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Coexistence of Neuromyelitis Optica Spectrum Disorder, Sjogren's Syndrome And Tuberculosis In A Postpartum Female-A Case Report

Case Report

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Abstract

Patients with NMOSD coexisting with both sjogren's syndrome and tuberculosis are very rare. There are no specific guidelines for the treatment of these patients especially during pregnancy. We report a case of AQP4 seropositive Nmosd associated with Sjogren syndrome and tuberculosis in a postpartum female and is treated with rituximab. In 2017 a 25 year female presented with quadriparesis and is diagnosed as AQP4 positive NMOSD. She is treated with I.V.methyl prednisolone. 1 year later she became pregnant and is asymptomatic during pregnancy and at 2 months postpartum she developed paraparesis. She is treated with i.v.methyl prednisolone and plasmapheresis and started on Azathioprine. At the same time she is diagnosed with Sjogren's syndrome. In 2021 Azathioprine is changed to MMF in view of pancytopenia. In 2022 she developed pulmonary TB and used ATT for 6 months. She discontinued MMF at that time. After completing ATT she conceived for the second time, remained asymptomatic throughout pregnancy and at 3 months postpartum she again developed quadriparesis. She is treated with rituximab with significant improvement of symptoms. For treating relapsing NMOSD coexistent with Sjogren syndrome and tuberculosis in the postpartum period, rituximab may be a good choice.

Keywords: Neuromyelitis Optica Spectrum Disorder; Sjogren Syndrome; Tuberculosis; Rituximab

Abbreviations

AQP4-Aquaporin 4; ATT-Anti TB treatment; MMF-Mycophenolate Mofetil; NMOSD-Neuromyelitis Optica spectrum disorder; TB-Tuberculosis.

Introduction

Neuromyelitis optica (NMO) also known as Devic syndrome is a rare relapsing auto-immune disease of the central nervous system (CNS) which is sometimes found in association with other autoimmune disorders, including SS. Based on the revised criteria by Wingerchuck et al, a diagnosis of NMO can be made in the presence of both absolute and two of three supportive criteria [1]. The absolute criteria include optic neuritis and myelitis; while the supportive criteria are magnetic resonance imaging (MRI) evidence of a contiguous spinal cord lesion (3 or more segments in length), MRI brain non-diagnostic for multiple sclerosis and serological evidence of NMO-IgG or aquaporin 4 (AQP4) antibodies. NMO spectrum disorders (NMOSD) includes a wide range of neurologic conditions that express NMO antibody and share features with NMO but do not meet the strict diagnostic criteria specified previously [2].

Case Presentation

A 27-year-old female presented with recurrent neurological deficits i.e.

1ST EPISODE IN 2017 – weakness of all four limbs with bladder symptoms and sensory loss below neck. MRI showed long segment myelitis in cervical region. Serum aquaporin 4 antibody is positive. patient is treated with I.V methylprednisolone pulse therapy for 3 days. patient had complete improvement in 2 months. patient was on oral steroids for 1 year and stopped once she became pregnant.

2ND EPISODE IN OCTOBER 2019 – paraparesis, reduced sensation below thorax and urge incontinence. The patient was 2 months postpartum during the onset of symptoms. MRI showed T2 hyperintense lesions in dorsal spinal cord. She received I.V methylprednisolone for 3 days and large volume plasmapheresis -5 sessions and then started on oral azathioprine. At the same time patient is diagnosed with Sjogren's syndrome – lip biopsy proven. Concluded as NMO-Sjogren Overlap.

3rd EPISODE IN APRIL 2020 - 1month history of bilateral painful nonprogressive blurring of vision. VEP showed P100 prolongation in both eyes. She is treated with i.v.methyl prednisolone and large volume plasmapheresis and continued on azathioprine.

IN 2021 –Routine investigations revealed pancytopenia. Azathioprine was stopped in view of pancytopenia. Planned for serum TPMT testing and to start azathioprine in low dose but patient refused for the test. Mycophenolate mofetil is started for long term immunomodulation.

IN JANUARY 2022 – Patient developed pulmonary tuberculosis and used ATT for 6months. She has no contact history of tuberculosis. She didn't take MMF at this time. 1 month after completing ATT she conceived for the 2nd time.

4TH EPISODE IN 2023 – stiffness of all 4 limbs with no weakness or sensory involvement or bladder involvement. She is 2 months postpartum during this time.

On Examination patient is conscious and oriented and vitals are stable. HMF normal. Visual acuity 6/18 with fundus pale disc. Other cranial nerves are intact. Motor system increased tone in all limbs with power 5/5. DTR 2+. Sensory system-touch pain and temperature -intact in all dermatomes. Vibration and proprioception intact bilaterally. MRI showed altered signal intensity with thickening of both optic nerves and altered signal intensity in the cord extending from cervico-medullary junction to C7. Patient is treated with I.V methylprednisolone for 5days and given 1gm of rituximab. Patient improved after rituximab injection and is relapse free till now.

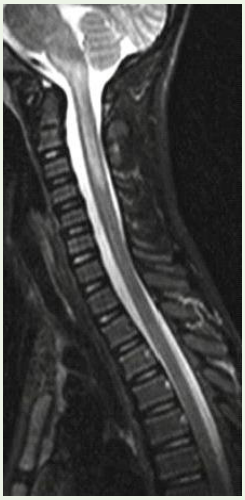

Figure 1: MRI T2 weighted sequence of the cervical region showing contrast enhancement from cervico-medullary junction to C4.

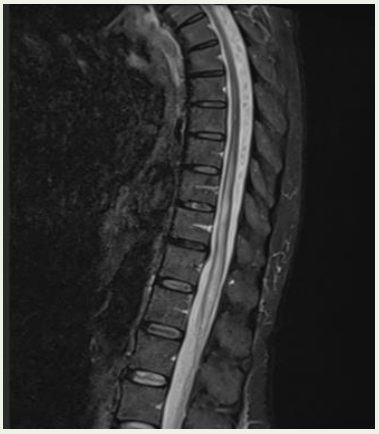

Figure 2: MRI spine showing T2 hyperintensity in the distal spinal cord for >3 vertebral segments

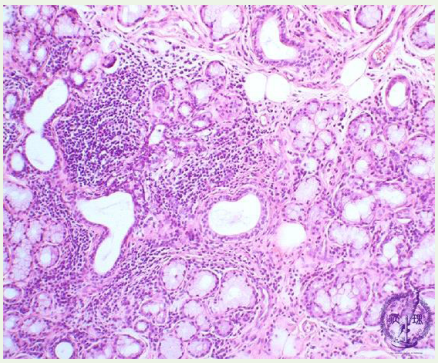

Figure 3: Lip biopsy showing focally infiltrated lymphocytes around interlobular ducts of minor salivary glands.

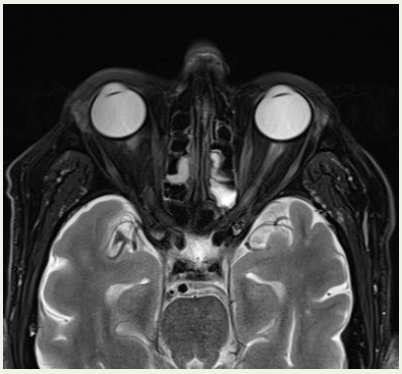

Figure 4: MRI brain with orbital cuts showing thickening of bilateral optic nerves (left > right).

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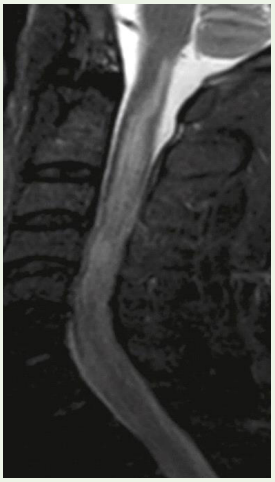

Figure 5: MRI Cervical spine Sagittal STIR-Longitudinally extensive T2 hyperintense lesion from cervicomedullary junction to c7.

Discussion

NMOSD is a relapsing neurological illness that has been sometimes reported in association with primary SS and SLE. NMO is characterized by recurrent episodes of myelitis and optic neuritis and most patients have a unique antibody against NMO IgG/ AQP4. Patients who have NMO IgG/AQP4 antibodies have frequent relapses compared to those who do not have the antibodies [3].

Relapsing NMO carries a poor prognosis, thereby validating the need for early testing and treatment. It has been reported to have a 5 year survival rate of approximately 80% [4]. The cumulative effect of recurrent attacks renders a debilitating outcome, including permanent disabilities requiring assistive devices for ambulation and monocular blindness [4]

NMO and NMOSD are rare diseases with no consensus on treatment guidelines. The treatment options are empiric and based on disease severity ranging from plasmapheresis, corticosteroids, and rituximab to oral agents such as azathioprine, MMF and methotrexate. Early initiation of systemic steroids alone or in combination with other immunosuppressive therapies is most often used. In our patient with relapsing disease course, combination of steroids and azathioprine/MMF is used.

One established treatment of acute NMOSD relapses during pregnancy is intravenous administration of 1 g methylprednisolone/ day over five consecutive days. For non-pregnant NMOSD patients, currently favoured long-term treatment includes azathioprine (AZA) and rituximab (RTX). Case reports describe bone marrow suppression with anaemia and severe lympho- and pancytopenia in infants after maternal AZA exposure. Evidence suggests that RTX treatment carries a higher risk of premature births. As RTX treatment during pregnancy also transiently depletes B cells in newborns measuring B lymphocyte levels of fetuses is advised if exposed to RTX after the first 20 weeks of pregnancy. Overall, careful risk-benefit analysis of stopping or continuing immunosuppressive treatment is necessary in patients planning families. Treatment with mycophenolate mofetil should stop 6 weeks; methotrexate, 12 weeks; and mitoxantrone, 6 months before attempting to conceive [5].

Conclusion

we report this case due to its rarity of occurrence. There are no specific guidelines for the treatment of NMOSD coexistent with Sjogren syndrome and tuberculosis especially during pregnancy or post-partum period. Rituximab might be used in such patients with less side effects during pregnancy or lactation not only to improve the disease symptoms but also to reduce the risk of relapses. Further studies are needed to establish guidelines for new treatment options.

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