University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2018, Vol. 4(2)

A case of recurrent optic neuritis with transverse myelitis in a young boy

KALAIVANI
Department of Ophthalmology,MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract : Recurrent optic neuritis is associated with demyelinating disorders like multiple sclerosis or with transverse myelitis (neuromyelitis optica) and it is important to differentiate between the two as the treatment and prognosis differ. A 13 year old boy with with recurrent bilateral optic neuritis was treated with intravenous methyl prednisolone followed by oral prednisolone for the first 2 episodes. He developed optic neuritis with transverse myelitis 3 months after the first episode. His MRI brain was normal. MRI spine was abnormal for more than 3 segments. He was treated with azathioprine and his vision improved. This case stresses the importance of the correct diagnosis and appropriate management of bilateral recurrent optic neuritis.

Keyword : Recurrent optic neuritis, Transverse myelitis, Neuromyelitis optica

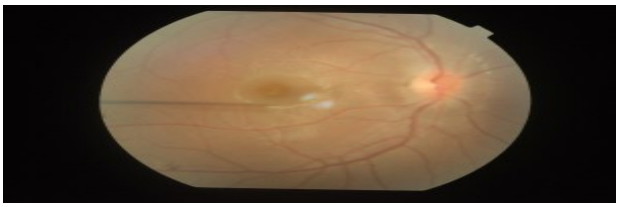
Introduction

Recurrent optic neuritis is associated with demyelinating disorders like multiple sclerosis or with transverse myelitis (neuromyelitis optica) and it is important to differentiate between the two as the treatment and prognosis differ. We here present a case of recurrent optic neuritis and discuss the management options.

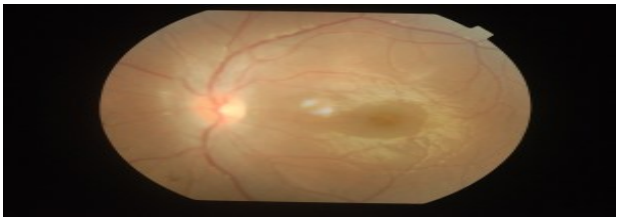
Case Description

A 13 year old boy presented with gradual painless decrease of vision over the course of 2 days in the left eye. On examination, he had a visual acuity was 6/6 in the right eye and 6/36 in the left eye. Anterior segment examination was unremarkable except for the presence of relative afferent pupillary defect (RAPD) in the left eye. Intraocular pressure was 18 mm Hg in both the eyes. Fundus examination of the left eye showed clear media with hyperemic disc with blurred margins and attached retina. Colour vision was defective in the left eye. A presumptive diagnosis of left optic neuritis was made and the boy was given intravenous methyl prednisolone 1 gm for 3 days followed by tablet prednisolone 1mg/kg body weight. His vision improved to 6/12 in the left eye and the patient was discharged. Two weeks later he had decrease of vision in the right eye and further worsening of vision in the left eye. He had a visual acuity of 1/60 in the right eye and HM in the left eye this time.

RAPD was present in the left eye. Fundus examination revealed hyperemic disc with blurred margins in both the eyes. The intraocular pressures were normal. He was diagnosed to have bilateral recurrent optic neuritis and was readmitted and given another course of intravenous methyl prednisolone 1 gm for 3 days followed by oral prednisolone 1 mg/ kg and was transferred to neurology department. His blood investigations were normal. MRI brain also revealed no abnormality. Oral steroids were gradually tapered and his vision improved to 3/60 in the right eye and 1/60 in the left eye and he was discharged. He developed high grade, intermittent fever, 3 months after the first episode, which lasted for about 1 week. It was associated with vomiting for 2 days and arthralgia. The body pain lasted even after the fever resolved. He developed numbness in both the lower limbs, 2 weeks after the fever started. It was over the foot initially but gradually extended upto the chest over a period of 3 days. He also developed weakness for standing, getting up for standing and walking from the next day. He also had difficulty in turning over. However, he did not have difficulty in using both his upper limbs or in raising the neck from the pillow. He had dribbling of urine followed by normal stream and constipation. There no abnormality of sweating. General examination was normal. The pulse rate was 50 per minute and and blood pressure 90/70 mm Hg. Examination of the cardiovascular and respiratory systems revealed no abnormality. Neurological examination was as follows. The higher motor functions were normal. Cranial nerve examination other than optic nerve was normal. Visual acuity in the right eye was finger counting close to face in the right eye and 1/60 in the left eye. Pupils were 3 mm, sluggishly reacting and fundus examination revealed hyperemic disc with blurring of disc margins in the right eye and optic atrophy in the left eye. Motor examination revealed normal bulk but increased tone. There was normal power of neck and upper limb, while the power was reduced for movements at hip, knee and ankle joints. Extensor plantar reflex was elicited on both sides. Pain, temperature and vibration sensations were decreased below the level of T5.

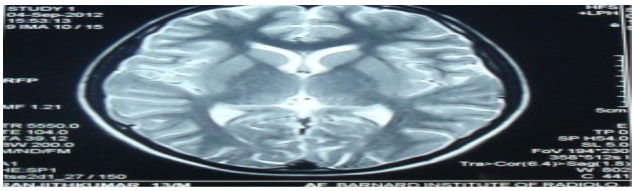


Fundus Photo of the Right Eye

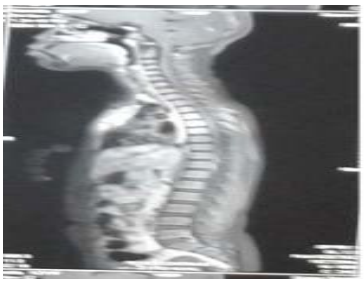


Fundus Photo of the Left Eye

A provisional diagnosis of bilateral recurrent optic neuritis with acute transverse myelitis due to demyelinating disease [multiple sclerosis (MS)] or neuromyelitis optica (NMO) was made. MRI brain with orbit was done which showed enlargement of left optic nerve with T2 flair hyperintensity mainly in the intracanalicular and intraorbital part. On contrast it showed enhancement suggestive of optic neuritis. MRI brain had no abnormalities. However, MRI of the spinal cord revealed T2 weighted hyperintensity from D5 to conus suggestive of NMO. He was started on oral azathioprine 2 mg/kg and put on close follow up. His vision improved to 6/60 in the right eye and was 1/60 in the left eye but his paraparesis persisted. He is on maintenance dose of azathioprine.



MRI Brain



MRI Spine

Discussion

Bilateral optic neuritis is usually seen in children and has a good visual recovery. DEMYELINATING OPTIC NEURITIS (MULTIPLE SCLEROSIS) It is an inflammatory demyelinating CNS disease characterized by relapsing and remitting course. There is an autoimmune attack on the myelin coating of the optic nerve, rather than axons themselves causing their loss. The saltatory conduction breaks down affecting vision. Myelin is phagocytosed by microglia and macrophages, subsequent to which astrocytes lay down fibrous tissue. A similar process affects white matter in brain, brainstem and spinal cord. No peripheral nerves involved. The recovery is also good. NEUROMYELITIS OPTICA (Devic's disease)

It is a rare inflammatory CNS demyelinating disease affecting optic nerve and spinal cord. There is involvement of optic nerve, spinal cord and ocular sympathetic system. There is a considerable debate whether NMO is a variant of MS or not. It affects both eyes simultaneously or sequentially, followed transverse myelitis days to weeks later. The pathology is more extensive, less sharply defined with greater destruction of axis cylinders. There are no white matter lesions on MRI as in MS.

There is CSF pleocytosis and elevated CSF protein. There is also the presence of aquaporin-4 antibody (AQP4-IgG). The visual recovery is poor and it usually requires long term immunosuppressives.

(1) The optic neuritis in NMO patients might be unilateral or bilateral;

(2) the interval over which patients developed the disease index events (optic neuritis and myelitis) has no diagnostic significance; (3) the majority of patients have a relapsing course; (4) the clinical manifestations are frequently severe; (5) cerebrospinal fluid (CSF) abnormalities are characterized by neutrophilic pleocytosis or a greater number of nucleated cells usually associated with negative oligoclonal bands; (6) abnormal signal on spinal cord MRI extend over 3 vertebral segments; and (7) brain MRI is either normal or showed unspecific lesions. These features could promptly help to distinguish NMO from MS and were used, therefore, for formulation of the 1999 diagnostic criteria for NMO1 The diagnostic criteria for NMO was revised2 in 2006 after the discovery of the NMO -specific antibody3 and of AQP4, as its targeted antigen4. It required the presence of Optic neuritis Acute myelitis

At least two of three supportive criteria

1. Contiguous spinal cord MRI lesion extending over 3 vertebral segments
2. Brain MRI not meeting diagnostic criteria for multiple sclerosis
3. NMO-IgG seropositive status Recently the spectrum of NMO has been expanded5 to include Limited forms of neuromyelitis optica Idiopathic single or recurrent events of longitudinally extensive myelitis (3 vertebral segment spinal cord lesion seen on MRI)

Optic neuritis: recurrent or simultaneous bilateral

Asian optic-spinal multiple sclerosis

Optic neuritis or longitudinally extensive myelitis associated with systemic autoimmune disease.

Optic neuritis or myelitis associated with brain lesions typical of neuromyelitis optica (hypothalamic,

corpus callosal, periventricular, or brainstem)

Some patients with aquaporin-4 autoimmunity present brainstem, hypothalamic or encephalopathy symptoms either preceding an index event or occurring isolatedly with no evidence of optic nerve or spinal involvement. On the opposite hand, other patients have optic neuritis or longitudinally extensive transverse myelitis (LETM) in association with typical lesions of NMO on brain MRI and yet are AQP4-IgG seronegative. An expanded spectrum of NMO disorders was proposed6 to include these cases. Currently, NMO is considered as a central nervous system (CNS) AQP4 channelopathy which causes variable damage predominantly to the optic nerves and spinal cord, although other CNS structures that highly express AQP4 may be also affected.

Differentiating MS and NMO

Feature	MS	NMO
Laterality	Usually unilateral	Usually bilateral
Visual loss	Less	More
Visual Recovery	More	Less
White matter lesions on Brain MRI	Usual	Rare usually resolving
Transverse myelitis	Rare	Spans > 3 spinal cord segments
Clinical involvement beyond spinal cord and optic nerve	Usual	Rare
Tissue destruction and cavitation	Less	More
CSF oligoclonal bands	Frequent	Rare
CSF protein content	Less	More
Treatment with DMDs	Effective	Ineffective, even worsening
Treatment with Immunosuppressives	Firstline	Firstline

Our patient had bilateral recurrent optic neuritis and acute transverse myelitis. MRI brain was normal
but MRI spine revealed involvement of more than 3 spinal cord segments. He was treated with azathioprine and his vision improved. This case stresses the importance of the correct diagnosis and appropriate management of bilateral recurrent optic neuritis.

References

1.Wingerchuk DM, Hogancamp WF, O'Brien PC, Weinshenker BG. The clinical course of neuromyelitis optica (Devic's syndrome). Neurology 1999;53:1107-14.
2.Wingerchuk DM, Lennon VA, Pittock SJ, Lucchinetti CF, Weinshenker BG. Revised diagnostic criteria for neuromyelitis optica. Neurology 2006;66:1485-9.
3.Lennon VA, Wingerchuk DM, Kryzer TJ, et al. A serum autoantibody marker of neuromyelitis optica: distinction from multiple sclerosis. Lancet 2004;364:2106-12.
4.Lennon VA, Kryzer TJ, Pittock SJ, Verkman AS, Hinson SR. IgG marker of optic-spinal MS binds to the aquaporin-4 water channel. J Exp Med 2005;202:473-7.
5.Wingerchuk D. M., Lennon VA, Lucchinetti CF, Pittock SJ, Weinshenker BG. The spectrum of neuromyelitis optica. Lancet Neurol 2007;6:805-15.
6.Lana-Peixoto MA, Callegaro D. The expanded spectrum of neuromyelitis optica – evidences for a new definition. Arq Neuropsiquiatr 2012;70(10):807-13

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities