Abstract Title

Is it Multiple Sclerosis or Neuromyelitis Optica Spectrum Disorder (NMSOD)?

Limit the body of the abstract to be 500 words or less. The abstract form does not accept graphs, charts, tables, etc. Abstract should have no names or institutions.

Introduction:

Neuromyelitis Optica Spectrum Disorder (NMSOD) is an uncommon antibody-mediated disease of the CNS. Untreated, approximately 50% of NMSOD patients will become wheelchair users and blind, and a third will have died within 5 years of their first attack. Recent advances in targeted immunotherapy have markedly changed the trajectory of this once debilitating illness.

Case Description:

A 45-year-old female with no known past medical history presented with progressive right-sided weakness. Her symptoms started with upper back and right arm pain. Subsequently, she experienced progressive right arm and leg weakness over the course of the next 3 days. Her exam exhibited distal right upper and lower extremity weakness (arm 3/5 and leg 1/5), hyperreflexia, and sustained clonus. Contrast cervical spine MRI revealed a longitudinally enhancing lesion extending from C3 to T1 segments. MRI of the brain and the remaining cord was normal. A lumbar puncture demonstrated 0 white blood cells, normal protein and IgG index, and five oligoclonal bands. She was initiated on intravenous solumedrol (1g/day for 5 days). This was followed by 5 plasmapheresis exchanges. Serological evaluation revealed a positive Aquaporin 4 Antibody (titer > 1:160). Following studies on anti-MOG antibodies, paraneoplastic antibodies, autoimmune and infectious markers were negative. Over the ensuing month, her neurological status improved in her extremities (arm 4+ and leg 2/5).

Discussion:

NMSOD is a rare autoimmune CNS demyelinating disorder. It involves relapsing inflammatory lesions typically affecting the optic nerves, spinal cord (longitudinally extensive transverse myelitis – LETM), and circumventricular organs, such as the area postrema, if involved produces unremitting vomiting. All these factors lead to significant morbidity and mortality due to blindness, quadriplegia, and respiratory failure. If untreated, outcomes have historically been much worse than for patients suffering from Multiple Sclerosis (MS) =. Oligoclonal bands in CSF are absent in 75-80% of NMSOD patients but may be present in some cases. MS and NMSOD have overlapping clinical presentations, but MS is more commonly associated with oligoclonal bands and supratentorial cerebral and short-segment spinal cord lesions. However, these differences are not strict as we now know that NMSOD can also cause both supratentorial and infratentorial lesions. NMSOD's spinal cord lesions are contiguous, affecting multiple spinal cord levels. Furthermore, it is associated with positive AQP4 in 75-80% of cases, which are known to be pathogenic and integral to disease pathogenesis. AQP4-IgG binding to aquaporin-4 channels on astrocyte foot processes triggers astrocytopathy through the activation of complement-mediated destruction. Thus, NMSOD, is predominantly a B-cell and complement-mediated disease, whereas the pathophysiology of MS involves both B and T-cells, with downstream activation of macrophages and microglia. Acute treatment of NMSOD involves high-dose IV steroids and plasma exchange, though there are no large clinical trials demonstrating long-term efficacy of these treatments. Long-term immunotherapy appears to be highly effective in reducing relapses and includes anti-CD5 Abs targeting the complement cascade and anti-CD19 Abs depleting the B-cell population producing AQP4, positively altering the course of this previously destructive and deadly condition.

Is it Multiple Sclerosis or Neuromyelitis Optica Spectrum Disorder (NMSOD)?

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INTRODUCTION

- NMSOD is an uncommon antibody-mediated disease of the CNS.
- Untreated, approximately 50% of NMOSD patients will become wheelchair-bound and blind, and a third will die with in 5 years
- Recent advances in targeted immunotherapy have markedly changed the trajectory of this once debilitating illnes

CASE DESCRIPTION

- A 45-year-old female with no significant PMH presented with progressive right-sided weakness.
- Symptoms started with upper back and right arm pain followed by progress weakness .
- PE exhibited R arm 3/5 and leg 1/5 weakness and bilateral clonus R > L
- MRI Head negative for infarct.

CLINICAL COURSE

MRI Cervical Spine	Longitudinal enhancing lesion extending from C3 to T1 as shown in figure A concerning for acute transverse myelitis
LP	0 WBC, normal protein and IgG index, and 5 oligoclonal bands.
Serology	Positive Aquaporin 4 Antibody titer > 1:160
Treatment	Started on high dose IV STERIOD for 5 days followed by plasmapheresis with physical therapy

POST DISCHARGE FOLLOW UP

- Significant improvement in PE. Current PE has little to no weakness in RUE & 3/5 strength in RLE.
- Received Rituximab and anti-CD 19 ab. FU MRI after 3 month as shown in figure B

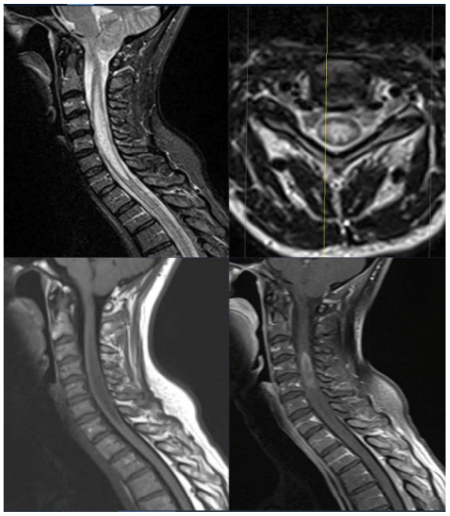

FIGURE A

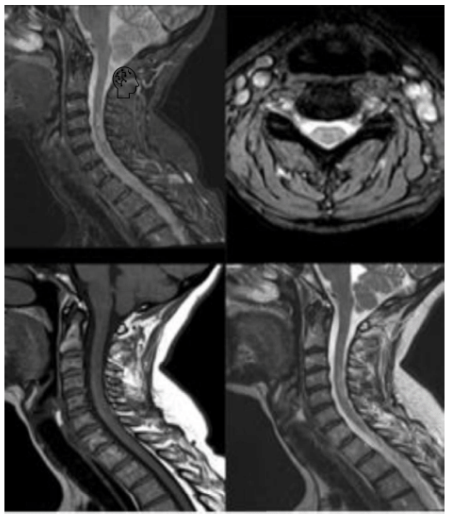

FIGURE B

TAKE HOME MESSAGE

1. **It's not always the stroke or MS.**
2. **Transverse myelitis has a wide etiology.**
3. **MS and NMSOD are similar in clinical presentation.**

DISCUSSION

- NMOSD is a relapsing inflammatory disorder affecting spinal cord, optic nerve and circumventricular organs.
- MS is more commonly associated with supratentorial cerebral and short-segment spinal cord lesions and oligoclonal bands.
- Oligoclonal bands in the CSF are absent in 75-80% of NMOSD patients but may be present in some cases.
- NMOSD is associated with positive AQP4 Abs in 75-80% of cases.
- NMSOD is a B-Cell and complement mediated disease whereas MS involves B and T cells .
- Acute treatment of NMOSD involves high-dose IV steroids and plasma exchange.
- Long-term immunotherapy appears to be highly effective at reducing relapses.
- These treatments include anti-CD5 Abs targeting the complement cascade and anti-CD19 Abs depleting the B-cell population producing AQP4, positively altering the course of this previously destructive and deadly condition.
- If untreated, NMOSD outcomes are typically worse than MS.

REFERENCES

1. Goldschmidt C, Galetta SL, Lisak RP, Balcer LJ, Hellman A, Racke MK, Lovett-Racke AE, Cruz R, Parsons MS, Sattarnezhad N, Steinman L, Zamvil SS, Frohman EM, Frohman TC. Multiple Sclerosis Followed by Neuromyelitis Optica Spectrum Disorder: From the National Multiple Sclerosis Society Case Conference Proceedings. Neurol Neuroimmunol Neuroinflamm. 2022 Oct 21;10(1):e200037. doi: 10.1212/NXI.0000000000200037. PMID: 36270950; PMCID: PMC9673749.
2. Huda S, Whittam D, Bhojak M, Chamberlain J, Noonan C, Jacob A. Neuromyelitis optica spectrum disorders. Clin Med (Lond). 2019 Mar;19(2):169-176. doi: 10.7861/clinmedicine.19-2-169. PMID: 30872305; PMCID: PMC6454358.
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