

Case Report

A Rare Case of Juvenile Xanthogranuloma in Thigh

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Abstract: Juvenile xanthogranuloma (JXG) is a chronic non granulomatous inflammatory reaction, non-Langerhans cell histiocytosis characterized by skin lesions that tend to be self-limited. This is a regressing or stabilizing disorder usually occurring during infancy and childhood and it is characterized by one or more cutaneous nodules, and less often by additional lesions in deep soft tissue and organs. We report a case of 14 years old young boy with a large nodular form of JXG presented in thigh.

Keywords: Juvenile xanthogranuloma, non granulomatous, inflammatory reaction

INTRODUCTION:

Juvenile xanthogranuloma is a chronic non granulomatous inflammatory reaction, non-Langerhans cell histiocytosis characterized by skin lesions that tend to be self-limited[1]. This is a regressing or stabilizing disorder usually occurring during infancy and childhood and it is characterized by one or more cutaneous nodules, and less often by additional lesions in deep soft tissue and organs[2]. In 1989, Tahan et al suggested that the term should be changed to xanthogranuloma, since between 15% and 30% of these lesions occur in individuals older than 20 years of age; however, the term, juvenile xanthogranuloma is still widely used[3]. Cutaneous manifestation of JXG includes asymptomatic red and brown papules or nodules, most often with a tinge of yellow. In rare cases, it can involve internal organs and muscles. Very few cases of deep seated JXG have been reported and, of those, most have been located in the trunk. We report a case of 14 years old young boy with a large nodular form of JXG presented in thigh.

CASE REPORT

A 14 year old otherwise healthy boy presented with a thigh mass discovered incidentally by the parents. On examination, the mass was painless and deep seated. Clinically it was diagnosed as Lipoma and advised for surgery. Post-surgical resected lesion measured 3.0·2.5·2.0 cm, and was well circumscribed. The cut surface of the lesion was homogeneous and tan-yellow. On microscopy, sheets of diffuse infiltrate of histiocytes, occasional multinucleated giant cells, lymphocytes, and a few eosinophils (Figure 1 and 2). Histiocytes were cytologically bland, with round to oval nuclei and regular nuclear contours. The cytoplasm of the histiocytes ranged from eosinophilic to finely vacuolated (Figure 3). Only occasional Touton giant cells with peripheral lipid in the cytoplasm were present. There was no evidence of caseation or central necrosis. The epidermis was spared. The special stains showed no acid-fast organisms, bacteria, or fungi. Therefore, the diagnosis of juvenile xanthogranuloma was made histopathologically.

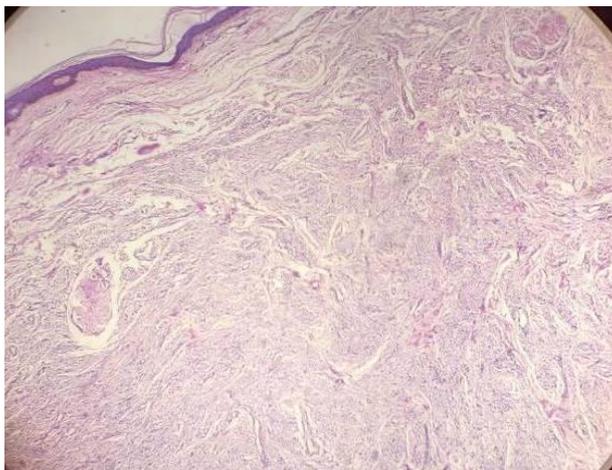


Fig-1: Microscopy shows a mass lesion seen in the dermis (H & E, 4X)

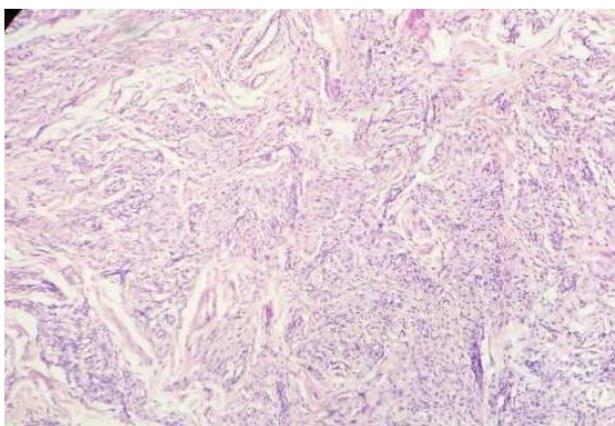


Fig-2: Microscopy shows proliferation of histiocytes arranged in sheets and nests. (H & E, 10X)

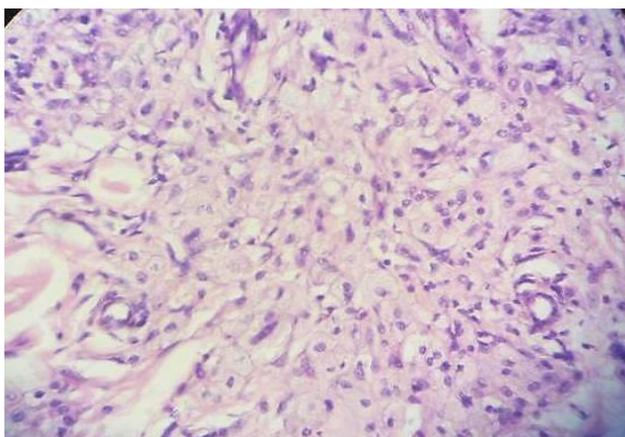


Fig- 3: Microscopy shows foamy histiocytes and mononuclear cells (H & E, 40X)

DISCUSSION

JXG is a benign cutaneous fibrohistiocytic lesion and a type of granulomatous process[3,4]. Although, the term, was coined by Helwig and Hackney

in 1954, the original description of this entity came from Adamson who described it five decades earlier as “congenital xanthoma multiplex[3,5]”In 40–70% of patients, JXG develops in the first year of life. In 5–

17% of cases, the skin lesions may appear soon after birth[6].The highest incidence of JXG in adulthood is observed in patients aged 20 to 30 years, but the disease is generally rare in adults. The disease most often affects male children, whereas in adults, there is no sexprevalence[6].

Even though the etiology is unknown, it is believed to result from disordered macrophage response to the nonspecific injury. Most authors presume that JXG is caused by granulomatous histiocytic reaction in response to undetermined stimuli, probably physical or infectious[7,8]Bergman et al. revealed that in monocyte-derived macrophages of adult patients with JXG, intracellular synthesis of cholesterol is enhanced[8].

Clinical manifestations in JXG include cutaneous or subcutaneous nodules which are found in infants under 1 year old, although there are rare cases in adults[8,9].Nodules are well-separated and of different size and colour, usually yellow or red-brown, measuring up to 1 cm in most cases. Nodules may be single (60–82% of patients) or multiple and most typically are located on the head, neck and upper trunk, but lesions may affect any site of the body such as lungs, liver, central nervous system, bones and endocrine glands with different systemic symptoms. Mucosal lesions associated with JXG are hardly ever observed[9].

Gianotti described two most frequent clinical types of JXG. A small nodular form with nodules that are well separated from the surrounding skin and measuring from 2 mm to 5 mm. Nodules in the large nodular form can reach the size of 1–2 cm[10].

The lesions of juvenile xanthogranuloma usually consist of a ill-defined mass lesion composing of homogeneous proliferation of histiocytic otherwise foamy type macrophages admixed with chronic inflammatory cells and few of the histiocytes are seen abutting on thin epidermis, some with attenuated, elongated rete ridges[6,10]. The lesional borders are usually poorly circumscribed. In our case, the margin of the lesion was also poorly circumscribed and did not involve the subcutaneous tissue. Along with the proliferation of foamy macrophages, chronic mononuclear inflammatory cells infiltrate with occasional plasma cells and eosinophils were also noted.

Immunohistochemistry plays a major role in the differential diagnosis between Langerhans cell histiocytosis (LCH) and JXG. JXG lesions usually label

strongly with markers CD68, factor XIIIa, HAM 56, Mac 387 and often anti CD4.S-100 protein, which is a marker for the diagnosis of LCH, is typically absent. According to few authors, neither factorXIIIa negativity, nor S-100 positivity should preclude the diagnosis of JXG[11,12].

The prognosis in patients with JXG without internal organ involvement is favourable. Spontaneous regression usually occurs within 6 months to 3 years[11].

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