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Case report

Localised lichen myxedematosus: an infrequent entity

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ABSTRACT

Lichen Myxedematosus (LM) is seen within the spectrum of primary cutaneous mucinoses. Primary cutaneous mucinoses is a heterogeneous group of skin disorders characterized by abnormal dermal deposition of mucin. Here, we present a case of a 35-year-old individual with pruritic skin-colored to hyperpigmented discrete papules. These papules were distributed over the neck, upper trunk, lower back, scalp, and forearms, arranged in a rippled pattern for the past 3 months. Various investigations, including hemogram, serum biochemistry, thyroid profile, lipid profile, viral markers, serum electrophoresis, and histopathological examination, were conducted. Based on history and thorough examination, the case was diagnosed as lichen Myxedematosus (papular variant). The patient received treatment with topical steroids and systemic antihistamines, showing moderate improvement.

Keywords: Lichen myxedematosus, Cutaneous mucinoses, Heterogeneous

INTRODUCTION

Lichen myxedematosus (LM), is primary dermal mucinoses. Cutaneous mucinoses have abnormal dermal deposition of mucin. Mucin, composed of glycosaminoglycans, hyaluronic acid, and dermatan sulfate, is a component of the extracellular matrix and is produced by fibroblasts. Its main function is to maintain the salt and water balance within the dermis, as it can absorb 1,000 times its own weight in water.[1] Cutaneous mucinoses are classified into primary and secondary mucinoses. Primary mucinoses are in which the dermal mucin deposition is main feature whereas secondary mucinoses has dermal mucin deposition as an additional histological feature and secondary phenomenon. The cause of its abnormal cutaneous deposition remains unclear, although diverse proposed physiopathogenic mechanisms exist, including paraproteinemia and serum factors that could promote mucin production within the skin.[2]

Scleromyxedema is classified as mucinoses, which are a heterogeneous group of dermatoses with a common denominator of pathological deposits of mucin. It is broadly classified into generalized, localized, and atypical.

Here in, we report a case of localised papular variant of scleromyxedema who presented with cutaneous lesions over scalp and upper body.

CASE REPORT

A 35 year old female resident of Vizianagaram, house wife by occupation came with complaint of itchy, skin colored raised lesions over the neck, upper trunk, lower back, scalp and forearms for past 3 months. Patient was apparently normal 3 months back, then she developed skin colored raised lesions initially over the neck then on the trunk, lower back, forearms and later over the scalp. There was no history of pain, difficulty in moving the joints. There was no history of drug intake prior to onset

of lesions. Patient had no history of diabetes and hypertension. There was no significant family history. Patient had no relevant personal history. Rest of the history was unremarkable. General and systemic examination showed no abnormalities. On dermatological examination, hyperpigmented papules arranged linearly over the abdomen with rippled pattern of distribution at places. Multiple skin coloured papules closely arranged over the upper midline of back (Figure 1 and 2) and over the paravertebral area were seen. Skin coloured shiny papules seen over the upper back. Based on the complete history and clinical examination, differentials considered were extensive milia, localised sclermyxoedema, follicular mucinosis, and papular sarcoidosis.

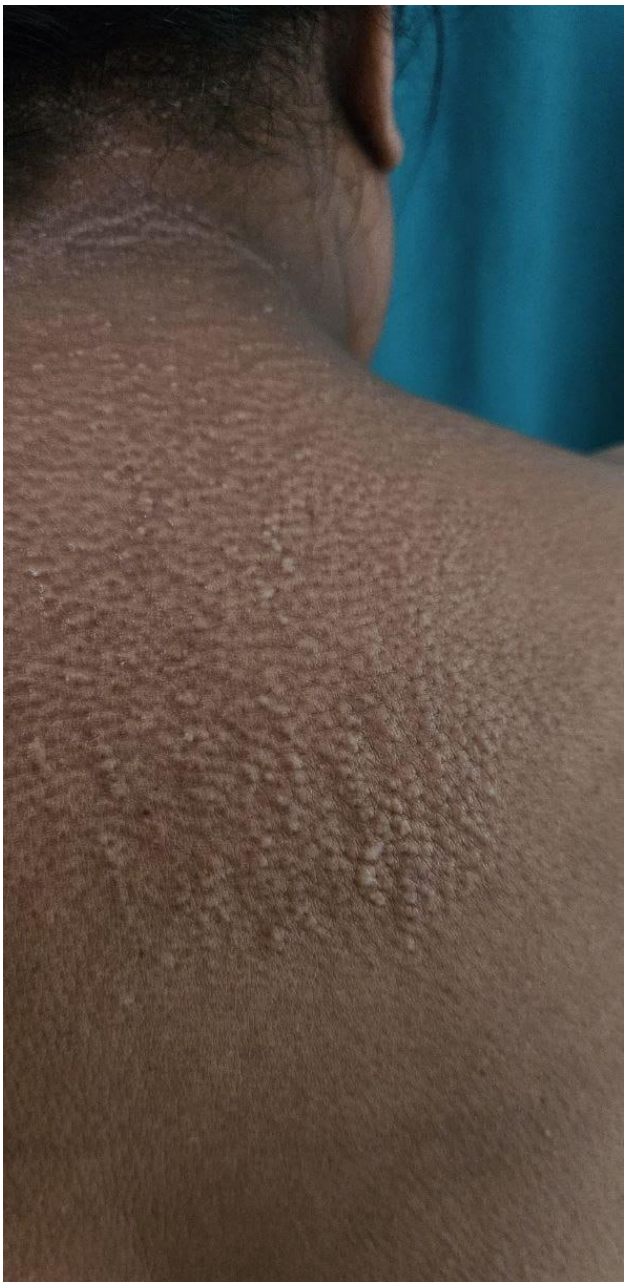

Figure 1: Multiple closely arranged papules with rippled pattern over back of upper trunk.

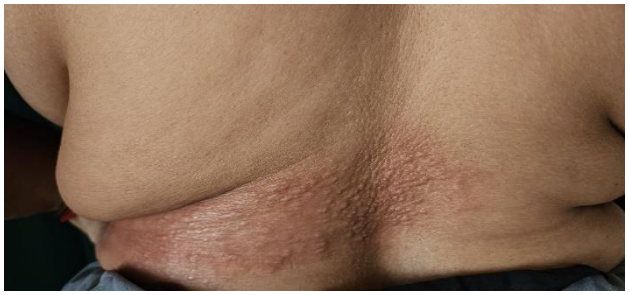

Figure 2: Multiple closely arranged papules with background of erythema on lower back.

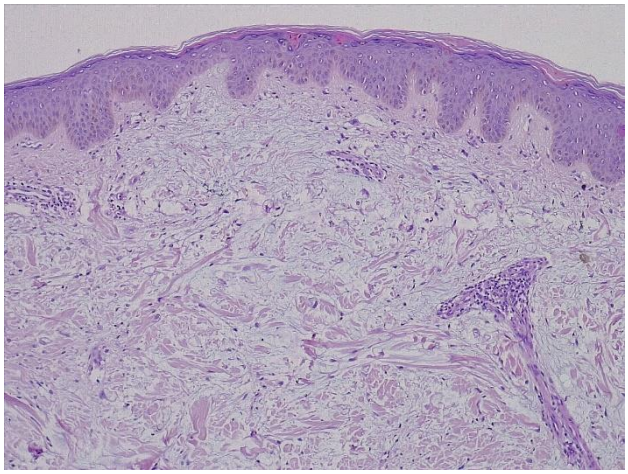

Figure 3: Histopathological image showing basket weave orthokeratosis, superficial dermis and deep dermis showing interstitial deposition of mucin (4x image).

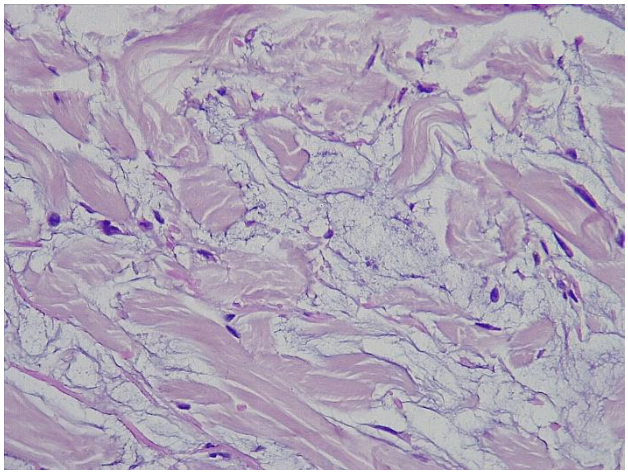

Figure 4: Abundant mucin deposition in superficial dermis.

A 4 mm punch biopsy was done from the lesion on back. Complete blood picture showed normal levels. Thyroid profile, lipid profile and urine examination were under normal limits. Biochemical tests including blood glucose, liver and renal function tests did not show any

abnormalities. Imaging studies including chest X-ray showed no abnormalities. Patient was seronegative for HIV, HBsAg, HCV. Serum electrophoresis showed no paraproteinemia. The histopathological examination (Figure 3 and 4) showed basket weave orthokeratosis, preserved granular layer, mild acanthosis and intact basal layer. Superficial dermis shows-abundant interstitial deposition of mucin along with foci of mild perivascular lymphocytic infiltrate. In deep dermis, mild interstitial deposition of mucin was seen which was suggestive of LM (papular mucinosis).

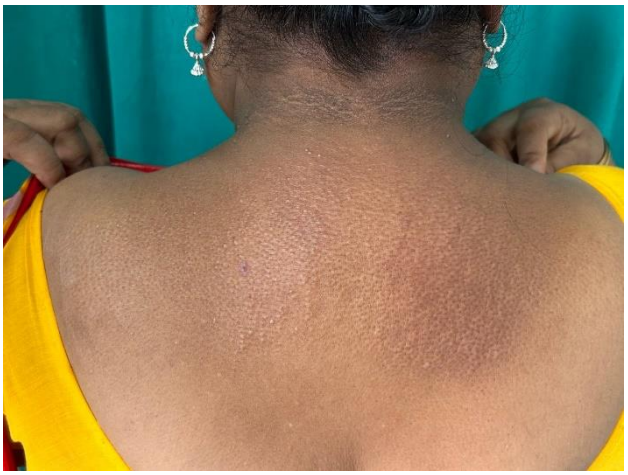

Figure 5: Post treatment - moderate improvement of lesions present.

The patient was treated with topical betamethasone dipropionate 0.05% cream over cutaneous lesions in the morning. Topical tacrolimus 0.1% ointment over the lesions in the night. For scalp lesions, halobetasol propionate 0.05% cream application was prescribed for night. Anti-histamines were also given. Pruritis subsided and moderate improvement to treatment was observed after 1month of treatment (Figure 5). Patient was being monthly monitored for the associated complications and is currently under follow up.

DISCUSSION

Papular mucinosis (PM), or LM, is clinically characterized as few to multiple, 2-5 mm, skin-colored, firm, waxy, dome - fashioned papules, some also can coalesce to form plaques, causing thickening and hardening of the skin.[3] The scalp and mucosa are rarely affected. In our case, the scalp is involved.

The etiology of PM isn't understood; but it is believed that PM takes location due to the proliferation of fibroblasts with fibrosis and excessive deposition of mucin (glycosaminoglycans) with inside the dermis. Paraproteins also play a function in scleromyxedema. Localized PM also can rise up due to community plasma cell dyscrasia, but distinctive triggering factors may be involved. Autoantibodies and cytokines along with interleukin-1, tumor necrosis factor-α, and transforming growth factor-β stimulate the fibroblast proliferation.[4] Histochemically, it is a heterogeneous mixture of acid glycosaminoglycans, which stain with alcian blue and toluidine blue.[7]

In classification for LM by Rongioletti et al distinguishes between three different subgroups of LM, each with different diagnostic criteria: LM includes two clinicopathologic subsets: a generalized papular and sclerodermoid form (also called scleromyxedema) and a localized papular form.[2]

Their diagnostic criteria for LM is papular or nodular eruption, mucin deposition with variable fibroblast proliferation, and without monoclonal gammopathy and thyroid disease.[10] They categorised localised LM into five subtypes: discrete papular LM, acral persistent papular mucinosis, self-healing papular mucinosis with adult and juvenile types, papular mucinosis of infancy, and nodular type."

However, not all cases of papular mucinosis fit these above two criteria. A third group of atypical or intermediate forms, which doesn't meet the criteria for either scleromyxedema or the localized form, includes: scleromyxedema without monoclonal gammopathy; localized form with monoclonal gammopathy and/or systemic symptoms; localized forms with mixed features of the 5 subtypes, and non specific cases.[4,5]

In present case, sclerodermoid appereance was absent. The classical clinical features and signs seen in generalized myxedematosus are leonine facies, donut sign (A central depression with raised margin over proximal interphalangeal joint) & sharpie sign (Deep furrowing evident on trunk, shoulder, limbs). Lesions were distributed over localised areas of body and no systemic involvement was seen favouring the diagnosis of LM.

The histopathological examination (HPE) of LM shows a triad of: diffuse mucin deposition, increased collagen deposition, and proliferation of irregularly arranged fibroblasts. The epidermis may appear normal or thinned in chronic lesions, secondary to pressure from underlying mucin. There are atrophied follicles and mild superficial perivascular lymphoplasmocytic infiltrate often present.

In scleromyxedema, a diffuse pattern of mucin deposition is observed. Mucin deposition in localized and generalized forms of LM may be found in the upper or mid-dermis. Due to morphological overlapping, differentiation between generalized and localized LM based only on histopathology is not always possible.[9]

Our case showed abundant interstitial deposition of mucin along with foci of perivascular lymphocytic infiltrate in superficial dermis and mild interstitial mucin deposition in deep dermis which favoured LM.

The treatment of PM is often challenging and may not always be effective. While scleromyxedema has shown some improvement with various therapies including Psoralen-ultraviolet A therapy (PUVA), oral glucocorticoids, retinoids, thalidomide, plasmapheresis, extracorporeal photopheresis, dermabrasion, carbon dioxide laser excision, melphalan, interferon-α, and intravenous immunoglobulin in isolated cases, treatment for PM remains less clear. Some cases of localized PM have responded successfully to topical tacrolimus and corticosteroids.[6]

In our case, successful treatment was achieved with topical corticosteroids and tacrolimus, resulting in visible improvement in lesion appearance, with ongoing follow-up.

Moreover, although our patient did not show evidence of multiorgan involvement in the workup, previous reports indicate that extracutaneous involvement can occur. Therefore, based on a review of systems, further laboratory tests and imaging studies should be considered.

CONCLUSION

The present case highlights a case of Localised Lichen myxedematosus who presented with classical skin lesions in rippled pattern in localised areas of body but without any comorbidities and systemic involvement.

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