CASE REPORT

Sjogren's Syndrome with Interstitial Lung Disease

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ABSTRACT

Sjogren's syndrome (SS) is an autoimmune disease affecting exocrine glands with known properties to cause chronic systemic multi-organ disease involvement. It produced the typical features of sicca syndrome, but due to insidious onset of the disease, patient may present with complications of the condition at the initial presentation. We present a case of primary Sjogren's syndrome (SS) who first presented with pulmonary manifestations and was subsequently diagnosed as Lymphocytic Interstitial Pneumonia (LIP). The patient was extensively investigated and received appropriate treatment modalities.
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INTRODUCTION

Sjogren's syndrome (SS) is a chronic autoimmune disease affecting exocrine glands, particularly the salivary and the lacrimal glands resulting in dryness of the mouth and eyes. It may manifest in variety of other disease (1). This disorder is characterized by lymphocytic infiltration to the glands. This causes gradual loss of function over time and produces the typical features of sicca syndrome like dryness of the eyes and mouth (2). However, SS has insidious onset, and the sicca syndrome may be absent during initial presentation of the condition. SS patients may instead present with the complications of the disease such as interstitial lung disease (ILD). Lymphocytic Interstitial Pneumonia (LIP) commonly occurs in autoimmune diseases, namely primary SS and Systemic Lupus Erythematosus (SLE) (3). We herein report a case of SS with initial presentation of respiratory symptoms and subsequently diagnosed to have ILD of LIP.

CASE REPORT

A 43-year-old non-smoking woman presented to our centre with history of persistent dry cough and haemoptysis for several weeks. The symptoms initially started at the age of 27 years old, where she would develop occasional coughing episodes and blood streak sputum throughout the years. She did not seek any medical advice despite the symptoms keep recurring about twice a year and lasted for few days per episode.

Since she had been having daily symptoms of frank haemoptysis with dry cough, she finally sought medical attention in 2018 after nearly 10 years of initial symptoms. Chest X ray was normal but CT scan thorax revealed lung cysts with traction bronchiectasis. She was subsequently treated for pulmonary tuberculosis (PTB). However, she had persistent symptoms while on treatment. All PTB evidence was negative, hence the treatment was stopped after one month. She defaulted her follow up citing that she could tolerate her symptoms.

The patient presented again in 2021 with worsening daily symptoms of haemoptysis and dry cough. She developed new symptoms which were dryness of the eyes and reduced salivation. She denied any other constitutional symptoms or any sign to suggest infection. Physical examinations revealed dry red eyes and dental caries. Lung auscultations revealed bilateral crepitations over lower and middle zones.

Suspicion of Sjogren syndrome was confirmed with positive Schirmer test and presence of positive anti-Sjogren Syndrome related Antigen-A (SSA/Ro) antibody. She had low titre positivity for Antinuclear Antibody (ANA) and Rheumatoid factor.

Her chest X ray was normal (Figure 1). The HRCT revealed evidence of traction bronchiectasis with segmental atelectasis and multiple centrilobular

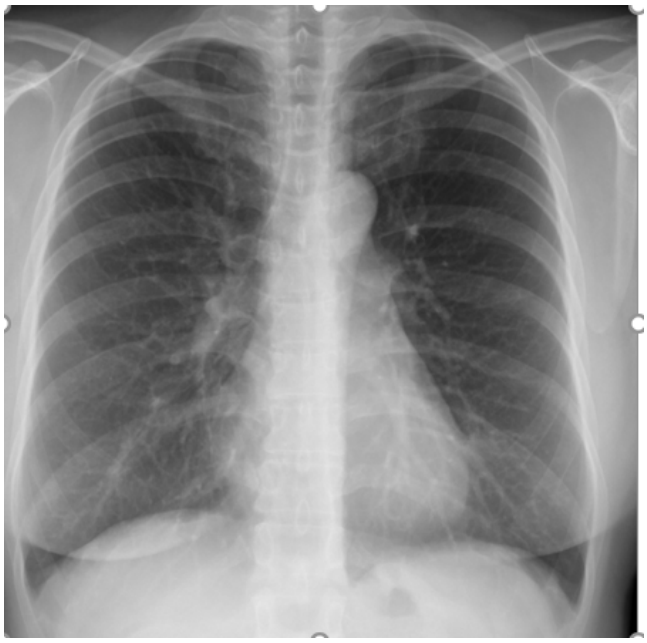

Figure 1: Normal chest X ray

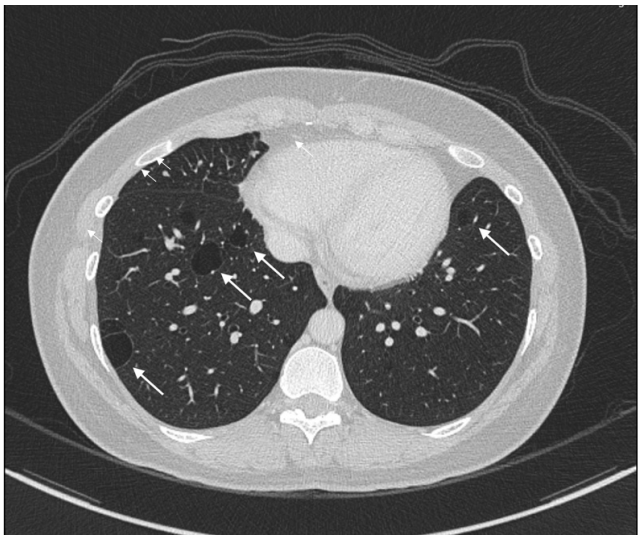

Figure 2: : Thorax CT-scan showing multiple scattered thin-walled lung cysts in bilateral lung fields (Arrow showing lung cysts

nodules surrounding the bronchiectasis (Figure 2). A full lung function test including DLCO was normal. Echocardiogram of the patient showed normal left ventricular ejection fraction with no evidence of pulmonary hypertension.

Multidisciplinary conference involving radiologist, rheumatologist and respiratory team was held, and she was eventually diagnosed with Sjogren Syndrome with Lymphocytic Interstitial Pneumonia (LIP). The diagnosis was based on clinical features, investigations results supported by the classic radiological image. The lung biopsy was not commenced in this patient as the bleeding risks of lung biopsy out-weighted the benefits. As her symptoms resolved, the patient was treated symptomatically. Her repeated spirometry was also normal.

DISCUSSION

Diagnosis of Sjogren Syndrome is made based on the 2016 American College of Rheumatology/ European League Against Rheumatism (ACR/EULAR) Classification Criteria for Primary SS. The criteria are applicable for patient who presented with at least one symptom of either ocular or oral dryness. The diagnosis can be made based on weighted scoring of salivary gland biopsy, anti-SSA/SSB antibody screening, Ocular Staining Score, Schimer's test or unstimulated whole saliva flow rate (3). SS has insidious onset of disease with various clinical manifestations which made diagnosis often difficult and delayed (4). Typical presentation of sicca syndrome may be absent during early disease. Patient has potential to develop more prominent extraglandular manifestations of SS and presents to healthcare setting with symptoms involving respiratory system, gastrointestinal system, and arthritis. Respiratory symptoms of dry cough and haemoptysis can either be the presentation of sicca syndrome or the manifestation of Lymphocytic Interstitial Pneumonia (LIP) in this patient.

Pulmonary disease is commonly associated with primary SS and may manifest in a variety of different clinical entities (3). As high as 20% of SS cases are associated with various respiratory symptoms. Subclinical manifestation of pulmonary involvement can be up to 50% of all SS patients based on CT-scan or bronchoalveolar lavage histopathological findings.

Incidence of dry cough with dental caries is known to be higher in patient with primary SS and LIP compared to SS patient with other types of ILD (4). Similar case of LIP presented with hemoptysis was reported in a 54 -year -old Japanese man (5). Possible explanation of hemoptysis is ruptured of scattered thin- wall cysts which is the characteristic nature of LIP by imaging.

Diagnosis of LIP is made on clinical and radiological findings, which is confirmed on lung biopsy and subsequent histological examination. High resolution CT (HRCT) of the thorax has been the gold standard in diagnosing cystic lung disease and to characterize the nature of the cyst. In LIP, centrilobular nodules with scattered thin-walled cysts as well as interlobular septal and bronchovascular bundle thickening are the characteristics CT findings of the condition (4). Lung biopsy is the most definitive test to confirm LIP, however subjecting patient for this procedure is not necessary from clinical standpoint. Long term complications that need to be monitored include lymphoma, bronchiectasis and secondary lung infection which may lead to end stage lung disease. It is estimated that 5% of patients with LIP develop lymphoma and the risk is higher for patient with SS (5). Clinical improvements can be expected in

primary SS & LIP with corticosteroids. However long-term survival rate remains guarded (5).

CONCLUSION

Primary SS presented with interstitial lung disease is a known entity. High degree of clinical suspicion with extensive investigations are necessary to get to the diagnosis and ensure proper initiation of treatment.

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