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Scleroderma-like capillaroscopic pattern in SLE is not a sign of overlap syndrome in both adults and children

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Nailfold capillaroscopy is a non-invasive imaging technique for morphological assessment of capillaries in the nailfold area and represents a key method for differentiation of primary and secondary Raynaud's phenomenon (RP) in rheumatic diseases. 'Scleroderma'-type microangiopathy is a reference pattern in rheumatology. It is accepted as a diagnostic criterion in systemic sclerosis (SSc) and is characterised by the presence of giant capillaries, haemorrhages and devascularisation.[1] Although a 'scleroderma' pattern is prevalent in SSc (70%–90%)[2 3] and dermatomyositis (63%–89%),[4] it may also be observed less frequently in other rheumatic diseases such as SLE and rheumatoid arthritis without features of overlap syndrome.[5] Ten years ago, it was proposed that 'scleroderma-like' capillaroscopic changes in SLE are a hint of subclinical overlap with SSc associated with anti-RNP antibody in adults and children.[6–8] The first report questioning the association between 'scleroderma-like' capillaroscopic changes in SLE and overlap syndrome with SSc with anti-RNP antibody positivity in the adult patient population was published in 2013.[9] The frequency of 'scleroderma-like' microangiopathy in the group under study was 13.3% and it presented with symptoms of secondary RP in all patients as well as with signs of vasculitis of digital vessels in half of the cases.[9] Later on van Roon *et al* reported similar results about the presence of 'scleroderma-like' capillaroscopic changes with a frequency of 17% without overlap with SSc and without significant clinical differences compared with patients with SLE without a 'scleroderma-like' pattern.[10] Notably, an association between 'scleroderma-like' microangiopathy and cutaneous digital lesions in adjacent areas was reported in patients with SLE and cutaneous lupus erythematosus (CLE) with digital skin involvement.[9 11 12]

Recently published reports by Schonenberg-Meinema *et al* also revealed the presence of a 'scleroderma-like' capillaroscopic pattern in patients with childhood-onset SLE without overlap with SSc and without anti-RNP antibody positivity.[13 14] Out of 41 patients with SLE with disease onset <18 years, a 'scleroderma-like' pattern was observed in 17.1% (7/41) of the cases without SSc-associated symptoms. Positivity of anti-RNP antibodies was not different in patients with and without a 'scleroderma-like' capillaroscopic pattern.[14] In a longitudinal study of 53 patients with childhood-onset SLE, a similar frequency of 'scleroderma-like' pattern was reported (18.9%). However, there was no association of the capillary pattern with disease activity and RP, though anti-RNP antibodies were detected significantly more frequently in patients with 'scleroderma-like' changes. During a 5-year follow-up after disease onset, patients with a 'scleroderma-like' pattern did not develop SSc symptoms, but more than half of them presented with SLE-related irreversible disease damage that could not be predicted by SLEDAI (SLE Disease Activity Index) at diagnosis or during the follow-up.[13] These observations indicate that scleroderma-like microangiopathy could be observed in both children and adults without the presence of overlap syndrome with SSc and without association with anti-RNP antibody.[5 9 10]

To sum up, a 'scleroderma-like' pattern is a relatively non-specific morphological finding that could be found in different rheumatic diseases other than SSc and SSc-associated overlap syndromes (eg, SLE in adults and children, dermatomyositis, rheumatoid arthritis).[5 9 10 13 14] Additionally, it could be observed in CLE as a local skin pathology without evidence of systemic vasculopathy. Interpretation of the diagnostic and prognostic significance of 'scleroderma-like' microangiopathy should consider the overall

context. Further studies of the discriminating features of microangiopathy in SSc and SLE in terms of evolution of microvascular pathology and staging are warranted.

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