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SJÖGREN'S SYNDROME PRESENTING FIRST TIME AS SENSORY NEURONOPATHY

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ABSTRACT

Primary Sjögren's is an autoimmune disease which mainly affects exocrine glands causing a sicca syndrome. Neurological manifestations are rarely seen in Sjögren's syndrome, but when present they are debilitating. The most frequent neurological manifestation are peripheral neuropathies especially sensory axonal neuropathy, painful sensory neuropathy etc. Dorsal root ganglionopathy is less frequently seen but is more handicapping. We present a case of a young female who presented first time with sensory neuropathy in absence of any other manifestations of of Sjögren's syndrome.

KEYWORDS: Sjögren's syndrome(s.s), Dorsal Root Ganglionopathy (DRG).

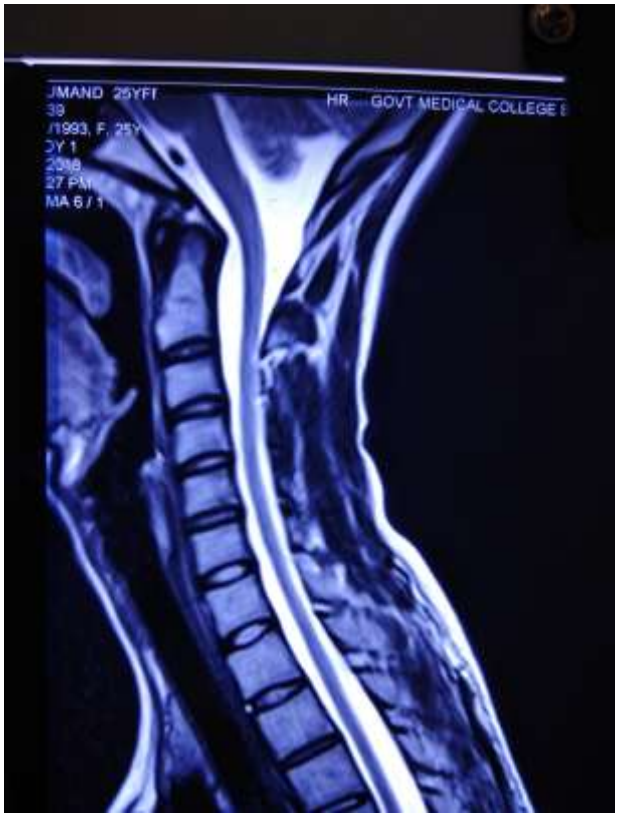
CASE REPORT

20 year old female presented to us with complaints of numbness and parasthesias of left foot and hand. The symptoms progressed over a year to involve all the limbs. There was difficulty in performing skilled activity like writing, buttoning and unbuttoning in dim light. Patient also had a tendency to sway to either side especially in the dark. There was no other symptom.

Examination of the patient revealed normal vital signs.

Systemic examination was normal. CNS examination revealed normal higher mental functions including consciousness, orientation, language and memory. Cranial nerve examination was normal. Motor bulk, tone, power of muscles were normal. The sensations for pain and temperature were normal but position and vibration sense were impaired in all the limbs. Deep tendon reflexes were all absent. Ataxia was present which worsened with closing eyes in darkness, Romberg's sign. Gait was high steppage. There were no cerebellar signs.

Her investigation revealed a normal complete blood counts, kidney function tests, liver function tests, blood glucose, sodium, potassium, calcium, phosphorus and uric acid levels. ANA, antiRO, antiLA, RF and CRP were positive. A shirmer test was done which was normal and a usg abdomen revealed no abnormality. Nerve conduction studies revealed non-stimulable sensory nerves but normal motor. Nerve conduction studies and MRI brain and spine was normal.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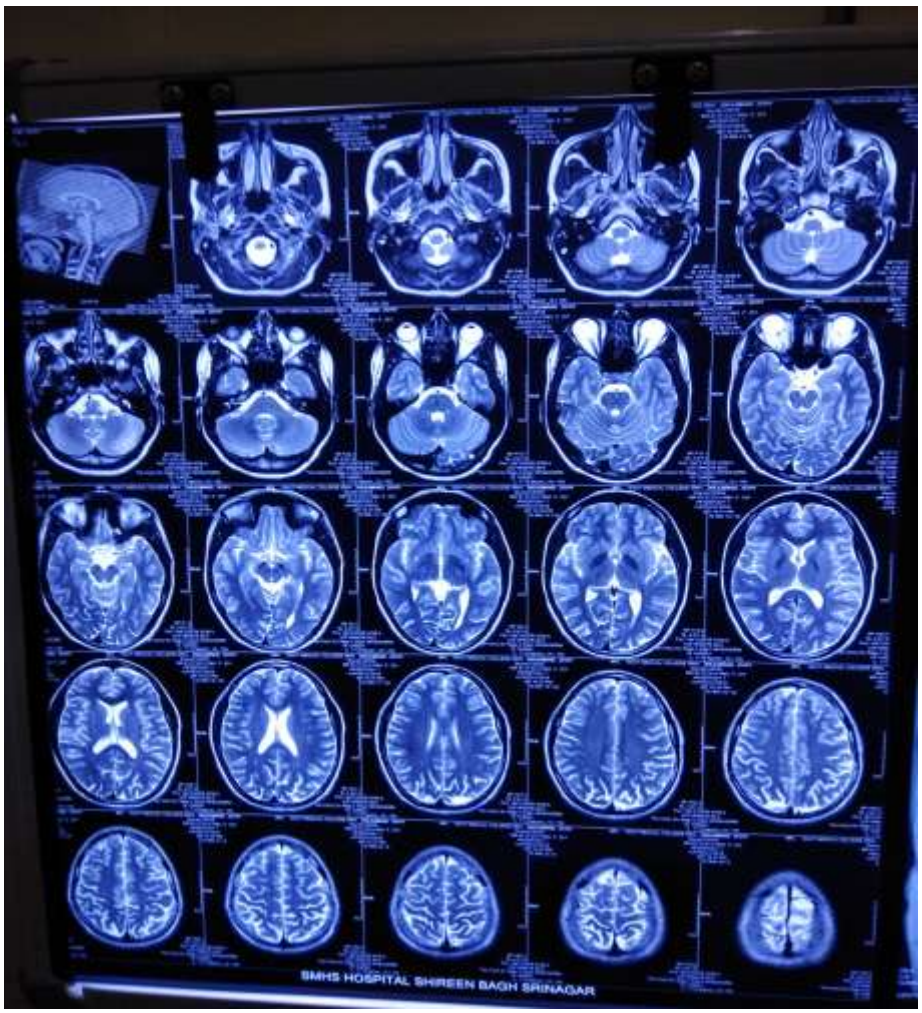
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DISCUSSION

Sjogren's Syndrome

Sjögren's syndrome is a chronic, slowly progressive autoimmune disease characterized by lymphocytic infiltration of the exocrine glands resulting in xerostomia and dry eyes. Approximately one-third of patients present with systemic manifestations; a small but significant number of patients develop malignant lymphoma. The disease presents alone (primary Sjögren's syndrome) or in association with other autoimmune rheumatic diseases (secondary Sjögren's syndrome). Sera from patients with Sjögren's syndrome often contain autoantibodies to non-organ-specific antigens such as immunoglobulins (rheumatoid factors) and extractable nuclear and cytoplasm antigens (Ro/SS-A, La/SS-B). Sjögren's syndrome is characterized by both lymphocytic infiltrations of the exocrine glands and B lymphocyte hyperreactivity. The majority of patients with Sjögren's syndrome have symptoms related to diminished lacrimal and salivary gland function. In most patients, the primary syndrome runs a slow and benign course. The initial manifestations can be mucosal or nonspecific dryness, and 8-10 years may elapse from the initial symptoms to full-blown development of the disease. The principal oral 8-10 years may elapse from the initial symptoms to full-blown development of the disease. Primary Sjögren's syndrome is diagnosed if (1) the patient presents with Eye and/or mouth dryness, (2) eye tests disclose keratoconjunctivitis sicca, (3) mouth evaluation reveals the classic manifestations of the syndrome, And/or (4) the patient's serum reacts with Ro/SS-A and/or La/SS-B autoantigens. Labial biopsy is needed when the diagnosis is uncertain or to rule out other conditions that may cause dry mouth or eyes or parotid gland enlargement.

Sensory neuronopathy also known as ganglionopathy are pure sensory neuropathies caused by dorsal root ganglia neural destruction,[1] which results in multifocal sensory deformities quite different from length dependent pattern of axonal neuropathies. Dorsal root ganglionopathies is Rare when compared to other neuropathies and its true prevalence is unknown. Several pathogenic mechanisms such as genetic susceptibility, drug related toxicity, infectious agents and autoimmune damage has been proposed. Other causes of dorsal root gangliopathy may be paraneoplastic syndrome (anti hu antibody positivity) inflammatory autoimmune disease, neurotropic viral infection, vitamin B12 deficiency, chemotherapeutic agents and pyridoxine toxicity. Autoimmune diseases such as Sjogrens syndrome and celiac disease[1,2] have been incriminated as other causes. The clinical features of sensory neuropathy are parasthesias, sensory ataxia, and difficulty in fine motor tasks due to impaired proprioception, reduced vibration sense, reduced or absent deep tendon reflexes and positive Romberg's sign. motor power is usually preserved but autonomic dysfunction may be found.[3,4] Electrophysiological studies reveal widespread reduced sensory potential amplitudes without a distal worsening gradient towards legs. Asymmetric response[5,6] may be observed. Motor nerve studies and distal motor amplitudes are normal. Somatosensory evoked potentials may reveal abnormal central conduction times which are due to degeneration of dorsal columns of the spinal cord. MRI has been used as a sensitive technique especially in long duration disease patients which may show hyperintense T2W lesion at posterior column and volumetric reduction in cervical cord resulting from dorsal root degeneration of their projections in the fasciculus gracilus and cuneatus.[7] The gold standard for diagnosis of sensory neuropathy is dorsal root ganglion biopsy but is rarely performed due to possible side effects. Sural nerve biopsy usually shows massive axonal loss and is not helpful in diagnosis.[8]

Most of the literature on Sjogren's syndrome focus on axonal neuropathy so that there is a limited data on Longterm outcome and therapeutic response on Sjogren's related sensory neuropathy. Due to rarity of this disease there are no randomized controlled trials and conclusions are saved as case reports or small case series in the literature. There are some reports about the efficacy of the immunosuppressive drugs, plasma exchange, intravenous immune globulin and anti TNF drugs with controversial effects.

CONCLUSION

Sensory neuropathy is a rare disease which can occur in Sjogren's syndrome patients. Its progression is somewhat heterogeneous but tends to be chronic, insidious and debilitating despite treatment. It may be the first presentation of Sjogren's syndrome and may create a lot of difficulty in establishing of underlying Sjogren's syndrome.

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