MULTICENTRIC RETICULOHISTIOCYTOSIS

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A 54-year-old male presented with multiple, itchy, reddish-brown colored papules on the face, arms and trunk of 8 months duration. Histopathological study of the skin biopsy specimen revealed features typical of multicentric reticulohistiocytosis.

Key word: Multicentric reticulohistiocytosis

Introduction

Multicentric reticulohistiocytosis (MR) is a rare systemic granulomatous disease of unknown cause and has a distinct histopathology. Less than 100 cases have been reported in the literature.[1] MR is characterised by polyarthritis of hands and large joints and nodular infiltration of the skin, bones, mucous and synovial membranes. An association with malignancy is evident in more than 20% of the patients. [1,2] Cutaneous lesions are nontender, reddish-brown nodules of variable sizes. They are found distributed on the nose, paranasal areas, hands, ears, forearms, neck and trunk in descending order of frequency. Half of the patients have mucosal involvement. Histopathology is confirmatory of the diagnosis.

Case Report

A 45-year-old male presented with reddish-brown, itchy papules of 3 to 5mm size, lying singly or in groups on the forehead, cheeks and neck of 8 months duration (Fig.1). Mucous membranes, palms and soles were free of lesions. He had occasional pain in the small joints of the hands. Systemic examination revealed no abnormality. Routine laboratory tests on blood and urinalysis were normal.

Blood sugar, urea, creatinine, LFT and lipoprotein profile were within normal limits. Skiagram of the hands and ultrasound of abdomen were normal. Rh factor, VDRL, TPHA and ELISA for HIV infection were negative. Histopathology of the skin revealed tight aggregates of large histiocytes with abundant PAS positive cytoplasm and vesicular nuclei in the dermis (Fig.2). Infiltrate of polymorphs and lymphocytes were present interspersed in between.

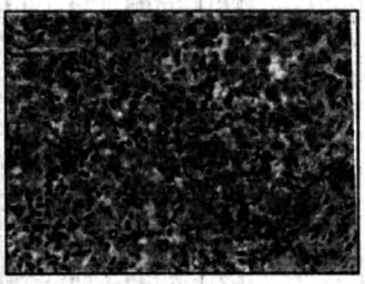

Fig1. Photomicrograph of skin showing aggregates of large histiocytes in the dermis (H&E x400)

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Discussion

The tern MR was introduced by Goltz and Waymon.Other names used to describe this disorder include lipoid dermatoarthritis, reticulohistiocytosis, giantcell reticulohistio - cytosis and normocholesterolaemic xanthomatosis.[3] The clinical course varies. The disease becomes quiescent in 5-8 years.[4] In some it remains stationary and in some within a few months to a few years may develop carcinoma of bronchus, stomach, breast and cervix.[5] Our patient had typical lesions of MR which was confirmed by histopathology though there were no significant ear nodules, joint or systemic involvement. There is no specific treatment for this condition. Various drugs like corticosteroids, antimalarials, salicylates, indomethacin, antimitotic compounds like nitrogen mustard, cyclophosphamide, and chlorambucil have been tried.[1,2] Aldridge et al reported a patient whose cutaneous lesions benefited from PUVA.[6] PUVA aggravated the itching and skin lesions in our patient. A few lesions on the face healed with minimal scarring and hyperpigmentation following liquid nitrogen cryotherapy. Intramuscular triamcinolone 40 mg once in 4 weeks was tried which showed symptomatic improvement and subsequent clearance of lesions on the face after 6 months. But new crops of lesions started appearing on the trunk and extremities. He has not developed any systemic complaints or joint involvement. Our patient may be in a relatively early stage of the disease or may represent a benign variant of the same.[3] This case is being reported for its rarity as 85% of cases reported earlier where in whites.[5]

References

1. Ginsburg WW, O'Duffy JD, Morris JL, et al. Multicentric reticulohistiocytosis: Response to alkylating agents in six patients, Ann Int Med 1989;3:384 - 388.

2. Moschella SL, Cropley TG. Diseases of mononuclear phagocytic system, In: Dermatology 3rd ed, editors Moschella SL, Hurley HJ WB Saunders company, Philadelphia,1992;1031 - 1141.

3. Rendall JRS, Vanhegan RI, Robb-Smith AHT, et al. Atypical multicentric reticulohistiocytosis with paraproteinaemia. Arch Dermatol 1977;113 : 1576 - 1582.

4. Hashimoto K, Pritzker MS. Electron microscopic study of reticulo histiocytoma. Arch Dermatol 1973; 107: 263 - 270.

5. Lesher, Jr JL, Allen BS. Multicentric reticulohistiocytosis. J Am Acad Dermatol 1984;11:713-723.

6. Alridge RD, Main RA, Daly BM. Multicentric reticulohistiocytosis and cancer. J Am Acad Dermatol 1984;10:296-297.