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CASE REPORT

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A Sjögren's syndrome case presenting with myelitis and without sicca symptoms

Myelit bulguları ile prezente olan ve sicca semptomları olmayan sjögren vakası

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ABSTRACT

Sjögren's syndrome (SS) is a chronic inflammatory disorder of exocrine glands with autoimmune etiology. Neurological symptoms occur in approximately 20% of patients with Sjögren's syndrome. Neurological symptoms may occur before the onset of sicca symptoms such as; dry eyes and dry mouth. The prevalence of peripheral and central neurological manifestations in Sjögren syndrome are about 15% and 5% respectively. CNS involvement is much less common than peripheral nervous system involvement. In this case, our patient presented symptoms of transverse myelitis; however, the serum Anti-Ro/SSA and anti-La/SSB antibodies were negative and there were no dry eyes and dry mouth.

Keywords: Sjögren's syndrome, Myelitis, Sicca symptoms

ÖZET

Sjögren sendromu (SS) egzokrin bezlerin kronik, inflamatuar ve otoimmün hastalığıdır. SS' da hastaların %20' sinde nörolojik bulgular gelişir. Nörolojik bulgular göz ve ağız kuruluğu gibi sicca semptomları ortaya çıkmadan önce de görülebilir. Periferik sinir sistemi tutulumu (PSS) %15, santral sinir sistemi (SSS) tutulumu %5 oranında görülür. SSS tutulumu, PSS tutulumuna göre çok daha az görülür. Bu vakada, transvers myelit ile prezente olan, serum anti Ro-SSA/anti La-SSB antikorları negatif olan ve sicca bulguları olmayan bir sjögren sendromu ele alınmıştır.

Anahtar Kelimeler: Sjögren sendromu, Myelit, Sicca semptomları

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Sjögren's syndrome (SS) is a chronic inflammatory disorder of exocrine glands with autoimmune etiology. Neurological symptoms occur in approximately 20% of patients with Sjögren's syndrome [1-2]. Neurological symptoms may occur before the onset of sicca symptoms (dry eyes and dry mouth) [3]. Anti-Ro/SSA and anti-La/SSB antibodies are present 50% to 70% of patients with Sjögren syndrome [4]. Patients with SS that have neurological symtoms, only 21% have anti-SSA or anti-SSB antibodies [5]. In this article, we report a patient with SS, presenting as myelitis without any sicca symptoms and without any Anti-Ro/SSA and anti-La/SSB antibodies in the sera of patient

CASE REPORT

A 30 year old man presented with left leg weakness about 1.5 year. On first examination, he had weakness in his left leg with muscle strength of 4/5 in proximal,3/5 in knee flexion and extansion and there was no weakness in distal muscles. The weakness progressed. After a few months left upper limb and bilateral foot weakness added with muscle stregth of 4/5 in distal muscles. He had areflexia at lower limbs and loss of joint sensation and vibration sense in lower limbs. There was decreased sensation to pinprick and light touch in left leg and left hand at 4th and 5th fingers. Also there was bilaterally decreased sensation with a stocking distrubition and . All of the additional neurological examination was normal. The result of routine laboratory studies were at normal levels, including complete blood cell count, liver function, kidney function, coagulation, thyroid, lipids, glucose, glycosylated hemoglobin, C-reactive protein and vitamin B12. Antibody test was negative for Ro/SS-A, La/SS-B, ANA,HIV, HCV and classical paraneoplastic antigens such as Anti-Yo, Anti-Hu, Anti-Ri, Anti- Amphiphysin, Anti- CV2.4, Anti-PNMA$_2$/Ta. Erythrocyte sedimantation rate (ESR) (3 mm/hour) and rheumatoid factor (<20 IU/ml) was normal on admission. Also serological tests for Hepatit B surface antigen, syphilis and brucella were negative. Analysis of cerebrospinal fluid examination revealed raised protein level (81 mg/dL) with normal chloride and sugar level and no cells. Urine and serum immunelectrophoresis were negative. Autoantibody to aquaporin-4 (NMO-IgG) was also negative. ENMG revealed sensorial axonal polyneuropathy. SNAP amplitude was extensively reduced in the nerves examined with relatively preserved conduction velocity. Thorax CT, upper and lower abdomen CT, thyroid USG, scrotal USG and PET done for cancer screening. Lomber MRI was done outer center and was normal. Cervical and thoracal MRI performed after admission. MRI showed signal changes with enhancing effect. Lesions were spanning the cervical and thoracic cord posterior column. Brain MRI was not done with the cause of the patient's claustrophobia . VEP was normal.

The presence of sensorial axonal polineuropathy and longitudinally extensive tranverse myelitis that spans the servical and thoracic spinal cord led us thinking about SS. The positive schirmer' test and lip biopsy supported the diagnosos of SS.

DISCUSSION

Sjögren's syndrome affect the nervous system in approximately 20% of cases. The prevalence of peripheral and central neurological manifestations in Sjögren syndrome are about 15% and 5% respectively [6]. Most common peripheral nervous system (PNS) presentations are axonal polyneuropathies (distal axonal sensory and sensorimotor), sensory ganglionopathy, motor neuropathy, small fiber neuropathy, multiple monuneuritis, trigeminal and other cranial neuropathies, autonomic neuropathies, demyelinating polyradiculoneuropathy [7]. The sensory neuropathies constitute the most frequent PNS complication [2]. Central nervous system (CNS) manifestations of Sjögren's syndrome may affect spinal cord, brain stem, optic nerves, cerebellum, and cerebral hemispheres. CNS involvment is much less common than peripheral nervous system involvement. Transverse myelitis is an inflammatory disorder of the spinal cord that presents acutely or subacutely. It may cause weakness, sensory loss and bowel and bladder involvement. Transverse myelitis in Sjögren's syndrome spans more than three levels of the spinal cord. Longitudinally extensive transverse myelitis that spans the cervical and/or thoracic spinal cord is the most common pattern in Sjögren's syndrome associated myelitis. Serum autoantibodies have low sensitivity in Sjögren syndrome associated neuropathy . But Sjögren syndrome with longitudinal myelitis, 85% of patients have positive serum Anti-Ro/SSA and anti-La/SSB antibodies [5].

Conclusion: In this case, our patient presented with symptoms of transverse myelitis. But the serum Anti-Ro/SSA and anti-La/SSB antibodies were negative and there were no dry eyes and dry mouth . Neurological symptoms may precede the onset of sicca symptoms. So, it is important to consider SS in differantial

diagnosis even if there is no sicca syndrome or even if autoantibadies are negative.

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