An NMO Presentation Resembling Extensive Multiple Sclerosis

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INTRODUCTION

Multiple sclerosis (MS) and neuromyelitis optica (NMO) are both central nervous system demyelinating diseases caused by autoimmunity. Classically NMO has simultaneous or sequential involvement of the optic nerves and spinal cord in a longitudinally extensive manner (greater than 3 segments) with less frequent involvement of the brain. In MS involvement of the spinal cord is limited to less than 3 vertebral bodies with significant brain involvement. Though Aquaporin 4 antibodies are sensitive and specific for diagnosing NMO these antibodies may be present in a small percentage of patients with multiple sclerosis. Given this overlap cortical involvement has been looked to as a distinguishing feature that is present in multiple sclerosis but not typically in NMO.

OBJECTIVES

To demonstrate a spectrum of demyelinating diseases that could have features of specific entities like MS or NMO with overlapping clinical, serological and radiological findings.

METHODS

This is a case report and review of the literature.

CASE RPEORT

This is a 70 year old Chinese female who has history of recurrent bilateral lower limb weakness attacks for almost 20 years starting when she was in China leading eventually to severe paraparesis with urinary and fecal retention. On her initial assessment in 2005, her MRI of the spinal cord showed STIR signal change involving a long segment of the thoracolumbar cord. Her MRI of the brain showed extensive white matter lesions involving cortical and subcortical regions with almost all the white matter tracts involved

She also had abnormal visual evoked potentials bilaterally, She had a positive NMO Abs with a titer of 1:7280, Her serologic work up was otherwise negative except she had +ve SSA and JO-1 which were not felt to be indicative of a rheumatologic disorder. The patient failed to improve radiologically and clinically on two types of Interferon, IV steroids, and was finally started on Imuran 75mg BID along with prednisone 15 mg daily.

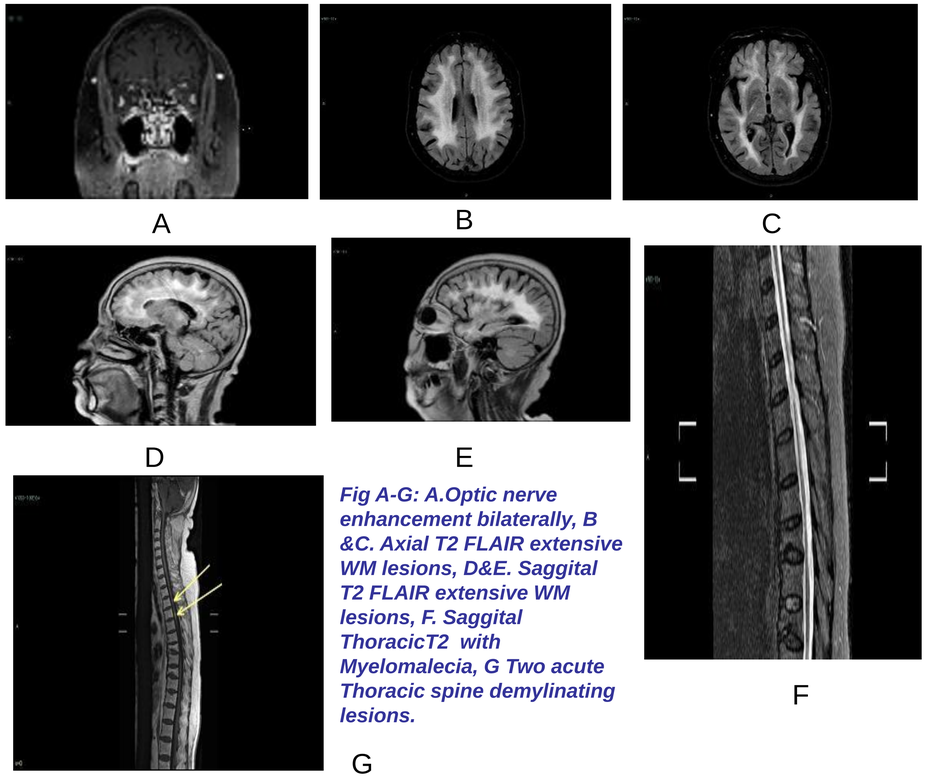

Fig A-G: A.Optic nerve enhancement bilaterally, B &C. Axial T2 FLAIR extensive WM lesions, D&E. Saggital T2 FLAIR extensive WM lesions, F. Saggital ThoracicT2 with Myelomalecia, G Two acute Thoracic spine demylinating lesions.

She was found to be positive for JC virus antibodies which raised concerns for starting Rituximab due the potential risk of PML. Her most recent exacerbation consisted of right optic neuritis which was confirmed by MRI of the orbit. She received a pulse treatment for steroids and was discharged in a stable condition with improvement in her vision in the right eye.

DISCUSSION

MRI characterization of MS or NMO has been postulated previously. More than once (1-6), it was stated as a first line method of distinguishing between the two conditions which is even more helpful than serological markers(2). Different criteria have been applied including the shape, location and size of the lesions (2,3). Most recently there was emphasis on whether any lesions are adjacent to the lateral ventricle or the inferior lateral wall. In another review it was noticed that NMO lacks cortical lesions which are present in relapsing remitting MS (1). Studies that looked at combined radiological techniques like spinal cord and brain MRI had a better way of establishing the character of these lesions and attributing them to one disorder (NMO) rather than the other (MS) (4,5,6). Having said that there were still cases where these criteria were not fulfilled and the presentations overlapped which leaves a category of overlapping phenotypes that share non-specific findings on imaging and lab tests.

CONCLUSION

Our case illustrates white matter involvement in demyelinating disease can be extensive with characteristics from more than one disease (NMO vs. MS). In the future more specific biological or radiological markers could be useful in distinguishing these presentations and guiding therapy.

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