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LETTER TO THE EDITOR

Sarcoidosis and Sjögren's syndrome: Mimicry or coexistence?

Dear Editor,

Sarcoidosis and Sjögren's syndrome (SS) are both multisystemic inflammatory diseases of unknown etiology. Although lungs and the tracheobronchial tree are the most common involvement sites in sarcoidosis it rarely affects the salivary and lacrimal glands. By contrast, SS mainly affects the exocrine glands, but it may also have systemic manifestations. Occasionally some features of sarcoidosis may mimic SS when parotid and/or lacrimal gland involvement is present. Although they are different clinical entities, there are several reports suggestive of sarcoidosis and coexisting SS in the literature [1,2].

A 58-year-old Caucasian woman presented with dry cough and exertional dyspnea. She had a 6-month history of dry mouth and dry eyes. There were no symptoms of arthralgia, myalgia, muscle weakness, rash, photosensitivity, oral ulcers, sclerosis, or Raynaud phenomenon. Physical examination was unremarkable except dryness of oral mucosa and bilateral basal crackles. Laboratory examinations revealed increased erythrocyte sedimentation rate and C-reactive protein. Routine hematological laboratory and blood chemistry values were normal. Chest radiograph showed bilateral hilar lymphadenopathy and lower zone reticular infiltrates. Chest computed tomography revealed multiple mediastinal and bilateral hilar lymphadenopathies in addition to bilateral pulmonary nodules with peribronchovascular involvement (Figure 1A, B). A mild restrictive ventilatory defect and mild reduction in carbon monoxide diffusion capacity was observed on pulmonary function tests. She underwent bronchoscopy. A transbronchial fine needle aspiration biopsy from the right lower paratracheal station revealed noncaseating granulomatous inflammation (Figure 1C). Serum angiotensin converting enzyme level was elevated remarkably. Antinuclear antibody and antibodies against SS-A/Ro were positive. Rheumatoid factor, and specific antinuclear antibodies against double stranded DNA, DNA topoisomerase-1, Smith, U1 ribonucleoprotein, Jo-1, and SS-B/La autoantigens were negative. Bilateral Schirmer's tear test yielded positive. Histopathological examination of minor salivary glands showed periductal lymphocytic infiltration (Figure 1D). The patient was diagnosed with coexisting SS and sarcoidosis. Treatment with prednisolon and hydroxychloroquine were started. During a 2-year follow-up, clinical symptoms and laboratory findings normalized. Radiographic findings regressed. Corticosteroids were tapered and stopped. Until now, the patient has been treated with hydroxychloroquine with no major symptoms and findings.

Our patient was considered as sarcoidosis on the basis of compatible clinical and radiological findings, and histopathological verification of noncaseating granulomas in mediastinal lymph node biopsy (Figure 1C). Elevated serum angiotensin converting enzyme also supported sarcoidosis diagnosis. By contrast, diagnosis of SS was based on dry eye and dry mouth symptoms, positive bilateral Schirmer's test, and the presence of characteristic autoantibody (anti-Ro/SS-A), in addition to evident lymphocytic sialoadenitis in salivary gland biopsy [3]. SS can present either alone as primary SS or secondary to an underlying connective tissue disease. In our patient, symptoms of articular involvement, rash, photosensitivity, oral ulcers, sclerosis, Raynaud phenomenon, myalgia, and muscle weakness were absent. Specific antinuclear antibodies except anti-Ro/SS-A were negative. Signs, symptoms, and laboratory findings suggestive of renal and hematological disorders were absent. In this context, rheumatoid arthritis, systemic lupus erythematosus, systemic sclerosis, mixed connective tissue disease, dermatomyositis, and polymyositis were excluded.

Sarcoidosis and SS share similar immunologic and histopathologic features. Making a differential diagnosis between these two diseases may be difficult in cases that present with common clinical features [4]. By contrast, it was shown that the frequency of sarcoidosis is higher in patients with SS than in the general population [2]. Immunologic profile with a high Antinuclear antibody,

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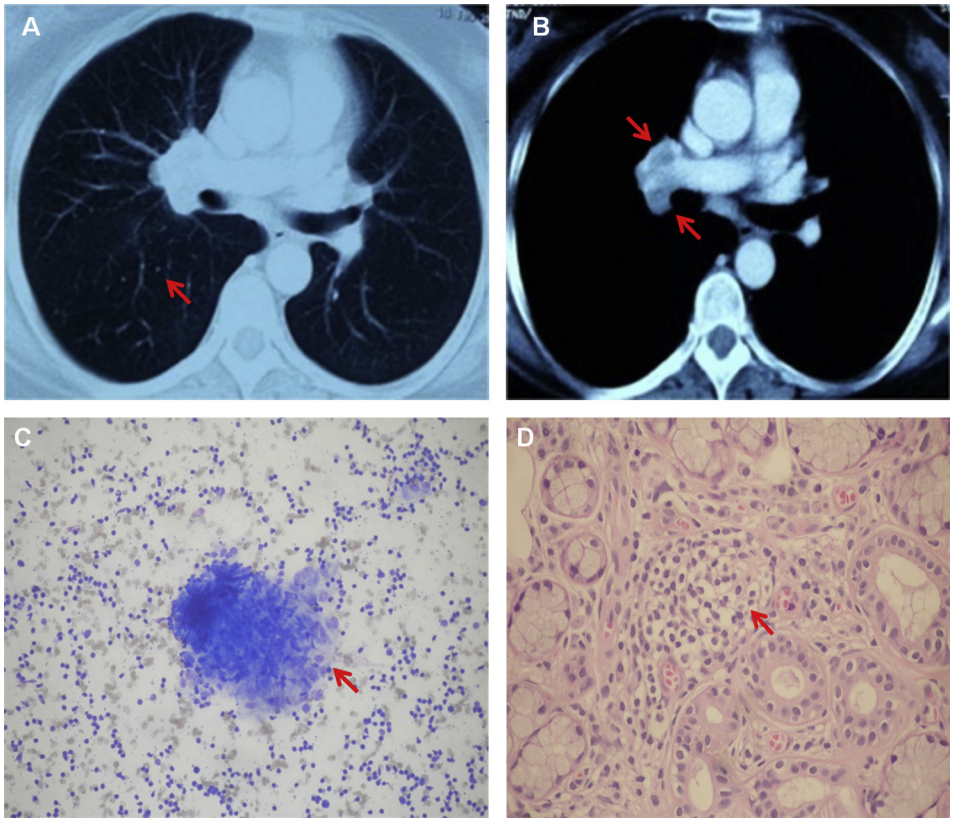

Figure 1. (A,B) Bilateral hilar lymphadenopathies and pulmonary nodules with peribronchovascular involvement on chest computed tomography. (C) Noncaseating granulomas in the paratracheal lymph node; transbronchial fine-needle aspiration, hematoxylin and eosin stain, original magnification ×100. (D) The connective tissue around the ducts is heavily infiltrated with lymphocytes, minor salivary glands, hematoxylin and eosin stain, original magnification ×100.

Rheumatoid factor, and anti-Ro/SS-A was shown to have an important role in differentiating between sarcoidosis mimicry or coexistence with SS in addition to biopsy.

We emphasize that both sarcoidosis and SS may coexist, as illustrated in our case. Analysis of autoantibodies may be very helpful in making differential diagnosis or deciding their coexistence. Additionally, the salivary gland biopsy may be highly discriminative in distinguishing between SS (focal sialadenitis) and sarcoidosis (granulomatous inflammation) [5]. Currently, the acceptance of sarcoidosis as an exclusion criterion for SS diagnosis may lead to underestimation of SS in these cases. A revision for exclusion criteria of SS diagnosis might therefore be necessary.

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