

Scleromyxedema: A rare case report with dermoscopic findings

Dear Editor,

Cutaneous mucinosis is a heterogenous group of disorders which are characterised by an abnormal mucin deposition in skin. Scleromyxedema, also known as generalised papular and sclerodermoid lichen myxedematosus, is a cutaneous mucinosis characterised by a generalised papular and sclerodermoid eruption, a triad of increased fibroblast proliferation, mucin deposition and fibrosis. Monoclonal gammopathy is commonly associated.

A 50-year-old Indian male presented with an insidious onset and gradually progressive eruption of erythematous papules associated with thickening of skin primarily over the face and a tender swelling in the central aspect of the face. The patient did not have a history of any systemic disease, nor did he have any other systemic complaints on enquiry.

On cutaneous examination, the patient had a diffuse erythematous, mildly edematous, tender plaque on centofacial

area involving the glabella, and root of the nose giving rise to leonine facies [Figure 1a]. The chin also had a diffuse infiltration with shiny surface and a background yellowish hue. There was infiltration and thickening of both ears with a shiny appearance of the overlying skin. Additionally, there were sheets of firm, waxy, lichenoid to brownish papules, with few making linear configuration over the back of the neck, trunk, and proximal extremities [Figures 1b and 1c]. Increased furrowing of skin on the back (Shar-Pei Sign) and abdominal areas was noted. Both proximal interphalangeal joints had tightening of skin with central depression and an elevated rim (doughnut sign).

Dermoscopic examination of the glabella (DermLite DL4 dermatoscope—3 Gen Inc., San Juan Capistrano, CA, USA—×10 magnification, polarised mode) [Figure 2] revealed localised yellowish brown and yellowish white roundish structureless areas with interspersed linear irregular vessels and few out of focus vessels and accentuated pigmentary



Figure 1a: Centofacial erythematous plaque with infiltration of forehead with glabella (leonine facies) and chin.



Figure 1b: Papular eruption over the back with furrowing of creases visible on lower back (Shar-Pei sign).



Figure 1c: Central depression over proximal interphalangeal joint with elevated rim (doughnut sign).

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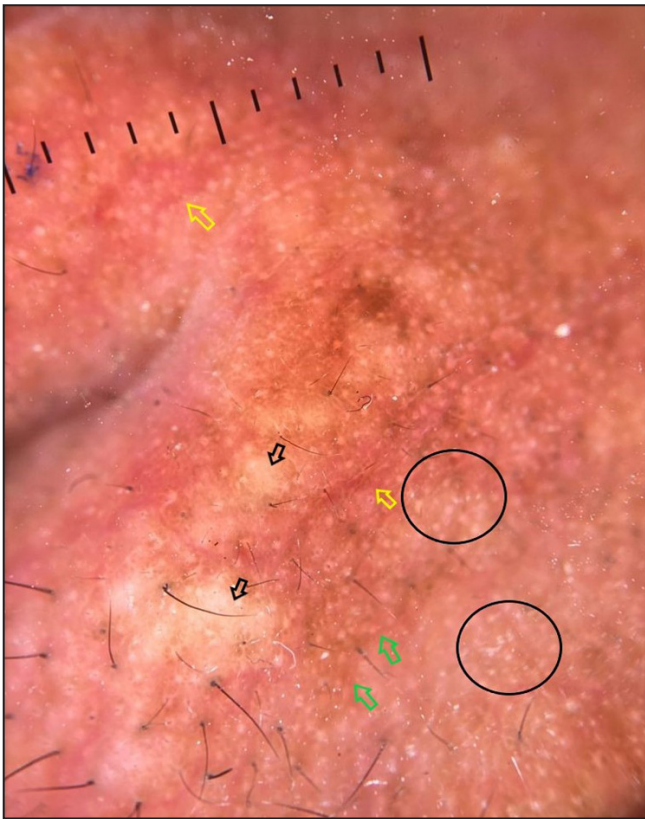


Figure 2: Dermoscopy of the glabella showing localised yellowish brown and yellowish white structureless area (black arrow), ill focused linear irregular vessels (yellow arrow) accentuated pigmentary network (green arrow), clustered whitish round homogenous areas leading to sago grain like appearance due to eccrine openings (black circles).

network at few areas. There was a relative alopecia with vellus hair showing peripilar scales and few white and black dots.

Dermoscopy of lesion on neck [Figure 3] showed glistening yellowish white round to oval areas with loss of pigmentary



Figure 3: Dermoscopy from papular lesions over the neck showing glistening yellowish white, round to oval areas (coalescence into a linear array is prominent with accentuation of pigmentation in the periphery).

network (few coalescing into a linear array with accentuation of pigmentation in the periphery).

Systemic examination of the patient including the sensorimotor examination was normal.

Serum protein electrophoresis with immunofixation showed an IgG monoclonal gammopathy. Chest X-ray showed mediastinal lymph node enlargement. Spirometry revealed a mild (30–50%) restrictive pattern but high resolution computed tomography (HRCT) of chest did not show any fibrotic changes. Electrocardiogram was low voltage with a mild left axis deviation.

Histopathological examination of the glabellar skin was suggestive of scleromyxedema, as shown [Figures 4a and 4b].

On the basis of the above findings, a provisional diagnosis of scleromyxedema was considered. Opinion from a hemato-oncologist as well as a rheumatologist was sought and the patient was advised a bone marrow aspiration and biopsy.

Strikingly, our patient did not show any signs or symptoms of systemic involvement at the time of diagnosis, but long term follow-up is still essential.

Previously, Mendes Bastos P *et al.* had described clusters of round white-ivory homogenous areas resembling rice grains, which they reported to correlate well to dermal fibroblast proliferation and collagen accumulation.¹ Dermoscopically, we were also able to notice yellowish-white round structureless areas which seem to correlate well with mucin deposition on histopathology along with pigmentary and vascular changes [Table 1].

Scleromyxedema is a chronic and unpredictable disease associated with monoclonal gammopathy and severe systemic implications including neurologic, haematologic and cardiologic involvement leading to a guarded prognosis. However, our patient had no clinically evident systemic involvement with presence of classic cutaneous lesions.

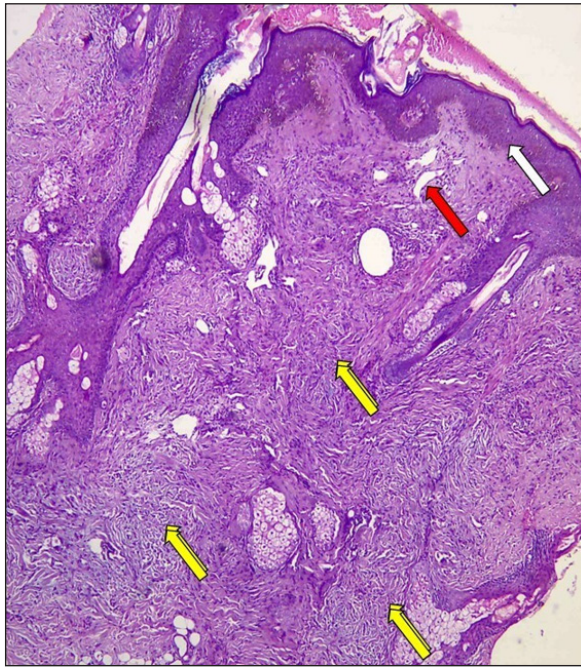


Figure 4a: White arrow displays epidermal acanthosis and hyperpigmentation of the basal layer which corresponds to the accentuation of pigment network. The red arrow displays presence of dilated capillaries which correspond dermoscopically to the telangiectasia and linear curved vessels. The yellow arrows suggest areas of deposition of mucinous material (Haematoxylin and eosin stain, × 10 magnification).

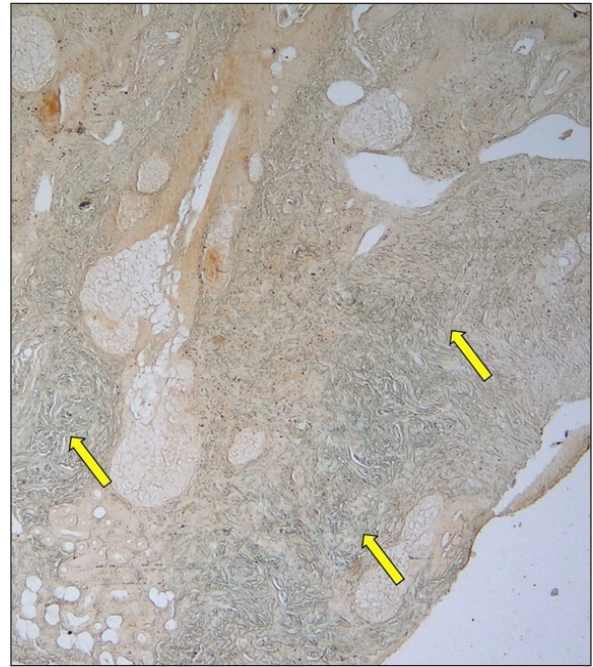


Figure 4b: There is presence of fibroplasia and bluish material in the mid and deep dermis denoted by yellow arrows, which on alcian blue stain displays bluish discoloration suggestive of mucin depositions (Alcian blue stain at pH 2.5, × 10 magnification).

Table 1: Correlation between the dermoscopic and histopathological features of scleromyxedema

Sr	Dermoscopy	Histopathology
1	Yellowish white structureless areas.	Interstitial fibrosis and mucin deposition.
2	Linear and irregular vessels.	Dilated capillary in the papillary dermis.
3	Accentuated pigmentary network.	Irregular epidermal acanthosis and basal layer hypermelanisation.
4	Erythema in the periphery.	Superficial peri vascular, peri appendageal infiltrate.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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