

Diffuse Indurated Skin

A healthy 60-year-old Chinese male presented with a 3-week history of progressive patchy discolouration, pain and swelling of bilateral lower limbs (left more than right), associated with constitutional symptoms of appetite loss. There was no preceding trauma, new drugs or topical applications. Clinical examination revealed extensive indurated skin over both lower limbs, extending to the inguinal fold on the left (Fig. 1). The tightness of the skin had precluded him from bending his left knee.

Magnetic resonance imaging (MRI) of the left lower limb showed diffuse skin thickening and a reticular pattern in the subcutaneous fat, in keeping with skin inflammation (Fig. 2). Blood tests were unremarkable and there was no paraproteinaemia. A malignancy workup, comprising of computed tomography (CT) of the thorax, abdomen and pelvis and endoscopic evaluations, was negative. Histology of a skin biopsy revealed mucin deposits in the dermis, thickened collagen bundles and fibroblastic proliferation.

The skin induration improved with topical application of 0.1% betamethasone valerate ointment but the patient subsequently defaulted follow-up.

What is the most likely diagnosis?

- A. Systemic sclerosis
- B. Scleromyxoedema
- C. Nephrogenic systemic fibrosis
- D. Acute lipodermatosclerosis
- E. Pretibial myxoedema

Discussion

It is important to differentiate between the various conditions (such as those in options A to E) presenting clinically with indurated and hardened skin, in order to correctly identify associated underlying diseases that may be life threatening. Scleromyxoedema is the generalised form of lichen myxoedematosus, an idiopathic cutaneous disorder characterised by the proliferation of fibroblasts with excess mucin deposits in the skin, sparing the mucous membranes. The generalised form may involve internal organs and can be fatal. It is often associated with monoclonal gammopathy, or other bone marrow malignancies,¹ although not in this case.



Fig. 1. Extensive indurated skin extending to the inguinal fold on the left lower limb.

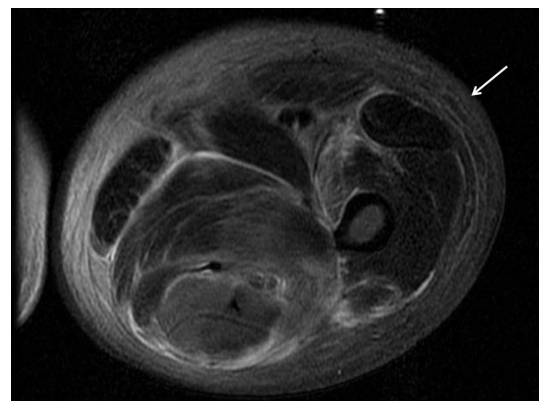


Fig. 2. Magnetic resonance imaging (MRI) images of diffuse skin thickening and a reticular pattern seen in the subcutaneous fat extending from the level of the upper thigh to the ankle.

Answer: B

The skin induration may resemble scleroderma of systemic sclerosis, but the latter is characterised by an excess production of collagen in the dermis, rather than mucin. There is no associated calcinosis or telangiectasia in scleromyxoedema.¹ Nephrogenic systemic fibrosis has almost identical histology features with scleromyxoedema, with excess mucin production and fibroblast proliferation, but occurs in patients with renal failure and may be precipitated by the use of gadolinium-containing contrast agents.¹

Lipodermatosclerosis is a type of panniculitis that typically affects patients with venous insufficiency. In the acute inflammatory state, patients often present with swelling, skin induration and hyperpigmentation over their lower limbs, with histology revealing a lymphocytic infiltrate and tissue necrosis in the subcutaneous fat layer.² In pretibial myxoedema, the areas affected are localised to the anterior-lateral shins, and is almost always associated with Grave's disease.³ The histological findings of pretibial myxoedema include increased mucin production and stellate-shaped fibroblasts, but there is no increase in the number of fibroblasts.³

Another much rarer condition that also causes diffuse indurated skin is scleredema, but it typically affects the upper body, and is associated with a history of a prior upper respiratory tract streptococcal infection, diabetes mellitus or monoclonal gammopathy. Histologically, scleredema is due to an excess production of both collagen and mucin in the dermis; the absence of fibroblast proliferation differentiates it from scleromyxoedema.¹

Management of scleromyxoedema includes the evaluation of internal organ involvement and underlying malignancies. Various therapies have been tried to varying success, such as intravenous immunoglobulin, thalidomide and corticosteroids.¹

Conclusion

Diseases resulting in hardening of the skin are characterised by an increase in collagen production, mucin production, and/or the number of fibroblasts in the dermis. These require to be differentiated, as there can be different associated systemic involvements and malignancies, and therefore varying treatment options and prognoses.

REFERENCES

1. Boin F, Hummers LK. Scleroderma-like fibrosing disorders. *Rheum Dis Clin North Am* 2008;34:199-220.
2. Kirsner RS, Pardes JB, Eaglestein WH, Falanga V. The clinical spectrum of lipodermatosclerosis. *J Am Acad Dermatol* 1993;28:623-7.
3. Holahan HM, Farah RS, Swick BL. Pretibial myxedema. *Cutis* 2014;94:60,73-4.

Vanessa ZY Lim,¹ MBBS, MRCP (UK), Hong Liang Tey,^{1,2} MBBS,
FRCP (Edin), FAMS

¹National Skin Centre, Singapore

²Lee Kong Chian School of Medicine, Singapore

Address for Correspondence: Dr Tey Hong Liang, National Skin Centre, 1 Mandalay Road, Singapore 308205.
Email: teyhongliang111@yahoo.com