International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Primary Sjogren's Syndrome Presenting as Neuromyelitis Optica Spectrum Disorder (NMOSD)

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ABSTRACT

Primary Sjogren's syndrome is a rare autoimmune disease characterised by keratoconjuctivitis sicca due to autoimmune destruction of exocrine glands. Neuromyelitis optica (NMO), is a severe, relapsing demyelinating disease that predominantly affects optic nerves and the spinal cord. NMO and related NMOSD have been found to be associated with Primary Sjogren's syndrome. Here we report a patient who presented as Neuromyelitis optica spectrum disorder (NMOSD) and on evaluation found to have Primary Sjogren's syndrome despite the absence of glandular symptoms. NMOSD as the initial presentation of Primary Sjogren's syndrome is less frequently reported in the literature; hence this case has been reported.

Keywords: Autoimmune disease, Neuromyelitis optica, demyelinating disease, optic nerves.

INTRODUCTION

Primary Sjögren syndrome (pSS) is an autoimmune disease characterized by lymphocytic infiltration of the exocrine glands and associated with circulating autoantibodies. Sjogren syndrome has been associated with various extraglandular manifestations including neurological dysfunction. [1] Neuromyelitis optica (NMO), also called Devic disease, is a severe, relapsing, autoimmune inflammatory and demyelinating central nervous system disease that predominantly affects optic nerves and spinal cord. The disorder is now recognized as a spectrum of autoimmunity targeting the astrocytic water channel aquaporin-4 (AQP4).

Neuromyelitis optica spectrum disorder (NMOSD) is a recently proposed unifying term for NMO and related syndromes. [2] 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 of NMO. An association between the NMO, NMOSD and primary Sjogren's syndrome has been reported in the literature. [3,4] Here in this case, the initial presentation was that of NMOSD and later on found to have primary Sjogren's syndrome. PSS with NMOSD as the initial manifestation is usually rare, hence we reported this case.

CASE HISTORY

A 29 year old female presented with acute onset of left eye pain associated with diminution of vision for 2days. She was diagnosed to have left eye optic neuritis and treated with intravenous methylprednisolone 1gm /day for 3 days followed by oral steroids (figure 1). Patient recovered completely over a period of 5 days. After 2 years, she developed a second episode involving pain over both the eyes with decreased vision. Following treatment with pulse methylprednisolone, her symptoms subsided but there was a residual impairment in vision of both eyes (6/12 &

6/9). She was evaluated for recurrent optic neuritis and was found to have positive anti aquaporin 4 antibodies. MRI brain revealed enhancement of both optic nerves with normal brain parenchyma suggestive of bilateral optic neuritis. CSF examination showed normal protein, glucose and cell counts with no oligoclonal bands. She was diagnosed as a case of NMOSD and treated with oral steroids and azathioprine.

Inspite of the treatment patient developed third episode 6 months later, where she presented with acute onset of both ocular pain with reduced vision. On examination, her visual acuity was 6/36 (right) & counting finger (left). Anterior segment examinations of both eyes were normal. Fundoscopic examination showed bilateral swollen and hyperemic disc. Blood investigations showed anaemia (Hb-9.6gm %), elevated ESR (35mm/hr), elevated CRP (24mg/l), normal renal and liver function tests. Serum electrolytes were normal. MRI Brain with screening of spine showed T2/FLAIR hyperintensities in bilateral optic nerves with normal spinal cord (figure 2). On further evaluation, her Antinuclear antibody (ANA) by immunofluorescence was positive (1:100 dilution) with speckled pattern. ANA (Immunoblot) showed positivity for SSA/ Ro52 & SSB/La. Anti dsDNA, Antineutrophil cytoplasmic antibody (p-ANCA & c-ANCA) and antiphospholipid antibodies were negative. Complements C3, C4 were normal. CSF examination showed normal protein, glucose and cell counts without oligoclonal bands. Patient denied of any sicca symptoms. However, ophthalmological evaluation showed bilateral positive Schirmer's test (less than five mm). Lip biopsy of minor salivary glands revealed evidence of multiple lymphocytic foci with focus score of 2 with normal salivary gland architecture, consistent with the diagnosis of Sjogren's syndrome; thus fulfilling the international criteria for diagnosis of Sjogren's syndrome. The patient was treated with intravenous methylprednisolone (1 gm/day) for five days followed by oral prednisolone and Mycophenolate mofetil. Patient responded well to the treatment with no further episodes. On follow up, her visual acuity was 6/24 in right eye and 6/18 in left eye.

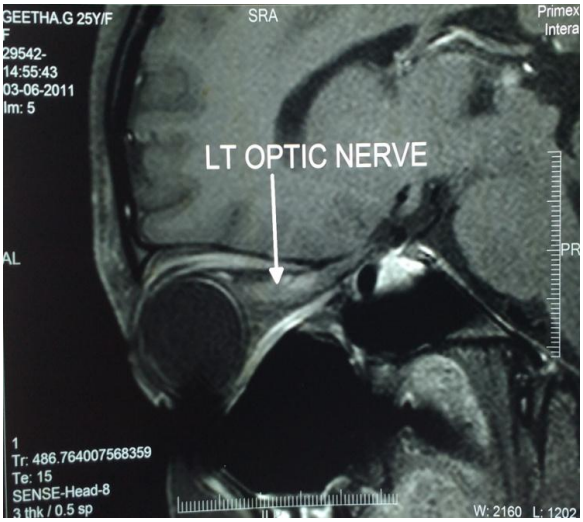

Figure 1: MRI Brain (T1) showing enhanced left optic nerve

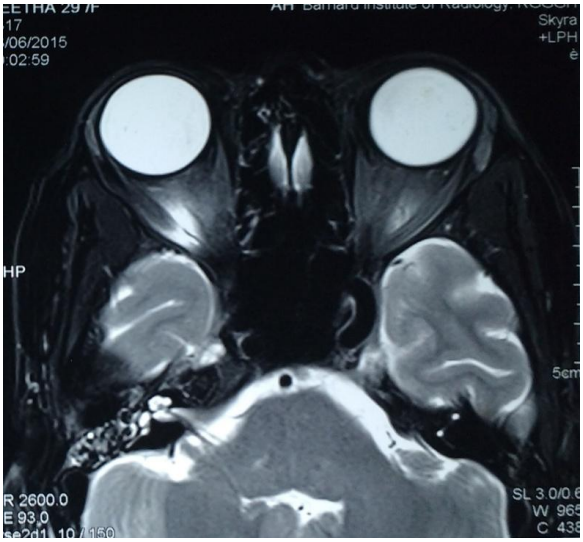

Figure 2: MRI Brain showing T2/FLAIR hyperintensities in bilateral optic nerves

DISCUSSION

Neurologic abnormalities are protean in primary Sjögren's syndrome, which include asymptomatic brain lesions on MRI to symptomatic brain lesions, meningitis, myelopathy, cranial neuropathy, sensorimotor polyneuropathy and mononeuritis multiplex. CNS involvement in patients with Sjögren's syndrome is much less common than peripheral nervous system involvement and ranges from 2% to 25%. [5] Spinal cord involvement in the form

of multiple sclerosis, progressive myelopathy, longitudinally extensive myelitis or transverse myelitis can occur. Anti-aquaporin-4 antibody targets a water channel expressed on astrocytes, have 70% sensitivity and 90% specificity for NMO. [6]

About 26 cases of overlap SS and NMO have been identified since 2011. [3] Delalande *et al.*, reviewed the neurologic manifestations in 82 patients with Sjögren's syndrome and found that 13 out of 82 (16%) patients had visual loss secondary to Optic Neuritis. [7] In another study by Gono *et al.*, three of 17 (18%) primary Sjögren's syndrome patients had Optic neuritis. [8] Upto 40% of NMO patients have a systemic autoimmune disorder, often systemic lupus erythematosus, Sjögren's syndrome, p-ANCA associated vasculitis, myasthenia gravis, Hashimoto's thyroiditis, or mixed connective tissue disease.

Our patient presented with recurrent episodes of optic neuritis without the evidence of myelitis, but positive anti aquaporin-4 antibody, hence diagnosis of NMOSD was made. Immunological evaluation showed positive anti SSA/Ro52 & anti SSB/La, though the patient denied sicca symptoms. A retrospective review suggested that up to 33% of patients who presented with primary SS involving the CNS did not have sicca symptoms at the time of presentation but developed them over a period of 5-year. [9] A review by kahlenberg JM et al., showed that, of the total patients with SS and NMOSD, 36% had evidence of optic neuritis and 85% of patients with optic neuritis had positive NMO antibody. [10]

A retrospective blinded serological survey support the evidence of coexisting NMOSD with positive NMO-IgG occurring with SS rather than as a complication of SS. [11] The first line treatment of acute attack of NMO or NMOSD is high dose intravenous methylprednisolone 1gm daily for 3-5 days followed by oral prednisolone. Azathioprine, rituximab, mycophenolate mofetil, and methotrexate can be used as maintenance therapy. [12] Plasmapheresis or cyclophosphamide may be considered if there is no clinical improvement with steroid therapy alone. [13]

CONCLUSION

This case report emphasis the coexistence of NMOSD with Sjogren's syndrome. Patients presenting with recurrent optic neuritis, even without the features of myelitis should be screened for anti NMO IgG/ AQP4 antibodies, to favour the diagnosis of NMOSD. Rheumatologic evaluation is necessary in these patients to rule out the coexistent Connective tissue disease for appropriate management and follow up.

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How to cite this article: Saranya. C. Primary Sjogren's syndrome presenting as neuromyelitis optica spectrum disorder (NMOSD). Int J Health Sci Res. 2019; 9(9):252-255.

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