



SHRINKING LUNG SYNDROME AS AN INITIAL MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

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BACKGROUND

Systemic Lupus Erythematosus (SLE) is a systemic autoimmune disease. The respiratory system can be involved in different ways, such as pleuritis (with or without pleural effusion), pneumonitis, interstitial disease, pulmonary hypertension or alveolar hemorrhage. Shrinking lung syndrome (SLS) is a rare manifestation in SLE.

CASE REPORT

35 years old woman, diagnosed with SLE in February 2017 (arthralgia, alopecia, malar rash, photosensitivity, oral ulcers, positive ANA 1:80 nuclear speckled pattern, anti-dhDNA 1:80 and serositis). She had a history of worsening exertional dyspnea, which began 1 year before the diagnosis of SLE, associated with pleuritic pain on the right hemithorax. Chest radiography showed bilateral reduction in pulmonary volume. Computed tomography (CT) of the chest revealed plate-like atelectasis at the right lung base, without signs of interstitial disease. Angiotomography showed no evidence of pulmonary thromboembolism. Spirometry revealed a moderate restrictive disorder with significant reduction of maximum inspiratory and expiratory pressures. Echocardiogram exhibited no abnormalities. Following the investigation, the patient was diagnosed with SLS and started a monthly infusion schedule of cyclophosphamide for 6 months, followed by maintenance with azathioprine.

CONCLUSION

The SLS is a rare cause of pulmonary involvement in the SLE. It consists of dyspnea, pleuritic pain and radiological examination with suggestive findings such as atelectasis at the pulmonary bases and uni or bilateral elevation of the diaphragmatic dome. Its pathophysiology is not well defined, but it has been suggested that diaphragm involvement secondary to fibrosis, alteration of its innervation and/or reduction of the expandability of the chest wall might play a role. Treatment is not well established, but case reports mention the use of corticosteroids, immunosuppressants, rituximab, xanthines and beta-agonists. It should be remembered as a diagnostic possibility to explain dyspnea in SLE patients without evidence of heart failure, anemia, pulmonary thromboembolism, interstitial lung disease, among others.