Pulmonary Involvement in a Patient with Secondary Sjögren’s Syndrome Due to Progressive Systemic Sclerosis

Münire GÖKIRMAK*, Oya KAYACAN**, Gülten DURMAZ**

* Department of Pulmoner Secondary Turgut Özal Medical Center, University of İnönü, MALATYA
** Department of Pulmoner Secondary Medical Faculty, University of Ankara, ANKARA

SUMMARY
Sjögren’s syndrome (SS) may occur as a distinct entity (primary SS), or in association with other rheumatic diseases (secondary SS). A 56 year-old housewife with progressive systemic sclerosis and secondary SS had also interstitial lung changes. The patient complained of dry eyes and dry mouth for years. A chest X ray revealed a reticulonodular pattern. The pulmonary involvement may be due to secondary SS or PSS, however it is not possible to establish this fact in the patients like the one presented here.

Key Words: Sjögren’s syndrome, progressive systemic sclerosis, interstitial lung disease.

ÖZET
Progresif Sistemik Skleroz ve Sjögren Sendromu Olan Bir Hastada Akciğer Tutulumu
Sjögren sendromu (SS) aynı bir antite olarak (primer SS) veya diğer romatizmal hastalıklarla birliktede (sekonder SS) ortaya çıkabilir. Progresif sistemik sklerozu (PSS) ve sekonder SS’u olan en esansımsız hastamızda aynı zamanda akciğerde interstisyal değişiklikler saptandı. Hasta yıllardır gözlerinde ve ağzında kuruluktan yakınmaktaydı. Akciğer grafisinde retikulonodüler patern mevcuttu. Pulmoner tutulumun sekonder SS’a veya PSS’a sekonder olabileceğini düşünüldü, ancak burada sızılan hastanın olduğu gibi böyle hastalarda bu durumu aydınlatmak mümkün olamaktadır.

Anahtar Kelimeler: Sjögren sendromu, progresif sistemik skleroz, interstisyal akciğer hastalığı.

Sjögren’s syndrome (SS) is an autoimmune disease characterized by exocrine gland involvement, particularly the lacrimal and salivary glands. It is known to occur as a distinct entity (primary SS) or in association with other rheumatic diseases (secondary SS), mainly rheumatoid arthritis (RA). The coexistence of SS and progressive systemic sclerosis (PSS) has been suggested by individual case reports, and the relationship of this disorder to scleroderma is investigated. However, because of the lack of well defined criteria for the diagnosis of SS, its prevalence in PSS has been reported to vary from as low as 1-2% to as high as 90% (1).
CASE REPORT

A 56 year-old housewife was admitted to an eye specialist with the complaints of dry eyes and burning sensation, after which she was evaluated in the immunology department. Since the patient’s radiological examination revealed a reticulonodular pattern, she was referred to our department of chest diseases and tuberculosis.

The patient’s complaints began 25 years ago, when hyperpigmentation occurred on her face and thoracic skin. She had had Raynaud’s phenomenon for years. She had caries of teeth during the second decade and began to use dental prothesis in her thirties. She had episodic parotid gland enlargement. She reported an inability to cry on emotion. During the last five years, she had difficulty on swallowing, especially the dry food. In addition she lost the sensation of smell and taste almost totally.

Twenty years ago the patient had undergone a partial gastrectomy.

On physical examination, she was afebrile and normotensive. Her radial pulse was 74/min, and arrhythmic. She had telangiectasia on her face. She couldn’t open her mouth totally and had angular cheilitis bilaterally. The skin of the extremities were dry and thick. There were flexion contractures on the joints of the hand and her fingers were edematous. There were bilateral rales on chest auscultation. The examination of other systems were normal.

The hematological, blood chemical and urine laboratory values were normal. An ECG showed occasional ventricular premature contractions. PPD was found to be 22 mm. ASO was negative, CRP and RF were found ++++. Shirmer test was positive. ANA was found to be positive with a homogeneous granular pattern while anti-dsDNA was negative. Serum immunoelectrophoresis disclosed an increase in IgG and IgA; IgM and complement levels were within normal limits. Anti-Scl 70 was negative.

The pulmonary function tests and arterial blood gases were within the normal limits while CO diffusion capacity was 52% of predicted.

DISCUSSION

Primary SS is associated with multiple autoantibodies, as anti-SSA and anti-SSB. It may have extraglandular manifestations in skin, vessels and the visceral organs.
The secondary SS which was also found in our patient who had an underlying PSS, does not differ histopathologically from the primary form. Alexandros et al compared the patients who had secondary SS with the CREST variant of PSS with patients with primary SS. The only difference was the more frequent enlargement of parotid gland in the secondary SS group (2). Our case also had occasional enlargement of parotid glands.

Secondary SS is seen primarily due to RA. But SLE, PSS, primary biliary cirrhosis, chronic active hepatitis, mixed cryoglobulinemia, hypergammaglobulinemic purpura, autoimmune thyroiditis may all cause secondary SS. The frequency of secondary SS due to PSS varies in different reports due to variable diagnostic criteria. Rasker et al, sought features of SS in 26 patients with PSS and in age and sex-matched control subjects. They found only one patient which showed the complete picture of SS with clinical and laboratory evidence of lacrimal and salivary gland involvement. But a number of patients and control subjects had various individual symptoms and signs of lacrimal and salivary disorders. Therefore they suggested that the association of SS with PSS is far less common than previously suggested (3). Alexandros et al reported 20.5% of the prevalence of SS in patients with PSS. In contrast to Rasker et al, they suggested that SS is common in PSS although the patients may lack prominent exocrine gland symptoms (1).

Primary or secondary SS both may cause multiple symptoms due to lacrimal and salivary gland involvement. Our patient had the complaints of dry eyes, burning sensation, inability of crying on emotion showing the lacrimal gland involvement. Shirmer test which is necessary for the diagnosis of involvement of eye in SS was found positive. The complaints of dry mouth, difficulty in swallowing, change of taste and episodic gland enlargement with the usage of dental prostheses indicated the involvement of salivary glands. The biopsy of labial salivary glands was in accordance with SS.

The pulmonary involvement in primary or secondary SS may occur as a lymphocytic alveolitis, lymphoid interstitial pneumonitis, interstitial fibrosis, pleuritis, pseudolymphoma or malignant pulmonary lymphoma (4-6). Secondary pulmonary hypertension may also be seen (7).

Vitali et al compared the lung involvement of SS between the patients with primary or secondary SS. They suggested that the lung involvement was more frequent and severe in patients with secondary form of the syndrome. They also found that in both forms, the lung function changes did not correlate with other clinical and serological parameters, except for a more severe impairment of diffusion capacity in patients with Raynaud’s phenomenon (6).

In patients like the one presented here, it is not possible to establish whether the pulmonary involvement is secondary to SS or PSS. PSS may also affect the respiratory system through various mechanisms. Dyspnea, the major symptom of lung involvement, may not be found in patients with objective evidence of lung disease (8). Our patient did neither have prominent respiratory symptoms despite evidence of an interstitial disease.

Lung involvement in PSS may be due to a variety of nonpulmonary etiologies. These include skin thickening of the thorax, scleromatous fibrosis of the diaphragm, and chronic aspiration from esophageal dysmotility and reflux. However, intrinsic lung abnormalities appear to be more common. Pulmonary fibrosis is the most common pathologic finding in the lung with the bases most strikingly involved. Pleural fibrosis and pulmonary hypertension may also be found (8).

Our case had an interstitial pattern with a decrease of CO diffusion capacity. Alveolitis may be the first step of this kind of pulmonary fibrotic process, as is hypothesised for other interstitial lung diseases. Gallium scanning, a noninvasive technique of monitoring alveolitis, was abnormal in more than half of the systemic sclerosis patients studied (8). Bronchoalveolar lavage, another method of evaluating alveolitis in interstitial lung diseases has been reported in a number of studies in systemic sclerosis. Generally BAL results have not correlated particularly well with
the degree of pulmonary involvement except for an association of neutrophils and macrophages with decreased DLCO (8). Silver et al performed BAL in 43 nonsmoking patients with systemic sclerosis and 49% of the patients were found to have alveolitis with an absolute increase in alveolar macrophages and granulocytes. They also found that, patients with persistent alveolitis had significantly greater reduction in pulmonary function over time than patients without alveolitis (9). The differential cytology of BAL did not reveal alveolitis in our patient.

In a report of İmecik et al, two patients with PSS and an interstitial pattern were investigated. One of the patients had also squamous cell cancer of the lung (10).

The majority of patients with pulmonary fibrosis do not have progressive deterioration, and therapeutic interventions may not be necessary. In patients with deteriorating disease, corticosteroids may be useful, especially when they are given during the alveolitis stage (8,9). The patients with deteriorating pulmonary symptoms and lung functions who have no evidence of alveolitis may be a candidate for D-penicillamine therapy, although dramatic improvement in severe restrictive disease should not be anticipated (8). Since our patient did not have prominent respiratory symptoms and no deterioration was recognized on follow-up examinations, she was instituted neither corticosteroids nor D-penicillamine therapy.

Pulmonary hypertension due to PSS has been attempted to be ameliorated by vasodilators, e.g. nifedipine. There are numerous reports suggesting the effectiveness of these drugs, either in the catheterization lab or transiently in the clinical setting. However, no study has shown a significant alteration in the dismal downhill course of pulmonary hypertension in PSS (8). Although it was not possible to perform right heart catheterization in our patient, nifedipine was instituted in order to relieve the symptoms of Raynaud’s phenomenon.

REFERENCES


Address for Correspondence:
Münire GÖKIRMAK, MD
Department of Pulmoner Secondary
Turgut Özal Medical Center
University of İnönü
44069, MALATYA