ORIGINAL RESEARCH PAPER

General Medicine

CASE REPORT-NEUROMYELITIS OPTICA/ DEVIC'S DISEASE: A RARE DISEASE

KEY WORDS: Neuromyelitis Optica; neuromyelitis optica –IgG ; optic neuritis; transverse myelitis

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ABSTRACT

Neuromyelitis Optica spectrum disorder or Devic's disease are autoimmune diseases that manifest clinically with 6 core symptoms, which include acute optic neuritis, transverse longitudinal segment myelitis, and acute brainstem syndrome.{1}Neuromyelitis optica is characterized by attacks of optic neuritis and longitudinally extensive transverse myelitis. It is often confused with multiple sclerosis. Early discrimination between NMO and MS is important because the two diseases have different natural histories and treatment regimens. Despite the absence of a definitive therapeutic strategy for NMO syndrome, methylprednisolone pulse therapy is recommended in acute phase. Treatment strategies in relapse phases are aimed at preventing relapses, and increasing evidences show a better clinical response of immunosuppressive therapy than immunomodulating therapy.

We report a case of NMSOD in a patient of age 55 , with transverse myelopathy as the initial manifestation. The patient presented with acute gait disturbance and hypoesthesia in right lower limb since 10 days . After thorough physical examination and various lab investigations and imaging, the patient received high dose methylprednisolone. The patient's serum was positive for anti-AQP4 antibodies .During the course of treatment , patient again presented with complaint of blurred vision in the ER after 1 year. Diagnosis of NMSOD was established. After combining methylprednisolone and immunosuppressant therapy , the patient's medical condition was stable and relapses were prevented.

CASE REPORT

A 55 year old female presented to ER with c/o diplopia, blurred vision in the left eye since 2 days and gait disturbance since 5 days. Beside these, she had rectal incontinence, urinary retention and numbness in lower extremities. Neurological examination revealed right lower limb power 2/5 and 3/5 in the left extremity. Decreased sensation was belowT7. Babinski's sign was positive bilaterally with clonus at the left ankle. Vitals of the patient were as follows -BP-150/100MMHG; SPO2-99% on room air. No h/o trauma reported. Past history is evident of spastic paresis in b/l lower limbs (with 2 episodes of relapses 2 months apart) a year ago for which patient was on steroid therapy and symptoms subsided after pulse therapy.

T2W images of the thoracic spine showed cord lesions with swelling as well as with contiguous hyperintensity. The CSF was negative for oligoclonal bands.The serum sample was positive for anti-AQP4 antibody. No optic nerve lesions were detected in the MRI. The diagnosis of Neuromyelitis Optica was established. She was started with steroid pulse therapy (methylprednisolone 250mg i/v 6 hrly for 5 days) and plasma exchange therapy. Her symptoms began to subside. Neuro Ophthalmology consult was done. On examination, B/L pale disc and left RAPD (Relative afferent papillary defect) and MRI Orbit revealed subtle reduced girth of left optic nerve close to orbital apex. MRI spine revealed LETM involving thoracic spine and associated cord swelling with no cord compression suggestive of non compressive myelopathy-Transverse Myelitis. Hence, from all of the above, NMO diagnosis was established. Neurological examination before discharge showed grade 4 strength in her lower limbs and she could walk with assistance.

DISCUSSION

Through this study, we highlight the complexity of diagnosing neuromyelitis optica and the treatment. This patient presented with typical symptoms of NMO but was diagnosed as a case of MS before she presented to us. Differential diagnosis between Longitudinal Extensive Transverse Myelitis [LTEM] that occurs in MS and NMO and SLE is important since different therapeutic regimens are required interferon treatment may cause a flare up of the diseases.[2] The course of NMO is often more acute. NMO IgG antibody marker is highly specific for this.

The current diagnostic criteria for NMO requires the presence of optic neuritis and acute transverse myelitis along with supportive criteria of aquaporin 4 seropositivity, normal brain MRI or not meeting criteria for MS and longitudinally extensive cord lesions extending over 3 or more segments. However, the prognosis for NMO varies. Hence, an early intervention with the appropriate treatment modality in patients with NMO decides on favourable outcome from an acute episode.

DECLARATION OF CONFLICTING INTERESTS

The authors declared no potential conflicts of interest with respect to research, authorship and/or publication of this article.

REFERENCES

[1] Wingerchuk DM, Banwell B, BENNETT JL, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. Neurology 2015;7:1-3

[2] Ko FJ, Chiang CH, Jong YJ, Chang CH, Neuromyelitis optica (Devic's disease) report of one case.1989;30(6):428-431.