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Sjögren's Syndrome Presenting as Remitting Seronegative Symmetric Synovitis with Pitting Edema (RS_3PE)

Remitting seronegative symmetric synovitis with pitting edema (RS_3PE) syndrome is characterized by symmetrical and acute synovitis, pitting edema, the absence of rheumatoid factor, increased acute phase reactants, lack of bony erosions on radiography, and benign and short clinical course. Half of all patients with Sjögren's syndrome experience arthritis during the disease course. We here describe the first case of Sjögren's syndrome presenting as RS_3PE. She had swelling in knees, ankles, and wrists. After then the swelling spread to her lower legs, feet, face, and both hands. She was admitted to another hospital and was suspected of lupus or rheumatoid arthritis. Three months later, she had dry mouth and had lower lip biopsy. She was admitted to this hospital due to development of swelling in face and lower legs for 3 days. On physical examination, she had pitting edema in both hands and feet dorsum. Laboratory test showed elevated erythrocyte sedimentation rate, positivity of rheumatoid factor, anti-nuclear antibody, and anti-Ro antibody. There was no erosion in the hands radiography. Schirmer's test and lip biopsy was compatible with Sjögren's syndrome. She was diagnosed RS_3PE and Sjögren's syndrome. She was begun with prednisolone and her symptoms improved gradually.

Key Words : *Sjogren's Syndrome; Synovitis; Edema; RS_3PE*

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INTRODUCTION

In 1985, McCarty and colleagues reported a series of patients characterized by symmetrical and acute synovitis, marked pitting edema, the absence of rheumatoid factor (RF), increased acute phase reactants, lack of bony erosions on radiography, and benign and short clinical course (remitting seronegative symmetric synovitis with pitting edema, RS_3PE syndrome) (1, 2). Since this original description, numerous additional cases have been reported. Pitting edema, one of the main characteristics of RS_3PE, may also be noted in connective tissue disorders, such as systemic sclerosis, polymyositis, and mixed connective tissue disease (MCTD) (3). During the course of Sjögren's syndrome, 50% of all patients experience episodes of arthritis. Articular symptoms and signs include arthralgia, morning stiffness, intermittent synovitis and chronic polyarthritis (4). However, there has been no report of RS_3PE in Sjögren's syndrome until now, and we herein describe the first case of Sjögren's syndrome presenting as RS_3PE.

CASE REPORT

A 35-yr-old woman was admitted because of swelling in the face and lower legs in April 2001. She had been healthy until the previous December when she developed fever, swelling, and pain in both knees, ankles, and wrist joints. Then the swelling spread to her lower legs, feet, face, and hands. The swelling was worse in the morning and after walking. She experienced intermittent pain in the knees, ankles, and wrists for a few days. The morning stiffness lasted less than 30 min. She was admitted to another hospital and was suspected to have lupus or rheumatoid arthritis. In March 2001, she developed dry mouth and underwent lower lip biopsy. Three days prior to admission in April, the swelling in her face and lower legs was aggravated. Her past medical history included artificial fertilization in 1996, but she denied oral ulcer, photosensitivity, alopecia, dry eye, or Raynaud's phenomenon. She had experienced intermittent lumbar pain for the previous few years. On physical examination, she had a puffy face but no swelling or tenderness in the parotid gland. She had swelling and pitting edema in her hands (Fig. 1A), lower legs, and the dorsum of the feet (Fig. 1B), but there was no associated tenderness. However, she did have swelling in both wrist and left 2nd proximal interphalangeal joint and tenderness in both metacarpophalangeal joints and the right knee joint. There was no tenderness in the lumbar spine or buttock. The straight leg raising test was negative. Extremity muscle power was intact. Laboratory tests gave a white cell count of 4,330/μL, hemoglobin 11 g/dL, platelet 172,000/μL, Westergren erythrocyte sedimentation rate 27 mm/hr (reference range 0-9), serum protein 5.9 g/dL, albumin 2.9 g/dL, aspartate aminotransferase/

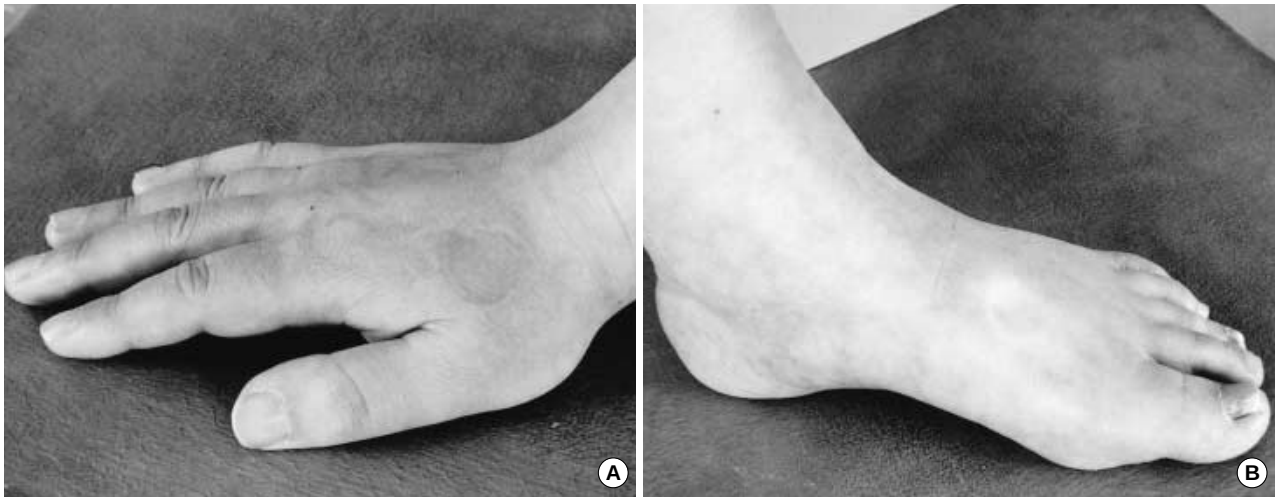

Fig. 1. Swelling and pitting edema of both hands (A) and the dorsum of the feet (B).

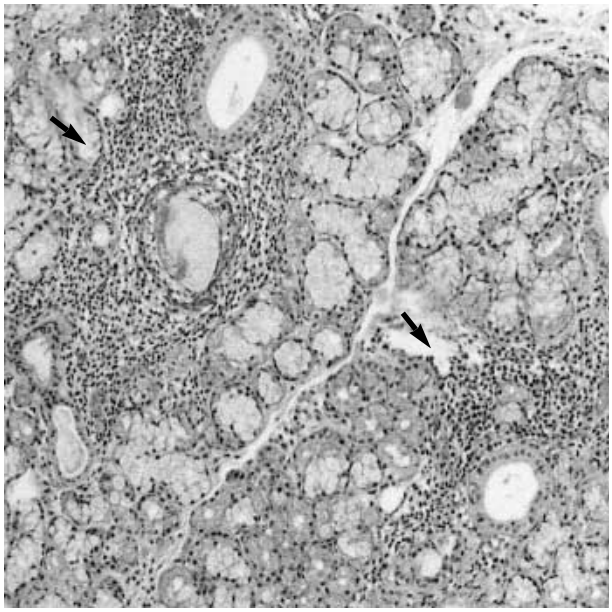

Fig. 2. Minor salivary gland biopsy showing moderate lymphocytes infiltration and 2 foci (arrows) of more than 50 lymphocytes (H&E, ×100).

alanine aminotransferase 27/13 IU/L (reference range <40/<40), and creatinine 0.8 mg/dL. Urine analysis was normal. Serologic tests indicated RF 54.4 IU/mL (reference range 10-30), antinuclear antibody (ANA) 1:320 in a speckled pattern, positive anti-Ro antibody, and weakly positive anti-ribonucleoprotein (RNP) antibody. The tests were negative for anti-La antibody, anti-ds DNA antibody, HLA-B7 and B27. Thyroid function test was normal. There were no abnormal radiography findings in the hands, lumbar spine and pelvis. Bone scintigraphy revealed the absence of any active lesion of bone or joint. Minor salivary gland biopsy showed moderate lymphocytes infiltration and two foci of more than 50 lymphocytes infiltration (Fig. 2). The result of Schirmer's test was compatible with Sjögren's syndrome (<5/5 min in both eyes). Therefore, the patient was diagnosed with Sjögren's syndrome presenting as RS3PE and she was given prednisolone at 20 mg/day. Her symptoms showed gradual improvement and 5 months after discharge, the prednisolone treatment was tapered to 10 mg/day.

DISCUSSION

The present case exhibited the characteristic features of the RS3PE syndrome: acute symmetrical polysynovitis with pitting edema of the dorsum of the hands and feet and negative radiologic finding. However, the presence of RS3PE syndrome is rare in young woman and its association with rheumatoid factor positivity is also uncommon. Although the presence of HLA-B7 has been noted in 9-75% of RS3PE cases (5, 6), HLA-B7 was negative in the present case. We suspect that the presence of autoantibodies such as ANA and RF is a systemic autoimmune process of Sjögren's syndrome. Because her arthritis was not present for at least 6 weeks and morning stiffness did not last for at least 1 hr before maximal improvement, she was not satisfied with the criteria for the classification of rheumatoid arthritis. Differential diagnoses in this patient included spondyloarthropathies (7, 8), systemic sclerosis, mixed connective tissue disease (MCTD), and systemic lupus erythematosus (3). She did not complain of inflammatory back pain. Her pelvis AP radiography and bone scintigraphy showed normal findings. Although she presented hands edema and anti-RNP antibody, which might suggest MCTD, she did not exhibit myositis, Raynaud's phenomenon, or acrosclerosis. The titer of anti-RNP antibody was low for the criteria of MCTD. The patient did not have clinical features of lupus, except ANA positivity. In Sjögren's syndrome, 50% of patients experience

episodes of arthritis during the disease course. Articular symptoms and signs include arthralgia, morning stiffness, intermittent synovitis, and chronic polyarthritis. Joint radiography of the hands usually do not reveal erosive changes (4, 9). There has been no previously published report of RS_3PE in a patient with Sjögren's syndrome. Many authors have reported that RS_3PE appears to be an early manifestation of rheumatic diseases because the underlying diseases sometimes require several years to manifest and need to be followed for a long time (10, 11). In the present case, RS_3PE might have occurred incidentally or as an early manifestation of Sjögren's syndrome. In summary, we herein described the first case of Sjögren's syndrome presenting as RS_3PE. It is suggested that the possibility of Sjögren's syndrome should be taken into consideration for the differential diagnosis of patients with RS_3PE.

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