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Case Report

Sjogren's Syndrome Presenting as Longitudinally Extensive Transverse Myelitis

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Abstract

Longitudinal extensive transverse myelitis (LETM) is defined as a spinal cord lesion that spans three or more contiguous vertebral segments in length. We present the case of a 30 year old woman who presented with weakness and dysaesthesias of both lower limbs along with urinary retention and constipation. She had history of joint pains, dry eyes and mouth for the past 3 years. Paraplegia with hyperreflexia and an extensor plantar reflex was evident on neurological examination. Cerebrospinal fluid was acellular, with mildly raised protein (60 mg/dl) and sugar of 40 mg/dl. MRI of the spinal cord revealed longitudinally extensive diffuse hyperintensities extending from C4 to conus medullaris. MRI of the brain was normal. Visual evoked potential (VEP) was abnormal. Antibodies for neuromyelitis optica (anti-aquaporin-4 antibody) were negative. Patient turned out to be positive for antinuclear antibodies and anti SS-A antibodies. Schirmer's test was positive and patient was diagnosed as a case of Sjogren's syndrome with LETM. The patient was given corticosteroids and intravenous immunoglobulin (IVIg) following which she showed partial recovery. LETM is a characteristic feature of neuromyelitis optica (NMO) but such spinal lesions can also occur in systemic autoimmune diseases like Sjogren's syndrome.

Keywords: Longitudinal Extensive Transverse Myelitis; Sjogren's Syndrome; Neuromyelitis Optica; NMO Spectrum Disorders

Introduction

Longitudinally extensive transverse myelitis (LETM) is defined as a spinal cord lesion that extends through three or more contiguous vertebral segments. LETM has been most commonly associated with neuromyelitis optica (NMO). However, systemic autoimmune diseases like Sjogren's syndrome (SS), systemic lupus erythematosus (SLE), antiphospholipid syndrome, inflammatory conditions like Behcet's disease, sarcoidosis, multiple sclerosis (MS), and infectious diseases can also cause LETM and thus are important differential diagnosis [1].

Case Report

Our case is a 30-year-old woman who presented with weakness and dysaesthesias of both lower extremities. Weakness was insidious in onset and gradually progressive over a period of 20 days, associated with urinary retention and constipation. She also had band like sensation at the level of nipples. The patient had a prior history of joint pains, dryness of eyes and mouth for the past 3 years. Neurological examination revealed paraplegia with hyperreflexia and an extensor plantar response. There was a sensory level at T4. Cerebrospinal fluid was acellular, with raised protein (60 mg/dl), and a sugar of 40 mg/dl. MRI of the spinal cord (Figure 1) showed longitudinal extensive lesions extending from C4 to the conus medullaris. MRI of the brain (Figure 2) was normal. Visual evoked potentials (VEP) showed prolonged P-100 latencies bilaterally. Anti-aquaporin-4 antibodies for neuromyelitis optica (qualitative) were negative. Antinuclear antibodies (ANA), Anti-SSA antibodies were positive and anti-double stranded DNA (ds DNA) antibodies were negative. Schirmer's test in both eyes was also positive (3 and 4 mm). A final diagnosis of LETM secondary to SS was made. The patient was treated with intravenous methyl prednisolone I gram daily for five days. As there was no clinical improvement she was initiated on intravenous immunoglobulin 2 gram/kg over five days. Following this she showed partial recovery of symptoms and she was thereafter discharged on oral prednisolone 50 mg daily. Follow-up MRI scan 2 weeks later of the spinal cord showed disappearance of the high signal intensities.

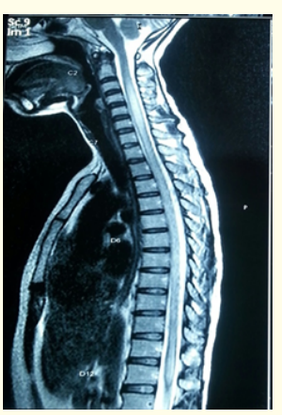

Figure 1: MRI spine: T2 sagittal images showing multiple, confluent high signal intensities within the spinal cord, extending from the third cervical vertebra to the conus medullaris.

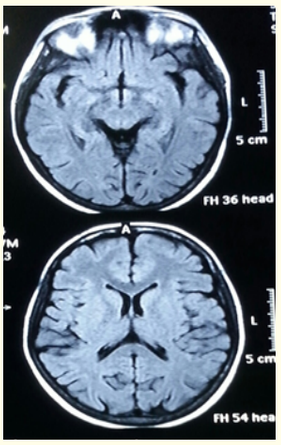

Figure 2: Normal MRI Brain, fluid inversion recovery (FLAIR) sequences.

Discussion

LETM is a characteristic feature of NMO (positive for NMO-IgG antibodies). Apart from NMO, systemic autoimmune disorders such as SS and SLE can present as or with LETM (negative for NMO-IgG antibodies). In some patients, NMO and systemic autoimmune disorders, particularly SS or SLE, coexist and have been grouped under NMO spectrum disorders (NMOSD) (positive for NMO-IgG antibodies) [2]. This has been seen in a retrospective study by Pittock., *et al*. where in group 1 out of 153 patients with NMOSD, NMO-IgG was detected in 66.7%, ANA in 43.8%, and SSA antibodies in 15.7%. In group 2, 5 out of 14 patients with NMOSD were NMO-IgG positive and 49 controls of SLE/SS in group 1 and 2 were NMO-IgG negative. The study concluded that NMOSD with seropositivity for NMO-IgG occurring with SS/SLE is an indication of coexisting NMO rather than a vasculopathic or other complication of SS/SLE [3]. SS can be associated with neurological complications, and often as the first manifestation. Teixeira et al. in his study detected neurological involvement in 28% of patients with primary SS [4]. Delalande., *et al.* [5] in his study concluded that neurological complications are the first symptom of SS in 81% patients. Kahlenberg [6] reported a case of SS with NMOSD as the first presentation. Our case was diagnosed primarily as SS with LETM and was NMO-IgG negative. So far few cases have been reported [7-9] where LETM was the first presentation of SS.

Conclusion

Thus, we propose that all patients presenting with LETM who are negative for NMO-IgG antibody must be investigated for SS as the treatment modalities are different in cases with LETM due to NMO and versus those secondary to SS. Differentiating the two conditions, therefore, has practical clinical implication.

Conflict of Interest

On behalf of all authors, the corresponding author states that there is no conflict of interest.

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