CASE REPORT

Subhyaloid Hemorrhage in a Case of Devic's Disease

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ABSTRACT

A 14-year-old boy was admitted for paraplegia, acute urinary retention, and a sudden reduction in the visual acuity of both eyes. Fundus examination revealed bilateral optic neuritis with large subhyaloid hemorrhage in left eye. Cerebrospinal fluid examination showed mild pleocytosis and absence of oligoclonal band. Magnetic resonance imaging (MRI) revealed hyper intensity in T2-weighted images along the spinal cord. No abnormality was detected in brain MRI. Visual evoked potentials were suggestive of bilateral optic neuropathy. He received intravenous pulse corticosteroids (methyl prednisolone) for 3 days followed by oral prednisolone. There was improvement in both the visual acuity and the systemic manifestations. We diagnosed the case as neuromyelitis optica (NMO) based on the examination and investigation findings. NMO or Devic's syndrome is an uncommon clinical syndrome associating unilateral or bilateral optic neuritis and transverse myelitis. Subhyaloid hemorrhage, as an ophthalmic feature of NMO, has not been reported till date. We report this case so as to alert the clinician about this unusual presentation of NMO.

Keywords: Neuromyelitis optica, optic neuritis, subhyaloid hemorrhage, transverse myelitis

INTRODUCTION

The term "neuromyelitis optica (NMO)" ("Devic's syndrome") refers to a syndrome characterized by optic neuritis and myelitis.[1] Patients with multiple sclerosis (MS), acute disseminated encephalomyelitis, systemic lupus erythematosus, and Sjögren syndrome, viral and bacterial infections can also present with similar neurological impairments. But more commonly no underlying cause can be found.[2]

Neuromyelitis optica is a rare syndrome in Western countries, constituting <1% of demyelinating disease.[3,4] Its incidence worldwide is around 5/100,000. The disease is much rarer in India but more common in Japan and East Asia.[5]

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Ocular manifestations in NMO are usually in the form of unilateral or bilateral optic neuritis or optic atrophy. Subhyaloid hemorrhage, generally seen in anemia, thrombocytopenia and proliferative diabetic retinopathy, has not been reported as a feature of Devic's disease.[6] Childhood cases of NMO constitute a rare clinical entity with possible poor visual and motor outcome. The report by Gokce *et al*.[7] suggested possible poor motor and visual outcome while several other studies have reported good outcome in cases of childhood Devic's.[8-11]

CASE REPORT

A 14-year-old Indian boy was referred to the ophthalmology outpatient department for painless reduction in vision of 3 days duration. He had been on admission in the medical ward for flaccid weakness of lower extremities and urinary retention. There was a history suggestive of upper respiratory tract infection of 1-week prior to the onset of the neurological problems. There was no history suggestive of connective tissue disorder or trauma. There was no significant family history and no history of exposure to any toxins. His best-corrected visual acuity (BCVA) in right eye (RE)

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was hand movement and in left eye (LE) was light perception with an accurate projection of rays in both eyes. Anterior segment examination showed relative afferent pupillary defect in the LE. Fundus examination of LE revealed blurring of the disc margin with disc hyperemia, peripapillary hemorrhage, and a large subhyaloid hemorrhage involving the macula and inferior quadrants [Figure 1a]. RE pupillary reaction was sluggish with normal fundus examination [Figure 1b]. Neurological examination revealed weakness of all four limbs with bilateral positive Babinski sign. Examination of the cranial nerves and cognitive functions were within normal limits. Cerebrospinal fluid (CSF) examination showed clear fluid with, protein concentration of 52 mg/dl and sugar level of 80 mg/dl. There was mild pleocytosis (>40 cells/mm^3) and increased immunoglobulin G. No oligoclonal band was detected. Visual evoked potential (VEP) showed prolonged latency and decreased amplitude of both eyes. T2-weighted sagittal section magnetic resonance imaging (MRI) of the spine showed long segment T2-hyperintensity involving central gray matter and enhancing in T1-contrast images suggestive of myelitis [Figure 2]. Venereal Disease Research Laboratory test, human immunodeficiency virus-enzyme linked imunosorbent assay and viral studies on serum and CSF were negative. Antinuclear antibodies and antibodies to double stranded-DNA were not found. On the basis of clinical findings and MRI picture, we have not come across any such difference of visual acuity outcome in literature,moreover severe visual loss in LE could be due to subhyaloid haemorhage. He was treated with intravenous methylprednisolone (1 g/day for 3 days) followed by oral prednisolone in tapering doses over 2 weeks. Follow up at 1 week, this was the finding seen in our case, subsequently he was lost in follow up. There was complete improvement of his urinary symptoms and limb weakness. However, he was subsequently lost to follow-up.

DISCUSSION

The first case of optic neuritis and myelitis demonstrating inflammatory changes in the spinal cord and optic nerves was reported by Jacob Augustus Lockhart Clarke in 1865.[12] In general, NMO is sporadic, although there are a few case reports of familial occurrences.[13] The clinical course of NMO is described as monophasic or polyphasic. The former type occurs mainly in the pediatric age group and follows a fulminant course with varying degrees of recovery. Polyphasic course is characterized by episodes of relapses and also remissions. NMO predominantly affect smiddle aged adults and it is rare in paediatric age group. If it occurs in paediatric age group, age of onset is usually like the one seen on this case.[14,15]

Cases can present with either visual loss or myelopathy. In most cases, involvements of the spinal cord and optic nerves occur within 3 months of each other.[16] Occasionally, optic nerve and spinal cord symptoms begin simultaneously as in our case. The absolute diagnostic criteria as given by Wingerchuk *et al*.[16] were: (1) Optic neuritis (2) acute myelitis (3) no evidence of clinical disease outside the optic nerve or spinal cord. Our patient also fulfilled the major supportive criteria ([1] negative brain MRI at onset[2] spinal cord MRI with signal abnormality extending over 3 vertebral segments[3] CSF pleocytosis of >50 white blood cell/mm^3 or >5 neutrophils/mm^3) and the minor supportive criteria ([1] bilateral optic neuritis[2] severe optic neuritis[3] severe, fixed, attack-related weakness in one or more limbs).[16] Either one or both eyes may be involved, and the extent of myelitis is variable. Prodromal symptoms like fever, myalgia, headache or sore throat are present in about one-third of cases.[15,16] Visual loss is usually due to optic neuritis or optic atrophy. In the present case, severe visual loss in LE was probably due to a combination of optic neuritis, and a massive subhyloid hemorrhage whereas in

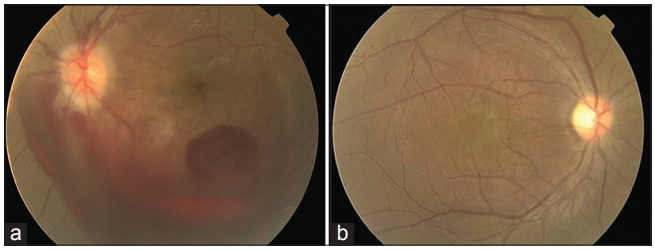

Figure 1: (a) Fundus photograph of left eye showing hyperemic disc with blurred margins, peripapillary hemorrhage, and large subhyaloid hemorrhage. (b) Right eye showing normal fundus

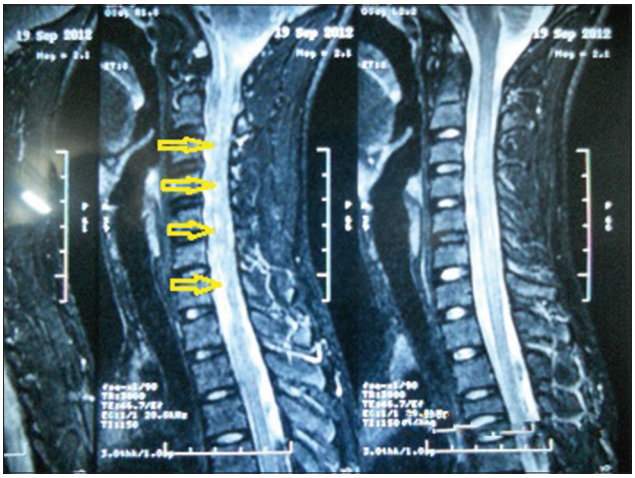

Figure 2: Cervico-thoracic spinal cord magnetic resonance imaging in the sagittal plane T2-weighted image showing increased signal intensity extending from cervicomedullary junction up to the level of T4 vertebra with mild expansion of the cord, involving central gray matter, and enhancing in T1 contrast images suggestive of myelitis

RE was due to retrobulbar neuritis. Retinal and subhyaloid hemorrhages may occur in a case of optic neuritis as reported by Gupta *et al*.[17] The forward dissection of severe peripapillary hemorrhages was probably the cause of subhyaloid hemorrhages in our case, whereas the scattered retinal hemorrhages resulted from the central retinal vein compromise due to the optic disc swelling. Features such as the presence of polymorphonucleocytes or eosinophils, absent oligoclonal bands in the CSF, a normal brain MRI scan, and abnormal VEPs should be evaluated carefully for impending optic nerve involvement. The spinal cord symptoms in NMO are not different from those of other causes of myelitis.[15] If cerebral and brainstem findings are present, a search for alternative etiologies should be done. Oligoclonal bands are reported to be present less frequently than in typical cases of MS.[14,16] Gadolinium enhanced T2-weighted images of MRI spinal cord shows areas of increased signal intensity involving several sections of the spinal cord. The transverse myelitis in NMO is distinct from that seen in MS. In NMO, the transverse myelitis is longitudinally extensive, spanning more than three vertebral bodies in length. In MS, spinal cord lesions usually are more discrete and involve one or two spinal cord segments.[16] Usually, the optic neuritis in NMO is bilateral and retrobulbar and results in severe vision loss, worse than that seen in patients with MS.

Glucocorticoids are typically used to treat cases acutely and may be beneficial.[13,16] Our case also showed dramatic response to pulse steroid therapy. Plasma exchange may be tried in patients who do not respond to glucocorticoids.[18] Interferons and sometimes immunosuppressant drugs are used with the hope that further relapses will be prevented, but prospective data in support of their efficacy are lacking.[14,16]

Pediatric Devic's is a rare clinical entity. Treatment with corticosteroids and immunosuppressant therapy prevents long-term sequelae with excellent visual and neurological prognosis.[9]

Our report indicates that papillitis with subhyaloid hemorrhage can be the presenting feature in a case of pediatric NMO. Timely diagnosis and prompt management may achieve a good visual and neurological outcome.

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