Pictorial Quiz

A 58-year-old lady with cough and breathlessness

Saurabh Mittal, Karan Madan

Department of Pulmonary Medicine and Sleep Disorders, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence: Dr. Karan Madan, Department of Pulmonary Medicine and Sleep Disorders, All India Institute of Medical Sciences, New Delhi - 110 029, India. E-mail: drkaranmadan@gmail.com

A 58-year-old lady presented to pulmonary medicine outpatient department with complaints of cough and shortness of breath for 5 months. Cough was dry in nature and there was no diurnal variation. She had exertional shortness of breath which was gradually worsening over the last 5 months. There was no history of hemoptysis, chest pain, or constitutional symptoms. She had polyarthralgias involving both large (knee, elbow) and small joints (hands and wrists) which was not associated with morning stiffness or joint swelling. The patient also had a history of dryness of eyes and mouth for 4 years. There were no significant occupational exposures and the patient was a lifetime never-smoker. A history of hypothyroidism was present for the last 3 years which was well controlled with treatment.

On respiratory system examination, bilateral fine crackles were audible. Rest of the general physical and systemic examination was unremarkable. On eye examination, Schirmer's test was positive objectively confirming the presence of dry eye. On further evaluation, anti-Ro and anti-La antibodies were found positive in high titers. Anti-thyroid peroxidase antibody titers were also elevated. Pulmonary function test demonstrated mild restrictive defect (forced vital capacity 68% predicted) with diffusion impairment (Diffusion capacity for carbon monoxide (DLCO) 57% predicted). High-resolution computed tomography (HRCT) scan of the thorax was performed and is shown in Figure 1.

QUESTION

What is the likely diagnosis?

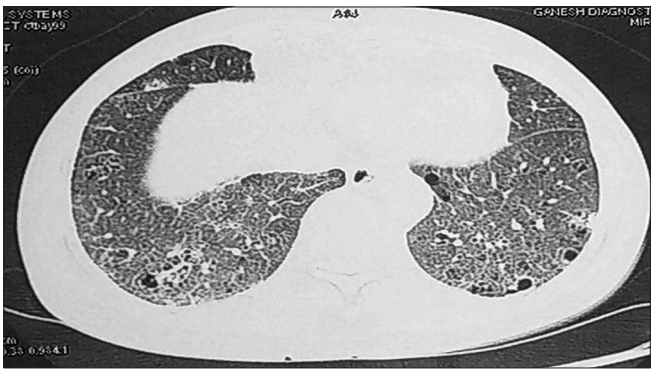

Figure 1: High-resolution computed tomography scan of thorax showing diffuse bilateral lung cysts along with centrilobular nodules and diffuse ground-glass opacities

Access this article online

Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/lungindia.lungindia_278_17

How to cite this article: Mittal S, Madan K. A 58-year-old lady with cough and breathlessness. Lung India 2019;36:70-1.

ANSWER

Lymphocytic interstitial pneumonia (LIP) associated with Sjogren's syndrome (SS).

The lung window sections of the thoracic HRCT scan demonstrate bilateral diffuse cysts with extensive ground glass opacities (GGOs) along with centrilobular nodules. Patient was advised cryoprobe lung biopsy for definite diagnosis but did not consent for the procedure. Treatment with oral corticosteroid therapy was initiated.

DISCUSSION

SS is an autoimmune disorder, diagnosis of which is frequently delayed due to nonspecific clinical presentation. SS is characterized by lymphocytic infiltration of exocrine glands which results in dryness of the mouth and eyes. Other organs such as joints, lungs, gastrointestinal tract, and blood vessels can also get involved in the disease process. Pulmonary manifestations in SS include airway involvement (bronchiolitis and bronchiectasis), interstitial lung disease (ILD), and lymphoproliferative disorders such as pulmonary lymphoma.[1] Clinically significant lung disease occurs in 9%–20% while HRCT abnormalities can be detected in 35%–50% of patients. Autoimmune thyroiditis is a frequent association.

Pulmonary involvement in SS can occur before the overt manifestations of the systemic disease. The most common pattern of ILD in SS is nonspecific interstitial pneumonia comprising about 45% of cases while LIP occurs in only 10%–15% of cases. Other patterns include organizing pneumonia, usual interstitial pneumonia (UIP), and acute interstitial pneumonia.[2] Almost 28%–30% of cases with LIP have associated SS. In LIP, lung is diffusely infiltrated by polyclonal B- and T-lymphocytes and plasma cells with lymphoid follicle formation. On HRCT, thickened broncho-vascular bundles, nodules of varying sizes, and ground-glass opacities (GGOs) are common. In LIP, cysts are commonly seen involving the lung parenchyma bilaterally and diffusely as compared to peripheral pleural-based cysts seen in UIP.[3] Cysts in pulmonary Langerhans cell histiocytosis tend to spare the lung bases, and nodules/cysts in varying stages of evolution are common. In lymphangioleiomyomatosis, diffuse involvement with bilateral lung cysts occurs in the absence of accompanying GGOs and nodules. The prognosis of patients with LIP is favorable with stabilization of lung functions after initiation of corticosteroid therapy. Other immunosuppressive agents such as azathioprine, cyclophosphamide, and rituximab have also been shown to be useful.[4,5]

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Kreider M, Highland K. Pulmonary involvement in Sjögren syndrome. Semin Respir Crit Care Med 2014;35:255-64.
2. Manuel R-C, Pilar B-Z, Raphaèle S, Hendrika B, Simon J B, Thomas D, *et al*. Characterization of systemic disease in primary Sjögren's syndrome: EULAR-SS Task Force recommendations for articular, cutaneous, pulmonary and renal involvements. Rheumatology (Oxford) 2015;54:2230-8.
3. Swigris JJ, Berry GJ, Raffin TA, Kuschner WG. Lymphoid interstitial pneumonia: A narrative review. Chest 2002;122:2150-64.
4. Seror R, Sordet C, Guillevin L, Hachulla E, Masson C, Ittah M, *et al*. Tolerance and efficacy of rituximab and changes in serum B cell biomarkers in patients with systemic complications of primary Sjögren's syndrome. Ann Rheum Dis 2007;66:351-7.
5. Deheinzelin D, Capelozzi VL, Kairalla RA, Barbas Filho JV, Saldiva PH, de Carvalho CR. Interstitial lung disease in primary Sjögren's syndrome. Clinical-pathological evaluation and response to treatment. Am J Respir Crit Care Med 1996;154(3 Pt 1):794-9.