perimetry revealed constriction of the visual field in the right eye, without any specific pattern. Laboratory investigations showed a normal erythrocyte sedimentation rate (ESR) and albumin: globulin ratio. The chest X-ray was normal. The Mantoux test and the autoimmune profile (LE cell, antinuclear antibody, and rheumatoid factor) were negative. The Cerebrospinal fluid (CSF) examination was normal and there was no evidence of oligoclonal bands (OCBs) in serum or CSF. Visual evoked responses revealed an increased P100 latency in the right eye (114 m sec) and were normal in the left eye. The brainstem auditory evoked response (BAER) and MRI of the brain and optic nerves (gadolinium enhanced) were normal [Figures 1-4]. Previous investigations including chest x-ray and MRI brain (performed in 2003) were normal. Perimetry performed in previous three episodes showed variable field loss, which did not conform to any specific pattern.

Patient received intravenous methylprednisolone, followed by tapering doses of oral steroids. Pain subsided within two days and vision improved over two weeks to 6/12 in the right eye. At present, patient is maintained on oral deflazacort (1 mg on alternate days), and has been asymptomatic since June 2006.

Discussion

CRION is a recurrent, steroid dependent optic neuritis, without evidence of any additional neurological deficits, sarcoidosis or systemic autoimmune diseases. Kidd et al., in their report of 15 patients, described a syndromic

**Abstract**

Chronic relapsing inflammatory optic neuropathy (CRION) is a recently described recurrent optic neuropathy which is steroid responsive. Several features distinguish this entity from optic neuritis associated with demyelinating disorders and connective tissue diseases. The severe degree of visual loss, persistence of pain after onset of visual loss, and recurrent episodes are unique to this disorder. We describe here a patient who presented with recurrent episodes of painful visual loss, followed by resolution of deficits, over a period of ten years. He was diagnosed as isolated optic neuritis conforming to features of CRION.

**Key Words**

Recurrent optic neuritis, steroid responsive
presentation of subacute optic neuropathy, prominent pain, prompt response to systemic corticosteroids, and relapse on steroid withdrawal.\textsuperscript{[1]} These patients had involvement of both optic nerves with the latency (between attacks), varying from days to 14 years. The features that differentiated this group from the classical demyelinating optic neuritis, included persistence of pain after onset of visual loss, the higher degree of visual loss, and frequent relapses. None of these patients had OCBs in the CSF and their MRI of the brain was normal. Out of 30 optic nerves, 19 imaged in these 15 patients showed abnormalities on MRI including thickening, high signal intensity, or enhancement.\textsuperscript{[1]}

Myers \textit{et al.} have described corticosteroid dependent optic neuritis, not associated with demyelinating disease.\textsuperscript{[2]} In their report of 48 patients, 32 had evidence of SLE, sarcoidosis, or other systemic autoimmune disease. Our patient had multiple attacks of steroid responsive painful optic neuropathy, each associated with significant visual loss, over a period of ten years. He has not developed any symptom suggestive of sarcoidosis or other systemic autoimmune disease, and the laboratory investigations for these entities were also negative.

The clinical presentation of our patient was similar to those described by Kidd \textit{et al.}, in terms of recurrent episodes of subacute onset of visual defect, significant pain and an excellent response to oral steroids.\textsuperscript{[1]} In individuals with the first episode of isolated optic neuritis (ON), clinical features such as, greater degree of visual loss, prominent pain that persists following the acute phase, and a total resolution of symptoms with steroids may be indicative of a benign course. However, a complete diagnostic work-up for demyelination, autoimmune disease, and other possible causes is mandatory in these patients. The possibility of CRION should be considered only in patients with recurrent and isolated ON. Although, no specific laboratory or clinical criteria have been described for the diagnosis of CRION, clinical indicators, including higher degree of visual loss, persistence of pain after visual loss is established, and relapse on steroid withdrawal is helpful. CRION should be alluded to only after exclusion of demyelination and other autoimmune disorders.

The importance of identifying these patients has therapeutic implication as CRION is highly responsive to steroids and may mandate long-term steroid use to maintain remission. Till date, there are no systematic studies evaluating the duration and intensity of immunosuppression required in patients with CRION; the data is purely observational, based on personal experience. The experience in our patient indicates that long-term and low dose corticosteroids are an effective and safe option.

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Source of Support: Nil, Conflict of Interest: Nil