

## Nailfold microangiopathy in dermatomyositis and systemic sclerosis: what is different in long term follow-up?

Dermatomyositis (DM) is a chronic inflammatory disorder involving mainly the skin and muscles. Systemic Sclerosis (SSc) is an autoimmune rheumatic disease of unknown etiology characterized by vascular alterations and progressive tissue fibrosis of multiple organs.

Peripheral microangiopathy is typical of SSc, as well as frequently observed in DM, and it can be easily recognized by nailfold videocapillaroscopy (NVC), a non invasive and safe technique. The characteristic NVC pattern named “scleroderma-pattern” that has been typically described in SSc, has also been reported in other “scleroderma spectrum disorders”, including DM. Only few studies described the main NVC changes over time in this last clinical condition.

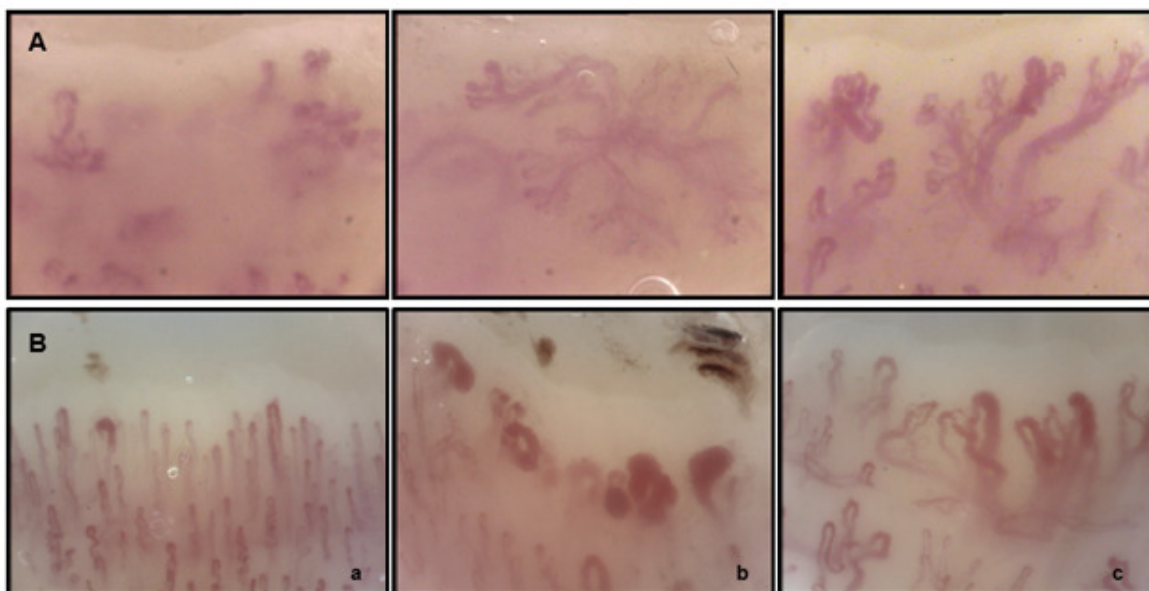


Fig. 1. A: Nailfold videocapillaroscopic pictures during the 3 year follow-up in a dermatomyositis patient (scleroderma-like pattern). B: Example of progression of nailfold videocapillaroscopic patterns during follow-up in a systemic sclerosis patient (a = Early, b = Active, c = Late pattern). Magnification 200x.

The aim of our retrospective study was to describe the main NVC changes in patients with DM, during a three year follow-up, and to investigate possible differences in NVC findings at first visit (first capillaroscopy) in comparison with patients affected by SSc matched for sex, age and disease duration.

Twenty-four DM (mean age  $54 \pm 15$ SD years; disease duration  $4 \pm 5$  years; 4 males and 20 females) and twenty-four SSc patients (mean age  $55 \pm 13$  years; disease duration  $4 \pm 5$  years; 4 males and 20 females) matched for age and disease duration at first NVC have been retrospectively evaluated. The following qualitative parameters have been assessed by NVC: enlarged loops (irregular or homogeneous increase of capillary

diameter  $\geq 20$  but  $< 50$   $\mu\text{m}$ ), giant capillaries (homogeneously dilated loops with a diameter  $\geq 50$   $\mu\text{m}$ ), microhaemorrhages (dark masses attributable to hemosiderin deposit), ramified capillaries (abnormally shaped capillaries, branching, bushy, interconnected, originating from a single capillary, expression of angiogenesis), disorganization of the vascular array, capillary density (number of capillaries per linear millimetre on first row). The microangiopathy evolution score (MES) was also calculated.

In this study nineteen out of 24 DM patients (79%) showed a NVC “scleroderma-like pattern” at baseline.

Comparing at baseline DM with SSc patients, the giant capillary and microhaemorrhage scores were significantly higher in SSc than in DM patients ( $p=0.04$ , and  $p=0.05$  respectively), while capillary density, ramification and disorganization scores were higher in DM than in SSc patients ( $p=0.05$ ;  $p=0.002$ ;  $p=0.004$ , respectively). The absolute number of ramified capillaries was significantly higher in DM patients ( $p=0.002$ ), while the absolute capillary number was significantly higher in SSc patients ( $p=0.05$ ) at baseline (Tab. 1).

No statistically significant variations of all the capillaroscopic scores was observed during the 3 year follow-up in DM patients (Fig. 1A), differently from SSc which microangiopathy is notoriously progressive (from the “Early” to the “Active” and finally to the “Late” pattern) (Fig. 1B).

By comparing DM patients with or without anti-Jo-1 antibody positivity, no statistically significant differences of the scores of the main capillary parameters was observed at baseline between the two groups. In our study, a relevant angiogenesis is a distinctive NVC feature in patients with DM: the comparison between DM and SSc patients at baseline demonstrated a particularly higher prevalence of giant capillaries and microhaemorrhages in the latter disease, as well as higher frequency of ramified capillaries (angiogenesis) and loss of capillaries in the former.

| NVC parameters                                    | DM              | SSc             | p     |
|---|-----------------|-----------------|-------|
| Dilated capillaries (score)                       | 2.33 $\pm$ 0.70 | 2.08 $\pm$ 0.58 | 0.15  |
| Giant Capillaries (score)                         | 1.00 $\pm$ 0.72 | 1.42 $\pm$ 0.58 | 0.04  |
| Microhaemorrhages (score)                         | 0.67 $\pm$ 0.56 | 1.00 $\pm$ 0.66 | 0.05  |
| Capillary number (score)                          | 1.54 $\pm$ 0.72 | 1.12 $\pm$ 0.85 | 0.05  |
| Capillary abnormal shapes / ramifications (score) | 1.79 $\pm$ 1.02 | 0.79 $\pm$ 1.02 | 0.002 |
| Capillary disorganization (score)                 | 1.67 $\pm$ 0.82 | 0.83 $\pm$ 1.01 | 0.004 |
| Absolute number of capillaries                    | 5.62 $\pm$ 1.95 | 6.83 $\pm$ 2.18 | 0.05  |
| Number of normal capillaries                      | 1.58 $\pm$ 2.22 | 2.08 $\pm$ 1.79 | 0.16  |
| Number of abnormal shapes / ramifications         | 1.92 $\pm$ 1.06 | 0.88 $\pm$ 1.08 | 0.002 |

Tab. 1. Comparison of nailfold videocapillaroscopic (NVC) parameters at baseline in whole dermatomyositis (DM) and systemic sclerosis (SSc) patients. Means  $\pm$  standard deviations are reported.

In conclusion, both SSc and DM may display similar nailfold microvascular abnormalities that can be detected by NVC. However, DM and SSc display quite different nailfold capillary features: this opens the door

toward a possible identification of a DM capillaroscopic pattern (different from the well known scleroderma pattern of SSc), which may be of great relevance for the early diagnosis of connective tissue diseases.

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## **Publication**

[Long-term follow-up of nailfold videocapillaroscopic changes in dermatomyositis versus systemic sclerosis patients.](#)

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