



Case Report

# Bilateral juvenile xanthogranuloma of the buttocks - An unusual location: Case report

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## Abstract

Juvenile xanthogranuloma (JXG) predominantly occurs in infancy and childhood. Juvenile xanthogranulomas (JXGs) are asymptomatic; benign; self-healing; self-limiting red, yellow, or brown papules and nodules composed of histiocytic cells. Most common site of occurrence is skin of the head and neck region and trunk but occurrence at rare sites such as nasal cavity, limbus of the eye, iris of the eye, anterior abdominal wall and tibia had been reported in literature. JXG is the most common form of non-Langerhans cell histiocytosis. Approximately 35% of cases of juvenile xanthogranuloma (JXG) occur at birth, with as many as 71% of cases occurring in the first year. The mean age at presentation is 22 months. Most JXGs resolve by age 5 years. Despite the term juvenile in the disease name, 10% of cases manifest in adulthood. Most of the lesions are solitary. Juvenile xanthogranuloma is a histiocytic inflammatory disorder capable of presenting as different histological patterns. The classic form consists of sheets of foamy histiocytes and numerous multinucleated Touton-type giant cells containing no to very few mitotic figures. We present a case of large juvenile xanthogranuloma (JXG) in a 7 year old child involving an unusual location and bilateral involvement of both the buttocks.

## Key words

Juvenile Xanthogranuloma (JXGs), Touton giant cell, Benign, Non-Langerhans Histiocytosis.

## Introduction

Juvenile xanthogranuloma (JXG) is a benign histiocytic process of uncertain histogenesis,

characterized by red to yellow, single or multiple cutaneous nodules that are usually located on the head and neck or, less frequently, in other

areas [1]. Most tumors (40–70%) appear early in life, during the first year, although onset in adulthood is possible [2]. Adamson [3] in 1905 defined multiple yellow cutaneous papules as congenital xanthoma multiplex. JXG more often develops on the head and neck as solitary or multiple cutaneous lesions McDonald [4] reported a similar condition and referred it to as nevoxanthoendothelioma. In 1954, Helwig and Hackney proposed the name “juvenile xanthogranuloma” to define cutaneous lesions that arise from fibrohistiocytes [5]. In general, Juvenile Xanthogranuloma is a regressing or self limiting disorder usually occurring in infancy. It is characterized by one or more cutaneous nodules over the skin frequently involving head and neck region and less often by additional lesions in the deeper soft tissue or organs themselves. 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, toenail, palms, soles and lips [2].

### Case report

A 7 year old girl brought by her mother to the surgical outpatient department, with complaints of swellings in both the buttocks since a year. Patient’s mother noticed these swellings one year ago but they were small and painless. Recently in the last 8 months the swellings increased in size. There was no history of trauma or injections at the site of swellings. No complaints of discharge from the swelling. On examination, right side swelling was of size 5 x 4 x 2.5 cm. On the left side, size was 7 x 6.5 x 4 cm noted. Skin over the swelling was normal. Swelling was mobile, soft to firm. A clinical and cytological diagnosis of lipoma was made and patient was posted for surgical excision of both the swellings. Excised specimens were sent for the histopathological examination (HPE). Grossly, received two swellings, one is skin

covered measuring 5.5 x 4 x 3 cm, skin flap measuring 4.5 x 3.5 cm. Other swelling was measuring 7 x 6 x 4.5 cm (**Figure - 1**). Cut section of both the swellings showed homogenous solid orange-yellow appearance (**Figure - 2**).

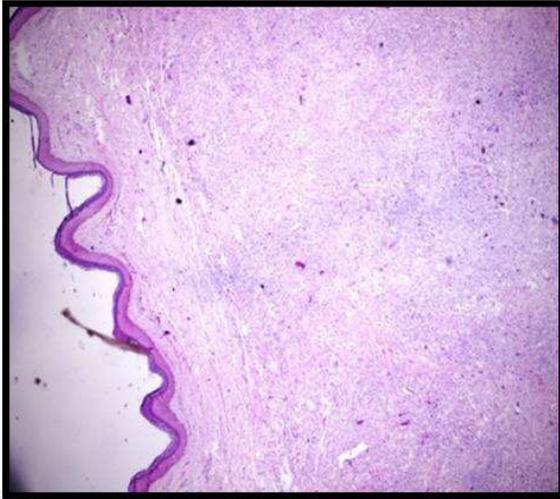
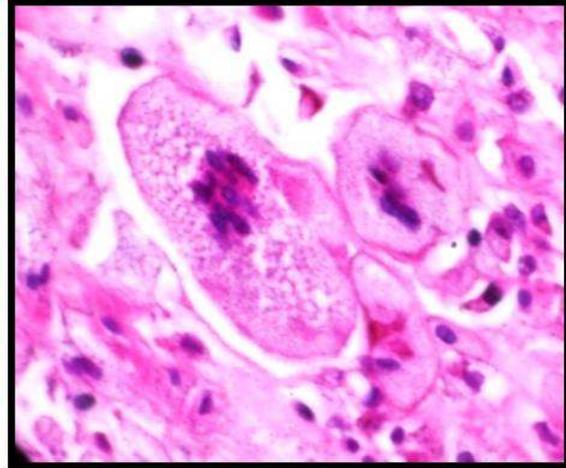
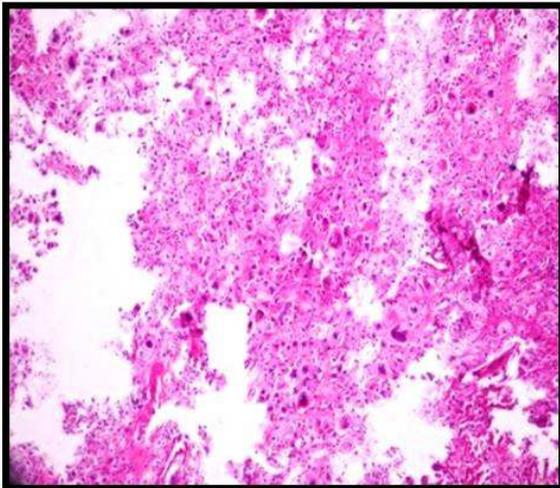
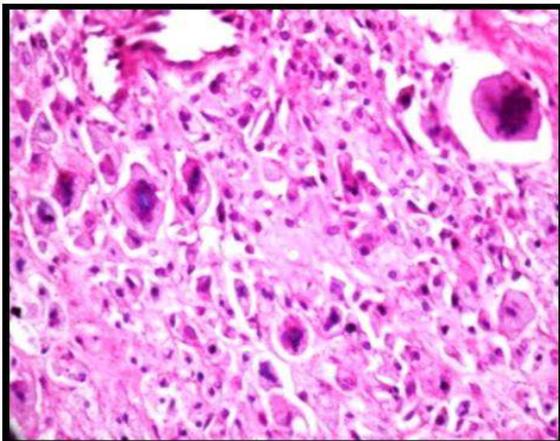
Microscopy showed stratified keratinized squamous epithelium (**Figure - 3**), thinned out at places with sub epithelial fibrocollagenous and fibro fatty tissue, dermal infiltrate consisting of lymphocytes, histiocytes, few eosinophils and many Touton giant cells (**Figure - 4, Figure - 5, Figure - 6**). Mitotic figures were 0-1/hpf.

**Figure – 1:** The excised gluteal swellings.



**Figure – 2:** Cut section of one of the swelling.



**Figure – 3:** Skin and subepithelial tumor tissue.**Figure – 6:** Touton giant cells. (100X View)**Figure – 4:** Low power view of the tumor tissue with lymphocytes, plasma cells, foamy histiocytes and Touton giant cells.**Figure – 5:** Touton giant cells. (High power view)

### Discussion

Juvenile xanthogranuloma (JXG) is a normolipemic, non-Langerhans cell histiocytosis characterized by yellowish cutaneous nodules that usually appear in early infancy and childhood, but may be present at birth or even in adulthood. Juvenile xanthogranuloma is usually asymptomatic. The most common locations for JXG are the head and neck, and solitary lesions are found in 60% to 82% of these patients. There are also examples of extracutaneous locations including the orbit, skull, liver, lung, spleen, kidney, brain, gastrointestinal tract, pancreas, and submandibular gland [6, 7]. Juvenile xanthogranuloma is a benign condition that usually regresses spontaneously within 3 to 6 years. Juvenile xanthogranuloma (JXG) occurs in whites approximately 10 times more frequently than in African Americans. In childhood, juvenile xanthogranuloma (JXG) occurs predominately in males (1.5: 1). Equal incidence occurs in adult males and females. Multiple cutaneous lesions occur predominantly in males (12: 1).

Adamson first reported JXG in the English literature in 1905 [3]. He presented a child who developed numerous yellow-white papules on the body in the first 2 weeks of life. He named



the entity as congenital xanthoma multiplex. In 1912, McDonough presented the first case review and renamed the condition nevoxanthoendothelioma [4]. In 1954, Helwig and Hackney again re-termed it as juvenile xanthogranuloma, reflecting its histopathologic appearance [5].

Histiocytosis-X or Langerhans granulomatosis mimic juvenile xanthogranuloma (JXG), both occur in early part of the life, self limiting and involute spontaneously. Histologically both exhibit giant cells and lipidization. But, the characteristic Birbeck granules in 50 % of the histiocytes in the Langerhans granulomatosis group have not been observed in Juvenile Xanthogranuloma (JXG). Additionally, inflammatory reaction is milder with few eosinophils in Juvenile Xanthogranuloma. One closer mimic of JXG is Fibrous Histiocytoma because both the entities have Touton giant cells, but JXG has more lymphocytic and eosinophilic infiltrate than that of Fibrous Histiocytoma.

JXG usually is a self-limiting disorder, and no definite treatment is required. The lesions most often regress over a period of 6 months to 3 years. Klemke, et al. [8] have demonstrated the successful use of CO<sub>2</sub> lasers in the treatment of multiple JXG lesions. In multiple lesions, complete surgical excision is the definite treatment of choice with negligible chances of recurrence.

### Conclusion

In conclusion, juvenile xanthogranuloma is a rare type of hyperplastic diseases of non-Langerhans histiocytes. JXG is most often a self-limiting disease that often spontaneously regresses. Lesions may resolve completely or may leave a residual atrophic or hyper

pigmented scar. The pathogenesis is unknown. When large and multiple lesions are present, surgical excision is treatment of choice.

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