

Case Report

A middle-aged woman with multiple itchy waxy papules: a case of scleromyxoedema

一名中年女仕之多處及瘙癢的蠟樣丘疹：硬化性黏液水腫一例

AWM Au 區慧明 and KC Lee 李景中

A 58-year-old woman presented with multiple itchy, skin-coloured, waxy papules over her face, back, chest and limbs for one year. The histology revealed papular mucinosis with marked fibrosis. Her thyroid function was normal and serum protein electrophoresis detected a kappa monoclonal band. Therefore, this was a case of scleromyxoedema.

在過去一年，一名五十八歲女仕的面部、背部、胸口及四肢各處，展現了多處瘙癢的肌膚色蠟樣丘疹。其皮損組織學檢查揭示了丘疹性黏蛋白沉積及顯著的纖維化。她的甲狀腺功能正常，而血清蛋白電氣泳則發現一 kappa 單株帶。因此，這一病例應為硬化性黏液水腫。

Keywords: Lichen myxoedematosus, papular mucinosis, scleromyxoedema

關鍵詞：黏液水腫性苔蘚，丘疹性黏蛋白沉積，硬化性黏液水腫

Case history

A 58-year-old woman presented with multiple itchy, skin coloured waxy papules over her face, trunk and limbs for 1 year. The onset of the skin

lesions was insidious. Initially, the lesions affected only the wrists and forearms, but then progressed to involve also the back and face. Concerning her past health, she had history of hypertension, and no history of drug allergy. She had consulted many doctors and was empirically treated with topical steroids and topical retinoid without obvious improvement.

Social Hygiene Service, Department of Health, Hong Kong

AWM Au, MBBS, MRCP

Department of Pathology, Queen Elizabeth Hospital, Hong Kong

KC Lee, FRCPath, FHKAM(Pathology)

Correspondence to: Dr. AWM Au

Yaumatei Dermatological Clinic, 12/F, Yaumatei Specialist Clinic, 143 Battery Street, Kowloon

Physical examination showed multiple small whitish to skin-coloured papules, ranging from one to few millimeters in diameter, with some coalescing into plaques, over the face, upper chest, upper back and bilateral arms (Figure 1).

Differential diagnoses included lichen nitidus, lichen amyloidosis, lichen myxoedematosus,

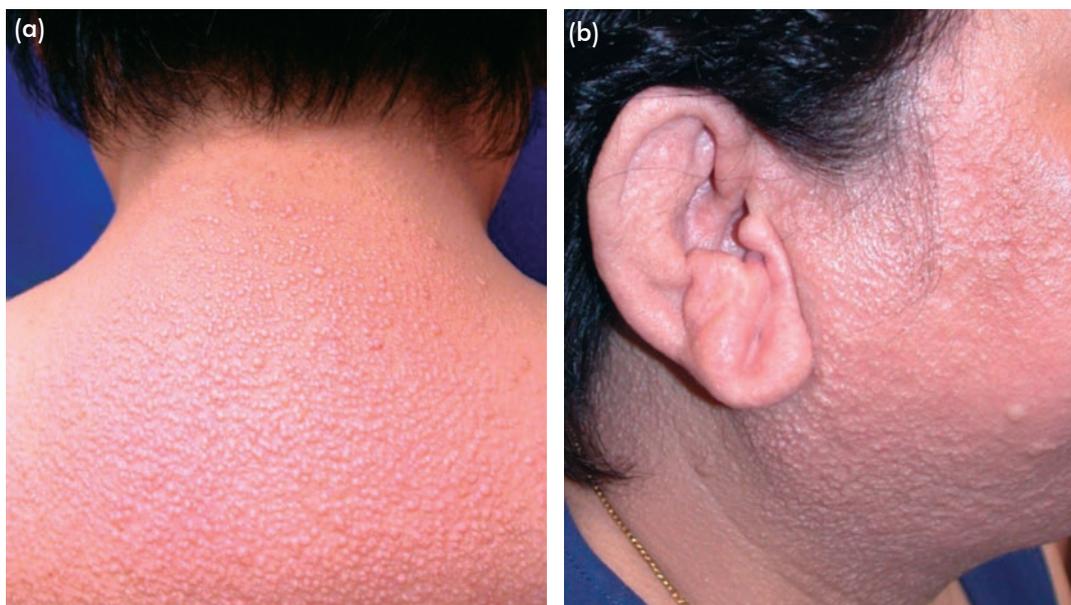


Figure 1. (a) Multiple small waxy papules over the neck and upper back. (b) Thickened earlobe and facial papules.

scleromyxoedema, scleroderma, scleredema, plane warts, eruptive xanthoma and eruptive syringoma.

Initial investigations which included blood tests for haemoglobin, white cell count, platelet count, liver and renal function, and thyroid function were all normal. ESR was 22 mm/hr. The fasting blood sugar was raised at 6.3 mmol/L. Serum IgG, IgA and IgM level were also normal.

Skin biopsy showed superficial dermal nodules causing slight elevation of the epidermis. The epidermis was otherwise unremarkable except for mild hyperkeratosis over the nodules. There was no evidence of interface change (Figure 2a). In a higher power view, irregularly arranged spindle cells were seen. The spindle cells were negative for histiocyte markers CD163 and PGM1, and were therefore most probably fibroblasts (Figure 2b). Mucin deposition within the papule was demonstrated by mucin stains (Figure 2c). The histological diagnosis of this case was papular mucinosis.

In view of the histological findings, further investigations were done. Malignancy screening

including carcinoembryonic antigen, alpha-fetoprotein, lactate dehydrogenase, chest X-ray, and stool for occult blood were negative. Thyroid function was normal. However, serum protein electrophoresis detected a kappa monoclonal band without immunoparesis. β_2 -microglobulin was 1.78 $\mu\text{g/ml}$ (normal < 1.43 $\mu\text{g/ml}$). The findings of relatively widespread skin involvement, mucinosis with increased fibroblasts and the presence of monoclonal gammopathy were most compatible with scleromyxoedema.

Discussion

Mucinosis is a heterogeneous group of disorders in which abnormal amount of mucin accumulates in the skin. There are two major groups of mucinosis. The first group is the primary mucinosis in which mucin deposition is the major histological feature and which results in distinctive clinical lesions. Examples of primary mucinosis include papular mucinosis, pretibial myxoedema, and scleredema, etc. The other group is the secondary mucinosis where mucin deposition is an associated finding. Examples are basal cell carcinoma, granuloma annulare and cutaneous T-cell lymphoma, etc.

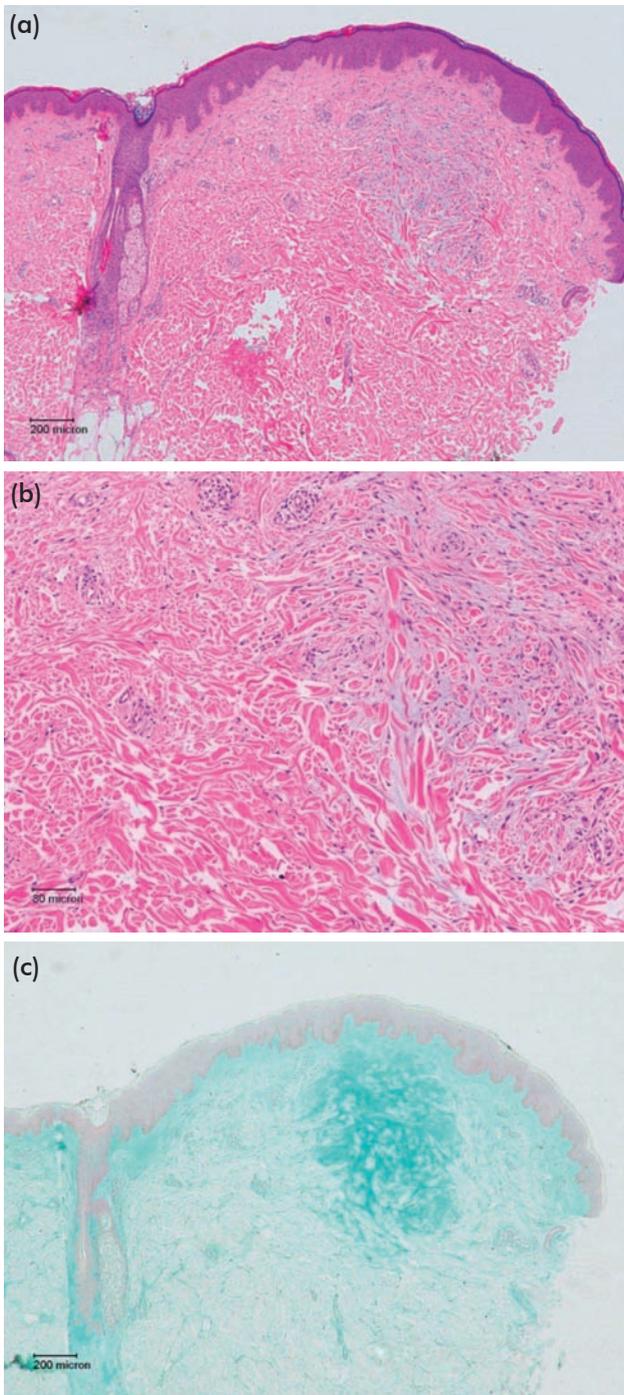


Figure 2. (a) Superficial dermal papule with mild hyperkeratosis but no interface changes. (b) Increased irregularly arranged fibroblasts with marked fibrosis. (c) Mucin deposition of dermal papule.

Papular mucinosis belongs to the group of primary cutaneous mucinosis. In literature, the term papular mucinosis and lichen myxoedematosus are often used indiscriminately.¹ It can be further classified into scleromyxoedema, a generalised lichenoid plaque form and other localised variants of lichen myxoedematosus. Scleromyxoedema should be distinguished from the localised variants because scleromyxoedema is frequently associated with monoclonal gammopathy and other systemic diseases such as myeloma which can be fatal, while the localised variants are usually chronic and do not run a disabling course.² Table 1 summarises the diagnostic criteria of scleromyxoedema.¹

Scleromyxoedema is an uncommon disease of unknown pathogenesis. It typically affects middle-aged adults without sex predilection. Clinically, the condition is characterised by a widespread symmetrical eruption of 2-3 mm, firm, waxy, closely spaced papules that are commonly arranged in a linear pattern, and are most commonly located on the hands, forearms, face, neck, upper trunk and thighs. The skin around the lesion is shiny and resembles scleroderma. The glabellar area is typically involved with deep longitudinal furrowing. Calcinosis and telangiectasia are always absent.³

Histologically, scleromyxoedema is characterised by a triad of microscopic features:⁴ (1) a diffuse deposit of mucin in the upper and mid reticular dermis; (2) an increase in collagen deposition; and (3) a marked proliferation of irregularly arranged fibroblasts.

Scleromyxoedema is almost always associated with paraproteinaemia. The monoclonal gammopathy is usually IgG with γ light chains. Although a mild plasmacytosis may be observed in bone marrow biopsies, less than 10% of patients with scleromyxoedema progress to multiple myeloma. Patients with scleromyxoedema can have a number of internal manifestations—muscular, neurologic, rheumatologic, pulmonary, renal and cardiovascular. Dysphagia, proximal muscle weakness due to myositis, disturbances of

Table 1. Diagnostic criteria of scleromyxoedema

Scleromyxoedema	Localised variants of lichen myxoedematosus
Generalised papular eruption and sclerodermoid features	Papular eruption (or nodules and/or papules due to confluence of papules)
Microscopic triad (mucin deposition, fibroblast proliferation, fibrosis)	Mucin deposition with variable fibroblast proliferation
Monoclonal gammopathy	Absence of monoclonal gammopathy
Absence of thyroid disorder	Absence of thyroid disorder

the CNS leading to unexplained coma, peripheral neuropathy, arthropathy, carpal tunnel syndrome, restrictive or obstructive lung disease, and a scleroderma-like renal disease may accompany or follow the cutaneous manifestations.³

There is no known effective treatment of scleromyxoedema. There have not been any well-designed clinical trials at the time of writing due to the rarity of the disorder. The commonly adopted approach is to treat the underlying paraproteinaemia or haematological malignancies.³ In patients with myeloma, high dose melphalan followed by stem cell support has been used successfully.⁵ Other chemotherapeutic agents (e.g. cyclophosphamide, methotrexate, chlorambucil, 2-chlorodesoxyadenosine) have been tried with similar results.⁶ However, the use of these drugs might be complicated by bone marrow suppression and severe sepsis.

Systemic corticosteroids are also often used but with limited effects. Topical corticosteroid and intralesional steroids have been tried with some success. Anecdotal reports from case series found improvement with the use of topical and intralesional hyaluronidase, PUVA, UVA1, systemic retinoids, electron-beam radiation, IVIg, plasmapheresis, extracorporeal photochemotherapy, dermabrasion, and topical dimethyl sulfoxide without treating the

underlying disease.⁵ Granulocyte colony-stimulating factor proved beneficial in one patient with idiopathic neutropenia, as did cyclosporine in a second patient. Interferon- α has led to paradoxical effects, with both improvement and worsening of the disease.³

This patient was referred to the medical unit for further investigation of underlying haematological diseases such as myeloma. Meanwhile, she also sought treatment from herbalist for symptomatic relief.

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