International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Pediatric Onsetneuromyelitis Optica: A Rare Case Report and a Brief Review of Literature

Dr. Dilruba Shareef Abdurahman

Junior Resident in Radiology, Travancore Medical College, Kollam, Kerala, India

Abstract: ***Neuromyelitisoptica (NMO) is an autoimmune demyelinating disorder for which the aquaporin-4 (AQP4) water channels are the major target antigens.Descriptions of new clinical and radiologic features in seropositive patients have expanded the spectrum of NMO, and the term NMO spectrum disorder (NMOSD) has been adopted. NMOSD is now included in a widening list of differential diagnoses. Aquaporin-4 receptor (AQP4) and myelin oligodendrocyte glycoprotein (MOG) antibody testing along with MRI whole spine with brain is used for diagnosis and evaluation of neuromyelitisoptica (NMO) and NMO spectrum disorder (NMOSD).***

Keywords: NMO, NMOSD, AQP4

1. Introduction

Neuromyelitis Optica (NMO) also called Devic disease is an autoimmune demyelinating disease induced by a specific auto-antibody, the NMO-IgG.NMO preferentially affects the optic nerve and spinal cord.Brain lesions do occur and often are distinct from those seen in MS.Demyelination of the spinal cord looks like transverse myelitis, i.e. often extensive over 4 -7 vertebral segments and the full transverse diameter.NMO IgG is a specific biomarker for NMO. Female: male = 9:1[1] Although the first symptoms of NMOSD commonly occur between 32 and 45 years of age [2], pediatric and elderly patients have been reported.

Pediatric-onset NMOSD is classified as onset of first NMOSD symptoms before 18 years of age. It represents 3%–5% of all NMOSD cases, and the first clinical events are usually optic neuritis (50%–75%) or longitudinally extensive transverse myelitis (LETM) (30%–50%) [2]

We present a confirmed case of pediatric onset NMO from Travancore Medicity, Kollam.

15yr old male presented clinically with proximal thigh pain, unable to void urine and difficulty in walking since 2 days.

On clinical examination, grade 4 power in bilateral lower limbs and bladder palpable.

His lab investigations were unremarkable except for elevated TLC (15100cells/cmm)

He was advised MRI whole spine with brain screening.

His sample was sent for NMO IgG antibody testing to neuroimmunology lab, Amritha Hospital Kochi which later came to be positive.

The patient was referred to Neurology department at SreeChitraTirunal Institute for Medical Sciences for further treatment once the diagnosis was confirmed.

MRI whole spine with Brain Screening

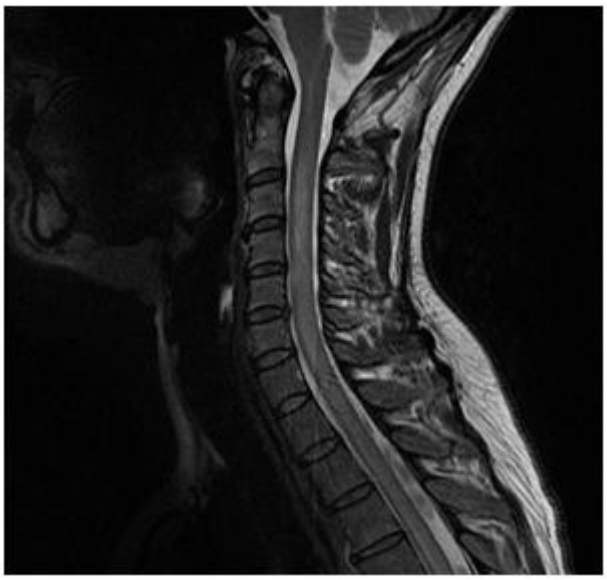

Figure 1: The spinal cord shows a longitudinally extensive lesion, with high T2 signal involving almost the entire cord.

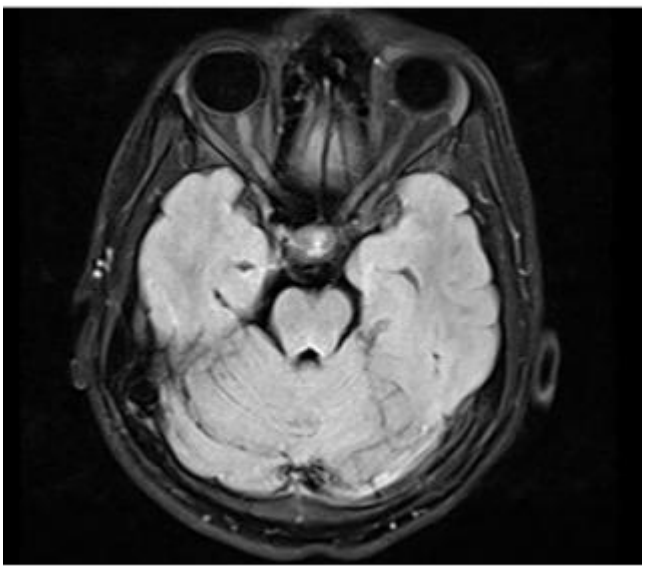

Figure 2: T2 Flair axial image of the brain shows hyperintensity of the right optic nerve.

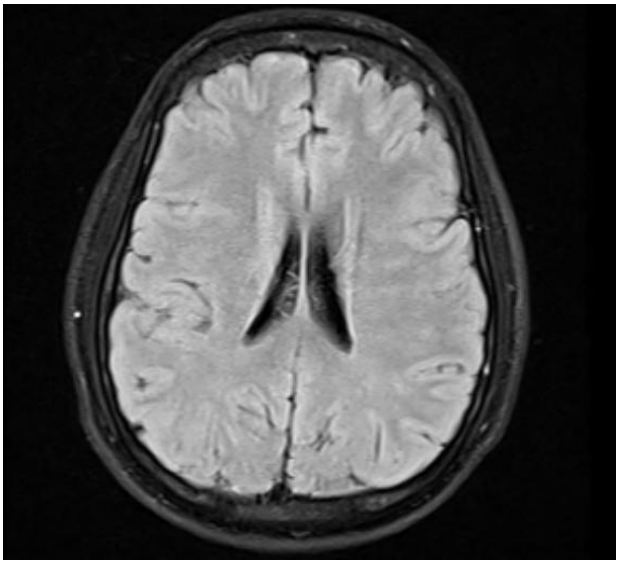

Figure 3: T2 Flair axial image of the brain shows periventricular patchy hyperintensities in bilateral frontal lobes.

2. Discussion

Clinical Features

The core clinical characteristics of NMOSD are distinguished by the locations of the CNS lesions: optic nerves, spinal cord, area postrema, brainstem, diencephalon, and cerebrum. Optic nerve involvement typically manifests as bilateral optic neuritis involving the optic chiasm with severe vision loss. Complete acute spinal cord syndrome is a classic clinical manifestation of spinal cord lesions. Intractable nausea, hiccups, and vomiting are related to area postrema syndrome. Patients with diencephalic involvement may have narcolepsy, anorexia, inappropriate diuresis, hypothermia, and hypersomnia. In brainstem involvement, oculomotor dysfunctions, long tract signs, and ataxia can be seen [3].The clinical manifestations and prognoses are distinct in seropositive and seronegative NMOSD. AQP4-IgG–seropositive patients usually have more severeclinical attacks, worse outcome, more relapses (81%–91%), higher female-to-male ratio, and more frequent coexisting autoimmune disorders compared with AQP4-IgG–seronegative patients [4].

Radiographic Features

MRI is the modality of choice.

Orbits

- Optic nerves appearing hyperintense and swollen on T2 weighted sequences and enhancing on T1 C+
- Bilateral optic nerve involvement and extension of the abnormal signal posteriorly as far as the chiasm is particularly suggestive of NMO [5]
- Atrophy of the optic nerves with associated hyperintensities on T2 weighted sequences may be seen in chronic stages of the disease [6]

Brain

These can be divided into four categories:[7]

1) Lesions which mirror the distribution of aquaporin 4 in the brain, which is particularly found in the periependymal regions abutting the ventricles:
   a) Periventricular (hemispheric) confluent smooth sessile white matter involvement (unlike MS, there are usually no Dawson's fingers)
   b) Periaqueductal grey matter
   c) Hypothalamus/medial thalamus dorsal pons/ medulla
   d) corpus callosum
      - Multiple callosal lesions with heterogeneous signal leading to a marbled pattern[8]
      - The splenium may be diffusely involved and expanded
2) Deep punctate white matter lesions
3) Corticospinal tract involvement by extensive longitudinal lesions.[9]
4) Larger >3 cm diameter hemispheric white matter lesions

Spinal cord

Spinal cord involvement is extensive, with high T2 signal spanning at least three vertebral segments, often many more (known as a longitudinally extensive spinal cord lesion)[7]. Cord swelling is usually present in the acute phase. Bright spotty lesions are a specific feature of NMO. It consists of marked T2 hyperintense (higher than CSF) and T1 hypointense foci in the central grey matter.[10]

Treatment and Prognosis

Treatment of NMO is evolving, with immunosuppression (e.g. anti-CD20 monoclonal antibody rituximab) appearing effective [5]. It is important to distinguish NMO from MS as the treatment not only is different but treating a patient with NMO with MS-specific therapies (e.g. beta-interferon or natalizumab) can actually lead to its exacerbation. Patients with a relapsing course have a poorer prognosis [4]:

Differential Diagnosis

- Multiple sclerosis
- Susac syndrome: involves the central portion of the corpus callosum
- Neuro-Behçet: mesodiencephalic involvement is typical
- Primary angiitis of the CNS (PACNS)
- Acute disseminated encephalomyelitis (ADEM): grey-white matter involvement with a more tumefactive appearance
- Amyotrophic lateral sclerosis (ALS): bilateral corticospinal tract involvement is more symmetrical

3. Conclusion

The discovery of a disease-specific antibody along with its typical MRI features have enabled the establishment of NMO as a distinct disease entity in the spectrum of CNS demyelinating disorders which was once thought of as a variant of MS. NMO is now regarded as a unique disease entity with significantly different prognostic and treatment implications.

Researches are progressing and in the future this will enable earlier disease detection and the development of more

targeted treatment strategies for this debilitating neurological disorder.

References

[1] Nakamura M, Misu T, Fujihara K, Miyazawa I, Nakashima I, Takahashi T, Watanabe S, Itoyama Y. Occurrence of acute large and edematous callosal lesions in neuromyelitisoptica. MultScler. 2009 Jun; 15(6):695-700. doi: 10.1177/1352458509103301. Epub 2009 May 12. PMID: 19435750.

[2] NeuromyelitisOptica Spectrum Disorders: Spectrum of MR Imaging Findings and Their Differential DiagnosisBrunaGarbugio Dutra, Antônio José da Rocha, Renato Hoffmann Nunes, and Antônio Carlos Martins Maia JúniorRadioGraphics 2018 38:1, 169-193

[3] Sorte, Danielle &Poretti, Andrea & Newsome, Scott &Boltshauser, Eugen&Huisman, Thierry &Izbudak, Izlem. (2015). Longitudinally extensive myelopathy in children. Pediatric Radiology. 45. 244-57. 10.1007/s00247-014-3225-4.

[4] Wingerchuk, D., Lennon, V., Pittock, S., Lucchinetti, C., Weinshenker, B. (2006). Revised diagnostic criteria for neuromyelitisoptica. Neurology. https://doi.org/10.1212/01.wnl.0000216139.44259.74

[5] Barnett Y, Sutton IJ, GhadiriM et-al. Conventional and Advanced Imaging in NeuromyelitisOptica. AJNR Am J Neuroradiol. 2013; . doi:10.3174/ajnr.A3592 - Pubmed citation

[6] BrunaGarbugio Dutra, Antônio José da Rocha, Renato Hoffmann Nunes et-al. NeuromyelitisOptica Spectrum Disorders: Spectrum of MR Imaging Findings and Their Differential Diagnosis. (2018) RadioGraphics. 38 (1): 169-193. doi:10.1148/rg.2018170141 - Pubmed

[7] Sarbu N, Shih RY, Jones RV et-al. White Matter Diseases with Radiologic-Pathologic Correlation. Radiographics. 2016;36 (5): 1426-47. doi:10.1148/rg.2016160031 - Pubmedcitation

[8] Nakamura M, Misu T, Fujihara K et-al. Occurrence of acute large and edematous callosal lesions in neuromyelitisoptica. Mult. Scler. 2009;15 (6): 695-700. doi:10.1177/1352458509103301 - Pubmedcitation

[9] Kessler RA, Mealy MA, Levy M. Treatment of NeuromyelitisOptica Spectrum Disorder: Acute, Preventive, and Symptomatic. Current treatment options in neurology. 18 (1): 2. doi:10.1007/s11940-015-0387-9 - Pubmed

[10] Wingerchuk DM, Lennon VA, Lucchinetti CF et-al. The spectrum of neuromyelitisoptica. (2007) The Lancet. Neurology. 6 (9): 805-15. doi:10.1016/S1474-4422(07)70216-8 - Pubmed