MALIGNANT ATROPHIC PAPULOSIS

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Malignant atrophic papulosis (MAP) is a rare disorder and only a few cases have been reported in blacks and Asians. A 38- year-old male with typical cutaneous lesions of MAP which were confirmed histopathologically is reported.

Key words: Malignant atrophic papulosis

Introduction

Malignant atrophic papulosis (MAP) is a rare fatal endovasculitis of unknown cause affecting skin, gut and CNS.[1] Crops of asymptomatic, numerous, erythematous papules, nodules with porcelain-white central atrophy and telangiectatic borders appear slowly for years anywhere except face, palms and soles.[2,3] Skin lesions are followed by gut lesions within 3 years.[3] In some cases it is confined to the skin.[4] Histopathologically typical chronic non ulcerative lesions show thin atrophic epidermis, complete dermal fibrosis with acellularity, and mild perivascular cuffing/obliteration of dermal vessels. Abundant mucin is seen in early lesions and only towards margins in late plaques.[5] Fibrinolytic activity is absent only in lesions but present normally in peripheral healthy skin.[6] MAP is thought to be a hypercoagulation syndrome with inhibited fibrinolysis.[7] Recently lymphocyte-mediated vasculitis was reported in the pathogenesis of MAP.[8]

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Case Report

A 38-year-old man developed two asymptomatic, atrophic plaques, one on the left upper arm (20x15cm) and the other on the left lower leg (15x15) and was diagnosed as a case of leprosy at the periphery and treated with MDT. He took regular therapy for 11/2 years without any relief and numerous new lesions erupted slowly over the trunk, limbs and face during MDT. There was no history of constitutional symptoms or systemic involvement including pain abdomen. In addition to above two big atrophic plaques, innumerable variable-sized scars were seen on the limbs, trunk and face. Most scars were depressed, porcelain white with telangiectatic borders, while others were depressed pigmented scars. Five, firm, asymptomatic, nontender, erythematous nodules with typical porcelain-white centres, finely wrinkled surface and ill-defined margins were seen on the trunk. Multiple, bilateral asymmetrical, 2-6 mm, erythematous shiny papules, some with pinpoint porcelain-white centres covered lateral side of cheeks, rim of pinnae, and neck. General physical and systemic examinations were normal except splenomegaly (2 cm

below costal margin). Eyes were normal. Histopathologically diagnosis of MAP was confirmed as marked atrophy of epidermis, necrosis and acellular dermis in wedge shaped pattern was seen along with endovasculitis and mild perivascular mononuclear infiltration. Routine laboratory tests on blood and urine were normal. LFT, RFT serum electrolytes, and x-ray of the chest were normal. L.E cell phenomenon was negative. Ultrasonogram of abdomen confirmed splenomegaly.

Patient was treated with aspirin (1050 mg) and dipyridamole (150 mg) daily for 10 weeks. No new lesion appeared after starting therapy.

Discussion

Probably this is the first case of MAP in the Indian literature. Face was predominantly affected in our patient but in earlier literature, involvement of face was not observed. MAP is considered a fatal disease but some patients may survive for long as disease remains localized only to skin[4], and in our patient also lesions were limited to skin since 2 1/2 years. Therapy with aspirin and dipyridamole was effective as telangiectasia disappeared and new lesions did not erupt after it.

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