Dermatological Quiz

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A 50-year-old woman complained of one year’s history of insidious onset of this mildly itchy skin rash around her upper back (Fig. 2a), proximal limbs (Fig. 2b) and her face extensively. The skin rash was associated with weight loss. Complete blood picture, liver and renal function tests were normal except a raised ESR of 70mm/hr. Skin biopsy at the papular lesion showed increased mucin deposition within the dermis with no epidermal change. There was an increase in irregularly arranged spindle cells in the dermis identified as dermal fibroblasts with intervening dermal fibrosis.

Questions:
1. What is your provisional diagnosis or differential diagnoses?
2. What other important specific tests you would like to order for investigating her weight loss?
3. How will you manage this patient?

Fig 2: Skin lesion at (a) upper back and (b) right upper arm

1. This middle aged woman presented with this quite asymptomatic extensive multiple shiny waxy a few millimetre small discrete whitish to skin-coloured papules which coalesced into larger papules/ small plaques affecting her face, upper back and limbs. The clinical differential diagnoses should include various forms of cutaneous mucinosis, lichen amyloidosid, scleredema of Buschke, scleroderma, eruptive xanthoma, eruptive syringoma and generalised myxoedema. Coupled with the history of systemic upset and histology showing increased dermal mucin deposition and fibroblast proliferation, the clinicopathological diagnosis of scleromyxoedema, which is a generalised form of cutaneous mucinosis, can be made.

Primary cutaneous mucinosis is a heterogenous group of disorders in which an abnormal amount of mucin accumulates in the skin with unknown pathophysiology. The widespread distribution form, as demonstrated in this patient, is known as scleromyxoedema. It presents with generalised shiny waxy small discrete whitish to skin-coloured papular eruptions with or without sclerodermoid features. Histologically, this rare disease is characterised by a triad of a) diffuse deposits of mucin in the dermis b) an increase in collagen deposition c) a marked proliferation of dermal fibroblasts arranged irregularly.

2. Scleromyxoedema is almost always associated with paraproteinaemia. The monoclonal gammopathy is usually IgG with gamma light chains. Less than 10% of scleromyxoedematous patients progress to multiple myeloma. As a result, a malignancy screening especially with immunoglobulin level and serum protein electrophoresis confirmed the presence of monoclonal gammopathy in this patient. Bone marrow aspiration did not show any myeloma changes.

3. There is no well proven effective treatment for scleromyxoedema because of the rarity of the disease. The commonly used approach is to treat the underlying paraproteinaemia or haematological malignancies which may alleviate the cutaneous mucinosis. In patients with confirmed myeloma, monthly melphalan combined with various drugs such as thalidomide, systemic steroids, and/or autologous haematopoietic stem cell transplantation are employed. A similar strategy has begun to be adopted for treating scleromyxoedema. However, the risks of marrow suppression and sepsis should be carefully weighed against the benefits. Other modalities which have been reported effective anecdotally include topical and intralesional hyaluronidase, PUVA, UVA1, systemic retinoids, electron-beam radiation, IVLg, plasmapheresis, extracorporeal photochemotherapy, cyclosporine, and granulocytes colony-stimulating factor.

Answer to Dermatological Quiz

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