Case Report on Neuromyelitis Optica

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Introduction

Neuromyelitis Optica (NMO) is a severe autoimmune inflammatory demyelinating disease of the central nervous system. Magnetic resonance (MR) imaging has become an important tool for diagnosing NMOSD, particularly for recognizing seronegative patients (AQP4-IgG negative) and patients whose serologic AQP4-IgG status remains unknown.

Case Report

Thirty-three years old female complained of sudden onset of slurring of speech and unsteadiness during walking. The patient had a history of fever before two weeks, followed by vomiting, nausea, and hiccups. No comorbidities. No significant family history or menstrual history. The patient was conscious. Vitals were stable. Higher mental functions, cranial nerves examination were normal, Hypotonia and Power-0/5 in both lower limbs. B/L plantar extensor, all other reflexes were present. Spine and cranium - normal. Other systems examinations were normal.

Investigations

Complete hemogram, renal function tests, liver function tests, electrolytes were within normal limits. CRP- 12mg/dl. Urine examination normal, Viral markers- negative, HSV-1 and two negatives, SARS Covid19 RT PCR negative, Aquaporin 4 antibody and Anti-MOG antibody negative. CSF examination- no significant findings, also negative for oligoclonal bands, Japanese encephalitis, HSV. ANA profile negative. ECG, CT chest/abdomen- normal, ECHO-normal. MRI spine- Transverse myelitis involving >3 vertebral length. MRI Brain with Contrast- Demyelination involving Pons, midbrain, bilateral middle cerebellar peduncles, and B/L frontoparietal ischemic demyelination changes. Blood, CSF, and urine culture were negative. Serum ACE levels normal, HLA B51/B5 - negative. Fundus examination-normal.

Based on 2015 IPND Criteria for NMOSD, this patient is diagnosed as Neuromyelitis Optica spectrum disorder with AQP4-IgG–negative or unknown status.

Management

The patient responded to Inj.Methyl prednisolone for seven days and later changed over to Tab Prednisolone. Advised follow-up for fundus examination and tapering of prednisolone dose.

Conclusion

This case highlights the necessity of MRI as an important role in diagnosing Neuromyelitis optica spectrum disorder with Aquaporin negative status.

References

1. Wingerchuk DM, Banwell B, Bennett JL, Cabre P, Carroll W, Chitnis T, de Seze J, Fujihara K, Greenberg B, Jacob A, Jarius S, Lana-Peixoto M, Levy M, Simon JH, Tenembaum S, Traboulsee AL, Waters P, Wellik KE, Weinshenker BG; International Panel for NMO Diagnosis. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. Neurology. 2015 Jul 14;85(2):177-89. doi: 10.1212/WNL.0000000000001729. Epub 2015 Jun 19. PMID: 26092914; PMCID: PMC4515040.

2. Matiello M, Schaefer-Klein J, Sun D, Weinshenker BG. Aquaporin 4 expression and tissue susceptibility to neuromyelitis optica. JAMA Neurol. 2013 Sep 1;70(9):1118-25. doi: 10.1001/jamaneurol.2013.3124. PMID: 23857303.

3. Waters P, Jarius S, Littleton E, Leite MI, Jacob S, Gray B, Geraldes R, Vale T, Jacob A, Palace J, Maxwell S, Beeson D, Vincent A. Aquaporin-4 antibodies in neuromyelitis optica and longitudinally extensive transverse myelitis. Arch Neurol. 2008 Jul;65(7):913-9. doi: 10.1001/archneur.65.7.913. PMID: 18625857.

4. Marios C Papadopoulos, Verkman. Aquaporin 4 and Neuromyelitis Optica. Lancet neurology.2012 June;11(6): 535-544. doi:10.1016/S1474-4422(12)70133-3

5. Jarius S, Wildemann B. The history of neuromyelitis optica. J Neuroinflammation. 2013 Jan 15;10:8. doi: 10.1186/1742-2094-10-8. PMID: 23320783; PMCID: PMC3599417.

6. Jarius S, Wildemann B. AQP4 antibodies in neuromyelitis optica: diagnostic and pathogenetic relevance. Nat Rev Neurol. 2010 Jul;6(7):383-92. doi: 10.1038/nrneurol.2010.72. PMID: 20639914.

7. Jarius S, Paul F, Franciotta D, Waters P, Zipp F, Hohlfeld R, Vincent A, Wildemann B: Mechanisms of disease: aquaporin-4 antibodies in neuromyelitis optica. Nat Clin Pract Neurol 2008, 4:202-214

8. Wingerchuk DM: Neuromyelitis optica: new findings on pathogenesis. Int Rev Neurobiol 2007, 79:655-688

9. Wingerchuk DM, Lennon VA, Luchinetti CF, Pittock SJ, Weinshenker BG: The spectrum of neuromyelitis optica. Lancet Neurol 2007,6:805-815

10. Jarius S, Paul F, Franciotta D, Ruprecht K, Ringelstein M, Bergamaschi R, Rommer P, Kleiter I, Stich O, Reuss R, Rauer S, Zettl UK, Wandinger KP, Melms A, Aktas O, Kristiferitsch W, Wildemann B: Cerebrospinal fluid findings in aquaporin 4 antibody positive neuromyelitis optica: results from 211 lumbar punctures. J Neurol Sci 2011, 306:82-90