PRİMER SJÖGREN SENDROMU OLAN HASTADA EROZİV ARTRİT: BİR OLGU SUNUMU

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ÖZET

Primer Sjögren sendromu, sıklıkla ağız ve göz kuruluğu ile seyreden bir kronik otoimmun ekzokrinopatidir. Ağız içi bezleri ve göz yaşı bezleri dışında, nadir de olsa diğer ekzokrin bezler de etkilenebilir. En sık kas-iskelet sistem tutuluşu ile karşımıza çıkmaktadır. Artralji, sabah tutukluğu ve romatoid artrite benzer kronik inflamatuvar poliartrit, eklem bulgularını oluşturmaktadır. Romatoid artrit'ten farklı olarak, Sjögren sendromunda sabah tutukluğu ve hareket kısıtlığı daha hafif olup, el ve el bilek deformasyonları görülmektedir. Romatoid artrit'ten ayıran en önemli özellik ise, direk grafi ve/veya magnetik rezonans görüntülerde, eklemlerde eroziv değişikliklerin olmamasıdır. Bu bildiride, primer Sjögren sendromu tanısı almış hastada, eroziv artrit rapor edilmiştir.

Anahtar sözcükler: Primer Sjögren sendromu, eroziv artrit, magnetik rezonans görüntüleme

Erosive arthritis in a Patient with Primary Sjogren's Syndrome: A Case Report

SUMMARY

Primary Sjogren's syndrome (SS) is an autoimmune exocrinopathy characterized by dry eyes and dry mouth. Exocrine glands other than salivary and lacrimal glands may be affected less frequently. The most common mode of presentation is musculoskeletal system involvement. Articular signs and symptoms include arthralgias, morning stiffness, and chronic polyarthritis that resemble those seen in rheumatoid arthritis (RA). Compared with RA, the arthritis tends to be more relapsing and remitting, and stiffness is less marked. The distinction from RA is that, there is not any erosive changes neither on direct radiography nor magnetic resonance imaging (MRI). We report a patient of primary SS presented with erosive arthritis.

Key words: Primary Sjogren's syndrome, erosive arthritis,magnetic resonance imaging

Sjogren's syndrome (SS) is a chronic, autoimmune disease, characterized by inflammation of the exocrine glands, leading to impaired function. SS is manifested by keratoconjunctivitis sicca (dry eye), xerostomia (dry mouth) and other extraglanduler abnormalities [1] Primary SS is defined as the presence of the above manifestations without additional connective tissue diseases. Secondary SS involves an association with rheumatoid arthritis (RA), systemic lupus erythematosus (SLE) or other connective tissue diseases [2]. Systemic manifestations are seen frequently in SS patients and may include both general constitutional symptoms such as fatique, myalgias and arthralgias, as well as other organ involvement. Arthritis occurs in about 30% of patients with SS, and it may resemble the arthritis of RA. Compared with RA, the arthritis tends to be more relapsing and remitting, and stiffness is less marked. They are often rheumatoid factor (RF) positive, many are initially misdiagnosed as having RA [3]. In contrast to RA, radiographs of the hands usually do not reveal erosive changes. We report a case of primary SS presented with erosive arthritis.

CASE REPORT

A 47-year-old female admitted to our Rheumatology Department for complaints of dry eyes, dry mouth, arthralgias, arthritis and morning stiffness persisting for 5 years. She described joint pain and morning stiffness of more than one hour, particularly at bilateral wrist, metacarpophalangeal (MCP), proximal interphalangeal (PIP), elbow joints and swelling in 2nd, 3rd MCP joint of the right hand. In her past medical history, she had been diagnosed as primary biliary cirrhosis (PBC) five years ago and had been given treatment with ursodeoxycholic acid. On physical examination, she had tenderness in MCP, PIP joints of both hands and sweling in 2nd, 3rd MCP joints of the right hand. Laboratory results included: polyclonal hypergammaglobulinemia on serum protein electrophoresis; positivity of IgM RF (RF:243IU, N<40). Erythrocyte sedimentation rate (ESR) was 110/h and C-reactive protein (CRP) was 12,04mg/dl (N<0,5 mg/dl). She had normochromic and normocytic anemia with hemoglobine of 11.2 g/dl. Serum alkaline phosphatase was 310 U/l (N<260 U/l), gama glutamyl transpeptidase was 56 U/l (N<32 U/l) whereas aspartate and alanine transaminase levels

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were normal. Urinalysis was also normal. Serologic tests for hepatitis B surface antigen and antibody for hepatitis C were negative. Abnormal thyroid hormone levels were found: free T3: 2,6 pg/ml (N:1.8-4.2 pg/ml), free T4: 0,3 pg/ml (N:0.8-1.8 pg/ml), thyroid stimulating hormone (TSH): 19,4 IU/ml (N:0.4-4.0 IU/ml). Anti-thyroglobuline antibody was positive at 1:1280 titer and anti-microsomal antibody was positive at 1:6400 titer. Thyroid ultrasonography showed the presence of diffuse heterogen goiter. These findings were consistent with Hashimoto's thyroiditis. The immunological investigation showed the positivity of antinuclear antibodies (ANA) as 1:5120 cytoplasmic and 1:10240 granular pattern, anti-mitochondrial antibody (AMA) at a titer of 1:1280, anti-Ro antibody at titer 304 U/ml (N<180 U/ml). Assays for anti-dsDNA, Critidia, C3, C4, anti-La, cryoglobuline, LKM, GPC, ASMA, and anti-cyclic cytrulinated peptid antibody (anti-CCP) were negative. Ophtalmologic examination was performed: Schirmer test was 5 mm in both eyes., tear break-up time was 5 sec./8 sec. Sialometry was 0.5ml/15min (normal>1.5 ml/15min.) Minor salivary gland biopsy was performed and grade 4 sialoadenitis was found according to Chisholm grading criteria (4). These results proved that the patient had primary Sjögren's syndrome according to American-European criteria [5] in addition to the diagnosis of PBC and Hashimoto thyroiditis.

Chest X-ray and the pulmonary function tests were normal. Radiographs of the hands showed suspected erosion on right hand 3rd MCP joint. A magnetic resonance imaging (MRI) study of the wrist and both hands was obtained (Figure 1). Erosive changes of the 3rd metacarpal head and synovitis on 3rd MCP joint of the right hand was reported. The patient was prescribed prednol 4mg/day, methotrexate 10mg/week, folic acide 5mg/day, hydroxychloroquine 200 mg/day, NSAIDs and tears substitute. At the follow-up control, artralgias, arthritis and morning stiffness improved. ESR and CRP decreased to normal values.

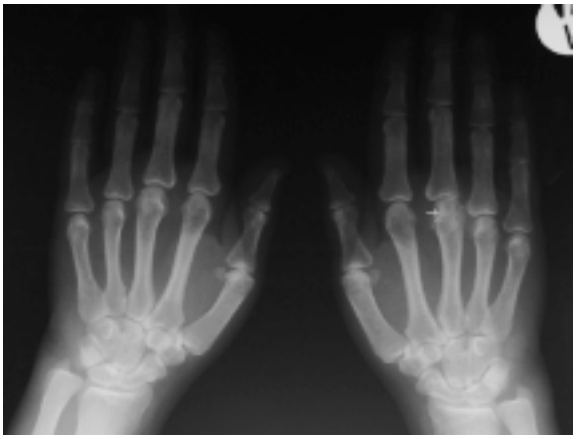

Figure 1. Plain radiography of the hands showed suspicient erosion of the 3rd MCP joint of the right hand.

DISCUSSION

We report a case; a female with primary SS overlapping with PBC and Hashimoto thyroiditis who had erosive arthritis demonstrated by MRI. In our current knowledge, there is some anectodal report in literature about erosive arthritis in patients with primary SS. Boutroutry et al. Reported that may be impossible to distinguish between patients with early RA and those with SLE and primary SS by means with MRI [5]. Arthritis occurs in about 30% of patients with SS, and it may clinically resemble the arthritis of RA. In contrast to RA, radiographs of the hands usually do not reveal erosive changes. In our patient, direct radiographic examination of hands showed suspicient erosive lesion. We performed MRI and demonstrated the erosive changes on right hand 3rd MCP joint unusually seen in primary SS patients.

Erosive disease on imaging is usually consistent with RA. Our patient might be diagnosed as overlap of RA and secondary SS rather than primary SS with erosive arthritis. According to the revised American-European criteria, to define a patient as secondary SS, anti-Ro and/or anti-La positivity is not needed [6]. Our patient had high titers of ANA and anti-Ro positivity mostly indicative of primary SS. If this was a RA and secondary SS overlap, more severe erosive changes on hands would be expected in such a patient with five years history of arthritis, high titers of RF and who had not taken any DMARDs therapies. RF is a predictor factor for more erosive and agressive disease in RA . It is also well known that erosive changes generally occur in the first two years of RA. Anti-CCP antibody was found negative in our patient. The specificity of anti-CCP antibody for RA was shown to be higher than 90% in several studies [7]. The diagnostic sensitivity of the test varied between 41-87.6%. In conclusion,Our patient is a rare case of primary SS with erosive arthritis reported in literature. The distinction between primary SS and RA/secondary SS is critical in these patients for the determination of the follow-up and treatment strategy.

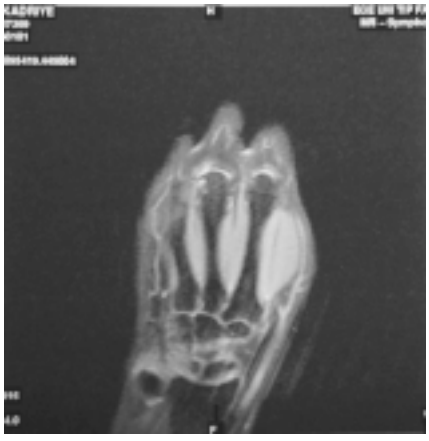

Figure 2. MRI of the right hand showed erosive changes of the 3rd metacarpal head and synovitis on 3rd MCP joint.

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