Neuro myelitis Optica without visual loss in an adolescent boy: A case report

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Abstract

Neuro myelitis Optica (Devic disease) is a chronic, uncommon demyelinating disorder in children, characterized by recurrent episodes of acute or sub-acute optic neuritis and myelitis and occasionally relapsing course. We report a report a case of an adolescent boy, 11year old with multiphasic Neuro myelitis Optica (NMO) without visual loss along with positive CSF oligoclonal bands and positive NMO- IgG antibody. The child had responded to very well to high dose methyl prednisolone and Rituximab therapy.

Key Words*:* Demyelinating, Neuro myelitis Optica, Anti-NMO Antibody

Introduction

Neuro myelitis Optica, NMO (Devic disease) and neuro myelitis Optica spectrum disorders (NMOSD) are demyelinating inflammatory disorders of the CNS often characterized by severe immune-mediated demyelination and axonal damage (1). NMO principally involves optic nerve and spinal cord. Acute attacks of bilateral sequential optic neuritis with visual loss and or longitudinally extensive transverse myelitis (LETM) seen mainly in young females with relapsing course (2). Few cases of NMO with positive anti-NMO IgG are reported in children, that too in boys.

Case Report

We report, a 11 year developmentally normal boy presented with weakness of all four limbs (Rt> Lt) of 14 days & difficulty in passing urine of 5 days duration. Weakness was progressive

leading to right complete quadriplegia over 2weeks with significant pain in both limbs. He had blurring of vision with intact vision. Except tachypnoea and distension of bladder, the child was hemodynamically stable. There was no history of fever, weight loss, rash, joint pain, seizures, headache, altered sensorium, hearing impairment, trauma & history of contact. He had two similar episodes in the last 6 months, approximately 2 months interval lasting for 7 days and improved with short courses of steroids. First episode was right hemiplegia while second episode was right side followed by left side weakness with slurring of speech. In both the episode, there was no visual loss or altered sensorium.

At admission, the child had RR-36 /min, regular, HR-108/m, BP- 100/66 mm Hg, SpO2- 97 % in room air, without automatic instability. There was no pallor, lymphadenopathy, clubbing or oedema. GCS was 15/15 with intact cranial nerves. Hypotonia was present with power of 1/5 in right upper and lower limbs and 2/5 in left upper and lower limbs. DTR were brisk in all the 4 limbs with ankle clonus and bilateral extensor plantar reflex. Sensory examination was normal. Respiratory exam reveals increased rate & efforts with shallow breathing. Other systems were normal.

Initial impression of demyelinating disorder & Transverse Myelitis was made keeping recurrent paralytic episodes with incomplete recovery with bladder and respiratory muscle weakness and normal sensorium in mind. Baseline CBC, CSF, renal and liver functions, lipid profile, HPLC and routine urine were normal. X ray spine did not reveal any lytic or compression lesion. MRI brain (Apr 2019) revealed T2 flair hyper intensity in medulla with normal spine. Repeat MRI brain was done during present admission and showed extensive medullary hyper intensities and long segment demyelination in cervical region (C2-C7) with normal cuts of optic nerve.(Image:1 & 2) Ophthalmic evaluation was normal. CSF for oligoclonal bands (OCB), Anti-Aquaporin-4 IgG (AQP4) antibodies were positive, but Anti-myelin associated oligo -glycoprotein (anti MOG antibodies) was negative. A diagnosis of NMO without optic neuritis was made. Child was treated with 5 days IV methyl prednisolone pulse therapy followed by standard oral prednisolone. Subsequent to the positive report of

anti NMO antibody, he was given 4 doses of Rituximab 1 gm per dose every 2 weeks. The child responded very well presently ambulatory with increased power in all limbs (> 4/5) without residual bladder dysfunction after 6 months.

Fig: 1 MRI Brain: Extensive Medullary demyelination

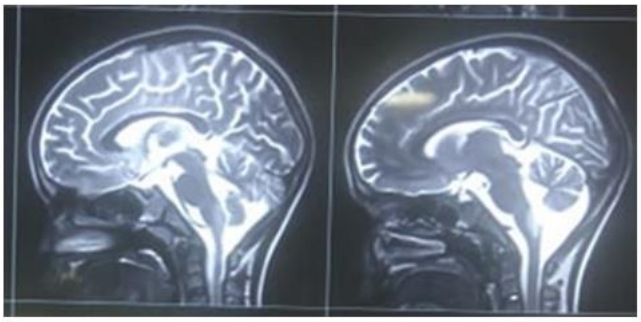
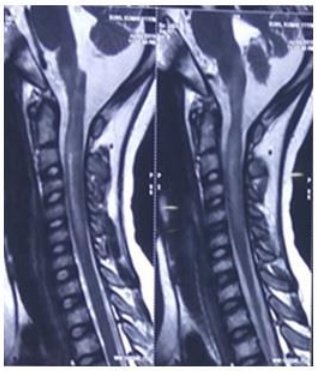


Fig 2: MRI Spine: Showing long segment demyelination in cervical region (C2-C7)



Discussion

NMO is an autoimmune demyelinating disorder and aquaporin-4 (AQP4) water channels are the major target antigens. NMO and NMOSD can be differentiated by the presence or absence

of disease specific AQP4 antibody (1). NMOSD is identified by involvement of optic nerves, spinal cord, area postrema, brainstem and diencephalon (3). Optic neuritis can be unilateral (4). Our patient had biphasic presentation, the first phase being encephalopathy and the second with myelitis and encephalopathy. Brain-stem syndromes due to medullary involvement of area postrema, presenting as either GI or oculomotor dysfunction, deafness, facial palsy, and vertigo. Acute neurogenic respiratory failure and death may occur, and it is more common in anti AQP4 IgG positive patients (5).

Other atypical features like progressive course, rapid nadir (infarction), continual worsening over 4 weeks, partial TM without LETM may be there with typical findings consistent with MS (5.6). AQP4 Antibody is disease-specific with high specificity (91-100%) and varying sensitivity (83-91%). Anti- MOG antibodies may be positive in some cases (7). Other tests, e.g. ANA, DNA, Anti-ds DNA antibodies, lupus anticoagulant, anti-phospholipid antibodies, ANCA, para-neoplastic antibodies (anti-CV2/CRMP5 and anti-Hu) may needs to be done if NMO tested is negative (8). Acute episodes of NMO are usually treated with IV methylprednisolone 1g for 5 days or plasma exchange, intravenous immunoglobulin, cyclophosphamide or oral prednisone. Gradual tapering of prednisolone over a 6–12 months should be done to prevent recurrences (9). Rituximab, 375 mg/m^2 weekly infusions for 4 doses or 2 doses of 1 gm each at 2 weeks interval followed by 6 monthly doses is better alternative to prevent relapses (10).

Conclusions

NMO though less common in children, typical presentation with or without visual loss particularly in absence of altered sensorium in boys may be a presenting symptom. To prevent disability, early recognition and appropriate treatment is crucial for acute attacks, which will also decrease the relapses. In our case, preserved vision was odd to the definition of NMO, but the features were very much classical which leads to reach to a definitive diagnosis.

References

1. Dean M. Wingerchuk, William F. Hogancamp, Peter C. O'Brien, et al. The clinical course of Neuro myelitis optica (Devic's syndrome)**.** Neurology 1999;53;1107

2. Absoud M, Cummins C, Desai N, et al. Childhood optic neuritis clinical features and outcome. Arch Dis Child 2011; 96:860–862

3. Bruna Garbugio Dutra et al Neuro myelitis optica Spectrum Disorders: Spectrum of MRI imaging Findings and Their Differential Diagnosis. Radiographics.rsna.Org 169 - 170

4. Chitnis T, Ness J, Krupp L, et al. Clinical features of Neuro myelitis optica in children: US Network of Pediatric MS Centers report. Neurology. 2016;86(3):245-252.

5. Banwell B, Tenembaum S, Lennn VA, Ursell E, Kennedy J, Bar-Or A, et al. Neuro myelitis optica-Immunoglobulin in childhood inflammatory demyelinating CNS disorders. Neurology 2008; 70: 344-52.

6. Fragoso YD, Ferreira ML, Oliveira EML, et al. Neuro myelitis optica with onset in childhood and adolescence. Ped Neurol, 2014;50:66–68.

7. Lekha P. Neuro myelitis optica antibody (NMO-IgG) status in Indian patients with multiple sclerosis and allied demyelinating disorders. Neurology Asia. 2008;13:175-8.

8. Jan-MendeltTillema and Istvan Pirko. Neuroradiological evaluation of demyelinating disease. Ther Adv Neurol Disord (2013) 6(4) 249–268

9. Margherita Nosadini et all. Rituximab monitoring and redosing in pediatric Neuro myelitis optica spectrum disorder. Neuroimmunol, 2016;3:e18

10. Mahmood, N. A., Silver, K., Onel, K., Ko, M., & Javed, A. (2010). Efficacy and Safety of Rituximab in Pediatric Neuro myelitis optica. Journal of Child Neurology, 26(2), 244–2

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