Systemic sclerosis and sarcoidosis: an exceptional coexistence

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Dear Editor,

Systemic sclerosis (SSc) association with sarcoidosis is rare, and a few cases have been reported.

Sarcoidosis is a systemic inflammatory disease of unknown etiology, characterized by the formation of non-caseating granulomas in any organ, with the lung being the most affected. It can, on the one hand, mimic several autoimmune rheumatic diseases or, on the other hand, develop concomitantly with them. The association between SSc and sarcoidosis is rare and a few cases have been described in the literature.

We present the case of a 46-year-old man, smoker (30 pack/year) with occupational exposure to silica dust for 14 years. He was referred to our hospital due to a Raynaud's phenomenon of 1 year of onset accompanied by digital ulcers, already medicated with nifedipine 60 mg/day, with good response. He reported polyarthralgia affecting shoulders, knees and ankles. He also referred dry cough, exertional dyspnea and night sweats lasting 2 years. On physical examination highlighted the presence of digital pitting scars, sclerodactyly and microstomy. Chest auscultation didn´t reveal pathological noises and muscular strength was normal. Laboratory findings were positive for antinuclear antibody at a titer of 1:1000 with anti-centromere pattern, anti-centromere and anti-TIF1-Y antibodies. Periungueal videocapillaroscopy showed an active scleroderma pattern. He was diagnosed as having limited SSc. Given the positivity of anti-TIF1-Y antibody, a neoplastic screening was made. Chest-abdomen-pelvis computed tomography demonstrated diffuse lymphadenopathy (mediastinal, hilar and peri-oesophageal), without other relevant findings. A bronchoscopy with bronchoalveolar lavage were performed with a normal cell count, negative cytology for malignant cells and negative cultures including tuberculosis. A subcarinal lymph node was biopsied, which histologic evaluation revealed non-caseating granulomas and absence of neoplastic cells (figure 1). Because of the above findings, and the elevation of angiotensin-converting enzyme level (84 U/L), the diagnosis of thoracic sarcoidosis stage 1 was established. Finally, extra-thoracic involvement was excluded using PET/CT.

Several immune-mediated diseases have been reported in association with sarcoidosis, including rheumatoid arthritis, autoimmune thyroid disease, Sjogren's syndrome, and ankylosing spondylitis. Pulmonary findings of sarcoidosis and SSc can present similar clinical characteristics, contributing to the diagnostic complexity when they coexist in the same patient.

High-resolution computed tomography findings can help distinguish pulmonary involvement of both diseases, which remains crucial to the disease treatment and prognosis. SSc interstitial lung disease is typically a nonspecific interstitial pneumonia pattern with a greater proportion of ground-glass opacities and a lower degree of coarse reticulation in the lower lung zones. On the other hand, lung manifestations of sarcoidosis include bilateral hilar and mediastinal lymphadenopathy and also mid-to upper-lobe-predominant groundglass opacities[1,2].

This association, while rare, has been reported in the literature. Of thirty cases found, most of them involved female patients, with predominance of diffuse SSc subtype. The onset of SSc antedated the diagnosis of sarcoidosis in most of the cases[3-6]. Although its etiology remains unknown, it could be explained by common genetic, environmental and pathogenic mechanisms. It should be noted that in this case, there was previous exposure to silica, which is described as a potential etiological factor in both sarcoidosis and SSc (as exemplified by its overlap with silicosis - Erasmus Syndrome)[7,8].

This case highlights the possibility of the coexistence of these two diseases and also emphasizes the need for multidisciplinary monitoring of these patients. We can conclude that pulmonary involvement in patients with SSc may not always be associated with SSc itself, but rather with other diseases, namely sarcoidosis. Characterizing pulmonary involvement plays a key role by determining prognosis and response to treatment.

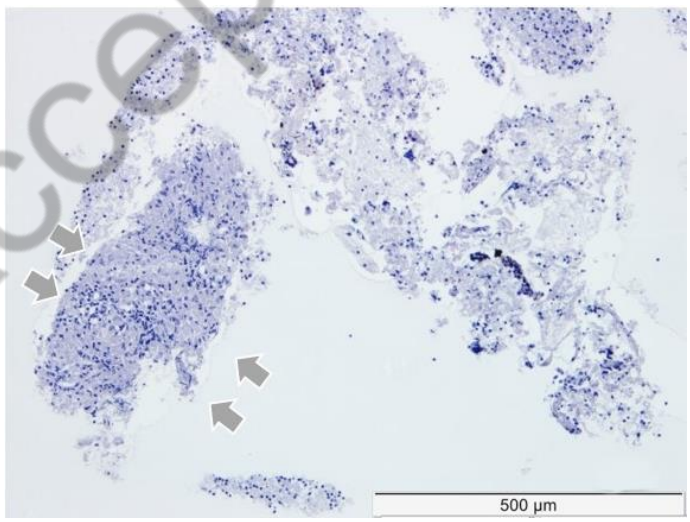

Figure 1. Hematoxylin-eosine stain, 400x. Columnar epitelial cells from the airway lining, scattered lymphocytes and macrophages were observed in the cytological exam. Importantly, an aggregate of histiocytes was identified (arrows).

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