Sjogren’s Syndrome Presenting with Interstitial Lung Disease

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Abstract
We report a patient of primary Sjogren’s syndrome presenting with interstitial lung disease. The clinical picture was dominated by respiratory symptoms leading to a delay in diagnosis.

INTRODUCTION
Sjogren’s syndrome (SS) is an autoimmune disease characterized by sicca symptoms, usually dry eyes and dry mouth. The disease may be primary (idiopathic) or secondary to other diseases like rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), systemic sclerosis (SSc), polymyositis or biliary cirrhosis. Respiratory involvement has been reported in primary SS in 9% to 75% of cases depending on the sensitivity of the variables studied and the criteria used to define the abnormality.1-4 We present here a patient with predominant respiratory symptoms, who on evaluation was found to have primary SS with interstitial lung disease (ILD). The relevant Indian literature has been reviewed.

CASE REPORT
A 52 year lady was referred to our hospital with complaints of fever, generalized weakness and dry cough of 1 year duration. She also complained of increasing breathlessness for the last 6 months. She had been diagnosed as chronic obstructive pulmonary disease and treated with bronchodilators without much relief. On examination, she was found to be dyspnoeic. Her respiratory rate was 26 per minute and pulse rate 92 per minute. She was normotensive but had pallor and extensive dental caries (Fig. 1). Examination of the respiratory system revealed presence of bilateral basal coarse crepitations suggestive of ILD. Other systems were within normal limits. Seeing her extensive caries, she was questioned about sicca symptoms. On direct questioning, she gave history of recurrent bilateral parotid swelling, decreased salivation and foreign body sensation in the eyes for the last 6 years. She denied any history of joint pains, Raynaud’s phenomenon, malar rash, oral ulcers, alopecia or photosensitivity.

Investigations revealed that her Hb was 11.1g/dl and ESR 40mm fall/1 hour. Her TLC, DLC, platelet count, urinalysis, blood sugar (fasting and post prandial), blood urea, serum creatinine, aminotransferases, serum calcium, phosphate and electrolytes were within normal limits. Schirmer’s test was positive (Right eye-3mm, Left eye-1mm). Her ANA (antinuclear antibody) was positive, speckled (>1:320), RF (rheumatoid factor) was positive (192 IU/ml). Anti –Ro and Anti-La antibodies were not done. Her ECG was normal. Her chest X-ray revealed presence of reticulonodular shadows in both lower zones bilaterally. Her CECT and HRCT chest showed honeycombing and advanced interstitial fibrosis (Fig. 2). Parotid scintigraphy revealed markedly reduced uptake by the gland. Her lip biopsy revealed periductal chronic inflammation with occasional acinar destruction suggestive of Sjogren’s syndrome. A diagnosis of primary SS with ILD was made and the patient managed with oxygen therapy, prednisolone, bronchodilators, artificial tears and dental care.
Respiratory involvement has been reported in both primary and secondary forms of SS. Desiccation of mucosa and changes in the bronchial glands, similar to the pathology in salivary glands, was described by Sjogren way back in 1933. The respiratory involvement in primary SS can be in the form of interstitial lung disease (ILD), small airways disease, xerotrachea, large airway obstruction, lymphoma/pseudolymphoma, bronchiolitis obliterans organizing pneumonia (BOOP) and pleural involvement. Pleural effusion is uncommon in primary SS and is seen more often in secondary SS. Occasionally, a patient may present primarily with respiratory symptoms and primary SS may come to the fore only after further evaluation, as happened in our patient.

A review of 343 patients with classic SS (both primary and secondary) seen at Mayo clinic revealed pulmonary involvement in 31 patients (9%). ILD was seen in 13 of the 31 patients (42%). However, 7 of these patients had concomitant RA making it difficult to ascertain whether the ILD was due to SS or RA. Other studies dealing with primary SS only have reported ILD in 8-25% cases. The involvement of trachea and large airways can result in dry cough. Patients may be wrongly treated for asthma, chronic bronchitis, or even tuberculosis, as happened in our patient who was given a label of chronic obstructive pulmonary disease.

We could come across only 2 series on primary SS from India, one from Lucknow comprising 26 patients and another from Chennai comprising 36 patients. None of the Indian series have reported pulmonary abnormalities, perhaps because of the small cohort size.

Pulmonary lesions, while not being rare in primary SS, often get overlooked. In patients with ILD, taking a history of sicca symptoms may provide a valuable clue to the underlying disease as illustrated in this case report.

REFERENCES