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Reproducibility of the scleroderma pattern assessed by wide-field capillaroscopy in subjects suffering from Raynaud's phenomenon

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Abstract

Objectives. The aim of this work was to study inter- and intra-observer agreement for the diagnosis of scleroderma pattern by wide-field capillaroscopy.

Methods. Images were taken from 50 patients known to have SSc and 50 controls consulting for RP who did not have SSc. These images were rated simultaneously by 11 experienced vascular medicine physicians as scleroderma pattern or not. Two weeks later, 7 of the 11 observers again rated the same images.

Results. Inter-observer agreement was almost perfect between the 11 observers (κ 0.86 ± 0.01), and the proportion of concordant observations was 79% (70–87). When each observer was compared with the reference, agreement was also almost perfect: κ coefficient 0.92 ± 0.03 and proportion of concordant observations 79% (70–87). Intra-observer agreement was also almost perfect: median κ coefficient 0.94 (0.78–0.96) and median proportion of concordant observations 97% (89–98).

Conclusion. Excellent inter- and intra-observer agreement was obtained in experienced vascular physicians for the diagnosis of capillaroscopic landscape by wide-field nailfold capillary microscopy.

Key words: nailfold capillaroscopy, systemic sclerosis, Raynaud's phenomenon, reproducibility

CLINICAL SCIENCE

Rheumatology key message

- Inter-observer and intra-observer agreement is high for scleroderma pattern in capillaroscopy.

Introduction

SSc is characterized by specific capillary abnormalities that can be assessed by videocapillaroscopy, occur early in the disease and often precede the diagnosis of SSc. The sclerodermic pattern is characterized by giant capillaries, decreased capillary density, avascular areas and disorganization of capillary bed architecture [1]. These abnormalities collectively form the scleroderma pattern, and capillaroscopy has been included in the recent EULAR/ACR definition of SSc [2]. The sclerodermic pattern is found in 82–99% of patients with confirmed SSc

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[3–7]. It is highly predictive of the diagnosis of SSc in the coming years in a patient suffering from RP [8, 9]. It has been proposed that the absence of ANAs and scleroderma capillaroscopic pattern is sufficient to rule out the diagnosis of subsequent SSc [9].

Thus, the place of capillaroscopy is crucial in the early diagnosis of SSc. Le Roy and Medsger [10] proposed that a capillaroscopic scleroderma pattern in a patient with Raynaud's is sufficient to consider the diagnosis of SSc. However, little data are available concerning the reproducibility of the diagnosis of the capillaroscopic scleroderma pattern, so we decided to perform a scleroderma pattern reproducibility study among experienced vascular physicians using wide-field nailfold videocapillaroscopy.

Methods

Selection of images

Images were selected from the first 50 patients with RP and SSc included in the multicentre SCLEROCAP study (373 SSc patients followed up for 3 years in order to validate the prognostic classifications of SSc) and from 50 controls who consulted for RP and who were not diagnosed as having SSc. There were 46 subjects with primary RP, 3 with RA and 1 with Buerger's disease. Acquisition of images was performed on eight fingers (all fingers except thumbs) by using a wide-field videocapillaroscopy device (×100 magnification, Perimed, Järfälla, Sweden). The images (1/finger) were recorded on a disk, which was then blinded by a research assistant and given to experienced vascular physicians not aware of the patient's identity or status for SSc.

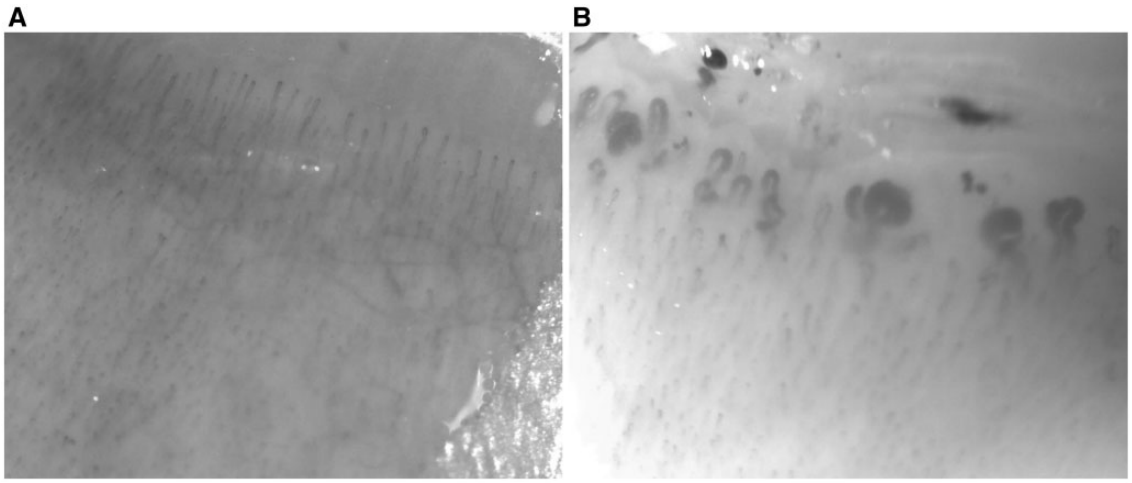
Measurements

Images from the four fingers of the left hand were inserted into a slide, and images from the four fingers of the right hand were inserted into another slide. Images from both right and left hands were projected simultaneously to 11 experienced vascular physicians (named thereafter observers) in a meeting of our microcirculation research group (2 December 2016). For each patient, the observers had to assess the presence or absence of a scleroderma landscape and provide a binary answer (scleroderma pattern, no scleroderma landscape). Scleroderma pattern was defined as a combination of giant capillaries, capillary loss, avascular areas and disorganization of the capillary bed architecture. A giant capillary was considered to be a capillary whose diameter was above 50 μm on at least one of its branches; capillary loss was defined as capillary density below 7/mm; avascular areas were defined as areas with <3 capillaries/mm; and disorganization was defined as lack of parallelism of capillaries (Fig. 1). We defined a reference rating for each patient as the answer given by at least 9 of the 11 observers or by general consensus in the event of at least three discordances. In 3 cases, only 8 observers gave a similar rating and the pictures from these 3 subjects were rated by all 11 observers. The same slides of the 50 patients and 50 controls were sent 2 weeks later to 7 of the 11 observers in order to assess intra-observer agreement.

Statistical analysis

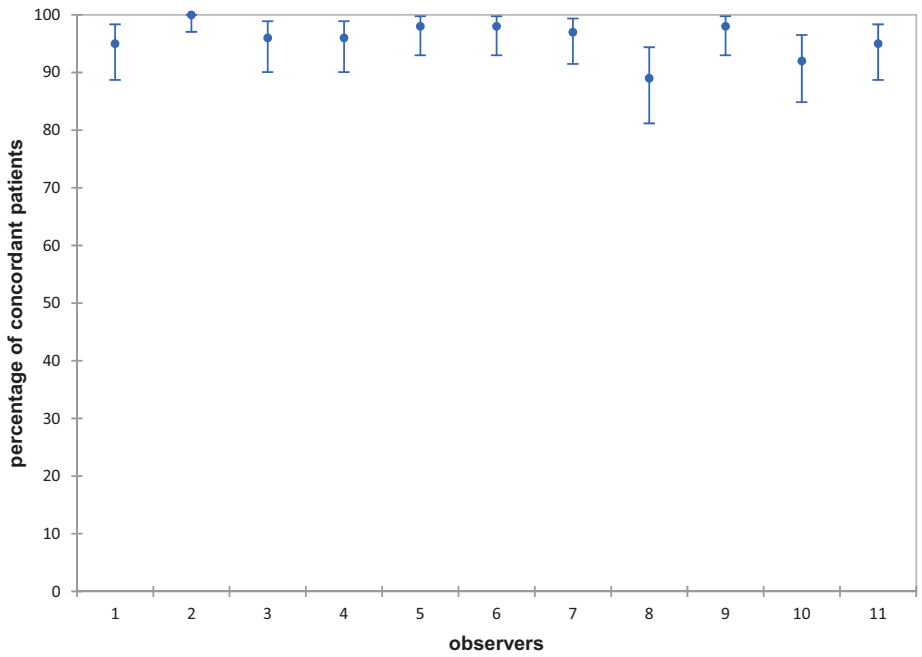
Inter- and intra-observer agreement was assessed by Fleiss' κ coefficient, and the Landis and Koch interpretation of κ values was used [11]: 0.2–0.4: fair; 0.4–0.6: moderate, 0.6–0.8: substantial; >0.8: almost perfect. The observed proportion of concordant observations (p0) was also calculated. Agreement was also calculated for each observer with the reference. XL stat software was used for statistics (Addinsoft, Paris, France).

Fig. 1 Normal and sclerodermic patterns in capillaroscopy



(**A**) Normal capillaroscopy: regular alignment of capillary loops, normal capillary density, no giant capillary.
(**B**) Scleroderma pattern: giant capillaries, capillary loss, haemorrhages, architectural disorganization.

Fig. 2 Agreement of each of the 11 observers with the reference



Results

Finally, 44 of the 50 SSc patients were found to have the scleroderma pattern (88%). Inter-observer agreement was almost perfect between the 11 observers (κ 0.86 ± 0.01), and the proportion of concordant observations was 79 (70–87). When each observer was compared with the reference, agreement was also almost perfect, with a κ coefficient of 0.92 ± 0.03 and a proportion of concordant observations of 79 (70–87) (Fig. 2). Intra-observer agreement was again almost perfect among the 7 observers who rated the images twice: the median κ coefficient was 0.94 (0.78–0.96), and the median proportion of concordant observations was 97 (89–98) (supplementary Table S1, available at *Rheumatology* Online).

Discussion

In a recent EULAR study, morphological abnormalities of the capillaries were not reproducible between either experienced observers or novices, resulting in a κ coefficient of below 0.50 [12]. However, the abnormalities studied were only morphological and did not take into account capillary loss, avascular areas or alteration of the capillary bed architecture, so the study did not truly address scleroderma landscape. Good inter-observer reproducibility has been reported otherwise for the diagnosis of scleroderma pattern in Raynaud's patients, with a proportion of agreement of 90% and a κ coefficient of 0.60 [13]. Overbury *et al*. [14] found moderate agreement between primary care physicians and an expert in capillaroscopic abnormalities (κ = 0.50, 95% CI: 0.49, 0.55). Our results show better inter-observer agreement. This may be due to technical differences in devices, since we used wide-field videocapillaroscopy with ×100 magnification: narrow-field capillaroscopy (×200) was used in the former studies [15]. We believe that it is easier to assess the diagnosis of scleroderma pattern by using wide-field capillaroscopy because the global architecture of the capillary bed is easier to assess. In the present study, we focused on the truly useful information for physicians, that is, the presence of a scleroderma pattern or not.

Reproducibility issues have been recently reported for the use of Maricq and Cutolo's classifications in predicting prognosis [16], so we found it useful to assess the reproducibility of the scleroderma pattern, the main information provided by capillaroscopy. Our results suggest very good reproducibility of wide-field nailfold videocapillaroscopy for the diagnosis of scleroderma pattern in the hands of experienced vascular physicians. Caution is required before transposing these results to less experienced observers, in whom further validation is needed. The diagnosis of scleroderma pattern is very reliable, and this supports the inclusion of capillaroscopy in the diagnosis criteria of SSc.

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Supplementary data

Supplementary data are available at *Rheumatology* Online.

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