Clinical Vignette

Rheumatology 2017;0:1
doi:10.1093/rheumatology/kex225

RNA polymerase III–positive systemic sclerosis in a patient with circumscribed morphea

A 51-year-old male with an 11-year history of stable circumscribed plaque morphea presented with a progressive, pruritic skin rash poorly responsive to topical corticosteroids and oral antihistamines. Physical examination demonstrated multiple shiny, indurated patches with scattered hypopigmentation on the dorsal arms, hands, chest, posterior neck and under the abdominal pannus in a band-like fashion. The fingers, feet and face were spared. Skin biopsy was consistent with morphea. ANA, ENA (including Scl-70) and centromere antibodies were negative. Narrow-band ultraviolet B therapy was initiated for generalized morphea. At 3 months he reported worsening skin sclerosis of the fingers and inflammatory arthritis involving the hands and wrists with 'cold-induced' hand pain. Physical examination showed new facial telangiectasias and sclerodactyly. Nailfold videocapillaroscopy showed features characteristic of SSc. An upper extremity arterial study confirmed vasospasm (consistent with RP) with occlusive vasculopathy in multiple digital vessels bilaterally. The patient scored 14 points on the ACR/EULAR classification criteria for SSc [1].

Morphea and SSc are clinically distinct diseases, with sclerodactyly, nailfold capillary abnormalities and RP typically absent in morphea. There are rare reports of coexistence, but whether they are two ends of the same disease remains controversial [2]. This patient had circumscribed morphea followed 11 years later by generalized morphea and, <6 months later, classic SSc. RNA Polymerase III antibody was positive, which is typically associated with severe and rapidly progressive skin involvement. Methotrexate was initiated for treatment.

Funding: No specific funding was received from any bodies in the public, commercial or not-for-profit sectors to carry out the work described in this article.

Disclosure statement: The authors have declared no conflicts of interest.

Kendra D. Watson[1] and Ashima Makol[2]

[1]Department of Dermatology and [2]Division of Rheumatology, Department of Internal Medicine, Mayo Clinic College of Medicine and Science, Rochester, MN, USA

Correspondence to: Ashima Makol, Division of Rheumatology, Department of Internal Medicine, Mayo Clinic, 200 1st Street SW, Rochester, MN 55905, USA.
E-mail: makol.ashima@mayo.edu

References

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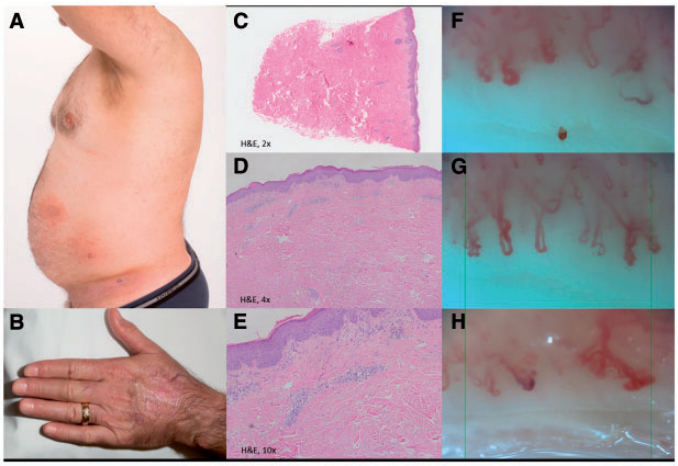
Fig. 1 Progression of morphea to SSc with late SSc pattern of microangiopathy on nailfold videocapillaroscopy



(**A**) Stable plaque morphea of the left upper abdomen with (**B**) new tense shiny plaques visible under the abdominal pannus and dorsal hand. (**C** and **D**) Skin biopsy demonstrated pandermal sclerosis and (**E**) vacuolar interface changes with mixed perivascular lymphoplasmacytic inflammation. Nailfold videocapillaroscopy (Optilia, 200×) showed (**F**) pericapillary oedema, giant capillaries and capillary microhaemorrhage and (**G**) decreased capillary density with (**H**) avascular areas and neovascularization.