Case Report

Neuromyelitis optica: a demyelinating disease

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ABSTRACT

Neuromyelitis optica (NMO) also known as Devic’s disease is a demyelinating disease. It involves the central nervous system and predominantly affects the spinal cord and optic nerves. It is characterised by episodes of optic neuritis and/or transverse myelitis. It is often confused with multiple sclerosis. NMO can occur rarely in children but cases of pediatric NMO need specific consideration owing to possible poor visual and motor outcome. We report a case of nine year male child admitted with history of fever followed by sudden weakness of body, sudden decline in visual acuity and involuntary passage of urine. The symptoms were reverted after the use of methylprednisolone. Clinical outcome was favourable with disappearance of sphincter troubles, improvement of visual acuity and progressive disappearance of motor troubles.

Keywords: Neuromyelitis optica, Optic neuritis, Multiple sclerosis, Methylprednisolone

INTRODUCTION

NMO is a heterogenous demyelinating disease of central nervous system characterised by recurrent and simultaneous inflammation and demyelination of optic nerve (optic neuritis) and the spinal cord (myelitis). Although it predominantly affects middle aged adults, pediatric cases of NMO constitute a distinctive clinical entity. Although it may be confused with Multiple Sclerosis (MS), it has a specific diagnostic criteria and unique pathologic features. Early discrimination between NMO and MS is important because the two diseases have different natural histories and treatment regimens.

CASE REPORT

A nine year male child presented with history sudden weakness of both lower limbs, sudden loss of vision in both eyes and involuntary passage of urine since last six days. There was history of fever with headache in last 10 days. Fundoscopy revealed optic disc swelling retinal hemorrhage. Ophthalmology evaluation was suggestive of bilateral severe optic neuritis.

Neurological examination revealed weakness of both lower limbs (without spontaneous mobility), deep tendon reflexes were exaggerated, reduced vibratory sensitivity in lower limb and pain when the lower limb were palpated. Bilateral plantar extensors were noted. Other cranial nerves and cognitive functions were normal.

Laboratory evaluation revealed normal hemoglobin, white blood cell counts was raised having neutrophilic predominance. Analysis of cerebrospinal fluid (CSF) revealed a white blood cell count of 55 cells, protein 44 mg/dl, and sugar 79.4 mg/dl. No oligoclonal bands in CSF. Magnetic Resonance Imaging (MRI) revealed abnormalities of spinal cord signal with hypo intensity in T1-weighted images and hyper intensity in T2-weighted images along the spinal cord. However, the cerebral region was normal. NMO-specific serum autoantibody
against the water channel aquaporin-4 (Aqp4) was sent which came out to be positive.

A diagnosis of NMO was made and the patient was treated initially with intravenous methyl prednisolone (30 mg/kg/day for five days) under cover of intravenous ceftriaxone as there was history of fever. Five pulses of intravenous methylprednisolone were given and patient improved considerably. He was advised to continue oral steroids. He was started on azathioprine after 20 days and was advised for Pulse methyl prednisolone therapy to be repeated. At the time of discharge from the hospital, there were no sphincter troubles, improvement of visual acuity and progressive improvement in motor function with weight bearing on his legs with some limping.

DISCUSSION

NMO is a demyelinating disorder characterised by monophasic or polyphasic episodes of optic neuritis and/or transverse myelitis. An association of spinal cord and optic nerve disorders was first reported in early 19th century.⁴ Although the inflammation may also affect the brain, the lesions of NMO are different from those seen in multiple sclerosis.⁵

NMO can occur in children but pediatric NMO needs specific consideration owing to poor visual and motor outcome.⁶ NMO is more common in women and girls than in men and boys, with female comprising over two-thirds of patients.⁷

NMO is associated with IgG antibodies to the aquaporin-4 water channel.⁸ But it is not clear why the attack on and depletion of spinal cord and optic nerve aquaporin-4 results in disruption of the myelin sheath in Central nervous system, but pathologic examination of the autopsy sample revealed both anti-aquaporin-4 IgG deposits and B cells in the spinal cord and optic nerves of NMO patients. In 2004, NMO-IgG (currently known as Anti-Aquaporin IgG) was first described⁹ leading to the distinction between positive and negative cases.

NMO was previously thought as a variant of MS but, it is different from MS.⁴,⁵ NMO usually spare the brain in the early stages in contrast to those in MS. And at initial stage of NMO, brain MRI is normal.² However, few patients may be present with brain symptoms with brain lesions as their first manifestation and develop recurrent brain symptoms without optic neuritis or myelitis.³

The diagnosis of NMO is based on clinical symptom. The main symptoms are loss of vision and spinal cord function. Optic neuritis may manifest as decreased visual acuity, visual field defects or loss of colour vision. Spinal cord involvement may lead to muscle weakness, decreased sensations, loss of bowel and bladder control. Diagnostic criteria of NMO includes the presence of acute optic neuritis and myelitis with at least two of the three supportive criteria, which consist of spinal cord MRI lesion extending over three vertebral segments, brain MRI lesion, which does not meet the diagnostic criteria for MS and NMO-IgG seropositive status.³ Detection of NMO-IgG antibody distinguish it from other demyelinating diseases. The NMO-IgG autoantibody is 76% sensitive and 94% specific for the diagnosis of NMO.³ CSF examination plays an important role in making the diagnosis and it is usually suggestive of pleocytosis and presence of oligoclonal bands in about 15-30% of NMO cases.⁶

There is no cure for NMO, but symptoms can be treated. Some patients recover, but many are left with visual and motor impairment, which can be severe. For acute attacks, intravenous methylprednisolone is used.⁷ If acute attacks does not respond to methylprednisolone, then plasmapheresis is the effective treatment which helps in removing the antibody.³ Immunosuppressive drugs like azathioprine, rituximab in addition with steroids are also helpful in decreasing the frequency and severity of attack.⁸ Few studies have reported a beneficial role of glatiramer acetate in NMO.⁹

Therefore, we conclude that the possibility of NMO should always be considered whenever clinical symptoms such as those observed in our patient are present. NMO is a rare, devastating disease affecting both visual and neurological system. The prognosis in NMO is generally poor. Some measure of improvement appears in a few weeks, but residual signs and disability may persist, sometimes severely. Future immunomodulatory intervention may augment the benefits of rehabilitation. Further long term studies on NMO primarily focusing on children are needed.

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