Dermoscopy and reflectance confocal microscopy for the diagnosis of scleromyxedema

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Key words: confocal microscopy; dermoscopy; primary mucinosis; scleromyxedema.

CLINICAL PRESENTATION

A 53-year-old woman presented with a 12-month history of slowly growing small, firm, pruritic papules on the dorsa of both hands, elbows, and forearms (Fig 1, *A*). The patient was otherwise well, with no other skin findings or extracutaneous complaints.

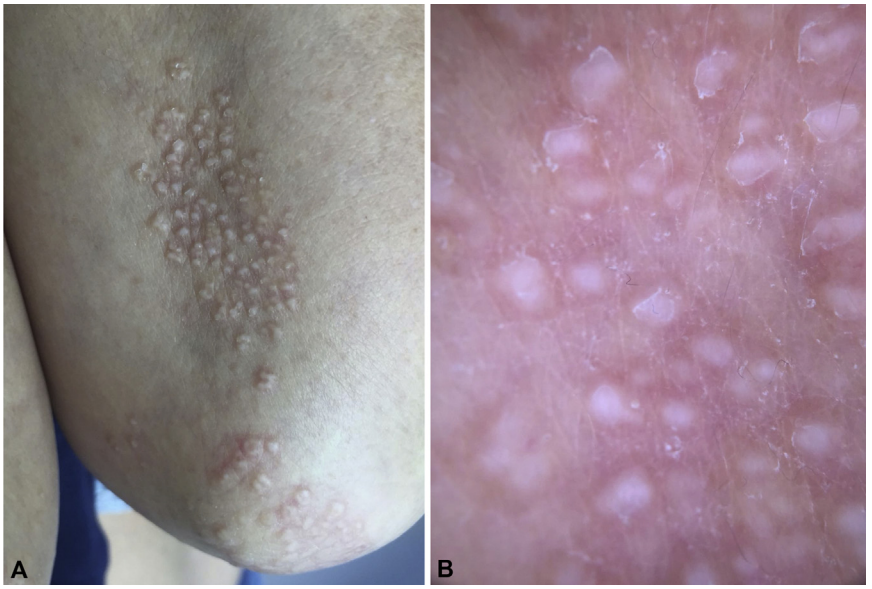

Fig 1. Scleromyxedema, clinical and dermoscopic presentation. **A**, Multiple, firm, pruritic papules are seen on the left elbow and forearm. **B**, Dermoscopy shows roundish and clustered white-ivory homogenous areas similar to rice grains in between normal skin.

DERMOSCOPIC APPEARANCE

Dermoscopy found clusters of round white-ivory homogenous areas resembling rice grains (Fig 1, *B*).

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CONFOCAL MICROSCOPY APPEARANCE

Reflectance confocal microscopic presentation is detailed in Fig 2.

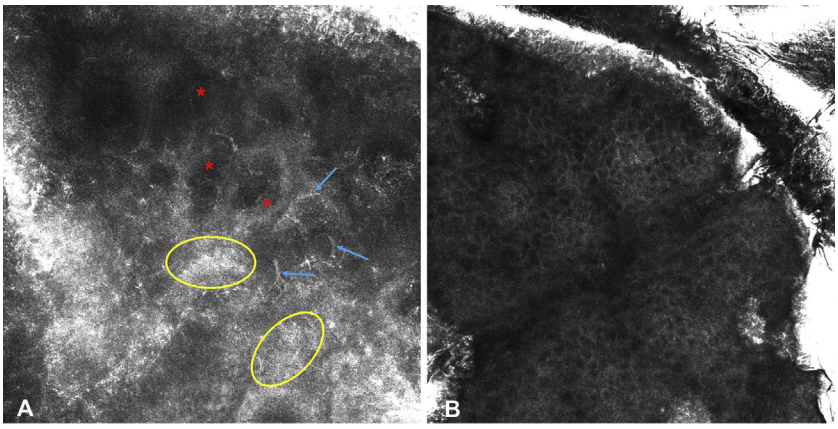

Fig 2. Scleromyxedema, reflectance confocal microscopy appearance. **A**, Basic image (0.5 × 0.5 mm) at dermoepidermal junction and superficial dermis level enables the observation of multiple randomly distributed stellate and bright structures (*blue arrows*) mixed with highly refractile thickened fibres (*yellow circles*), together with dark areas surrounding enlarged dermal papillae (*red asterisks*). **B**, Normal honeycomb pattern is observed at epidermal layer (basic image, 0.5 × 0.5 mm).

HISTOLOGIC DIAGNOSIS

Histologic examination found interstitial Alcian blue–positive mucin deposits in the reticular dermis, thickened collagen bundles, and increased fibroblastic proliferation (Fig 3).

Laboratory studies excluded viral chronic infections and thyroid dysfunction, but serum electrophoresis found a monoclonal gammopathy of the IgG type, with λ light chains. A diagnosis of scleromyxedema was made, and the patient was started on intravenous immunoglobulin treatment.

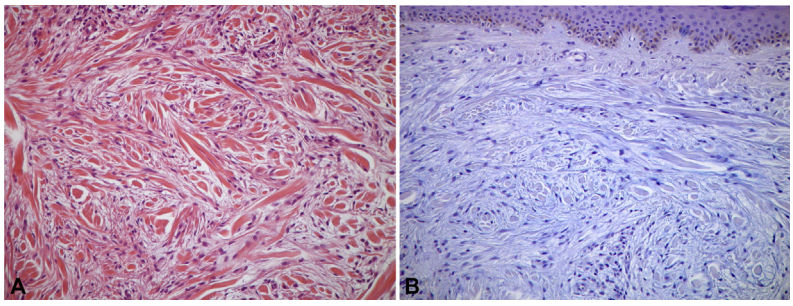

Fig 3. Scleromyxedema, histopathological examination. **A**, Thickened collagen bundles and increased fibroblastic proliferation. **B**, Interstitial Alcian blue–positive mucin deposits in the reticular dermis. (**A**, hematoxylin-eosin stain; original magnifications: **A** and **B**, ×200.)

DISCUSSION

The role of noninvasive imaging techniques has been described for the diagnosis of several inflammatory skin diseases.[1] Scleromyxedema is a rare primary mucinosis the diagnosis of which has relied on proper clinicopathologic correlation. Its presenting symptoms are usually cutaneous.[2]

Confocal features of dermal stellate cells, bright fibres, and dark areas corresponded well to scleromyxedema classical triad of marked fibroblast proliferation, increased collagen deposition, and diffuse mucin deposits, respectively. Dermoscopic rice grain–like structures also correlate well to dermal fibroblast proliferation and collagen accumulation.

As new insights were provided using dermoscopy and reflectance confocal microscopy, we showed its potential not only to increase diagnostic awareness for scleromyxedema, but also to avoid mistreatments and delays in further relevant diagnostic investigations.

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