<https://doi.org/10.47660/CBR.2020.17338>

FOUR CORNERS AND STRAIGHT EDGE SIGNS: SPECIFIC TOMOGRAPHIC FINDINGS IN THE EVALUATION OF INTERSTITIAL LUNG INVOLVEMENT IN CONNECTIVE TISSUE DISEASES

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BACKGROUND

The lungs are often affected by diffuse connective tissue diseases (DCTD). In recent years, the concept of interstitial pneumonia with autoimmune features (IPAF) has become part of the differential diagnosis of patients with tomographic or histological findings of interstitial lung disease and incomplete data for a specific DCTD.

CASE REPORT

Female, 45 years old, referred for rheumatologic evaluation with diagnostic hypothesis of interstitial pulmonary disease with autoimmune findings, due to a diagnosis of pulmonary fibrosis with nine years of evolution, currently on long term oxygen therapy, associated with a positive antinuclear antibody (ANA) (1:320 dense fine cytoplasmic) and anti-SSA (174). She denied any other symptoms suggestive of DCTD, such as skin lesions, mucous membranes, arthralgia, muscle weakness or alopecia. Other serological tests for DCTD were negative. She had a normal ophthalmological evaluation, normal salivary gland biopsy and refers a subjective Reynaud phenomenon. Esophagogram revealed mild dilation, hypotony and hypokinesia. Echocardiogram showed a PSAP of 80 mmHg and significant dilation of the right ventricle. Chest computed tomography (CT) demonstrated a pattern of usual interstitial pneumonia (UIP) with signs of four corners and straight edge. The peri-ungual capillaroscopy was normal. Based on the characteristics of visceral involvement, the diagnosis of systemic sclerosis sine scleroderma in the form of type I sine scleroderma was established.

CONCLUSION

The four corners sign describes a pattern of fibrosis that disproportionately affects the anterolateral regions of the upper lobes and areas of the posterosuperior segment of the lower lobes, while the straight edge sign refers to a reticular pattern confined to the pulmonary bases with a clear and abrupt limit with the normal parenchyma surrounding it, better observed in the coronal plane (Fig. 1). The four corners sign has a sensitivity of 28.6% and specificity of 97.6% for UIP related to systemic sclerosis, while the straight edges sign has a sensitivity of 25% and specificity of 94% for UIP associated with DCTD. The identification of these signs on chest CT, despite the low sensitivity, is very useful, especially in patients initially classified as having pulmonary interstitial disease associated with autoimmune finding.

Figure 1. “Four corners” and “straight edge” signs.