Original Research Paper

Volume-9 | Issue-10 | October - 2019 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Medicine

# A RARE ASSOCIATION OF NEUROMYELITIS OPTICA SPECTRUM DISORDER WITH SJOGREN'S SYNDROME: A CASE FROM WEST INDIA

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**ABSTRACT** Sjogren's syndrome(SS) is a chronic systemic disease characterized by inflammation and dysfunction of exocrine glands.A wide spectrum of neurologic manifestations ranging from asymptomatic brain lesions on MRI(3) to symptomatic brain lesions, meningitis, myelopathy, sensorimotor polyneuropathy and mononeuritis multiplex may accompany primary SS.NMO is relapsing neurological illness that has been sometimes reported in association with primary SS and SLE.Early diagnosis and treatment are critical in decreasing the possibility of irreversible severe neurological dysfunction.

**KEYWORDS :** Sjogren's Syndrome, Neuromyelitisoptica Spectrum Disorder (nmosd)

## INTRODUCTION:

Sjogren's syndrome(SS) is a chronic systemic disease characterized by inflammation and dysfunction of exocrine glands. Up to 65% of primary SS patients can experience extra glandular features including pulmonary, gastrointestinal, hematologic and neurologic disorders[1]. Symptomatic central nervous system involvement can be in form of myelopathy, optic neuritis, seizures, and cognitive dysfunction.NMO spectrum disorders (NMOSD) includes a wide range of neurologic conditions that express NMO antibody and shares features with NMO but do not meet the strict diagnostic criteria[2]. We report a rare case of SS complicated by neuromyelitisoptica spectrum disorder (NMOSD).

## CASE REPORT:

A 29-year-old female presented to us with complaints of fever with chills and loose stools. She was vitally stable on presentation. In the past, she had complain of diminished in vision in left eye 10 years back which got resolved after taking treatment details of which is not known. 2 years down the line, she developed bilateral parotitis which was painful and associated with dryness of eye and mouth. For that she consulted rheumatologist and she was diagnosed to have Sjogren's syndrome on basis on clinical and laboratory findings (Table 2) and was put on immunosuppressants. After 3 years, she again started developing diminished vision in left eye. She was diagnosed to have optic neuritis in left eye on MRI and treated with corticosteroids. In current visit, her fever workup was found to be normal. Two days after admission, she started developing weakness in all four limbs with headache. The symptoms got worsened rapidly in several hours to the point that she had difficulty with ambulation. The patient's neurological examination was remarkable for hypertonia, decreased power (Lower limb 0/5; Upper limb3/5), hyperreflexia along with sensory loss below umbilicus.

Gadolinium contrast enhanced magnetic resonance imaging (MRI) of the brain showed multiple confluent altered signal intensity lesions in subcortical and periventricular deep white matter of bilateral frontoparietal and occipital lobe, body of corpus callosum, left cerebral peduncle and left anterior pons (Fig 1&2) and spine screening showed few enhancing linear intramedullary lesions at C3-C4 level, D7 level and D10-D11 level(Fig 3). Her CSF routine examination was normal with raised IgG index and negative OCB(Table-1). Her serum NMO was sent and was found positive. So, diagnosis of NMOSD was made. Injection methylprednisolone 1 gm was given for 5 days followed by oral prednisone. She was discharged with improvement in limb weakness (Lower limb 2/5; Upper limb5/5) on oral prednisone with instructions to taper to maintenance dose of 20 mg over 2 weeks.

***Table 1: Summary of NMOSD tests***

| TEST | RESULT | NORMAL RANGE |
|---|---|---|
| IgG CSF | 29.90 mg/dl | 0-3.4 mg/dl |
| IgG Total Serum | 1140 mg/dl | 700-1600 mg/dl |
| CSF IgG Index | 0.940 | 0.28-0.66 |
| IgG Albumin Ratio | 0.87 | 0.09-0.25 |
| NMO1 (Aquaporin 4) Ab Serum | Positive | |
| MOG2 Ab Serum | Negative | |
| Oligoclonal Bands (CSF) | No Band | |
| Oligoclonal Bands (SERUM) | No Band | |

1-Neuromyelitis Optics; 2-Myelin Oligodendrocyte Glycoprotein

***Table 2: Summary of autoimmune workup***

| TEST | RESULT | NORMAL RANGE |
|---|---|---|
| ANA BY IF | 1:100 NUCLEAR SPECKLED +4 | |
| SS-A Ro | 76 AU/ML | <1 AU/ML: Negative |
| SS-B La | 80 AU/ML | <1 AU/ML: Negative |
| Scl-70 | <1 AU/ML | <1 AU/ML: Negative |
| Ds-DNA Ab | <1 AU/ML | <1 AU/ML: Negative |
| Centromere | <1 AU/ML | <1 AU/ML: Negative |
| U1 Sn RNP | <1 AU/ML | <1 AU/ML: Negative |
| Ro-52 | <1 AU/ML | <1 AU/ML: Negative |
| JO-1 | <1 AU/ML | <1 AU/ML: Negative |

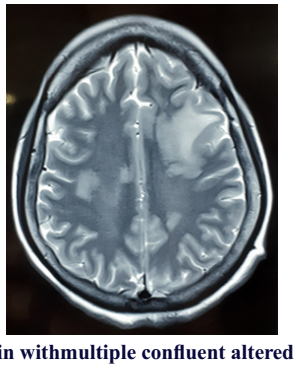

**Fig 1: MRI Brain withmultiple confluent altered signal intensity lesions**

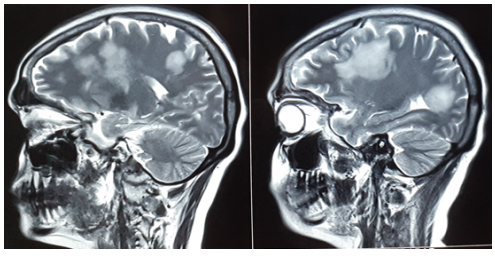

**Fig 2: MRI Brain sagittal view**

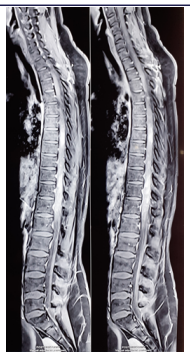

**Fig 3: MRI T2 weighted sequence few enhancing linear intramedullary lesions at C3-C4 level, D7 level and D10-D11 level**

## DISCUSSION:

Sjogren's syndrome (SS) is an autoimmune disease in which there is mononuclear infiltration of exocrine gland and salivary and lacrimal glands resulting in xerophthalmia and xerostomia, also known as sicca syndrome. Blood tests are usually positive for SSA/Ro or SSB/La antibodies. Rose Bengal test, Schirmer test, and saliva gland/lip biopsy can be done to support diagnosis.

Neurological involvement could be observed in approximately 20–25% of cases of SS. Neurological manifestation of SS involves central and peripheral nervous system. These range from asymptomatic brain lesions on MRI[3] to symptomatic brain lesions, meningitis, myelopathy, sensorimotor polyneuropathy and mononeuritis multiplex. Neurological involvement can be focal or multifocal central lesions to dementia and conditions that mimic MS such as neuromyelitis Optica (NMO) or NMSD. NMO is relapsing neurological illness that has been sometimes reported in association with primary SS and SLE. NMO is characterized by recurrent episodes of optic neuritis and myelitis and most patients have antibody against NMO IgG/AQP4. NMO is characterized by two absolute criteria; (1) optic neuritis and (2) acute myelitis plus 2 of the 3 following supportive criteria: (1) brain MRI not meeting criteria of MS, (2) spinal cord MRI extending over three or more vertebral segments, and (3) NMO-IgG positive. There have been reports of SS complicated by NMSD including long extended spinal cord lesion (LESCL). Min et al. and Estiasari et al. reported that the rates of central nervous system involvement in SS patients were with LESCL positive for the anti-AQP4 antibody in East Asia 42% and 18%, respectively[7,8]. However, SS complicated by NMO seems to be rare in non-East Asian regions. Kolfenbach et al. reviewed English literatures and found 21 cases of overlap SS and NMO with additional 5 more cases of their own[4]. Etiology of association between SS and NMO is still unclear. However, there is a retrospective blinded serological survey to support the evidence of coexisting NMO in NMSD with positive NMO-IgG occurring with SS rather than as a complication of SS[5]. The first line treatment of acute attack of NMO is high dose intravenous methylprednisolone 1000mg daily for at least three to five days [6]. Plasmapheresis or cyclophosphamide may be considered if there is no clinical improvement with steroid therapy alone. Azathioprine, rituximab, mycophenolate mofetil, methotrexate, prednisone, or mitoxantrone can be used as maintenance therapy[6].

## CONCLUSION:

This is a case of NMOSD associated with connective tissue disease The case supports the association between NMOSD and SS. Early diagnosis and treatment are critical in decreasing the possibility of irreversible severe neurological dysfunction. Awareness of the possibility of CNS disease is important because of the serious nature of CNS complications and because some are treatable with immunosuppressive medications. Our patient with SS who presented with NMOSD benefitted from early recognition and institution of appropriate therapy.

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