

Paediatric neuromyelitis optica

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Objective: To report a case of paediatric Neuromyelitis Optica (NMO)

Method: Case report

A 14-year-old girl presented with blurring of vision in her right eye for 4 days, with associated acute bilateral lower limb weakness and urinary incontinence. At presentation, visual acuity for her right eye was counting fingers and 6/9 in her left eye, with positive right relative afferent pupillary defect. Light brightness and red saturation in her right eye were reduced to 50% as compared to the left eye with right central visual field defect. The anterior segments and funduscopy of both eyes were normal.

On systemic examination, there were upper motor neuron signs and segmental sensory loss from T1-L2. Motor power for both upper and lower limbs were reduced with hyperreflexia. Blood investigation for Aquaporin 4 Ab was positive. Magnetic Resonance Imaging of the spine showed longitudinal extensive transverse myelitis.

She was given intravenous methylprednisolone 1 gram daily for 5 days followed by a tapering dose of oral prednisolone. She completed 5 cycles of plasmapheresis. Subsequently, she had near complete recovery with normal lower limb power. Her right eye vision had improved significantly to 6/9. She completed 3 cycles of Rituximab infusion.

Conclusion: This case report highlights the importance of early diagnosis in paediatric neuromyelitis optica and prompt treatment which may improve the prognosis of the disease.

Conflicts of interest: The authors report no conflicts of interest.

Keyword: Paediatric, Neuromyelitis optica, Immunomodulatory, Plasmapheresis

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Introduction:

Paediatric onset of neuromyelitis optica (NMO) is found in approximately 4% of reported cases.¹ Early differentiation of NMO from other childhood demyelinating disorders is crucial for institution of appropriate therapy and proper counselling to family and patient.²

Here we describe a case of a young patient with Neuromyelitis Optica and to emphasize the

early diagnosis and prompt treatment that may improve the prognosis of disease and prevent disability.

Method: Case report

A 14 year old girl presented with acute onset of right eye blurring of vision for 4 days, which was associated with bilateral lower limb weakness and urinary incontinence. She had no fever, upper respiratory tract infection symptoms, loss of appetite or loss of weight.

At presentation, visual acuity on the right eye was counting fingers and left eye vision was 6/9 with positive relative afferent pupillary defect on the right eye. Light brightness and red saturation were reduced to 50% as compared to

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the left eye with right central visual field defect. The anterior segments of both eyes were normal. Fundoscopy showed normal optic disc in both eyes with no optic disc swelling.

On systemic examination, there were upper motor neuron signs and segmental sensory loss from T1-L2. Muscle power for both upper and lower limbs were reduced 2-3/4 with presence of hyperreflexia. Babinski reflex was positive.

Magnetic Resonance Imaging of the spine showed long segment spinal cord hyperintensity with predominant central distribution from lower medulla to T11 with spinal cord edema at C2-C6 level consistent with a longitudinal extensive transverse myelitis (Figure 1). However, the author was unable to comment on the optic nerves due to metal artefacts from dental braces that obscured both orbits. No brainstem lesion seen. Blood investigations for Aquaporin 4 Ab was positive. CSF results show remarkably high immunoglobulin G (IgG) with no evidence of infection. Other autoimmune laboratory tests such as C3, C4 and rheumatoid factor (RF) were normal (107, 25, <11 respectively), anti-ds DNA was negative and antinuclear factor (ANF) was positive

She was given intravenous methylprednisolone 1gram daily for 5 days followed with a tapering dose of oral prednisolone over two months duration. She completed 5 cycles of plasmapheresis. Plasmapheresis was done in this case in view of her extensive spinal cord involvement and not much improvement of her vision after 5 days of IV methylprednisolone. Subsequently, she had near complete recovery with return of lower limb power. Her right eye vision improved significantly to 6/9. She completed 3 cycles of Rituximab infusion. She had no relapse for the past 2 years and currently on tablet Gabapentin 600mg tds for her neuropathic pain. Repeated MRI spine 1 year after the diagnosis showed resolution of the long extensive transverse myelitis. (Figure 2)

Discussion:

Neuromyelitis optica (NMO), otherwise known as Devic Disease, is an uncommon clinical syndrome of central nervous system inflammatory demyelination, comprising of optic neuritis and transverse myelitis.² It is a rare disease with most reported cases were in adult. The median age of children diagnosed

with NMO ranges from 10 to 14 years and the youngest reported being a 23-month-old boy³. Female preponderance is observed in both the adult and pediatric age groups.³ It is important to differentiate NMO from other diseases such as multiple sclerosis (MS) and acute disseminated encephalomyelitis (ADEM) as there are differences in treatment and prognosis.

NMOSD is stratified into two types: NMOSD with AQP4-IgG (NMOSD-AQP4); and NMOSD without AQP4-IgG or with unknown AQP4-IgG status. According to the new diagnostic criteria, NMOSD-AQP4 refers to patients (1) who have at least one core clinical characteristics of NMOSD in either optic neuritis, acute myelitis or area postrema syndrome (2) who were tested positive for AQP4-IgG; and (3) in whom alternative diagnoses are excluded.⁴

In our case, the patient presented with unilateral retrobulbar optic neuritis and transverse myelitis. Positive serology for AQP4-IgG along with extensive transverse myelitis aided us in making the diagnosis. Typically, cases in children are preceded with infection but our patient did not show any sign and symptom of infection prior to her presentation. As compared to MS, NMO is more common in paediatric patient and may have more profound vision loss, myelitis and intractable nausea and hiccup due to association with area of postrema lesion.⁵ While ADEM is typically an isolated event and can be the first manifestation of multiple sclerosis in children. Most children with ADEM initially present with fever, meningeal signs, and acute encephalopathy.⁶ MRI findings also differ between these three entities. Cacciaguerra L et al proposed that fulfillment of at least 2 of the 5 of the following conditions identified NMOSD with 91% specificity and 82% sensitivity: absence of juxtacortical/cortical lesions, absence of periventricular lesions, absence of Dawson fingers, presence of LETM, and presence of periependymal lesions following the lining of the lateral ventricles.⁷ In patients with such features, AQP4 serum testing should be used to help in diagnosis and to guide treatment.

The clinical course of NMO is variable. It may occur either as a severe monophasic illness or associated with varying degrees of recovery or polyphasic courses characterized by relapses and remissions² The mainstay of NMO therapy are immunomodulatory and immunosuppressive

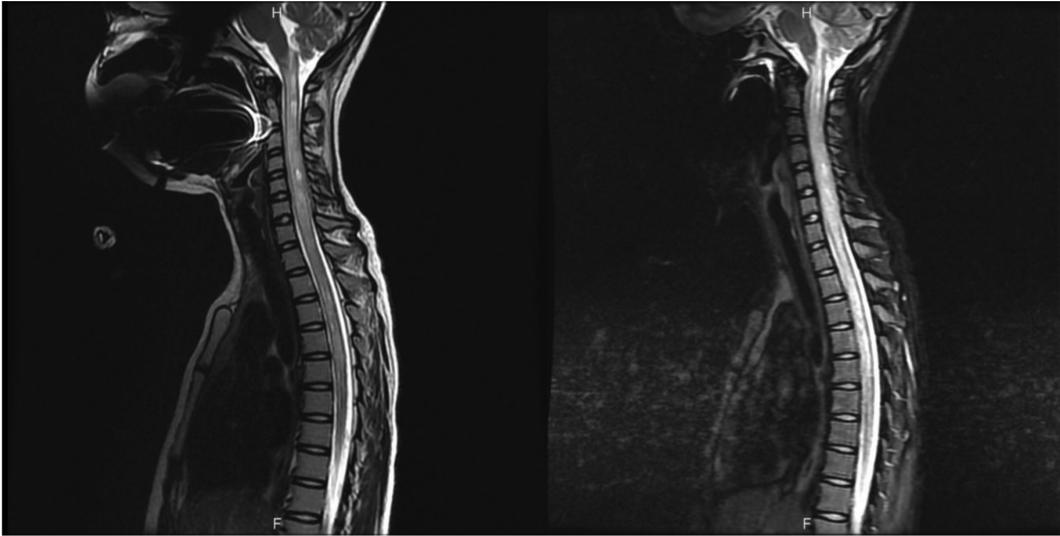


Figure 1: T1W and T2W Sagittal MRI shows diffuse long segment ill-defined patchy with high signal intensity within the spinal cord from lower medulla to T11 level and swollen spinal cord from C2 to C6 level.

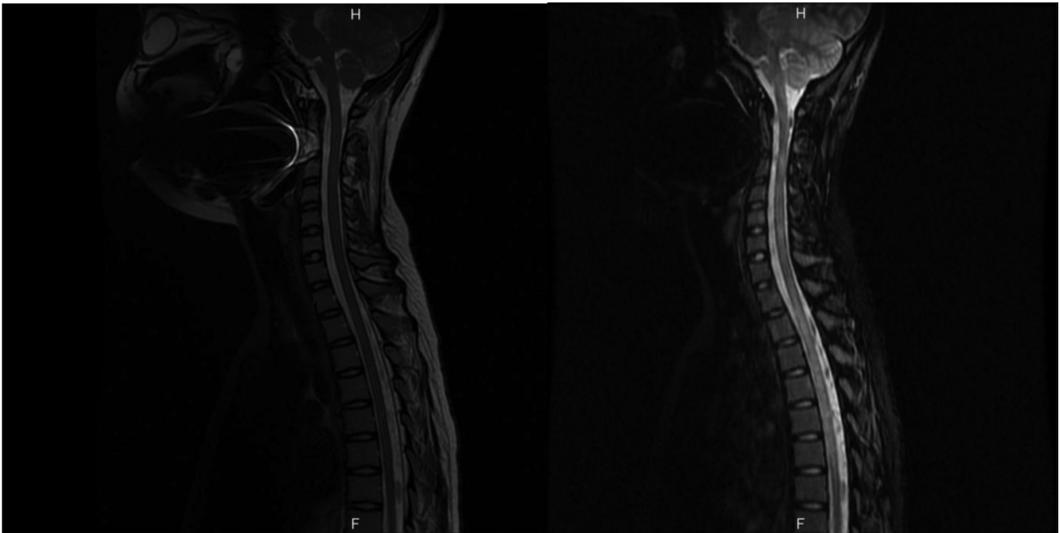


Figure 2: Previously seen patchy cord enhancement and T2W high intensity from C1 to T10 is no longer seen. Residual small hyperintense focus in anterior spinal cord at T11 and T12 is similar in appearance.

drugs to treat the acute attacks as well as prevention of neurological complications and rehabilitation. The first treatment typically given to a patient with NMO is high-dose IV methylprednisolone to reduce disease activity and further progression.

Plasmapheresis following IVMP therapy effectively removed anti-AQP4 antibodies and is accompanied by a substantial improvement in the neurological disability of patients with NMO. Study done by Su hyun Kim et al showed plasmapheresis resulted in marked reduction in serum anti-AQP4 antibody levels in patients with seropositive for anti-AQP4 antibodies.⁸ Previous studies involving patients with CNS inflammatory disease found that starting plasmapheresis early (within 15 to 20 days) is the most important predictor of a favorable response to the procedure⁹

Immunomodulatory drug used in this case is Rituximab. Rituximab is a human monoclonal antibody directed against CD20 that induces B-cell depletion when administered in vivo. It selectively depletes the humoral component of the immune system making it more effective for treating NMO.¹⁰

Absoud M et al reported that AQP4-Ab positivity is associate with early recurrence and visual impairment while physical disability in AQP4-Ab negative relapsing cases¹¹. Thus, early AQP4-Ab testing may allow prompt immunomodulatory treatment to minimize disability.

Our patient is among the 10 -27 percent of patients with a monophasic course¹². The 5-year survival rate is reported to be 90% for patients with a monophasic course of disease and 68% for relapsing patients¹³. She responded very well with treatment without any relapse in the last 24 months of diagnosis. She continued to show good vision and was free from any neurological symptoms 2 years after the initial presentation.

Conclusion:

This case report highlights the importance in early diagnosis of paediatric neuromyelitis optica and prompt treatment will ensure better prognosis of disease and prevent disability.

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