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

Sevdalina Nikolova Lambova

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. Although a ‘scleroderma’ pattern is prevalent in SSc (70%–90%) and dermatomyositis (63%–89%), it may also be observed less frequently in other rheumatic diseases such as SLE and rheumatoid arthritis without features of overlap syndrome. 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. 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. 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. 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. 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.

Recently published reports by Schonenberg-Meinem 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. 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. 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. 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.

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). 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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**ORCID iD** Sevdalina Nikolova Lambova http://orcid.org/0000-0002-1321-6770

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