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Scleroderma-overlap syndromes: capillaroscopy, laboratory, and clinical manifestations and follow-up compared to scleroderma patients

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Abstract

Introduction: Overlap syndrome (OS) is a group of systemic connective tissue diseases (CTDs) that meet the criteria of two or more CTDs. In this study, we evaluated clinical, laboratory, and capillaroscopic manifestations of patients with scleroderma OS (SSc-OS) and its subgroups and follow-up progression compared to patients with limited SSc (LcSSc).

Material and methods: In a 10-year cross-sectional study, we evaluated 135 adult patients (70 with SSc-OS and 65 with LcSSc) with the same skin score for their baseline and follow-up clinical, laboratory, high-resolution chest tomography (HRCT), echocardiography, and nailfold capillaroscopy data and compared them.

Results: Of the 135 patients, 70 had SSc-OS, including 45 (64.3%) cases of SSc-SS (Sjögren's syndrome), 11 (15.7%) of SSc-RA (rheumatoid arthritis), 9 (12.9%) of SSc-myositis and 5 (1.7%) of SSc-SLE (systemic lupus erythematosus), and 65 had LcSSc. Lung and heart involvement and pulmonary arterial hypertension (PAH) did not differ between the two groups ($p > 0.05$). Musculoskeletal involvement and non-specific pattern of capillaroscopy were higher ($p = 0.035$ and $p = 0.001$), and digital ulcer (DU) and scleroderma patterns of capillaroscopy were lower in the SSc-OS group ($p = 0.000$).

No significant relationship was found between capillaroscopic patterns and organ involvement in the two groups (p-value > 0.05). In the follow-up (3.71 ±2.63 years), new DU and progression of lung involvement ($p = 0.002$) and the progression in capillaroscopic patterns was lower in SSc-OS ($p = 0.000$). In the follow-up, new DU was not seen in the SSc-OS, with lower progression of lung involvement, skin score, and capillary damage.

Conclusions: In SSc-OS patients, the most common subgroup was SSc-SS. Scleroderma OS was associated with lower major organ involvement and capillaroscopy progression than LcSSc. Major organ involvement in patients with SSc-OS was significantly lower than in LcSSc patients. In the follow-up, new DU was not seen in the SSc-OS with lower progression of lung involvement, skin score, and capillary damage.

Key words: scleroderma-overlap, scleroderma, nailfold capillaroscopy, interstitial lung disease, pulmonary arterial hypertension.

Introduction

Overlap syndromes (OS) are systemic autoimmune diseases in which diagnostic criteria of at least two or more connective tissue diseases (CTDs) are fulfilled [1]. The most usual associations include systemic sclerosis (SSc), rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), dermatomyositis (DM) or polymyositis (PM), and primary Sjögren's syndrome (pSS) [1]. Synovitis and myositis were reported to be more common in

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patients in future studies. The fourth limitation of our study was not assessing other SSc-related autoantibodies (i.e. anti-RNA polymerase III, anti-Pm/Scl or anti-fibrillarin).

Therefore, the need for studies on a larger number of patients with longer duration and other OS groups is recommended as it seems that the presence of overlap manifestations makes the major organ manifestations less severe with a better follow-up progression.

Conclusions

In SSc-OS patients, the most common subgroup was SSc-pSS. The DU, cardiopulmonary, GI involvement, and PAH in patients with SSc-OS were significantly lower than in the LcSSc patients. Musculoskeletal involvement such as arthritis was more commonly seen in SSc-OS patients, especially in the SSc-RA subgroup. The progression of skin score was significantly lower in the SSc-OS patients; also, non-specific patterns in capillaroscopy were significantly higher than in the LcSSc patients, and the evidence of progression and changes in capillaroscopic patterns was significantly lower in the SSc-OS than the LcSSc group. In follow-up, new DU and the evidence of progression of lung involvement and skin score were significantly lower in the SSc-OS than LcSSc patients.

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All data and materials of this manuscript are available from the corresponding author on reasonable request.

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