JUVENILE XANTHOGRANULOMA

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Summary

A solitary skin lesion of juvenile xanthogranuloma appearing at the age of five years is reported.

KEY WORDS: Juvenile xanthogranuloma, Nevoxanthoendothelioma.

Juvenile xanthogranuloma, a term that has replaced the old term nevoxanthoendothelioma, is a rare, benign, self-limited disease of infants and children. It is characterised by mutiple, discrete, firm, 0.5 to 1 cm sized, reddish yellow papules or nodules which erupt suddenly during the first few months of life. The usual course of the skin lesions is spontaneous remission in one or two years. The blood lipid values remain normal, but manifestations may develop in the eyes, muscles, heart, testis, stomach and salivary glands[1]. The mature lesions non histological study, reveals granulomatous infiltration in the dermis consisting of histiocytes, lymphocytes, eosinophils, foam cells and giant cells of both foreign-body type and Touton's type. The presence of giant cells having a perfect 'wreath of nuclei' is quite typical of juvenile xanthogranuloma[2].

Many cases of juvenile xanthorgranuloma have been reported in the olden literature as 'peculiar forms' of xanthomas[3,4]. Mc Donagh in 1922 coined the term nevoxantheondothelioma for this disease[5]. His patient presented with skin lesions since birth. It was presumed that the lesions were due to proliferation of the endothelial cells of capillaries and subsequent xanthomatous degeneration[6]. The view that it is a true xanthoma is discarded now because the lesions do not always contain fat and there is no relation to a disordered lipid metabolism. Till recently, it was considered as an abortive, monosymptomatic, purely cutaneous form of Hand-Schuller Christian disease. A strong point against any relationship between juvenile xanthogranuloma and histiocytosis is the absence on E/M examination of Langerhan's granules in the histiocytes in juvenile xanthogranuloma as opposed to their presence in approximately half of the histiocytes in the three forms of histiocytosis. Juvenile xanthogranuloma is now generally regarded as an independent

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entity, as a reactive granuloma of unknown cause. Here, we are reporting a case of juvenile xanthogranuloma in a young girl.

Case report

A six years old girl was seen in the Dermatology section of Medical Hospital, Kottayam in October 1982 with an asymptomatic nodule on the abdominal wall. It started one year before as a small yellowish papule which gradually increased in size. Examination revealed a well circumscribed, pinkish yellow, non tender, firm nodule of $2 \times 1\frac{1}{2}$cm size on the upper part of anterior abdominal wall (Fig 1). There were no other skin lesions. A detailed ophthalmologic examination did not reveal any abnormality. System review was normal.

Investigations

Routine investigations on blood and urine did not show any abnormality. Blood VDRL was non reactive. Serum cholesterol was 160mg%. Serum Triglyceride 80mg%, SGPT 24 IU/L, Serum bilirubin 0.6mg%, total serum protein 7gm% (albumin 4gm%, globulin 3gm%) and alkaline phosphatase 8KA Units. X.ray of chest and bone survey showed no abnormality, ECG was normal. The skin lesion was excised under local anaesthesia. Histology revealed diffuse granulomatous infiltration of dermis consisting of histiocytes, lymphocytes, eosinophils and foam cells. Numerous foreign body giant cells and Touton giant cells (Fig 2) were seen. Giant cells with perfect 'wreath of nuclei' also were present (Fig 3). Epidermis appeared normal excepting for flattening of the rete. Follow up of the case for six months did not show any evidence of recurrence.

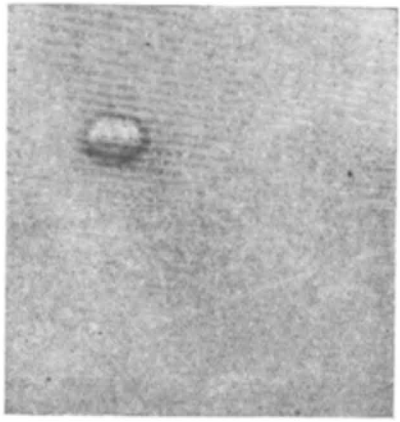

Fig. 1 A solitary lesion of Juvenile xanthogranuloma on abdominal wall.

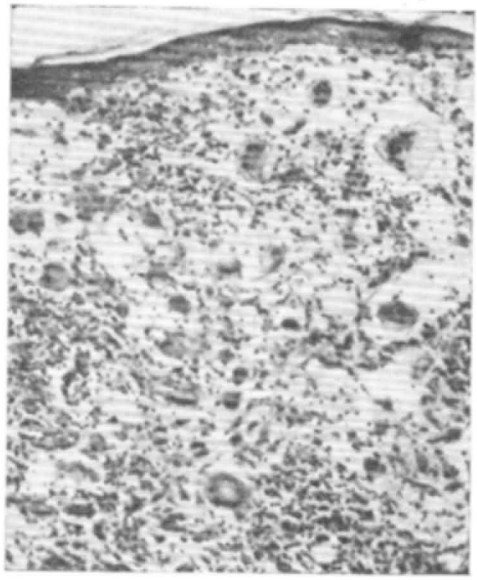

Fig. 2 Histology - (Low power) Shows diffuse granuloma of dermis consisting of histiocytes, lymphocytes and giant cells - Foreign body type, Touton type and those having a perfect 'wreath of nuclei.'

Discussion

Cases of juvenile xanthogranuloma are only rarely reported in dermatological literature from India[7,8]. Usually the lesions are multiple and appear during the first few months of life. Our case presented with a solitary nodule and the onset was at 5 years of age. A detailed examination

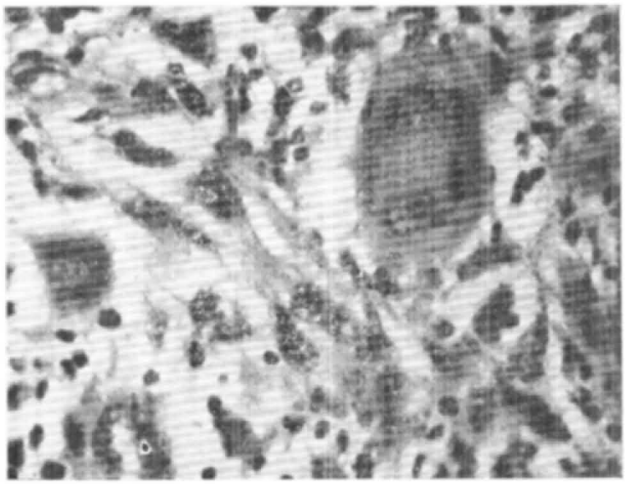

Fig. 3
Note a large giant cell having a perfect 'wreath of nuclei'.

of the eyes and other systems did not reveal any abnormality. The serum lipid values were normal. The giant cells having a perfect wreath of nuclei quite typical of juvenile xanthogranuloma were abundant in the dermis. The presence of these giant cells excluded the possibility of dermatofibroma with lipidization to which juvenile xanthogranuloma may closely resemble both clinically and histologically.

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