Generalized eruptive histiocytoma

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ABSTRACT

We are reporting a 62 year old male, who over a period of 1 year, developed a symmetric eruption of hundreds of brownish papules, with spontaneous regression of some lesions. The clinical and histopathological findings were compatible with the diagnosis of generalized eruptive histiocytoma.

Key words: Histiocytoma, histioctytosis, elderly male

INTRODUCTION

Generalized eruptive histiocytoma (GEH) is an unusual, benign disorder belonging to the group of non-Langerhans'-cell ('non-x') histiocytoses.[1] It is a papular, non-lipidic, self-healing histiocytosis affecting mainly adults.[2] The first adult case was described by Winkelmann and Muller in 1963.[3] Herein, we report a case of this rare entity in old age for the first time from the Asian subcontinent.

CASE REPORT

A 62-year-old man was presented with a history of developing small number of papular lesions on his trunk 12 months previously. He then slowly but progressively developed hundreds of elevated, brownish papular lesions elsewhere on his body. These lesions were neither painful nor itchy. Although some papules resolved spontaneously, new lesions continued to develop. There were no precipitating factors and no other systemic problems.

He had no abnormalities on general medical examination. There was no lymphadenopathy or hepatosplenomegaly on palpation. Cutaneous examination showed a widespread papular eruption on the face, trunk and proximal parts of the arms and legs [Figures 1 and 2]. The scalp, palms and soles were spared. Mucous membranes were not involved. The individual lesions were non-confluent, hemispherical and skin colored to brown papules, ranging from 3 to 8 mm in size; some displayed central umblication, clinically simulating molluscum contagiosum. The patient mentioned spontaneous regression of some lesions and focal residual hyper pigmentation was present at the sites of resolution. The eruption was asymptomatic.

Investigations revealed routine hematology, biochemistry, immunoglobulin, autoantibodies and lipids to be normal. Histopathalogical study of a biopsy from representative lesion showed a normal epidermis along with dermal infiltration by histiocytic cells, having vacuolated cytoplasm at places, intermixed with inflammatory cells. Perivascular arrangement of histiocytic cells was also evident in some areas [Figure 3]. Histochemical and ultrastructural studies could not be performed because of non-availability. The clinical picture, in conjunction with the histopatholgoical findings, suggested the diagnosis of GEH. Patient was planned to put on photochemotherapy but he refused to take it because of spontaneous resolution of few of the lesions.

Over a period of 4 years, our patient has continued to produce new lesions, a proportion of which have spontaneously regressed, but there has been no evidence of progression of disease beyond the skin till repeated follow-up of those 4 years.

DISCUSSION

GEH was first described by Winkelmann and Muller as a papular, non-xanthomatous, self-healing histiocytosis, predominantly affecting adults.[3,4] It is an extremely rare disease. The disorder may start at any age from 3 months

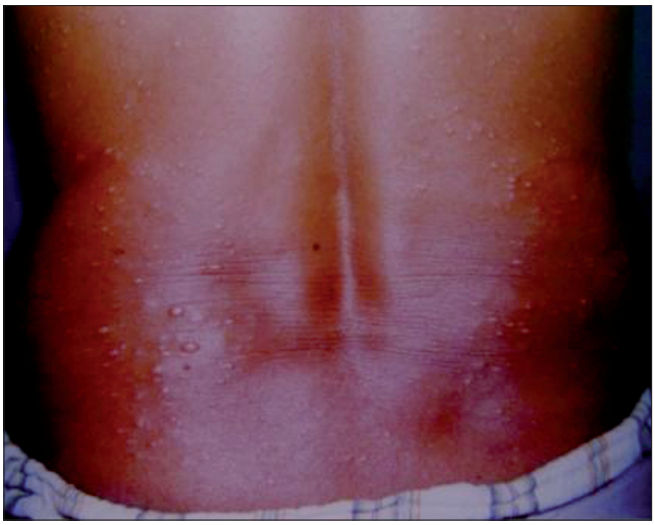

Figure 1: Widespread papular eruption on the trunk composed of non-confluent, hemispherical, skin colored to brown papules

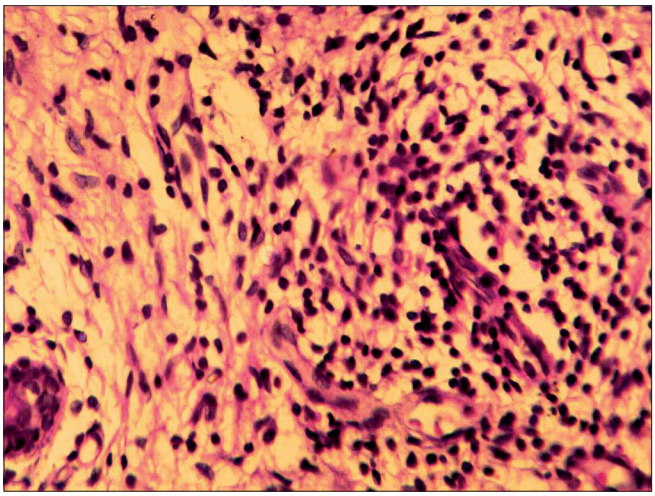

Figure 3: Biopsy of a lesion showing an unremarkable epidermis and a perivascular arrangement of histiocytic cells, having vacuolated cytoplasm at places, in the dermis (H&E, ×100)

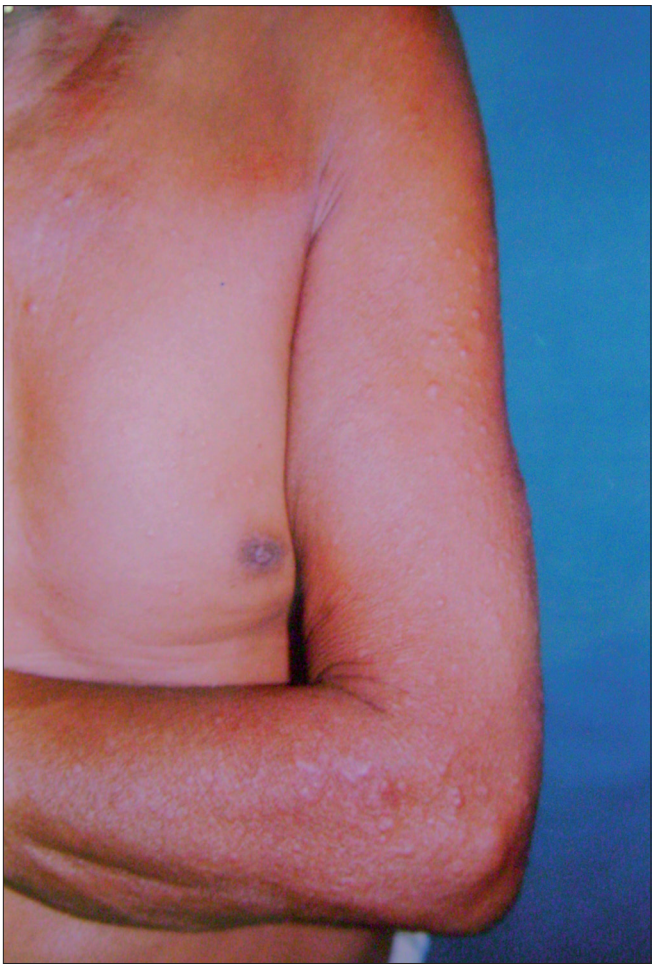

Figure 2: Generalized distribution of discrete, brownish papules on the arm

to 58 years.[2] Clinically, the skin lesions are asymptomatic, firm, erythematous or brownish papules measuring 3-10 mm in diameter. Clinical resemblance to molluscum contagiosum has been reported.[5] The lesions appear in successive crops and may be numerous without a tendency to grouping over the face, trunk and proximal parts of the limbs. Mucous membranes are rarely affected and visceral involvement has not been reported. Atypical GEH-type lesions have been reported in association with acute monocytic leukemia.[6] General health is always good and the disease has a limited course lasting a few years with eventual spontaneous regression of the lesions as happened in our case too. Histological examination shows a monomorphous histiocytic infiltrate in the upper and mid-dermis, devoid of foamy and giant cells. Histochemical and histoenzymatic studies show no intracellular accumulation of fat, mucopolysaccharides, glycogen or iron, but positivity for lysosomal enzymes.[1] Electron microscopic studies show that the histiocytic cells lack Birbeck granules but have cytoplasmic laminated bodies. At first presentation, the differential diagnosis may include angiomata, sarcoid, lymphoma, neurofibromatosis, urticaria pigmentosa and xanthomata. All of these would be differentiated and excluded by histology. Regarding treatment modalities for GEH, literature is silent but recent reports on use of photochemotherapy highlight complete resolution in one case[7] and relapse later on in another case.[8]

Our case demonstrates all the features of GEH and emphasizes the importance of clinical, histopatholgoical, immunohistochemical and ultrastructural examination in the investigation and diagnosis of histiocytic disorders.

The rarity of individual histiocytic disorders is such that only by pooling information will advances in classification, management and prognostic information be possible.

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