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NEUROMYELITIS OPTICA (DEVIC'S DISEASE) IN A 10 YEARS OLD BOY.

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ABSTRACT

Neuromyelitis optica (NMO) also known as Devic's disease is an acute demyelinating disorder combined with optic neuritis and transverse myelitis.A 10 years old boy presented in the ER with the complaints of fever and back pain for past 10 days and inability to walk for 6-7 days. He had developed urinary retention and constipation for the past 5 days along with abdominal distension. There was also blurring of vision in the left eye with only light perception and rapid afferent papillary defect was present while the right eye was normal. The provisional diagnosis was Transverse myelitis vsNeuromyelitisoptica (NMO). CSF revealed TLC 4, protein 54mg/dl, glucose 66mg/dl. Oligoclonal bands were negative. There was raised CPK. Visual evoked potential (VEP) showed prolongation of P100 latency along with amplitude loss, in left eye.His MRI spine showed extensive involvement of the spinal cord especially the cervical cord and there was no involvement of the brain. Left eye had optic neuritis.He was treated with steroid pulse therapy and later Plasmapharesis and was discharged home on azathioprine with no motor deficit but the visual loss was irreversible

KEY WORDS: optic neuritis, myelitis, Devic's disease

INTRODUCTION

Neuromyelitis optica (NMO) also known as Devic's disease is an acute demyelinating disorder comprising of optic neuritis and transverse myelitis.1 NMO predominantly affects middle aged adults, while case reports from the pediatric population have been increasing in past few years. A recent case series of pediatric NMO revealed strong female predominance2. NMO has specific diagnostic criteria and unique pathologic features when compared with multiple sclerosis (MS) 3. Pediatric NMO has comparatively poor visual and motor outcomes.1Traditionally, the term NMO was applied to those patients who experienced a monophasic event consisting of bilateral simultaneous optic neuritis and acute myelitis.4

CASE REPORT

A 10 years old boy presented in the ER with the complaints of fever and back pain for past 10 days and inability to walk for 6-7 days. He had developed urinary retention and constipation for the past 5 days along with abdominal distension. For the past two days he had been complaining of blurring of vision in the left eye. There was no history of any recent viral illness, vaccination or trauma. He had been to a hospital where he was catheterized and started on broad spectrum antibiotics for the past 4 days but there was no improvement they came to this hospital. His birth and development was normal and he was a student of class III. There was no history of any allergies or surgery. Immunization was up to date. Family history included two sisters and three brothers who were well and healthy. There was no family history for any neurological disease or unexplained visual loss. On examination the patient was fully alert, interactive and oriented. His anthropometric measures were on the 50th centile. He was afebrile, with stable vitals. There was no rash or lymphadenopathy. He was well hydrated and well nourished. His chest was clear with normal vesicular breathing while abdomen was full with slit like umbilicus and urinary bladder was palpable at the level of umbilicus, there was no shifting dullness or visceromegaly and gut sounds were audible. His GCS was 15/15 and higher mental functions were intact. Cranial nerves were intact. Tone was normal in all four limbs; power was 4/5 in both upper limbs and 1/5 in lower limbs. Deep tendon reflexes in upper limbs were +2 and in lower limbs +3. Planters were up going. Superficial reflexes were absent. On sensory examination pain, temperature, vibration and proprioception were intact. There were no signs of meningeal irritation. He was unable to sit and

complained of pain on movement. His left eye had only light perception and showed rapid afferent papillary defect while the right eye was normal
The provisional diagnosis was Transverse myelitis vsNeuromyelitisoptica (NMO). Complete blood count and basic metabolic workup was normal. Inflammatory markers (C- reactive proteins and Erythrocyte sedimentation rate) were within normal limits. CSF revealed TLC 4, protein 54mg/dl, glucose 66mg/dl. Oligoclonal bands were negative. There was raised CPK. Visual evoked potential (VEP) showed prolongation of P100 latency along with amplitude loss, in left eye.MRI revealed multiple abnormal T2 hyper intense signals showing post contrast enhancement almost along the entire spinal cord. These were predominately identified in the cervical region of the spinal cord along with swelling of the cord in this region. Overall appearance was suggestive of transverse myelitis. Bilateral globes and optic nerve appeared unremarkable. The grey matter and white matter appeared normalA diagnosis of Devic's disease was made and the patient was treated initially with intravenous methyl prednisolone (30 mg/kg/day for five days) followed by gradually tapered oral steroids. Condition of child remained static; therefore Plasmapharesis was done for 5 days. Azathioprine 2 mg/kg/day was added and limb physiotherapy started. Clinical outcome was favorable with significant reversible of power and function of limbs.

Discussion

By definition NMO is a monophasic or relapsing disorder of the optic nerves and the spinal cord, without evidence of white matter dysfunction of the brain, brainstem or cerebellum, with the exception of hypothalamic and lower brainstem dysfunction. Optic nerve and spinal cord dysfunction might be partial and it can be unilateral or even subclinical, with an abnormal visual evoked potential but no clinical signs. Partial forms of NMO are also called high-risk NMO, in which isolated transverse myelitis or optic neuritis occurs. The optico-spinal form of MS is quite similar to NMO. Hence, it appears to be a spectrum of NMO, with various degrees of involvement but course of NMO is more acute, sometimes fulminant. In contrastwith the optic neuritis of MS, NMO optic neuritis can be severe, fulminant and devastating with very poor prognosis.5Brain lesions have been observed in children; in one series 68% of NMO-IgG seropositive children and among them 45% had brain symptoms which corresponding to the MRI abnormalities6. In contrast to MS, attacks in NMO commonly spare the brain in the early stages. Hence, normal brain MRI is a common finding at the onset of NMO, so on follow-up scans must be performed periodically for development of later lesions in the course of the disease.4 Lesions in the Spinal cord are large, and extends over three or more vertebral segments in about 85-90% of patients and are mostly located in the cervical and upper thoracic region. The Current revised diagnostic criteria of NMO tells 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 consecutive vertebral segments, brain MRI lesion, which does not meet the diagnostic criteria for multiple sclerosis, and NMO-IgG seropositive status.Our patient had no systemic disorder or non-organ-specific autoimmune disorder or autoantibody. At the time of the left eye's involvement, our patient fulfilled the diagnostic criteria.7 CSF examination of our patient showed no pleocytosis. Oligoclonal bands of IgG in the CSF are frequently seen in MS and these are detected in 15-30% of patients with NMO.8 Oligoclonal bands were negative in our patient's CSF examination.A number of modalities have been tried but there is no proven treatment protocol as yet either in the acute attacks or in the long term remissions in NMO. Hence Intravenous corticosteroid therapy is the commonly preferred initial treatment for acute attacks.7 A total of 50% of the patients, those who are unresponsive to corticosteroid treatment may benefit from plasmapheresis.7 The efficacy of immunomodulatory therapies (Beta-interferon) have not been proven yet. However, immunosuppressive therapy (oral azathioprine, associated with or without oral steroids; intravenous immunoglobulin; Rituximab) is an accepted method to provide clinical remission of corticosteroid resistant NMO.9Our patient received both steroid pulse therapy and Plasmapharesis and was sent home on oral steroids and azathioprine. He became fully mobile with no neurological deficit in the lower limbs though his vision was lost in the left eye

Conclusion

NMO is a rare but severe devastating disease affecting vision and nervous system resulting in blindness and paraplegia in children. Hence an early intervention with the appropriate treatment modality in patients suspected with NMO decides on the favorable outcome from an acute episode

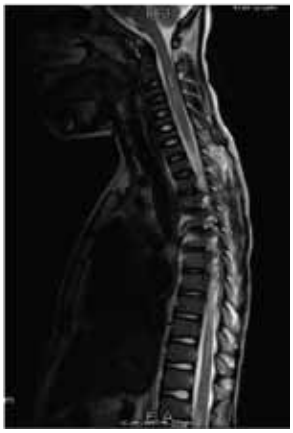

Fig. 1a

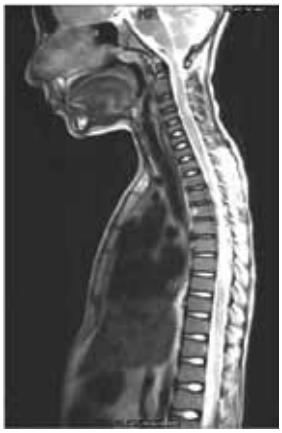

Fig. 1b

Fig 1a and b: Abnormal T2 hyper intense signals showing post contrast enhancement predominantly in the cervical region along with swelling of the cord

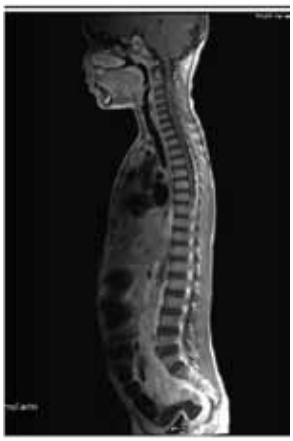

Fig. 2a

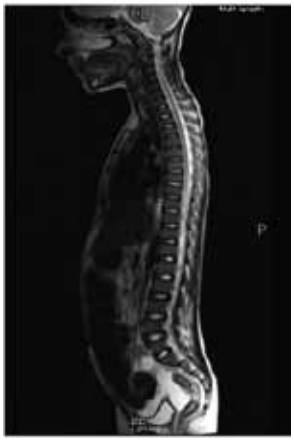

Fig. 2b

Fig 2a and b: The entire spine shows post contrast enhancement with marked hyper intense signals in the cervical cord

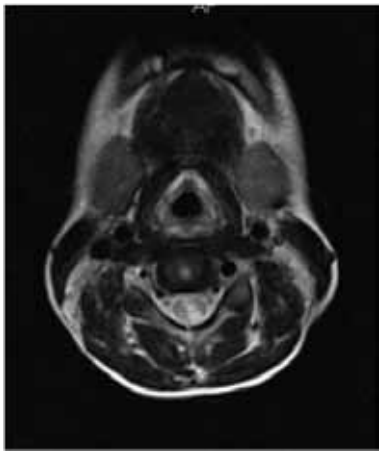

Fig. 3

Fig 3. Shows transverse section of the cervical spine with hyper intensities more marked posteriorly as compared to anterior cord

References

1. Saiz A, Zuliani L, Blanco Y, Tavolato B, Giometto B, Graus F. Revised diagnostic criteria for neuromyelitisoptica (NMO). Journal of Neurology. 2007;254(9):1233-7.
2. Tillema JM, McKeon A. The Spectrum of Neuromyelitis Optica (NMO) in childhood. Journal of Child Neurology.27(11):1437-47.
3. Barnett MH, Sutton I. Neuromyelitis optica: not a multiple sclerosis variant. Current opinion in neurology.25(3):215-20.
4. Wingerchuk DM, Lennon VA, Pittock SJ, Lucchinetti CF, Weinshenker BG. Revised diagnostic criteria for neuromyelitisoptica. Neurology. 2006;66(10):1485-9.
5. Mandler RN. Neuromyelitis optica, Devic's syndrome, update. Autoimmunity reviews. 2006;5(8):537-43.
6. McKeon A, Lennon VA, Lotze T, Tenenbaum S, Ness JM, Rensel M, et al. CNS aquaporin-4 autoimmunity in children. Neurology. 2008;71(2):93-100.
7. Wingerchuk DM, Lennon VA, Lucchinetti CF, Pittock SJ, Weinshenker BG. The spectrum of neuromyelitisoptica.The Lancet Neurology. 2007;6(9):805-15.
8. Bergamaschi R, Tonietti S, Franciotta D, Candeloro E, Tavazzi E, Piccolo G, et al. Oligoclonal bands in Devicâ€™s neuromyelitisoptica and multiple sclerosis: differences in repeated cerebrospinal fluid examinations. Multiple sclerosis. 2004;10(1):2-4.
9. Wingerchuk DM, Weinshenker BG. Neuromyelitis optica. Current treatment options in neurology. 2008;10(1):55-66.

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Author's contribution:

Farida Jan: Study concept and design, data collection, data analysis, manuscript writing, manuscript review
Amber Shabir: data collection, data analysis, manuscript writing, manuscript review
Shahnaz Ibrahim: data analysis, manuscript writing, manuscript review