Journal of Pakistan Association of Dermatologists. 2017; **27(3)**: 306-308.

PhotoDermDiagnosis

Generalized asymptomatic skin coloured papular eruption

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A 55-year-old man presented with 4-year history of multiple, asymptomatic, symmetric, discrete papules on forehead, nose, scalp, neck, beard area, behind the ears, dorsa of his hands, flexor aspects of forearms and back (**Figure 1**). The lesions started from his forehead, increased slowly in number and were persistent. They had no relationship to sun exposure. There was no history of other systemic involvement or any recent exposure to topical medications. No other family member was similarly affected.

Rest of the cutaneous and systemic examination did not reveal abnormal findings.

Laboratory investigations including complete blood count, ESR, liver enzymes, renal functions and thyroid profile were normal. Histopathological examination showed sparse superficial and deep, perivascular lymphocytic infiltrate. There were increased spaces between dermal collagen fibers due to increased connective tissue mucin deposition with focal areas of increased number of fibroblasts (**Figure 2**).

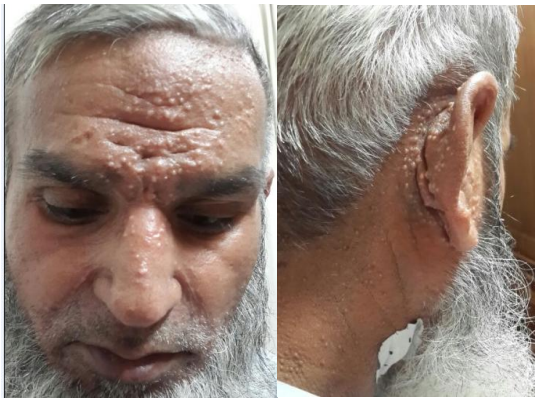

Figure 1 Multiple skin-colored papules on forehead and back of ears.

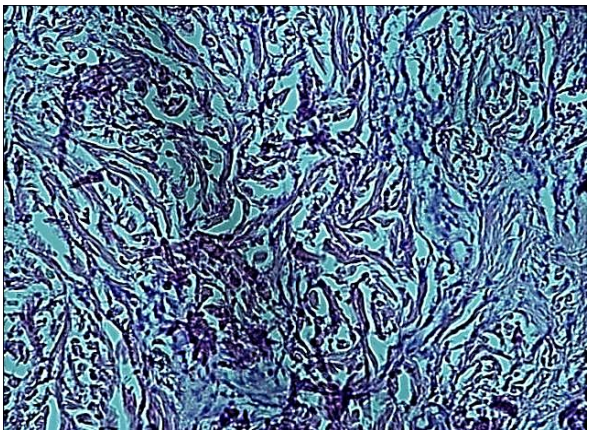

Figure 2 Increased spaces between with dermal collagen fibers.

What is your diagnosis?

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Diagnosis

Papular mucinosis (discrete papular lichen myxedematosus)

Discussion

Cutaneous mucinosis is a group of disorders characterized by abnormal deposition of mucin in the skin. Classification of cutaneous mucinosis can be done in several ways: either primary which is idiopathic and secondary in which underlying causes include endocrinopathies, connective tissue diseases and malignancies; by the pattern of distribution of the mucin which can be focal, diffuse or follicular; epithelial or dermal, the two types differing in their composition, as well as in the staining techniques required in order to identify them.[1]

Papular mucinosis (PM) or lichen myxedematosus, is a primary cutaneous mucinosis which is characterized by lichenoid papules, nodules or plaques due to dermal mucin deposition and fibroblast proliferation in absence of thyroid disorder.[2] The most accepted classification for papular mucinosis is by Rongioletti.[3] Papular mucinosis is classified into generalized and localized forms.[4,5,6]

Scleromyxedema is a generalized papular eruption with systemic implications characterized by sclerosis, paraproteinemia, and mucin deposition in the dermis.[7] Clinical features primarily include waxy, firm, 2-to-4-mm, flat-topped papules or dome-shaped papules. Papules often coalesce to form plaques. Less frequently, nodular, urticarial, or annular lesions may be appreciated. The most frequently affected areas are, dorsal aspect of the hands, face, elbows, and extensor surfaces of the extremities. Mucosae are not usually involved. The longitudinal folding of the glabella due to coalescence of papules gives rise to leonine face. There is also reduced mobility of the mouth, lips, joints of hands, arms and legs due to infiltration and sclerosis of skin. Systemic manifestations include proximal myopathy, inflammatory polyarthritis, central nervous system symptoms, esophageal aperistalsis, and hoarseness. Visceral involvement is associated with poor prognosis.

Localized PM is confined to a few sites and is not usually associated with sclerosis, paraproteinemia, systemic involvement and thyroid disease. In localized PM only few sites are involved. It is subclassified into four types; acral persistent PM, Discrete papular LM, cutaneous mucinosis of infancy and nodular LM. Discrete papular PM, the most common type of localized form occurs symmetrically over trunk and limbs with sparing of face and distal parts of extremities. Adults of both sexes are affected equally by papular mucinosis. The disease appears between 30-80 years of age.

Laboratory studies show an abnormal paraprotein in 90% of cases. Histologic examination shows mucin deposition in the upper and mid reticular dermis, increased fibroblast, dermal fibrosis and increased collagen deposition. It stains with alcian blue stain at pH 2.5 and is susceptible to hyaluronidase digestion.

Treatment of this condition is not much satisfactory.[8] Topical therapy is less or not effective. Clearance of lesions has been reported with cyclophosphamide[9] and melphalan, separately or in combination with prednisone. Retinoids, interferon-alpha, cyclosporine, calcineurin inhibitors, PUVA photochemotherapy, electron-beam therapy, intravenous immunoglobulins and dermabrasion have also been attempted. The overall prognosis for extensive disease is poor.

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