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PRESENTATION, DIAGNOSIS AND TREATMENT OF SHRINKING LUNG SYNDROME IN SYSTEMIC LUPUS ERYTHEMATOSUS: REPORT OF TWO CASES.

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BACKGROUND

Shrinking lung syndrome (SLS) is a rare disease and less known complication mainly associated with systemic lupus erythematosus (SLE). Its exact prevalence is unknown and the underlying pathophysiology remains unclear. SLS is characterized by progressive dyspnea, pleuritic chest pain, diaphragmatic elevation, reduction of lung volume without parenchymal abnormalities and restrictive ventilatory disturbance in pulmonary function tests (PFTs). The diagnosis is made with chest x-ray, thoracic high-resolution computed tomography (HRCT) and PFTs demonstrating restrictive ventilatory disorder and normal diffusing lung capacity for carbon monoxide (DCLO). Thoracic wall ultrasonography for assessment of diaphragmatic muscles is capable of measuring changes in diaphragm thickness during inspiration where lack of thickening diagnose diaphragmatic paralysis. Glucocorticoids (GC) are the most common method of treatment, possibly associated with immunosuppressive therapy, such as azathioprine, cyclophosphamide and rituximab. The GC dosage and treatment duration are still undefined, where each case needs an individualized approach. Moreover, theophylline and beta-agonists, alone or in combination with GC, have been also employed with the intent to increase diaphragmatic strength, but lack scientific evidence to support its benefits.

CASE REPORT

Case 1: L.S.V., female, 23 years old, diagnosed with SLE and APS, presented increasing difficulty in breathing, progressing to dyspnea at rest associated with pleuritic chest pain. Echocardiogram and thoracic HRCT, showed no evidence of disease activity or pulmonary embolism. Employed cardiopulmonary exercise test and PFTs demonstrated impairment of pulmonary volumes, severe restrictive ventilatory disorder with severe reduction of muscle strength. These findings were consistent with diaphragmatic paralysis and suggested the hypothesis of SLS. Patient received intravenous methylprednisolone pulse therapy (1 g daily for 3 days), followed by oral corticosteroids, with a significant clinical improvement.

Case 2: A.F.L., female, 24 years old, diagnosed with SLE and APS was admitted with right-sided ventilatory chest pain, secretive cough, and dyspnea on minimal exertion. In spite of treatment for pneumonia, the patient maintained pain and dyspnea. Searching for SLS was initiated by performing a chest x-ray with right diaphragmatic dome elevation and thoracic HRCT without pleural effusion or pulmonary parenchyma damage. Patient was submitted to PFTs with restrictive disorder and reduced total lung capacity. A thoracic wall ultrasound showed right diaphragm paralysis. Cardiologic etiologies were excluded by echocardiography. Patient had GC therapy optimized, causing gradual improvement of the symptoms.

CONCLUSION

A heightened awareness of SLS and its appropriate management is therefore required to decrease the morbidity associated with this rare syndrome.