LUPUS SCIENCE & MEDICINE™

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

Sevdalina Nikolova Lambova

To cite: Lambova SN. Scleroderma-like capillaroscopic pattern in SLE is not a sign of overlap syndrome in both adults and children. *Lupus Science & Medicine* 2022;**9**:e000749. doi:10.1136/lupus-2022-000749

Received 8 June 2022
Accepted 5 July 2022

Check for updates

© Author(s) (or their employer(s)) 2022. Re-use permitted under CC BY-NC. No commercial re-use. See rights and permissions. Published by BMJ.

Department of Propaedeutics of Internal Diseases "Prof Dr Anton Mitov", Faculty of Medicine, Medical University of Plovdiv, Plovdiv, Bulgaria

Correspondence to
Associate Professor Sevdalina Nikolova Lambova; sevdalina_n@abv.bg

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.

Contributors SNL is the sole author.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Not applicable.

Ethics approval Not applicable.

Provenance and peer review Not commissioned; externally peer reviewed.

Open access This is an open access article distributed in accordance with the Creative Commons Attribution Non Commercial (CC BY-NC 4.0) license, which permits others to distribute, remix, adapt, build upon this work non-commercially, and license their derivative works on different terms, provided the original work is properly cited, appropriate credit is given, any changes made indicated, and the use is non-commercial. See: http://creativecommons.org/licenses/by-nc/4.0/.

ORCID iD

Sevdalina Nikolova Lambova http://orcid.org/0000-0002-1321-6770

REFERENCES

1 van den Hoogen F, Khanna D, Fransen J, *et al*. 2013 classification criteria for systemic sclerosis: an American College of rheumatology/European League against rheumatism collaborative initiative. *Ann Rheum Dis* 2013;72:1747–55.
2 Bergman R, Sharony L, Schapira D, *et al*. The handheld dermatoscope as a nail-fold capillaroscopic instrument. *Arch Dermatol* 2003;139:1027–30.
3 Lambova S, Müller-Ladner U. Capillaroscopic findings in systemic sclerosis -- are they associated with disease duration and presence of digital ulcers? *Discov Med* 2011;12:413–8.
4 Bertolazzi C, Cutolo M, Smith V, *et al*. State of the art on nailfold capillaroscopy in dermatomyositis and polymyositis. *Semin Arthritis Rheum* 2017;47:432–44.
5 Lambova SN. Scleroderma-Like pattern in various rheumatic diseases. *J Rheumatol* 2020;47:942
6 Furtado RNV, Pucinelli MLC, Cristo VV, *et al*. Scleroderma-like nailfold capillaroscopic abnormalities are associated with anti-U1-RNP antibodies and Raynaud's phenomenon in SLE patients. *Lupus* 2002;11:35–41.
7 Donnarumma JFS, Ferreira EVM, Ota-Arakaki J, *et al*. Nailfold capillaroscopy as a risk factor for pulmonary arterial hypertension in systemic lupus erythematosus patients. *Adv Rheumatol* 2019;59:1.
8 Ingegnoli F. Capillaroscopy abnormalities in relation to disease activity in juvenile systemic lupus erythematosus. *Microvasc Res* 2013;87:92–4.
9 Lambova SN, Müller-Ladner U. Capillaroscopic pattern in systemic lupus erythematosus and undifferentiated connective tissue disease: what we still have to learn? *Rheumatol Int* 2013;33:689–95.
10 van Roon AM, Huisman CC, van Roon AM, *et al*. Abnormal nailfold capillaroscopy is common in patients with connective tissue disease and associated with abnormal pulmonary function tests. *J Rheumatol* 2019;46:1109–16.
11 Monfort J-B, Chasset F, Barbaud A, *et al*. Nailfold capillaroscopy findings in cutaneous lupus erythematosus patients with or without digital lesions and comparison with dermatomyositis patients: a prospective study. *Lupus* 2021;30:1207–13.
12 Lambova S. Capillaroscopic findings in systemic lupus erythematosus with cutaneous digital lesions. *Lupus* 2021;30:1696–7.
13 Schonenberg-Meinema D, Bergkamp SC, Nassar-Sheikh Rashid A, *et al*. Nailfold capillary scleroderma pattern may be associated with disease damage in childhood-onset systemic lupus erythematosus: important lessons from longitudinal follow-up. *Lupus Sci Med* 2022;9:e000572.
14 Schonenberg-Meinema D, Bergkamp SC, Nassar-Sheikh Rashid A, *et al*. Nailfold capillary abnormalities in childhood-onset systemic lupus erythematosus: a cross-sectional study compared with healthy controls. *Lupus* 2021;30:818–27.