UNUSUAL PULMONARY CYSTIC LESIONS IN A PATIENT WITH ANTI-PL-12 ANTISYNTHEASE SYNDROME – CASE REPORT

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BACKGROUND

Antisynthetase syndrome (ASS) is characterized by the presence of symptoms such as Raynaud's phenomenon, myositis, mechanic's hands, fever, commonly nonerosive arthritis of small joints, and interstitial lung disease. Interstitial lung disease is present in approximately 86% of patients according to some case series. The radiological patterns of pulmonary involvement are nonspecific interstitial pneumonia (NSIP), organizing pneumonia, acute interstitial pneumonia, or combinations of these patterns. So far, there are no descriptions in the literature about the presence of cystic lesions associated with the syndrome. In this report, we will describe a patient with lung dominant anti-PL-12 ASS who presents with lung cystic lesions.

CASE REPORT

Y.L.M., 21 years, woman, presented Raynaud's phenomena and inflammatory arthralgia, dyspnea on exertion and dry cough. The patient denied gastroesophageal reflux, dysphagia, muscle weakness or skin changes. She underwent basic laboratory tests, antibody panel, nailfold videocapillaroscopy, high-resolution chest tomography (HRCT) and pulmonary function test. The exams revealed normal blood count, normal renal and hepatic function. She presented positive ANA with a cytoplasmic dense fine speckled pattern 1:1280. Spirometry showed a severe pure restrictive disorder. The other antibodies requested in the first consultation (anti-Scl-70, anticentromere, anti-Ro, anti-La, Anti-Mi-2, anti-Jo-1, anti-dsDNA) were all negative. Nailfold videocapillaroscopy showed slightly disorganized capillary architecture, low capillary density, microhemorrhages suggestive of an early SD pattern. The HRCT showed a pattern of fibrotic NSIP associated with an unusual presence of thin-walled cysts. Initially, the patient was diagnosed with interstitial pneumonia with autoimmune features (IPAF) due to the presence of radiological interstitial lung disease not related to any defined connective tissue disease and serological and clinical evidence of autoimmunity. The patient underwent treatment with monthly cyclophosphamide pulses, evolving with partial improvement of symptoms. After seven cycles of cyclophosphamide, the patient abandoned the treatment. Subsequently, 4 years later, the patient returned to the hospital with symptoms of muscle weakness and worsening dyspnea in addition to fever and productive cough. On this occasion, pulmonary tuberculosis was diagnosed through bacilloscopy and sputum culture. The patient was treated for tuberculosis and, after a negative bacilloscopy and the infectious symptoms resolved, treatment with mycophenolate was gradually initiated. Nintedanib due to progressive decline in forced vital capacity and home oxygen therapy due to severe hypoxemia were started. Because of muscle weakness, a commercial panel (line blot) for antisynthetase antibodies was performed, which revealed positivity for anti-PL-12 antibody.

CONCLUSION

Interstitial pneumonia with autoimmune features can temporally evolve into a specific connective tissue disease. Commonly, ASS initiate without the complete and classic presentation, with subsequent appearance of the other symptoms of the syndrome. Specific phenotypes are related to specific antibodies and the presence of lung dominant disease are described in patients with anti-PL-12 antibody. Herein we describe a patient who presented a form of lung injury not described so far, at the best of our knowledge, characterized by the presence of cystic lesions associated to a NSIP pattern.

KEYWORD

Antisynthetase syndrome, Interstitial pneumonia, Lung cysts.