Case No. 16

Authors: Nicha Jenmanachaiyakun, M.D.
 Niorn Boonpuen, M.D.

Patient: A 48-year-old Thai woman from Bangkok

Chief complaint: Multiple whitish papules on the chest wall, upper back, both forearms, and both dorsum of hands and feet for 5 months

Present illness:
The patient gradually developed multiple asymptomatic whitish papules on the chest wall for 5 months. She was treated with topical corticosteroids at the clinic for 1 month without improvements. The lesions further developed on the upper back, both forearms, and both dorsum of hands and feet. The patient had no systemic symptoms.

Past history:
No history of trauma or exposure to chemical compound
No history of keloidal scar

Family history:
Neither similar skin lesions nor malignancy presented in her family members

Physical examination:
   General appearance: Good consciousness, not pale, no jaundice
   Normal CNS system
   No hepatosplenomegaly
   No lymphadenopathy

Dermatological examination:
   Multiple discrete flat-topped whitish papules on the chest wall, upper back, both forearms, and both dorsum of hands and feet.

Investigations:
CBC: Hb 11.5 g/dL, Hct 34.6%, WBC 6,000 cells/mm³, N 54%, L 36%, Mo 6%, Eo 3%, Plt 299,000 cells/mm³, MCV 76 fl BUN 11.9 mg/dL, Cr 0.9 mg/dL
LFT: Total protein 8.0 g/dL, Albumin 4.3 g/dL, Total bilirubin 0.35 mg/dL, Direct bilirubin 0.20 mg/dL, AST 18 U/L, ALT 12 U/L, ALP 59 U/L
TFT: Free T3 3.0 pg/mL (2.3-4.2), Free T4 1.1 ng/dL (0.9-1.8), TSH 6.602 µIU/mL (0.55-4.78)
Anti-HIV: negative, HCV Ab: negative, RPR: non reactive
Chest X-ray: WNL
Serum protein electrophoresis: Total protein 8.2 g/dL, A/G ratio 0.85 L,
Albumin 3.76 g/dL (3.2-5.0), Alpha1 globulin 0.21 g/dL (0.1-0.4), Alpha2 globulin 0.82 g/dL (0.6-1.0), Beta globulin 1.3 g/dL (0.6-1.3), Gamma globulin 2.12 g/dL (0.7-1.5)

Histopathology: Slide No. 59-0645 (right shoulder)
   Sections display unremarkable epidermis with hyperkeratosis. Neither spongiosis nor interface change is noted. No necrotic keratinocyte is present. The dermis shows a mild superficial perivascular infiltration of lymphocytes. No pigmented incontinence is seen. In addition, some fibroblastic proliferation is noted in the dermal interstitium with increased deposition of mucin demonstrated by alcian blue stain. The overall features are suggestive for lichen myxedematosus.

Diagnosis: Lichen myxedematosus (discrete papular subtype)

Treatment: Advice the patient that the disease rarely resolves spontaneously.

Discussion:
Lichen myxedematosus or papular mucinosis is an idiopathic cutaneous mucinosis.1 It is characterized by small, multiple, asymptomatic, firm, waxy papules (or nodules and plaques produced by the confluence of papules) usually on the upper and lower limbs and trunk.2 The skin is the only site of involvement. Lichen myxedematosus is not associated with sclerosis, paraproteinemia, systemic involvement or thyroid disease, in contrast to scleromyxedema (Table 1).3

Table 1. Diagnostic criteria of scleromyxedema versus lichen myxedematosus

<table>
<thead>
<tr>
<th>Scleromyxedema</th>
<th>Lichen myxedematosus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Generalized papular eruption and sclerodermitis features</td>
<td>Papular eruption (or nodules and/or plaques due to confluence of papules)</td>
</tr>
<tr>
<td>Microscopic triad (mucin deposition, fibroblast proliferation, fibrosis)</td>
<td>Mucin deposition with variable fibroblast proliferation</td>
</tr>
<tr>
<td>Monoclonal gammopathy</td>
<td>Absence of monoclonal gammopathy</td>
</tr>
<tr>
<td>Absence of thyroid disorder</td>
<td>Absence of thyroid disorder</td>
</tr>
</tbody>
</table>
The pathogenesis of lichen myxedematosus remains unknown. The incidence and prevalence rates are not known. The localized variants of lichen myxedematosus are subdivided into four subtypes: (1) a discrete papular form; (2) acral persistent papular mucinosis; (3) cutaneous mucinosis of infancy; (4) a pure nodular form (Table 2). Localized variants of lichen myxedematosus have been reported, in patients with HIV infection, hepatitis C viral infection, and also in generalized morphea, in patients with morbid obesity, subclinical hypothyroidism, and in the setting of toxic oil syndrome and L-tryptophan-associated eosinophilia-myalgia syndrome.

Table 2. Localized variants of lichen myxedematosus

<table>
<thead>
<tr>
<th>Characteristic of lesion</th>
<th>Discrete papular type</th>
<th>Acral persistent type</th>
<th>Cutaneous mucinosis of infancy</th>
<th>Nodular type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Characteristic of lesion</td>
<td>2-5 mm papules</td>
<td>Multiple ivory to skin-colored papules</td>
<td>Firm opalescent papules</td>
<td>Multiple nodules</td>
</tr>
<tr>
<td>Location</td>
<td>Limbs, trunk, Spare face</td>
<td>Dorsal aspect of the hands, extensor surface of the distal forearms</td>
<td>Neck, upper arms (especially the elbows), trunk</td>
<td>Limbs, trunk</td>
</tr>
<tr>
<td>Prognosis</td>
<td>Rarely resolve spontaneously</td>
<td>Lesions persist without systemic manifestations</td>
<td>No spontaneous resolution</td>
<td>No systemic symptoms</td>
</tr>
</tbody>
</table>

Histopathologically, in localized forms of lichen myxedematosus, fairly large amounts of mucin are present. Mucin accumulates in the upper and mid reticular dermis, and is associated with variably increased fibroblasts. Fibrosis is not marked and may even be absent. There is no significant increase in dermal collagen. Histology helps to distinguish localized variants of lichen myxedematosus from several papular eruptions that can have a similar appearance, such as granuloma annulare, lichen amyloidosis, lichen planus and other lichenoid eruptions.

We reported one case of lichen myxedematosus in which clinical features and histology fit into the diagnostic criteria. Our case is a discrete papular subtype. Discrete papular lichen myxedematosus (DPLM) is a very rare entity, which affects both genders, however it has more effect on males than females. A PubMed search of the literature for published cases of DPLM found reports of 14 cases (Appendix 1).

Since localized lichen myxedematosus is usually limited to the skin, it has a good prognosis and does not require any treatment. Wait-and-see approach is recommended. Topical application of corticosteroids, pimecrolimus or tacrolimus may be of some benefit. However, spontaneous resolution may occur, even in the setting of HIV infection.

References:


