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Case Report

Acral persistent papular mucinosis: A rare variant of cutaneous mucinosis

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Abstract Acral persistent papular mucinosis (APPM) is a distinctive form of dermal mucinosis not associated with systemic diseases. We report such a case in a sixty years old male who presented with few small papular lesions over both of his ears. These were managed successfully with excision and cryosurgery.

Key words
Papular mucinosis, acral persistent papular mucinosis, scleromyxedema, lichen myxedematosus

Introduction

Papular mucinosis is one of the cutaneous deposit diseases that presents as flesh-colored dermal papules mostly on the acral parts of the body. There is confusion with regard to the terminology of this entity in the literature. Localized form of the disease has been called papular mucinosis or lichen myxedematosus and generalized, confluent papular forms with sclerosis are known as scleromyxedema.[1,2] Although papular mucinosis is frequently used as a synonym for all three forms, but more appropriately it should be restricted to only mild cases. Acral persistent papular mucinosis is now considered to be a separate entity. This acral condition is rare, affects adults of both sexes equally and appears between ages 30 and 80. It is chronic and may be progressive. The primary lesions are waxy, 2-to-4-mm, dome-shaped or flat-topped papules. Frequently, they may coalesce into plaques or appear in a linear array. Less frequently, urticarial, nodular, or sometimes annular lesions may be appreciated. The dorsal aspect of the hands, face, elbows, and extensor portions of the extremities are most frequently affected. Mucosal lesions are absent. The coalescence of papules on the face, particularly of the glabella, results in longitudinal folding and may give the appearance of a leonine facies.[1-4] In scleromyxedema, large parts of the body may be involved; the skin shows erythematous, scleroderma-like induration that is accompanied by reduced mobility of the lips, hands, arms, and legs. Systemic manifestations have been described (myopathy, polyarthritis, esophageal aperistalsis, and hoarseness)

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and visceral involvement may be fatal. Laboratory studies in mild acral forms are usually normal but may show an abnormal paraprotein, usually of the IgG-8 type in scleromyxedema and more generalized forms of the disease. This underlying suggestion of a plasma-cell dyscrasia prompts examination of the bone marrow, which may be normal or show increased numbers of plasma cells or even myeloma.[1-3,5,6] Histologic examination of papular mucinosis shows dermal deposition of mucin, best depicted with alcian blue or colloidal iron stains. There is an increase in the number of fibroblasts and dermal fibrosis. Mild localized form may heal spontaneously.[7] Other treatment options include potent topical steroids, dermabrasion, cryosurgery and local surgical excision. In case of more generalized forms, clearance of lesions has been reported with melphalan and cyclophosphamide alone or in combination with prednisone. Both isotretinoin and etretinate have been associated with improvement. Interferon-alpha, cyclosporine, PUVA photochemotherapy, electron-beam therapy, IVIg have also been tried.[1,2,5,6,8,9] The prognosis is good in case of limited cutaneous disease but for extensive disease, it is generally poor. The purpose of reporting this case was to highlight this rare cutaneous deposit disorder.

Case report

A 60-year-old, otherwise healthy man reported with a two year history of 3-4, firm, skin colored papules on each ear. These were painless and non itchy. He did not notice similar lesions elsewhere over the body. On examination, 3-4, 0.1 to 0.4-cm, firm, skin-colored papules were noted on outer aspects of each ear (**Figures 1 and 2**). Laboratory investigations including; blood complete picture, serum glucose, serum cholesterol, liver and renal

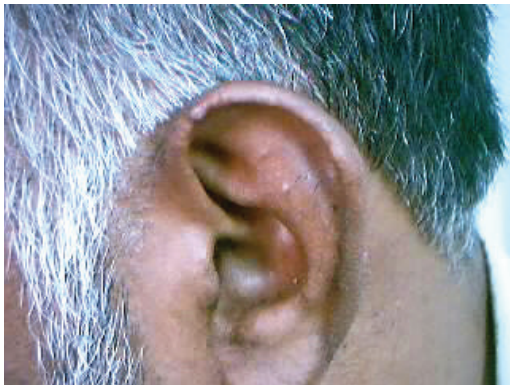

Figure 1 Small skin-coloured papules on left ear.

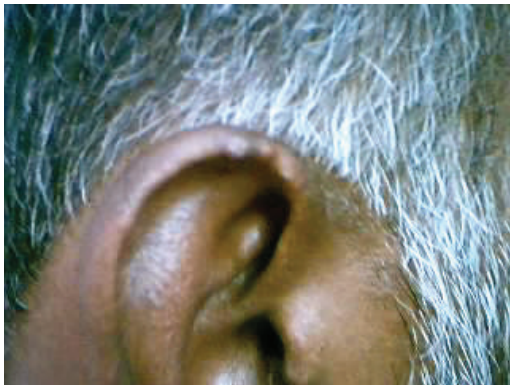

Figure 2 Small skin-coloured papules on right ear.

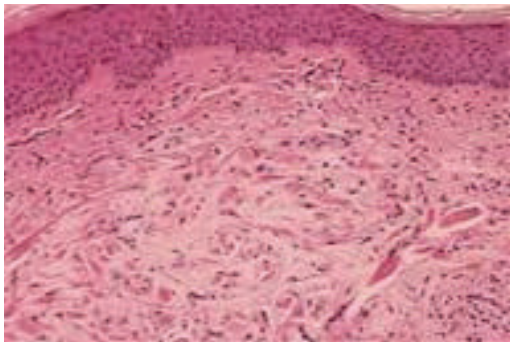

Figure 3 Histopathology showing upper and mid dermal mucin deposits separating collagen fibers and fibroblasts.

function studies were all within normal range. A serum protein electrophoresis was also normal. Excision biopsy of two larger lesions on both sides was done and it revealed upper and mid dermal deposits of mucin associated with a proliferation of plump fibroblasts (**Figure 3**). On confirmation of diagnosis, the remaining lesions were managed successfully with cryosurgery (by applying liquid nitrogen twice weekly for two weeks). There was no recurrence or new eruption during next six months.

Discussion

Acral persistent papular mucinosis is rather a rare benign entity, which occurs mostly on dorsum of the hands, face and ears.[1,4] In our case the disease was restricted only to the ears. The lesions were few and were symmetrically distributed. These were asymptomatic but patient was concerned about pebbly appearance of the superior margin of the ears and was little apprehensive about the nature of the lesions. After histological confirmation of the lesions, he was reassured about the benign nature of the disease. Generally, this is considered a benign form of dermal mucinosis that has not been associated with any other underlying disease, but there has been an occasional report of its association with paraprotienemia.[10] We did not find any such association in our case. Other forms of dermal mucinosis like, lichen myxedematosus and scleromyxedema have been clearly associated with multiple disorders.[2,5,6]

Conclusion

Acral persistant popular mucinosis should be considered a separate entity and must not be mixed with other mucinoses.

References

1. Jeremy E R. Papular mucinosis. *Dermatology Online J* 2001; **7**: 13.
2. Stephens CJM, McKee PH, Black MM. The dermal mucinoses. *Adv Dermatol* 1993; **21**:293.
3. Clark BJ, Mowat A, Fallowfield ME, Lee FD. Papular mucinosis. - is the inflammatory cell infiltrate neoplastic - the presence of a monotypic plasma cell population demonstrated by in-situ hybridization. *Br J Dermatol* 1996; **135**: 467-70.
4. Menni S, Cavicchini S, Brezzi A *et al*. Acral persistent papular mucinosis in two sisters. *Clin Exp Dermatol* 1995; **20**: 431-3
5. Howden SM, Herndon JH Jr, Freeman RG. Lichen myxedematosus: a dermal infiltrate disorder responsive to cyclophosphamide therapy. *Arch Dermatol* 1975; **111**: 1325-30.
6. Bata-Csorgo Z, Husz S, Foldes M *et al.* Scleromyxedema. *J Am Acad Derma*tol 1999; 41: 343-6.
7. Caputo R, Grimalt R, Gelmetti C. Self-healing juvenile cutaneous mucinosis. *Arch Dermatol* 1995; **131**: 459-61.
8. Tschen JA, Chang JR. Scleromyxedema: treatment with interferon alpha. *J Am Acad Dermatol* 1999; **40**: 303-6.
9. Liiter RK. Scleromyxedema: response to high-dose intravenous immunoglobulin (hdIVIg). *J Am Acad Dermatol* 2000; **43**: 403-6.
10. Borradori L, Aractingi S, Blanc F *et al*. Acral persistent papular mucinosis and IgA monoclonal gammopathy: report of a case. *Dermatology* 1992; **185**: 134-6.