

A Gentleman with Generalized Waxy Papules

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CASE SUMMARY

History

A 51-year-old gentleman presented with mildly itchy papules over the abdominal wall spreading to back, face and limbs for three years. He was given oral antihistamine, antibiotics and topical steroid by private doctors but improvement was not noted. There was no weight loss. Systemic symptoms including muscle weakness, dysphagia, nasal regurgitation, joint pain, Raynaud's phenomenon, dyspnoea and symptoms suggestive of peripheral neuropathy were all absent. He was on ramipril for IgA nephropathy and had a history of pulmonary tuberculosis in the past.

Physical examination

Multiple discrete waxy papules were present over the abdominal wall, neck, face, limbs and hands (Figure 1). The doughnut sign was noted at the proximal interphalangeal joints (Figure 2). There were neither vesicles nor pustules. Most of the skin papules were skin-coloured, with some lesions more erythematous and some pigmented. The buccal mucosa and nails were all normal. There were neither hepatosplenomegaly nor peripheral lymphadenopathy.

Differential diagnosis

The clinical differential diagnoses included eruptive syringoma, transient acantholytic dermatosis, Darier's disease, generalized granuloma annulare, lichen amyloidosis, lichen scrofulosorum and scleromyxoedema.

Investigations

The complete blood picture, liver and renal function tests, erythrocyte sedimentation rate (ESR)



Figure 1: Close up view of patient's arm showing the discrete waxy flesh colour papules



Figure 2: Discrete papules over dorsum of hand and the doughnut sign over the proximal interphalangeal joints

were normal. Other blood tests including creatinine phosphokinase (CPK), lactic dehydrogenase (LDH), thyroid function test, anti-nuclear factor and anti-ENA were also normal. Serum immunoelectrophoresis revealed an IgG λ paraprotein; IgA and IgM levels were not suppressed. Urine immunofixation also showed a faint IgG λ paraprotein. Chest X-ray showed granulomas in the left apex and fibrosis in both upper zones suggestive of old tuberculosis. An incisional skin biopsy was done on his left shoulder. It showed mild epidermal hyperplasia overlying dermal papules, which were composed of fibroblastic proliferation in mucin deposition. Pigmentary incontinence was apparent. Inflammatory infiltrate and sclerodermoid background were not noted. No amyloid deposit was noted. The histological features were compatible with scleromyxoedema.

Progress

He was given topical steroid but the lesions remained static. Dermabrasion was tried on the posterior neck and upper back of the patient. It resulted in flattening of the papules. He was referred to the physician for work up of his paraproteinaemia.

REVIEW ON SCLEROMYXOEDEMA

Definition¹

Lichen myxoedematosus is a disorder characterized by lichenoid papules, nodules and/or plaques due to mucin dermal deposition and a variable degree of fibrosis without thyroid dysfunction. It is further subdivided into three forms: generalized papular and sclerodermoid form (also called scleromyxoedema), a localized papular form and an atypical or intermediate form that does not meet the criteria for either scleromyxoedema or the localized form. Our patient has features compatible with scleromyxoedema. Diagnosis of scleromyxoedema should fulfill the following criteria: (1) generalized papular and sclerodermoid eruption; (2) mucin deposition, fibroblast proliferation and fibrosis; (3) monoclonal gammopathy; and (4) the absence of thyroid disease.

Pathogenesis

The exact pathogenesis is not known. It is suspected that paraprotein can directly stimulate fibroblast proliferation and mucin deposition. Previous study had shown sera from patients with lichen

myxoedematosus stimulated fibroblast proliferation in vitro, but removal of paraprotein did not affect fibroblast proliferation.² This suggested that another serum factor, not the paraprotein, was responsible for the fibroblast stimulation. Another group was able to show that sera from patients with lichen myxoedematosus stimulated hyaluronic acid and prostaglandin production by human fibroblast.³

Epidemiology

Scleromyxoedema is a rare disease. It usually occurs in middle-aged adults with no sexual predilection.

Clinical features^{1,4}

Cutaneous involvement manifests as a symmetric eruption of tiny closely spaced papules located at hands, forearms, face, neck, upper trunk and thighs. The skin nearby is shiny and oedematous. Erythema and a brownish discoloration may also be present in the involved areas. The glabella may be involved with deep longitudinal furrows. Skin stiffening with decreased mobility of mouth and joints may occur at a later stage. An elevated rim with a central depression due to skin thickening may be seen over the proximal interphalangeal joints of the hands ("doughnut sign").

Systemic involvement is usual in scleromyxoedema. Paraprotein is found in 83.2% of cases, mostly of the IgG λ subtype. Progression to multiple myeloma occurs in only 10% of cases. Other systemic involvement includes proximal myopathy, seronegative arthritis, Raynaud's phenomenon, dysphagia or nasal regurgitation secondary to gastrointestinal involvement, scleroderma-like renal disease, dyspnoea secondary to lung involvement and pulmonary hypertension. Central nervous system involvement includes confusion, dizziness, vertigo, dysarthria, ascending paralysis, seizures, syncope and coma. Peripheral neuropathy and carpal tunnel syndrome have also been reported.

Pathology²

Scleromyxoedema has a histological triad of diffuse deposit of mucin in the upper and mid dermis, increase in collagen deposition and a proliferation of

irregularly arranged fibroblasts. The epidermis may be thinned by the pressure of underlying mucin. A slight perivascular, superficial lymphocytic cellular infiltrate is often present.

Differential diagnosis

This condition has to be distinguished from generalized granuloma annulare, which can also be presented clinically with diffuse cutaneous papules and histologically with mucin deposition. In generalized granuloma annulare, the eruption can be arranged in a circular or arciform pattern and histopathologically, necrobiosis of collagen and a palisading or interstitial histiocytic infiltrate may be present. In localized lichen myxoedematosus, systemic involvement and monoclonal gammopathy are absent. It is also important to distinguish scleromyxoedema from the other primary cutaneous mucinosis such as dysthyroidotic mucinosis, papular-nodular mucinosis associated with lupus erythematosus, papular mucinosis of the toxic oil syndrome and the L-tryptophan associated myalgia-eosinophilia syndrome.

Treatment¹

As a rare disease, most reported treatment modalities were in the form of case reports or small series. The treatments reported to be useful for cutaneous disease include topical and intralesional hyaluronidase, topical and intralesional steroid, topical steroid with dimethyl sulfoxide, PUVA, Grenz ray, electron beam, dermabrasion and G-CSF. Treatments that have reported to be useful for cutaneous and systemic features include systemic steroid, oral retinoids, cyclosporin, melphalan, cyclophosphamide, interferon- α , intravenous immunoglobulin, autologous stem cell transplantation,⁷ plasmapheresis and extracorporeal photochemotherapy. In general, toxic drugs should be limited to patients who are disfigured, disabled or very ill.

Learning points:

Screening for the presence of paraprotein and thyroid function abnormalities should be performed in patients with scleromyxoedema.

References

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Web sites of Dermatology & Venereology in Hong Kong

The homepage of the Hong Kong Society of Dermatology & Venereology
<http://www.medicine.org.hk/hksdv/>

Hong Kong Dermatology & Venereology Bulletin
(Official Publication of the Hong Kong Society of Dermatology & Venereology)
<http://www.medicine.org.hk/hksdv/bulletin.htm>

Handbook of Dermatology & Venereology
(Published by Social Hygiene Service, Department of Health)
<http://www.hkmj.org.hk/skin/>

CME Online (Dermatology)
(CME Programme accredited by the Hong Kong College of Family Physicians)
<http://www.medicine.org.hk/cme/>

The Homepage of the Asian Dermatological Association
<http://www.medicine.org.hk/ada/>

