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Juvenile xanthogranuloma in adult: A rare case report

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Abstract
Juvenile xanthogranuloma (JXG) is a member of the non-Langerhans cell group of histiocytic proliferative disorders. It is a benign cutaneous fibrohistiocytic lesion most often affecting infants and young children. Juvenile xanthogranuloma is rare in adults, in whom the peak incidence is in the late twenties to early thirties and most adult patients have solitary lesions.
A 55 year-old male presented with multiple disseminated cutaneous eruption that started almost a decade ago on the trunk and 7 years ago on the face. The lesions were asymptomatic and followed a slowly progressive course. Cutaneous examination revealed numerous well demarcated, firm, rubbery brownish yellow non tender papulo-nodular lesions of different sizes over the face, neck, and front of the trunk. There were few lesions on the upper extremities, predominantly over the elbows. A differential diagnosis of eruptive xanthoma, xanthogranuloma, histiocytosis and neurofibroma was considered. Skin biopsies were taken and sent for histopathological examination. The diagnosis of multiple xanthogranulomas in an adult was made based on the clinical picture and histopathological findings. We describe a classic, rather an unusual clinical presentation in an adult highlighting the wide variation in presentation and the importance of considering this entity in the differential diagnosis of multiple benign eruptive lesions in an adult.

Keywords: Xanthogranuloma, cutaneous, adult

1. Introduction
Juvenile xanthogranuloma (JXG) is a member of the non-Langerhans cell group of histiocytic proliferative disorders[1]. It is a benign cutaneous fibrohistiocytic lesion most often affecting infants and young children. Classically, it presents as solitary nodule or multiple yellowish papules with most common location on head and neck followed by upper trunk. Juvenile xanthogranuloma is rare in adults, in whom the peak incidence is in the late twenties to early thirties and most adult patients have solitary lesions[2]. JXG was first reported by Adamson[3].
Although the head, neck and trunk are the most common sites for JXG, it can appear anywhere on the body, including the groin, scrotum, penis, clitoris, eyelid, toe nail, palms, soles and lips[4].
We describe a classic, rather an unusual clinical presentation in an adult highlighting the wide variation in presentation and the importance of considering this entity in the differential diagnosis of multiple benign eruptive lesions in an adult.

2. Case Report
A 55 year-old male presented with multiple disseminated cutaneous eruption that started almost a decade ago on the trunk and 7 years ago on the face. The lesions were asymptomatic and followed a slowly progressive course. The patient is a known diabetic since last 10 years andwasrecentlydiagnosedwithHypertensionandwas on antihypertensives since last1 year.
General examination of all vital systems was normal.
Cutaneous examination revealed numerous well demarcated, firm, rubbery brownish yellow non tender papulo-nodular lesions of different sizes over the face, neck, and front of the trunk. There were few lesions on the upper extremities, predominantly over the elbows.
A differential diagnosis of eruptive xanthoma, xanthogranuloma, histiocytosis and neurofibroma was considered. Skin biopsies were taken and sent for histopathologicalexamination.

3. Histopathology

Section studied showed well-circumscribed exophytic nodule. The nodule was separated from the flat epidermis by a clear zone formed mainly of histiocytes with other inflammatory cells and multinucleated giant cells (Fig.1). There were numerous multinucleated giant cells with classic Touton giant cells within the infiltrate (Fig.2).

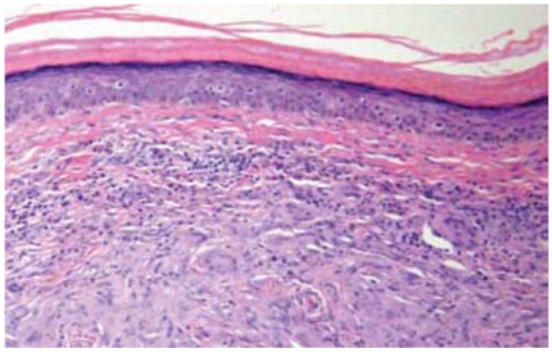

Fig 1:

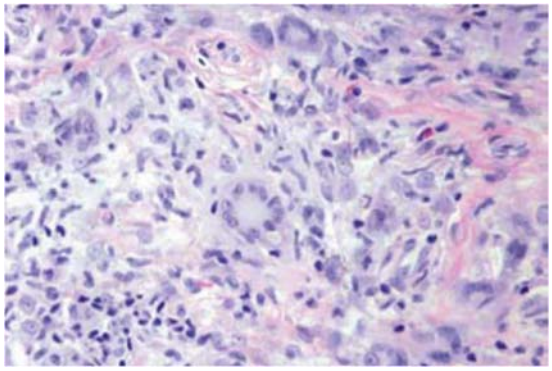

Fig 2:

The diagnosis of multiple xanthogranulomas in an adult was made based on the clinical pictureand histopathological findings.

Routine investigation including complete blood count, lipid profile, liver and renal function tests were within normal limits. Random Blood Glucose was 168mg/dl.

A peripheral smear examination was done and was found to be normal.

4. Discussion

JXG is a benign cutaneous fibrohistiocytic lesion and a type of granulomatous process, at times accompanied by lipid deposits. Adamson[3], who first described this lesion in 1905,defined single or multiple cutaneous nodules in infancy as congenital xanthoma multiplex.

The lesions were designated nevoxanthoendothelioma by Mc Donagh in 1912[5], who considered this disorder to be derived from endothelial cells. However, widespread recognition of an entity resembling JXG occurred in 1954[6]. Helwig and Macknay first coined the term juvenile xanthogranuloma in 1954[6] as a benign, asymptomatic and common self-healing disorder of non-Langerhans cell histiocytosis (LCH), affecting mostly infants, children and rarely adults. Eighty per cent cases appear in the first year of life and 20-30% cases present at birth[7].

Three main clinical forms are recognized: a small nodular/papular (2-5 mm); large nodular (5-20mm); and giant xanthogranuloma (more than 20 mm)[7]. But unusual clinical variants like mixedform, subcutaneous form, JXG *en plaque* have been reported recently[7].

JXG can be differentiated from xanthoma by the distribution of the lesion and the absence of lipid abnormalities. Three other differential diagnoses include molluscum contagiosum (pearly, dome-shaped papule with central umbilication), hemangioma and neurofibroma (firm lesion, associated caféau- lait spots)[8].

Histopathology is used to diagnose the presence of a non-LCH, but differentiation between the different subtypes is based mostly on immunohistochemistry and the clinical setting.

The basic histopathology of the non-LCH shows well circumscribed nodules with dense infiltrates of histiocytes. Those that involve the skin usually infiltrate the dermis. Giant multinucleate cells are variable in number and there is also a variable degree of predominantly perivascular and perilesional inflammatory cells. Touton giant cells (seen in 85% of cases ofJXG, in a recent series) [1], are characterized by a wreath of nuclei around a homogeneous eosinophilic cytoplasmic centre, while the periphery shows prominent xanthomatization. Electron microscopy has revealed a variety of nonspecific organelles including dense bodies, worm-like bodies and popcorn bodies, among others [9]. The cells of histiocytes andgiant cells are monocyte-macrophages in origin. They label strongly with macrophage markers such as CD 68 and HAM. On the other hand, S-100 protein immunoreactivity, which is a marker for the diagnosis of LCH, is typically absent [10,11].

Multiple xanthogranulomas are quite unusual in adults and that to occur in an eruptive manner isquite rare. It is important to recognize multiple adult xanthogranulomas, because of its goodprognosis and the absence of visceral involvement, therefore requiring no investigations or aggressive treatments, a very important observation made by Punithwavathy*et al.*[12]Surajit*et al.*[13] in their case report of adult onset xanthogranuloma.

It has been postulated that JXG represents a reactive granulomatous response of histiocytesto a yet unidentified stimulus. Evaluation for extracutaneous JXG is not indicated, unlessthere are symptoms or findings suggesting their presence, as they also disappear spontaneously[13].

It is our recommendation that adult form of xanthogranuloma should always be thought ofas a differential in any case of multiple eruptive disseminated papulo-nodular lesions in an adult.

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