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JUVENILE XANTHOGRANULOMA OF THE EAR: A RARE CASE REPORT

Dr Chandreyi Banerjee [1] , Dr Deepsikha Nandy [2]

[1] Specialist Medical officer, Swasthya Bhawan, Kolkata
[2]Post Graduate Trainee, Department of ENT Head Neck Surgery, Calcutta National Medical College & Hospital

Corresponding Author-
Dr Deepsikha Nandy
Phone: 8768791238

ABSTRACT:
Juvenile xanthogranuloma is a rare entity. It primarily, is a disease of infancy. It has been documented in its rarest form in an adult.
This is a case report of an adult-onset juvenile xanthogranuloma of both ears , proved by histopathology and IHC.

Key Words: Ear; Ear tumours; Juvenile xanthogranuloma; Skin lesions

INTRODUCTION

Juvenile xanthogranuloma (JXG) is a rare disorder, which belongs to the broad group of non - Langerhans cell histiocytosis[1] . Also it is the most common form of non-Langerhans cell histiocytosis[2] . Mostlty, JXG is a benign cutaneous fibrohistiocytic lesion that usually presents during infancy or early childhood.

Classically, it presents as solitary or multiple yellowish papules or nodules. The most common sites of involvement are the head, neck and trunk[3] .

From studies in recent years, however, it is clear that there are many clinical forms of JXG beyond this classic description. In such cases, the diagnosis may be more difficult to make without knowledge of the different possible clinical variants.

We describe a case of JXG with a rather unusual clinical presentation, and highlight the importance of considering this entity in the differential diagnosis of benign soft tissue tumours of the ear.

CASE PRESENTATION

A 22 year old boy was referred to the department of ENT & Head-Neck Surgery, Calcutta National Medical College & Hospital, with a diffuse fungating enlargement of both the ears (Figures 1 and 2), which was first noticed 2 months earlier as an area of redness.

The lesion initially started with a small reddish papule at the pinna, which later showed progressive growth along with appearance of some other similar lesions around it. It then took a form of fungating ear swelling with multiple diffuse brown-to-yellowish papulonodular eruptions. These eventually became quite prominent with occasional bleeding from the lesions. The boy was otherwise healthy.

The physical examination, revealed multiple yellow-brown, relatively well-demarcated papulonodular lesions with variable sizes (2–8 mm in diameter).

Figure 1

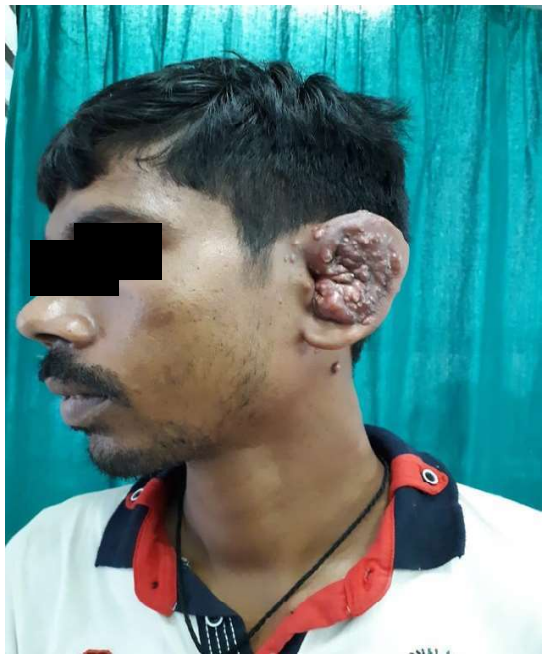

Figure 2

Figure.1 & 2 : Diffused fungating enlargement with multiple diffuse brown-to-yellowish papulonodular eruptions of right and left ear respectively.

Lesions were shiny, soft to elastic in consistency consistency present almost all over the pinna of both right & left ear [Figure 1 & 2].

The surface of some lesions were scaly. There was no vesiculation, erosion, or crusting. The mucous membranes, palms and soles,chest were unaffected and ophthalmologic examination was normal.

No other systemic involvement was noted. No other family members were affected.

The following differential diagnosis was made: lepromatous leprosy, tuberous xanthoma and xanthogranuloma.

Laboratory investigations, including routine hematological examination, liver and renal function test, were within normal range. Serum levels of lipids were not raised. The Ziehl-Neelsen stain for acid fast bacilli was negative.

Histopathological examination revealed dense granulomatous dermal infiltrates consisting of foam cells, multinucleated giant cells (mainly Touton type), histiocytes, lymphocytes, and a few eosinophils and neutrophils . The epidermis was thinned out without any grenz zone and inflammatory cells extended toward lower dermis to subcutaneous tissue (Figure 3 & 4).

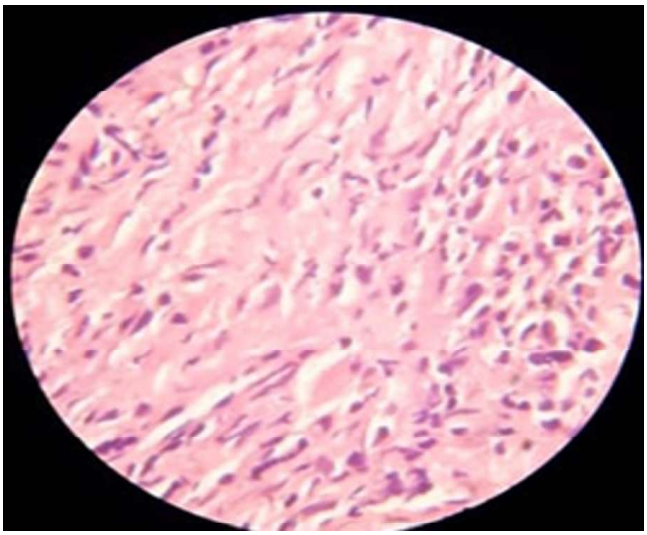

Figure 3

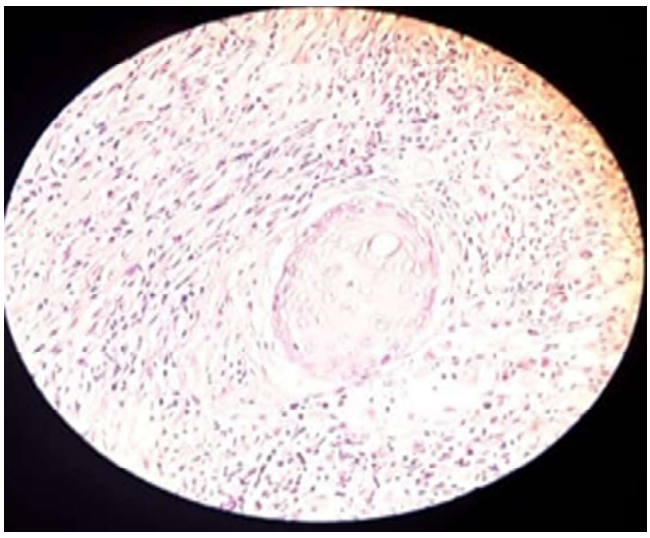

Figure 4

Figure.3 & 4.Microscopy shows foamy histiocytes and mononuclear cells Fig. 4. Shows a Touton giant cell.

The Cytology of the lesion confirmed that the histiocytes are non-Langerhans cells and led to the diagnosis of juvenile xanthogranuloma.. X-ray of chest and skull were normal. Ultrasonography of abdomen and pelvis showed no visceral involvement.

As there was a diffuse involvement with disfigurement, we planed to give some treatment to halt the disease process rapidly. We started isotretinoin 20 mg once daily after food. Patient was reviewed after one month and noted about 50% reduction of size of lesion . Same treatment was followed for another one month and noticed that most of the lesions had flattened with yellowish and hyperpigmented macules with decrease of size of lesion .

Repeat lipid profile and liver function test was done and no abnormalities were detected.

DISCUSSION:

Histiocytic disorders are broadly divided into two categories : Langerhans cell histiocytosis and non-'Langerhans cell histiocytosis.

Juvenile xanthogranuloma is classified under non-LCH and is also the most common of the non-LCH. It is a self-limiting and relatively uncommon benign cutaneous fibrohistiocytic lesion which can also affect deeper tissues and organs.

JXG is believed to result from a disordered macrophage response to a nonspecific tissue injury, resulting in a granulomatous reaction[3] .

JXG has been documented in many visceral locations such as lung, bone, testis, gastrointestinal tract, heart, eye and oral cavity[4] .

It can manifest as a multisystem disease [5]. Lesions can be single or multiple, well demarcated, rubbery , yellow to red papulonodules ranging upto 2 cm.

It is a disease of infancy and childhood which is rarely documented in adult. An association with neurofibromatosis type 1 (NF-1) and juvenile chronic myelogenous leukemia (JCML) has been reported[2].

Diagnosis is a combined approach of clinical and histopathological conclusion with IHC playing the vital role. Immunohistochemistry shows the lesions to be positive for factor XIIIa, CD68, CD163, fascin, and CD14 but negative for S100 and CD1. This can be used to differentiate these lesions from Langerhans cell histiocytoses[3] .

Most individuals with juvenile xanthogranuloma (JXG) are asymptomatic. Mostly it is the aesthetic and cosmetic reasons needing treatment.But can lead to ulcer formation and bleeding as in our case. As there was a diffuse involvement with disfigurement, we planed to give some treatment to halt the disease process rapidly.

We started isotretinoin 20 mg once daily after food. Patient was reviewed after one month and noted about 50% reduction of size of lesion . Same treatment was followed for another one month and noticed that most of the lesions had flattened with yellowish and hyperpigmented macules with decrease of size of lesion . Repeat lipid profile and liver function test was done and no abnormalities were detected.

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