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Sjögren Syndrome Simulating Relapsing Remitting Multiple Sclerosis Clinical Features: Case Report

Relapsing Remitting Multipl Skleroz Kliniğini Taklit Eden Sjögren Sendromu: Olgu Sunumu

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Summary

Sjögren syndrome (SS) is a chronic, inflammatory, autoimmune disease. It primarily presents with dry mouth and eyes (sicca symptoms) because it frequently affects exocrine, salivary and lacrimal glands. Neurological involvement in Sjögren syndrome is observed in approximately 20-25% of cases. Eighty seven percent of the neurological involvements are located in the peripheral nervous system and around 13% of the neurological involvements affect the central nervous system. Cerebral involvement presents a heterogeneous profile both in terms of localization (focal or diffuse) and progress of the condition (acute, progressive or reversible). The affected central nervous system can show clinical and radiological signs similar to multiple sclerosis (MS). A case with reported imbalance and difficulty in walking, who also showed MS-like lesions in magnetic resonance imaging and was previously diagnosed with Sjögren syndrome, is discussed. *(Turkish Journal of Neurology 2013; 19:145-147)*

Key Words: Multiple sclerosis, Sjögren syndrome

Özet

Sjögren sendromu (SS), kronik inflamatuvar otoimmün bir hastalıktır. Esas olarak ekzokrin bezleri etkileyip sıklıkla tükrük ve gözyaşı bezlerinin fonksiyonel etkilenmesine bağlı olarak göz ve ağız kuruluğu (sicca semptomları) ile ortaya çıkmaktadır. Sjögren sendromunda nörolojik tutulum olguların yaklaşık %20-25'inde görülür. Nörolojik tutulumların %87'si periferik sinir sistemi, yaklaşık %13'ü santral sinir sistemi tutulumu şeklindedir. Serebral tutulum gerek lokalizasyon (fokal veya difüz) gerekse tablonun gelişimi (akut, progressif veya reversibl) açısından heterojen özellikler gösterir. Santral sinir sistemi etkilenmesi multipl skleroz (MS) benzeri klinik ve radyolojik bulgular gösterebilir. Bu bildiride yürüme güçlüğü ve dengesizlik yakınmasıyla başvuran, beyin manyetik rezonans görüntülemede (MRG) MS benzeri lezyonlar olan ve yapılan araştırmalarda Sjögren sendromu tanısı alan bir olgu tartışılmaktadır. *(Türk Nöroloji Dergisi 2013; 19:145-147)*

Anahtar Kelimeler: Multipl skleroz, Sjögren sendromu

Introduction

Sjögren syndrome (SS) is a chronic inflammatory autoimmune disorder involving exocrine glands which often presents with functional disruptions in salivary and tear glands, leading to dry mouth and eyes (sicca symptoms) (1). Neurological involvement in SS is seen in 20-25% of the cases (2). Both peripheral and central nervous systems (PNS, CNS) may be affected in the neurological involvement. Peripheral nervous system is affected in 87% of such cases whereas CNS is affected in 13% of them. Cerebral involvement is generally heterogeneous both in terms of localization (focal or diffuse) and the progression (acute, progressive or reversible). The involvement of CNS may resemble multiple sclerosis (MS) clinical and radiological findings (5). In this report, we present the case of a patient with SS who consulted with walking difficulty and imbalance and who showed MS-like lesions in magnetic resonance imaging (MRI).

Case

A 50 year old, right-handed male consulted in our clinic with speech and walking difficulty, and balance problem. Walking difficulty had started a month ago and he started limping in addition to having fallen a few times. Slurred speech also started at

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this time. The severity of these complaints increased over time. He reported experienced impaired vision on the left side 20 years ago which resulted in complete loss of vision. After being diagnosed with MS 7 years ago following an vision impairment in the right eye, he was started on steroid treatment and recovered with partial loss of vision. He had been experiencing dry eyes and mouth for the past 10 years. For the last 3-4 years he also reported having joint paints. In his neurological examination, he was able to track the light with his left eye, There were also papillary atrophy and relative afferent pupil defect (RAPD) on the left side, bilateral dysmetria-dysdiadochokinesia, increased tendon reflexes, and bilateral Hoffman and Babinski signs were positive.

There were lesions in the periventricular deep white matter, cerebellar peduncle, pons and mesencephalon seen as isointense in the T1-weighted sequences, and hyperintense in the T2 and flair sequences in his contrast cranial MRI (Figures 1, 2). Patient's liver and renal function tests, RF, hepatitis panel, anti-HIV, hemogram, lung graphy and electrocardiography were normal. Protein C, protein S, antithrombin 3, homocysteine, vitamin B12, lupus anticoagulant anticardiolipin antibody, sedimentation tests were normal. Factor V Leiden mutation was negative. In the visual evoked potential test (VEP), P100 latency in the left eye was 172 ms, and 147 ms in the right eye. Oligoclonal band (OCB) test in cerepbrospinal fluid (CSF) was negative. Schirmer test was found positive. Minor salivary gland biopsy showed nonspecific findings. The patient was diagnosed with primary Sjörgen syndrome by the rheumatology clinic and started on steroid and hydroxychloroquine. In his 3-month follow-up, he reported relative improvement in his symptoms.

Discussion

Sjörgen syndrome is an autoimmune disorder with slow progression (6). It can be seen either as a primary syndrome or secondary to the progression of rheumatoid arthritis and scleroderma. The most prominent symptom is xerophthalmia (keratoconjunctivitis sicca, dry eyes) and xerostomia (dry mouth) (2,6), known as the sicca complex. Our patient showed the symptoms for sicca complex. Musculoskeletal involvement such as arthritis/arthralgia, Reynaud phenomenon, lung, kidney or liver involvement, gastrointestinal, endocrine or nervous system involvement or vasculitis can be the manifestations of an extraglandular involvement (2,6). Minor salivary gland biopsy is used to delineate any salivary gland involvement in SS. Detection of 50 or more perivascular or periductal lymphocytes adjacent to normal acini in the salivary gland biopsy is meaningful for the diagnosis (2,6). In our patient, salivary secretion test showed a rate of 0.5 ml<15 minutes. Salivary gland biopsy showed nonspecific findings. ANA, RF, anti-Ro (SS-A_, anti-La (SS-B) antibodies can be tested for (2,6). Our patient was positive for ANA and anti-Ro antibodies. Despite the fact that different diagnostic criteria were suggested to this day, there has been a consensus on the utility of the revised Europe criteria, also known as the American-European criteria (2). These diagnostic criteria include (7,8):

1. Dry eyes (xerophthalmia)
2. Dry mouth (xerostomia)
3. Positive Schirmer test or Rose Bengal test

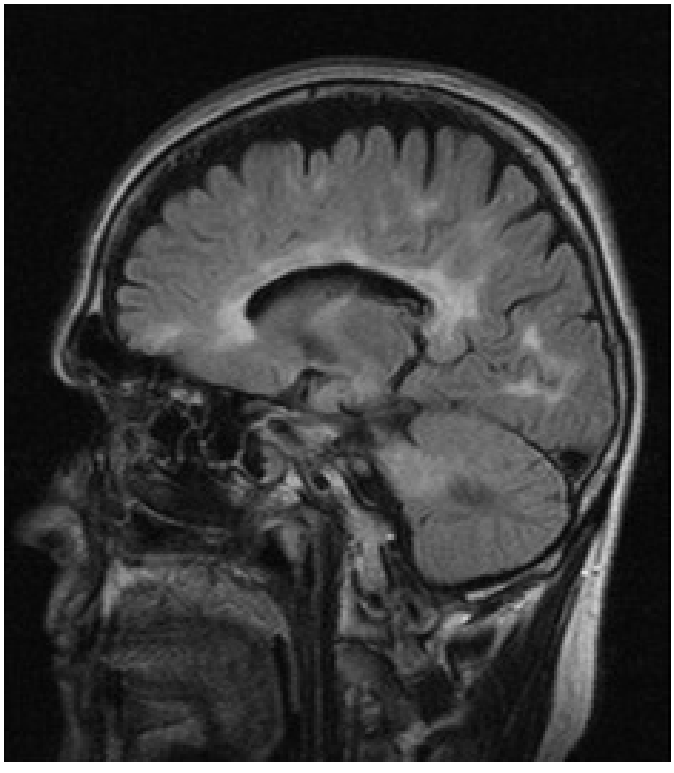

Figure 1. Wide-spread cortical and subcortical hyperintense lesions

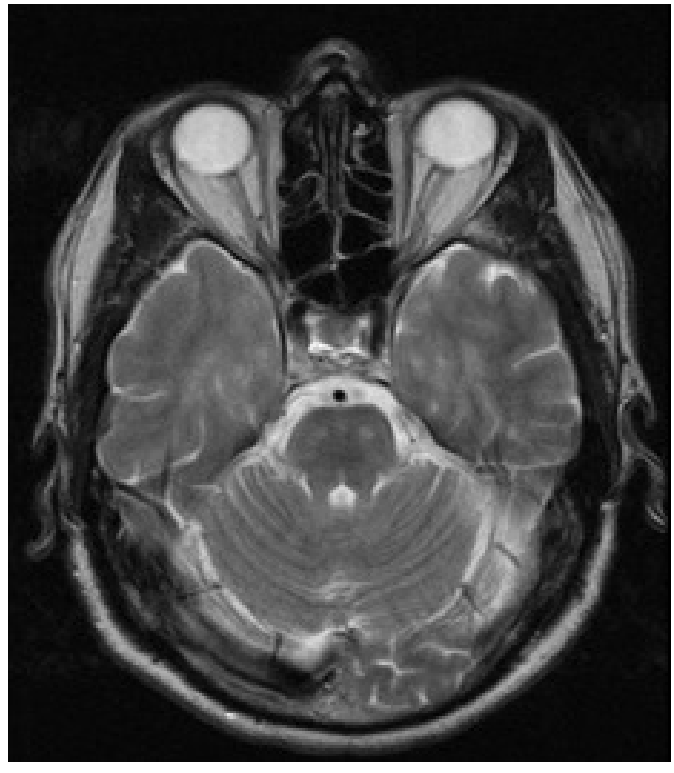

Figure 2. Hyperintense lesions in the brainstem

4. Presence of diagnostic histopathological findings in salivary gland biopsy

5. Other specific salivary gland anomalies (salivary gland scintigraphy, parotid scintigraphy)

6. Presence of Anti-Ro (SS-A), Anti-La (SS-B) antibodies.

Without any other associated disease, presence of 4 out of 6 or more of these criteria is an indicator of SS. Our patient was diagnosed with SS after showing 4 of these signs. Neurologically, cranial nerves, peripheral nerves and CNS can be involved (9-11). Neurological findings are observed in 20-25% of the patients (4). Optic neuropathy, acute or chronic myelinopathy, neuropathy, MS-like symptoms, encephalopathy, seizures or trigeminal, facial and cochlear nerve involvement are among some of the conditions that could be encountered (4,10,11). Our case presented with optic neuropathy and cerebellar findings. Magnetic resonance imaging is extremely useful in revealing the extent of cerebral dysfunction in SS patients. Most commonly, periventricular or subcortical white matter lesions are seen in T2 images (70-80%). In addition, demyelination-like hyperintensities that are suggestive of MS, sulci enlargement, ventricular dilation and rarely lesions of corpus callosum are seen. These MRI pathologies are seen in 80% of the SS patients with CNS involvement who show neurological signs (12,13). Our case also had lesions of the same type. Since these lesions can also be observed in vascular syndromes, vasculitis markers and thrombosis facilitating factors were evaluated and ruled out in the diagnosis. There is no wide-spread agreement on the treatment of SS cases with CNS involvement but intravenous pulse corticosteroid and cyclophosphamide are used in the treatment of progressive clinical neurological dysfunctions (14). We used pulse corticosteroid and observed visible improvement in the clinical findings. The clinical and cranial MRI findings in our case motivated the need for differential diagnosis with MS. Moreover, co-occurrence of MS and SS should be taken into consideration. Clinical and radiological findings that are suggestive of MS can also be observed in SS. The negative result of OCB test, dry eyes and mouth, ANA and Anti-Ro antibodies and Schirmer test positivity lead to the conclusion of SS with CNS involvement. In such cases, early diagnosis and proper treatment approach for SS carries crucial importance for the prognosis of the disease.

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