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Interstitial Lung Disease in a Patient with Primary Sjogren's Syndrome.

Sarang Patil[1]

[1]Junior Resident, Department Of Pulmonary Medicine, Dr. D. Y. Patil Medical College, Nerul, Navi Mumbai, Maharashtra, India.

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ABSTRACT

Sjogren's syndrome (SS) is a chronic inflammatory disease characterized by lymphocytic infiltrates in the exocrine glands, mainly the salivary and lacrimal glands. Interstitial lung disease (ILD) can be seen in 3 to 11 % of the patients with primary SS, often leading to life-threatening complications like ventilator failure and pulmonary hypertension. A sixty year old female presented with dry cough, low grade intermittent fever, and dryness of mouth and eyes. On examination, the patient was afebrile and all vitals were within normal limits. Oxygen saturation was at 96%. Breath sounds were heard equally bilaterally and infrascapular fine crepitations were heard on both sides. Pulmonary function testing had a mild restriction pattern and diffusing capacity of the lungs for carbon monoxide was reduced. Rheumatoid factor was raised and Schirmer's test showed wetting of less than 5 mm in 5 minutes. High resolution computed tomography revealed diffuse ground glass opacities and areas of consolidation in lung lobes bilaterally. Anti-nuclear antibody profile was positive for SS-A, SS-B and Ro 52 antigens. Findings of HRCT diagnosed the presence of interstitial lung disease. Symptomatic treatment was given for dry mouth and eyes and psoriasis. Steroid and immunosuppressive therapy was considered for interstitial lung disease. **Conclusion:** ILD in patients with primary SS may have numerous histopathologic patterns. Though evidence based treatment regimes have not been well established, early diagnosis and treatment is associated with improved lung functions and increased survival.

Keywords: Primary Sjögren's syndrome, interstitial lung disease, interstitial pneumonia

INTRODUCTION

Sjogren's syndrome (SS) is a chronic inflammatory disease characterized by lymphocytic infiltrates in the exocrine glands, mainly the salivary and lacrimal glands. It may present as a primary disease (primary SS) or in association with other autoimmune diseases such as rheumatoid arthritis, systemic lupus erythematosus, and systemic sclerosis (secondary SS). Extra-glandular organs like lungs may also be frequently involved and pulmonary involvement has been reported in 9 to 75% of the patients of primary SS.[1] Interstitial lung disease (ILD) can be seen in 3 to 11 % of the patients with primary SS,[2] often leading to life-threatening complications like ventilator failure and pulmonary hypertension. Here we present a case of interstitial lung disease in a patient with primary SS.

CASE REPORT

A sixty year old female presented to the outpatient clinic of our department with dry c ough and low grade intermittent fever for several months. She also complained of dry mouth and eyes since past 10 years and generalized weakness since 6 months. The patient was diagnosed with plaque psoriasis 5 years ago. A cream was prescribed which the patient stopped using after 6 months of diagnosis, without any further follow up. The patient had no history of any major illness, joint pains, infections, smoking or medications. On examination, the patient was afebrile and all vitals were within normal limits. Oxygen saturation was at 96%. No signs of pallor, icterus, cyanosis, clubbing, lymphadenopathy or edema were found. Breath sounds were heard equally bilaterally and infrascapular fine crepitations were heard on both sides. Chest x-ray is shown in [Figure 1]. Pulmonary function testing (PFT) had a mild restriction pattern and diffusing capacity of the lungs for carbon monoxide (DLCO) was reduced. The results of routine investigations like complete blood count, liver function and renal function were within normal limits. Peripheral smear showed normocytic normochromic anaemia without any poikilocytosis. No abnormalities were detected on iron and thyroid profile [Table 1]. Rheumatoid factor was raised (66.3 IU/ml) and Schirmer's test showed wetting of less than 5 mm in 5 minutes [Figure 2]. High resolution computed tomography (HRCT) revealed diffuse ground glass opacities and areas of consolidation in lung lobes bilaterally, which was suggested of nonspecific interstitial pneumonia

Name & Address of Corresponding Author
Dr Sarang Patil
Junior Resident,
Department Of Pulmonary Medicine
Dr. D. Y. Patil Medical College,
Nerul, Navi Mumbai, Maharashtra, India.

(NSIP) pattern. [Figure 3]. Anti-nuclear antibody profile was positive for SS-A, SS-B and Ro 52 antigens [Table 2]. According to the revised version of the European criteria proposed by the American - European Consensus Group the patient was diagnosed with primary Sjogren's syndrome. Findings of HRCT diagnosed the presence of interstitial lung disease. Symptomatic treatment was given for dry mouth and eyes and psoriasis. Steroid and immunosuppressive therapy was considered for interstitial lung disease.

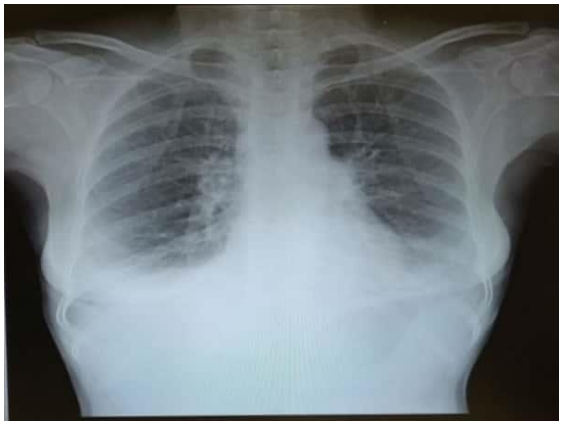

Figure 1: Chest x-ray of the patient

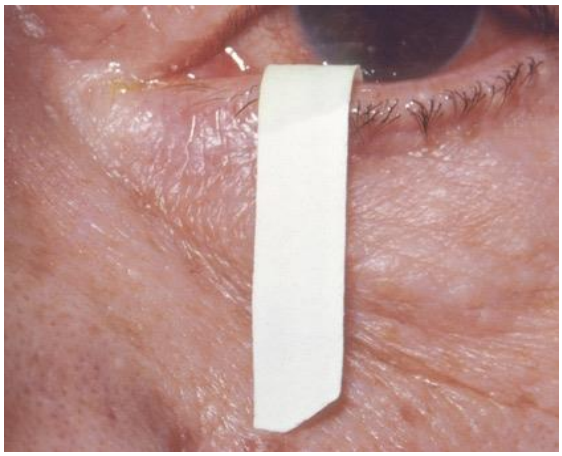

Figure 2: Schirmer's test showed wetting of less than 5 mm in 5 minutes

Figure 3: High resolution computed tomography of lungs showing diffuse ground glass opacities and areas of consolidation in lung lobes bilaterally

Table 1. Routine investigations of the patient

Investigation	Value
Hemoglobin	9.4 mg/dl
Reticulocyte count	0.5%
Thyroid profile	
Free Triiodothyronine	3.1 pg/ml
Free Thyroxine	17.6 ng/dl
Thyroid Stimulating Hormone	12.7 µIU/ml
Iron profile	
Total iron	129.4 µg/dl
Total iron binding capacity (TIBC)	297.3 µg/dl
Unsaturated iron binding capacity (UIBC)	167.9 µg/dl
Ferritin	55.2 ng/mL

Table 2. Antinuclear Antibody panel of the patient

Antigen tested	Observed value
SS-A	Positive
Ro 52	Positive
SS-B	Positive
nRNP/Sm	Negative
Sm	Negative
Scl	Negative
PM-Scl	Negative
Jo-1	Negative
Centromere protein B (CENP-B)	Negative
Proliferating cell nuclear antigen (PCNA)	Negative
Double stranded DNA	Negative
Nucleosome	Negative
Histones	Negative
Ribosomal P protein	Negative
Anti-mitochondrial M2	Negative

DISCUSSION

Interstitial lung disease is not an uncommon finding in SS and the patients are at risk for several different types of ILD, including NSIP, organizing pneumonia, usual interstitial pneumonia (UIP), lymphocytic interstitial pneumonia (LIP), cystic lung disease, and diffuse interstitial amyloidosis Though, lymphocytic interstitial pneumonitis has been classically described to be associated with SS, it may not be the most common subtype of ILD in SS. Lung biopsy results of Japanese Sjögren's patients showed that 61% had nonspecific interstitial pneumonitis, 12% had bronchiolitis and 12% had lymphoma.[4] Similarly high proportion of patients with nonspecific interstitial pneumonitis were shown in a series of Chinese patients by Shi et al and American patients by Parambil et al.,[5,6] There is scarce literature which details on the natural course of interstitial lung disease in patients with SS. Parambil et al reported 39% mortality during a median follow-up of 3 years. In a review of 17 patients with SS, Suda et al observed acute exacerbation in 6% of the patients.[7] A more benign clinical course with a 5-year survival of 83% was observed in the series of Japanese patients. However, it is difficult to reach a definite conclusion from these limited data and further studies are warranted. Although invasive diagnostic tests like bronchoalveolar lavage, transbronchial biopsy or surgical lung biopsy could not be performed in our patient, based on the imaging and clinical features, we suspect either NSIP or LIP to be the most likely histopathological pattern.

The HRCT findings in patients with SS-related pulmonary disease have been described by numerous authors previously.[8] Parambil et al reviewed the HRCT findings in patients of primary SS with ILD and observed a strong correlation with the underlying histopathologic pattern of lungs.[6] Most commonly seen radiological findings are ground glass opacities seen in 45%-92% of the patients and fibrotic honeycomb cysts seen in 13%-43%.[4] Furthermore, authors have suggested that the presence of multifocal cysts on HRCT should raise the clinical suspicion for SS related ILD.

There are no established treatment protocols for ILD in patients with primary SS patients. Previous investigators have hinted that the therapeutic response to steroid may vary with the histological findings in the lungs.[10] Though corticosteroid therapy has classically been the first-line therapy for ILD in pSS patients, Roca et al suggested that early management of alveolitis is necessary before irreversible alveolar-capillary damage occurs. Furthermore, patients who failed corticosteroid therapy have been reported to have a favourable outcomes with immunosuppressive therapy in combination with steroids.[5] Encouraging results have been shown with the use of oral cyclophosphamide and azathioprine.[9] Successful use of rituximab in steroid- resistant cases of ILD in primary SS was recently reported by a Gottenberg et al.[11]

CONCLUSION

ILD in patients with primary SS may have numerous histopathologic patterns. These varied histological types appear to have therapeutic implications as patients with UIP and amyloidosis diffuse type ILD respond poorly to steroids alone. Radiological patterns have been correlated with the underlying pathological subtype of ILD and the presence of multifocal cysts on HRCT scan is strongly associated with SS related ILD. Though evidence based treatment regimes have not been well established, early treatment initiation is associated with improved lung functions and increased survival.

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