Spontaneously resolving localized lichen myxoedematosus – adult onset variant

S V Paranavithane[1], K Satgurunathan[2]

Sri Lanka Journal of Dermatology, 2009, **13**, 32-33

Localized lichen myxoedematosus is a rare clinical entity which is not associated with paraproteinaemia. Spontaneous resolution is a recognized feature. Herein, we report a case of spontaneously resolving localized lichen myxoedematosus – adult onset variant to increase awareness of this rare entity.

Case report

A 53-year old male presented with mildly itchy thickening of skin over neck, ears, shoulders, upper back and chest for six weeks and thickened skin on face for two weeks.

He did not have a history of scre throat or sense of choking. On examination, yellowish and skin coloured papular induration was noted on neck, ears and upper trunk with an ill defined margin. Skin was movable over the induration. There was diffuse thickening of skin on face. He did not have tight skin on fingers.

The lesions on trunk and ears started to resolve spontaneously within two months of onset.

Histology of skin biopsy showed abundant mucin with soap bubble appearance and collagen strands in a lake of mucin. Collagen was not thickened or abnormal. Fibroblasts were not increased in number. There was a mild perivascular lymphocytic infiltrate.

Alcian blue stain was positive.

Basic investigations including fasting blood glucose were normal. Serum antistreptolysin O titre was less than 200 Todd units. Serum protein electrophoresis was normal. Thyroid functions were normal. Ultra sound scan of the abdomen showed mild hepatomegaly.

Discussion

The differential diagnoses in this type of clinical picture are, localized lichen myxoedematosus[1] and scleroedema.

There are clinical as well as histological differences between these two conditions. Clinically, lichen myxoedematosus comprises firm waxy lichenoid papular induration of skin with well defined borders usually occuring on face, hands and trunk. It may be pruritic. Insidious onset and persistent lesions are characteristic. The skin may be pinched above the induration.

In contrast, scleroedema is a diffuse non pitting woody induration of the skin, mainly localised to upper back and is not sharply demarcated from normal skin. It occurs suddenly and resolves spontaneously in most of the cases. The skin cannot be pinched in scleroedema. Patients with scleroedema experience a sense of choking.

Spontaneous resolution of localised lichen myxoedematosus is also a recognised feature. Histologically, in both conditions, abundant mucin in dermis, Alcian blue positivity at pH 2.5 (blue) indicating high levels of hyaluronic acid content, and dispersed collagen bundles are seen.

Lichen myxoedematosus has the hallmark feature of fibroblast proliferation, showing large stellate fiblroblasts. In scleroedema, fibroblast proliferation does not occur.

In our patient, spontaneously resolving localized lichen myxoedematosus[2] was diagnosed on clinical grounds, aided by the presence of abundant mucin in the dermis in histology. The typical histological features of lichen myxoedematosus such as, thickened, abnormal collagen and fibroblast proliferation[3,4] were absent in our patient. However, a mild perivascular lymphocytic infiltrate was noted.

Nevertheless, lichen myxoedematosus is described without these typical histological features by Rongioletti F and Rebora A in their updated classification of papular mucinosis, lichen myxoedematosus and scleromyxoedema[5].

Acccordingly, lichen myxoedematosus (LM) includes two main clinicopathologic subsets: A

[1]*Senior Registrar in Dermatology*, [2]*Consultant Dermatologist, Dermatology Department, National Hospital of Sri Lanka.*

generalized papular and sclerodermoid form (also called scleromyxedema) and a localized papular form. A third group of atypical or intermediate forms of LM, not meeting the criteria for either scleromyxedema or the localized form, is also described.

The diagnostic criteria for localized LM are as follows:[6]

(1) papular or nodular/plaque eruption,

(2) mucin deposition with variable fibroblast proliferation,

(3) the absence of both monoclonal gammopathy and thyroid disease.

Our patient fulfils all three of the above criteria for localized lichen myxoedematous and belongs to the variant known as "self healing papular mucinosis of juvenile and adult type".

References

1. Burns T, Breathnach S, Cox N, Griffiths C. Rook's Textbook of Dermatology: 7th Edition, Blackwell Science Publications 2004; **57**: 23-57.

2. Bolognia J, et al. Dermatology Mosby Publications. First Edition 2003; 647-50.

3. Fitzpatrick, et al. Dermatology in General Medicine, Hill Medical Publications, 7th Edition 2008; 536-7.

4. Pathology of skin, Weedon, Churchill Livingston Publications 2002; 408-11.

5. Rongioletti F, Rebora A. Updated classification of papular mucinosis, lichen myxoedematous and scleromyxoedema. *JAAD* 2001; **44**: 273-81.

6 Rongioletti F, Rebora A. Cutaneous mucinoses, microscopic criteria for diagnosis. *American Journal of Dermatolpathology* 2001; **23**: 257-67.