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Scleroderma-like Pattern in Various Rheumatic Diseases reply

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Dr. van Roon, *et al* reply

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Dr. van Roon, *et al* reply

To the Editor:

We thank Dr. Lambova for her interesting comment¹ on our recent article published in *The Journal*². We reported that a systemic sclerosis (SSc) or scleroderma-like capillaroscopic pattern is common in patients with Raynaud phenomenon, and can be frequently observed in patients with connective tissue diseases (CTD) other than SSc.

Dr. Lambova underlines the interesting point that a scleroderma-like capillaroscopic pattern could also be associated with presence of cutaneous digital vasculitis, which can be observed in a subset of patients with systemic lupus erythematosus (SLE)¹. In a previous study from Dr. Lambova's group, this pattern was observed in 2/4 patients with digital vasculitis³. Although this is a very interesting point, our retrospective database was not set up to answer this specific question. Only 30 patients from our study were classified as having SLE, of which 7 had cutaneous abnormalities of the fingers. In only 2/30 patients, the clinical diagnosis of digital vasculitis was made, of whom 1 had a scleroderma-like pattern and the other did not.

We agree with Dr. Lambova that a scleroderma-like capillaroscopic pattern is not specific for SSc, and as our study points out is frequently observed in other CTD. This has already been reported in the early studies on nailfold capillary microscopy⁴. A scleroderma-like capillaroscopic pattern is characterized by typical changes to the nailfold capillaries such as giant loops, neovascularizations, and capillary loss. Although morphologic changes are frequently observed in SLE, and some authors report associations with clinical disease variables and possibly activity, a scleroderma-like capillaroscopic pattern is relatively rare in SLE⁵. These abnormalities have been shown to be associated with vasculopathic changes in capillaroscopically guided nailfold biopsy studies, characterized by basal lamina thickening, perivascular cellular infiltrates, fibrosis, and edema in histology and electron microscopy⁶. Our study does support associations with vasculopathy in other organs, as confirmed by a recent study showing that a scleroderma pattern in SLE was associated with pulmonary arterial hypertension⁷. However, to the best of our knowledge, although several studies of nailfold capillaroscopy have been performed in SLE, none has addressed the association with digital vasculitis structurally.

A major challenge in studying digital vasculitis is that its diagnosis is based on clinical grounds; it is generally not demonstrated by objective imaging or histopathologic studies. Cutaneous lesions of the digits in SLE are polymorphous, and not always classified correctly. This is emphasized by a study that found histopathological confirmation of vasculitis in only a small proportion of SLE patients with digital lesions that were clinically diagnosed as vasculitis⁸.

We stress the comment of Dr. Lambova that a scleroderma-like pattern is associated with other CTD than SSc alone. However, although the hypothesis of a link of the pattern with digital vasculitis is a very interesting one, we cannot confirm it with our data, and strongly suggest that it be addressed in a well-designed prospective study.

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