Juvenile Xanthogranuloma

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Abstract

The histiocytoses are a heterogenous group of diseases that are characterized by the accumulation of reactive or neoplastic histiocytes in various tissues. Juvenile Xanthogranuloma is a member of the non-Langerhans cell group of histiocytic proliferative disorders. This present case report describes a juvenile xanthogranuloma in a twenty-month-old male baby.

Key words: Histiocytoses, Non-Langerhans cells, Juvenile xanthogranuloma

Introduction

Juvenile xanthogranuloma (JXG) is a rare disorder, which belongs to the broad group of non - Langerhans cell histiocytosis. The non-LCH are generally benign proliferative disorders but may have a systemic component as a major part of the disease.[1] Juvenile xanthogranulomas are benign tumours of histiocytic cells that occur predominantly in infancy and early childhood and spontaneously regress.[2] The classic presentation is that of successive eruptions in the head, neck, and upper trunk of initially red papules or nodules which later become yellow and finally brown flattened plaques or macules. They are firm and rubbery and large lesions may ulcerate. JXG represents an accumulation of histiocytes lacking Birbeck granules (non-Langerhans cells) which can be differentiated from Langerhans cells by specific staining techniques.[3] In mature lesions, a granulomatous infiltrate is usually present containing foamy cells, foreign body giant cells, and Touton giant cells as well as macrophages, lymphocytes and eosinophils.[4] It is a relatively rare cutaneous condition.[5] Here we report a case of 20 months old child with juvenile xanthogranuloma.

Case report

A 20 months old male baby was brought to our OPD by his parents with complaints of asymptomatic skin lesions in different parts of his body since the age of four months. The size of individual lesion and their number was increasing gradually since then. No history of vesiculation, discharge or ulceration of the lesions. The baby was delivered normally at term, after an uneventful pregnancy with no evidence of skin disease at birth. His developmental milestones were within normal limits. Examination revealed reddish brown to skin coloured firm non tender papules over both cheeks, both legs and right ear (fig-1,2). Few similar lesions were noted over the back. The size of individual papules ranged from 3 to 5 mm. No café-au-lait macules, organomegaly or bony defects were present. No history of similar illness in the family was noted. Complete blood count, haemoglobin level, peripheral smear and chest X-ray was sent. All these investigation were found to be normal. Skin biopsy was taken from a papule in right cheek which showed foamy macrophages in aggregates intermingled with lymphocytes, touton's giant cells were also noted (fig-3, 4) suggestive of Juvenile Xanthogranuloma.

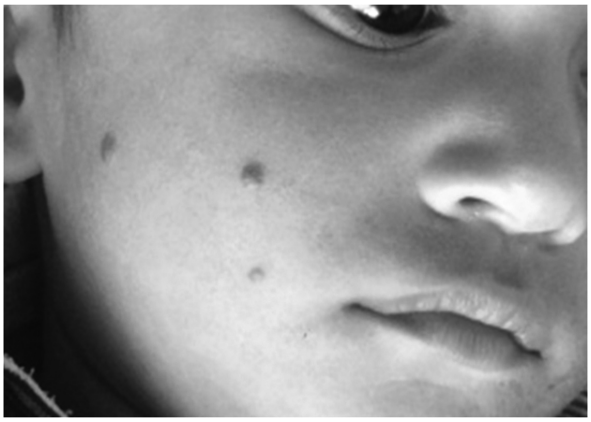

Figure 1: Redish brown papules over cheek

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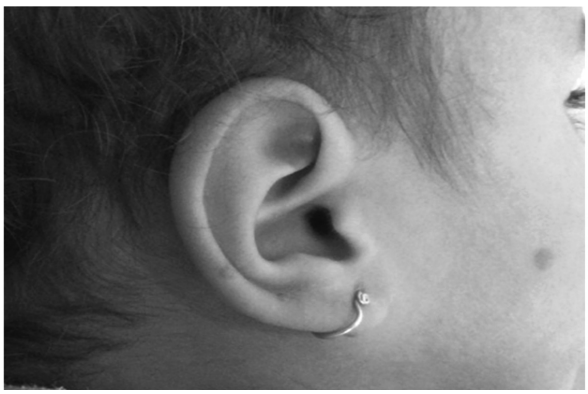

Figure 2: Skin coloured papule over ear

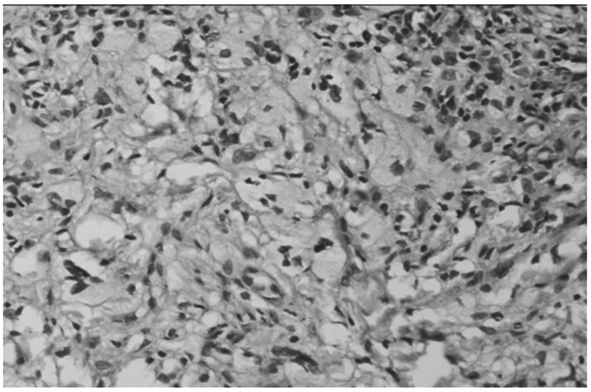

Figure 3: foamy macrophages

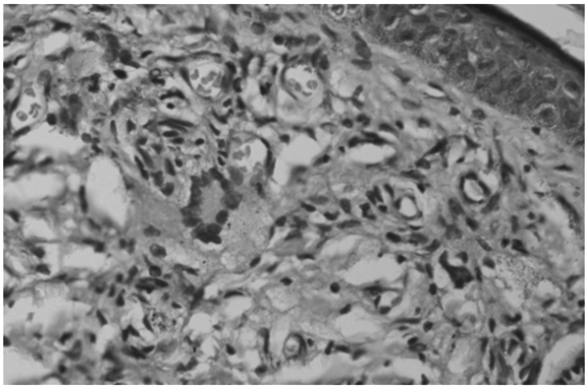

Figure 4: Toutons giant cell

Discussion

The histiocytoses are reactive or malignant conditions in which various tissues, including the skin, are infiltrated by cells of the monocyte macrophage lineage.[6] They are a heterogenous group of disorders classified by the Writing Group of the Histiocyte Society[7] into three classes:

Class 1: Langerhans cell histiocytosis

Class 2: Non-Langerhans cell histiocytoses

Class 3: Malignant histiocytoses

The non-LCH histiocytoses consist of a long list of diverse disorders. Juvenile xanthogranuloma is one of them. The first accurate report of JXG was in 1905 by Adamson who reported a child with numerous yellow papules over body developed in first two weeks of life.[8] The author termed these lesions congenital xanthoma multiplex. The name juvenile xanthogranuloma was introduced by Helwig and Hackney in 1954 based on the histopathologic picture.[9] The aetiology of JXG is unknown.[2]

The characteristic clinical features of JXG are its onset in infancy, sudden appearance of lesions and spontaneous regression. Most patients develop single lesion, but in others several lesions may develop and occasionally hundreds of lesions may be present.[2] The median age of presentation with solitary skin nodules is 2 years compared to 5 months for multiple skin lesions.[10] Large nodular variant with one or a few, red-brown to yellow nodules can be distinguished from the rare micronodular variant with numerous, small papules on the head and trunk and occasionally on the proximal part of the limbs.[11]

Systemic JXG is defined as involvement of two or more visceral organs in addition to multiple cutaneous and subcutaneous lesions. JXG with systemic involvement is a rare histiocytic disorder in which significant morbidity and occasional deaths may occur.[12] Other sites that may be involved are buccal mucosa and eyes (iris, ciliary body, and uvea, with possible blindness).[13] Rarely, systemic changes are seen in the lungs, liver, or spleen (hepatospleenomegaly), or very rarely in the pericardium or testes. JXG has been associated with other diseases including Juvenile chronic myelogenous leukemia, neurofibromatosis type 1, urticaria pigmentosa, insulin-dependent diabetes mellitus, aquagenic pruritus and possibly cytomegalovirus infection.[12]

The classic histologic findings of JXG include a nodular to diffuse collection of histiocytes with a finely vacuolated, foamy cytoplasm, round to oval nuclei with little pleomorphism and variable numbers of multinucleated Touton cells.[14] Three basic histologic patterns have been identified.[2] In an early JXG (EJXG), there is a dense monomorphic histiocytic compact sheet-like infiltration without lipidization with absence of multinuclear giant cells. In classic JXG (CJXG), there are foamy histiocytes with more abundant and distinctly vacuolated cytoplasm and variations of fully developed Touton giant cells. The third variety, transitional JXG(TJXG) has a predominance of spindle-shaped cells with foamy histiocytes and giant cells.

Due to its generally benign clinical course, the historical treatment for JXG has been surgical excision of a solitary skin lesion or observation alone, while waiting for spontaneous regression. In cases with significant multisystem involvement, chemotherapy regimens used to treat Langerhan cell histiocytosis may be effective. Regimens including both corticosteroids and a vinca alkaloid had the highest rate of complete response, partial response and stable disease[16].

Our case was diagnosed as xuvenile xanthogranuloma based on history, clinical findings and histology. Screening was done for systemic involvement and coexistent disorder by clinical examination and investigations but non were found. The patient was advised for followups to asses for spontaneous resolution of the lesions and also to monitor development of systemic involvement and other coexistent disease such as myelogenous leukemia.

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