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Sjogren's Syndrome Presenting as Longitudinally Extensive Transverse Myelitis. A case report - S Bashir Ahmad- Superspeciality Hospital GMC Srinagar

S Bashir Ahmad
Superspeciality Hospital GMC Srinagar, India

Introduction:

Sjogren's (SHOW-grins) syndrome is a disorder of your immune system identified by its two most common symptoms dry eyes and a dry mouth. The condition often accompanies other immune system disorders, such as rheumatoid arthritis and lupus.Neuromyelitis optica (NMO) is a rare relapsing auto-immune disease of the central nervous system which is sometimes found in association with other autoimmune disorders including Sjogren's syndrome. Sjogren's syndrome can be difficult to diagnose because the signs and symptoms vary from person to person and can be similar to those caused by other diseases. Side effects of a number of medications also mimic some signs and symptoms of Sjogren's syndrome.Tests can help rule out other conditions and help pinpoint a diagnosis of Sjogren's syndrome. We present the case of a middle aged female with Sjogren's syndrome (SS) and Neuromyelitis optica spectrum disorders (NMOSD) who had a rapidly declining neurological illness that responded to immunosuppressive therapy. Sjögren's syndrome (SjS, SS) is a long-term autoimmune disease that affects the body's moisture-producing (lacrimal and salivary) glands, and often seriously affects other organs systems, such as the lungs, kidneys, and nervous system. Primary symptoms are dryness (dry mouth and dry eyes), pain and fatigue. Other symptoms can include dry skin, vaginal dryness, a chronic cough, numbness in the arms and legs, feeling tired, muscle and joint pains, and thyroid problems. Skin dryness in some people with SS may be the result of lymphocytic infiltration into skin glands. The symptoms may develop insidiously, with the diagnosis often not considered for several years because sicca may be attributed to medications, a dry environment, or aging, or may be regarded as not of a severity warranting the level of investigation necessary to establish the presence of the underlying autoimmune disorder. Those affected are also at an increased risk (15%) of lymphoma. While the exact cause is unclear, it is believed to involve a combination of genetics and an environmental trigger such as exposure to a virus or bacterium. It can occur independently of other health problems (primary Sjögren's syndrome) or as a result of another connective tissue disorder (secondary Sjögren's syndrome). Sjögren's syndrome may be associated with other autoimmune diseases, including rheumatoid arthritis (RA), systemic lupus erythematosus (SLE) or systemic sclerosis. The inflammation that results progressively damages the glands. The disease was described in 1933 by Henrik Sjögren, after whom it is named; however, a number of earlier descriptions of people with the symptoms exist. Between 0.2 and 1.2% of the population is affected, with half having the primary form and half the secondary form. Females are affected about 10 times as often as are males. Though the disease commonly begins in middle age, anyone can be affected. Among those without other autoimmune disorders, life expectancy is unchanged. Diagnosis is by biopsy of moisture-producing glands and blood tests for specific antibodies. On biopsy there are typically lymphocytes within the glands. While Sjögren's syndrome is one of the most common auto-immune diseases, it has no specific and non-invasive diagnostic tests and treatment is directed at managing the person's symptoms. For dry eyes, artificial tears, medications to reduce inflammation, punctal plugs, or surgery to shut the tear ducts may be tried. For a dry mouth, chewing gum (preferably sugar-free), sipping water, or a saliva substitute may be used. In those with joint or muscle pain, ibuprofen may be used. Medications that can cause dryness, such as antihistamines, may also be stopped. The most specific extant diagnostic test requires lip

biopsy.Longitudinal extensive transverse myelitis (LETM) is defined as a spinal cord lesion that extends over three or more vertebrae, as seen on MRI of the spine. Our case is a 50 year gentleman who presented with history of severe radicular pain in the upper back followed by weakness of all four limbs for two months associated with paresthesias, urinary retention and constipation. Neurological examination revealed quadriparesis with sensory level at T1, hyperreflexia and extensor plantar response. MRI of the spinal cord showed longitudinally extensive T2 lesions /hyperintensities extending from C1 to the conus medullaris. Cerebrospinal fluid examination revealed mild lymphocytic pleocytosis (20 cells) with normal sugar (53mg/dl) and raised protein(61mg/dl). CSF staining, MTB-PCR, VDRL were negative. MRI Brain was normal. Anti NMO panel (anti-Aquaporin 4 antibody and anti- MOG antibody) was negative. ANA , anti-Ro/anti- SSA and anti-La/anti-SSB were positive and anti-ds DNA was negative. Schirmer test was positive in both eyes and lip biopsy revealed focal lymphocytic sialoadenitis. A final diagnosis of LETM secondary to Sjogren's syndrome was made. The patient was treated with intravenous methylprednisolone 1gm daily for five days. As there was no clinical improvement patient was initiated on plasmapheresis 250ml/kg in five sessions over a period of 10 days. Following this patient showed partial recovery in symptoms. LETM is a characteristic feature of neuromyelitis optica (NMO) but such spinal lesions can also occur in systemic autoimmune diseases like Sjogren's syndrome.