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## Solitary cutaneous nodule in a young boy: A diagnostic dilemma

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### Abstract

Juvenile xanthogranuloma (JXG) is a type of benign histiocytic proliferative disorders seen in first two decades of life. It is a relatively uncommon benign cutaneous fibrohistiocytic lesion. Clinically, it presents as a papule or nodule which can either be solitary or can be yellow-brown in colour and firm in consistency. Usually, it is located in the face and neck. Sporadically, it can involve internal organs which can lead to serious complications. Differential diagnosis of this entity should be kept in mind whenever young patients presents with solitary nodule as it is a benign condition and only treatment warranted with local excision of the lesion.

**Keywords:** Juvenile xanthogranuloma, fibrohistiocytic, benign, immunohistochemistry

### Introduction

Juvenile xanthogranuloma is a type of histiocytic proliferative disorders which may present at birth. However, it is more commonly seen in the first two decades of life. It is a relatively uncommon benign cutaneous fibrohistiocytic lesion <sup>[1]</sup>. Adamson reported it for the first time in the year 1905 <sup>[2]</sup>. Clinically, it presents as a papule or nodule which can either be solitary or can be yellow-brown in color and firm in consistency. Usually, it is located in the face and neck <sup>[3]</sup>. Sporadically, it can involve internal organs which can lead to serious complications <sup>[4]</sup>. Histologically, there is collection of histiocytes, foamy cells and touton giant cells. The lesions on skin generally follow a benign pattern with spontaneous resolution over the period. We present a case report of a 4 year old boy who presented with solitary red cutaneous nodule in the neck following minor injury.

### Case presentation

A 4-year-old boy presented with complaint of a swelling in the neck since past seven months. Over the period, the swelling gradually progressed and had increased in size from 4x4 mm to 1x1 cm over a span of 7 months. There was a history of minor trauma. No history of fever, pain or discharge was present. The child was born of full term normal vaginal delivery with uneventful postnatal course. He had no history suggestive of any developmental delay and was immunised for age. There was no history of similar illness in the past.

Examination revealed a well circumscribed papule in anterior midline of neck. It was 1 cm in diameter with a smooth surface. On palpation, it was a soft mass with regular margins and no fixity to underlying structures was noted. There was no clinical evidence of any lymphadenopathy. His blood investigations were within normal limits. No clinical anomaly was detected on his systemic examinations. The clinical diagnosis of Pyogenic granuloma was considered. The ultrasound neck revealed a well-defined oval hypoechoic lesion showing uniform low level internal echoes in the skin and subcutaneous plane in the midline submental region. Color doppler showed no flow within and around the lesion. The radiological findings were suggestive of Dermoid /epidermoid cyst. The patient underwent an excision biopsy.. Histological evaluation of the lesion revealed a bit lined by keratinised squamous epithelium. Dense dermal infiltrate of histiocytes was noted. These cells were seen extending into the subcutaneous fat. These cells were round to polygonal in shape with vesicular nucleus, prominent eosinophilic nuclei and foamy to eosinophilic cytoplasm. Background inflammatory infiltrates comprising predominantly of lymphocytes and plasma cells were also seen. Toutan type of giant cells were seen admixed with these cells. The number of mitosis was <1/10 hpf.

No atypical cells or any evidence of malignancy was present. Based on morphological findings, differential diagnosis of benign histiocytic proliferative lesion, Langerhans cell histiocytosis, Spitz nevus and rhabdomyosarcoma was considered. On histochemical evaluation the cells were immunoreactive for CD68 & vimentin. These cells were negative for S100, HMB45, desmin, myogenin & Cd1a. The Ki67 index was <1%. [Fig 1] Based on morphological findings and immunohistochemical findings the diagnosis of juvenile xanthogranuloma was made. The child post excision has been doing well and is on follow up.

### Discussion

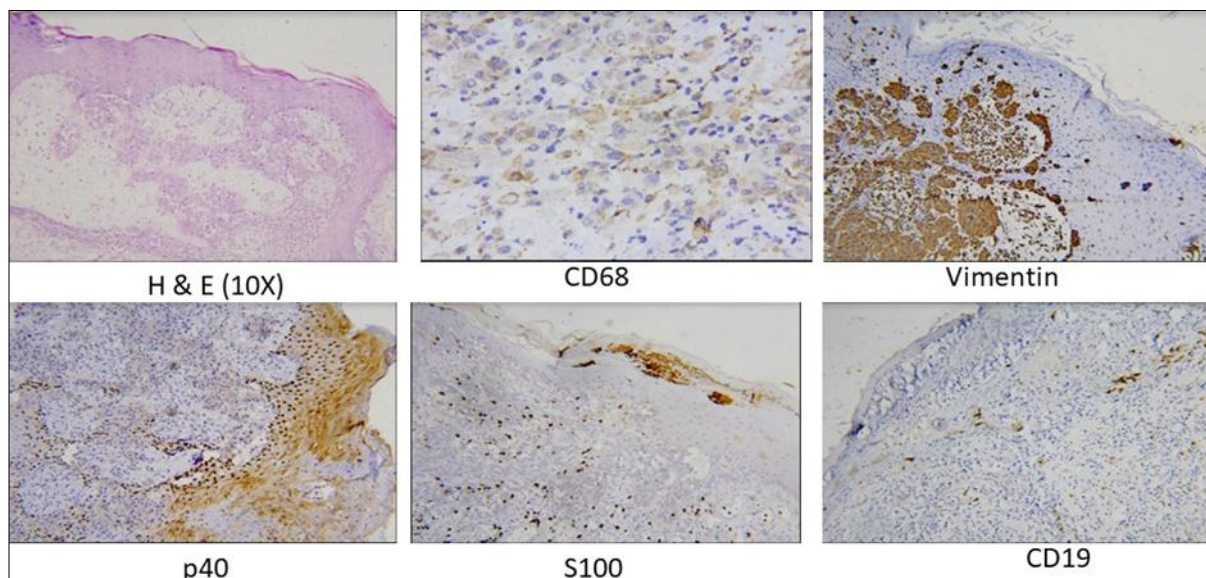
Histiocytic disorders are defined by their component cells. With appropriate clinical correlation, the cells which are CD1a+/Langerin+/S100+ can be ascertained as Langerhans Cell Histiocytosis (LCH). The ultrastructural studies are not recommended in these cases. The non-LCH disorders constitutes a spectrum of lesions which does not meet the phenotypic criteria of LCH. Juvenile xanthogranuloma is a benign cutaneous fibrohistiocytic lesion and a type of granulomatous process. The lesions may be accompanied by deposition of lipids. It was first reported by H.G. Adamson in the year 1905 [5]. The etiology is believed to be a non-specific insult leading to a disordered macrophage response [6]. The pathogenesis remains unknown. Infective aetiology or any physical factor may be one of the cause. Since the disease is clinically diagnosed and not always confirmed on biopsy, it can be misdiagnosed.

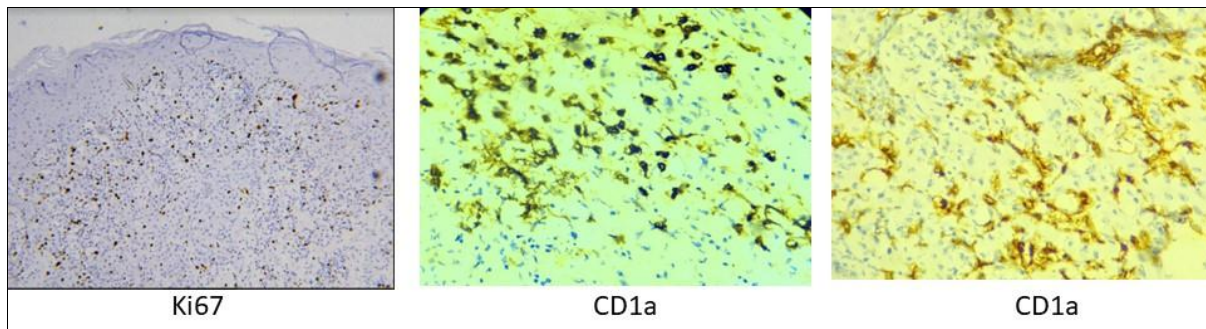
A solitary cutaneous lesion with or without any organ involvement is the most common presentation. The primary clinical feature is a papulonodular lesion which can be tan – orange in color and several millimeters in diameter, it may be single or multiple. The lesions may exceed >2 cm, which are termed as giant or macronodular lesions. The various specific forms have been described- Mixed, giant, subcutaneous, eruptive & plaque-type. The common sites of occurrence are the skin of head and neck, but these can sometimes occur on the trunk and extremities also. The lesion can be intramuscular as well which tends to be bigger than cutaneous ones. The eye is the most frequent extracutaneous location [7]. The most common ocular

presentation can be a asymptomatic iris tumor with signs of uveitis or unilateral glaucoma [8]. There may be ocular involvement without any cutaneous manifestation. Children younger than 2 years with multiple skin lesions are at greater risk of an ocular involvement [2]. Rarely, extracutaneous lesions have been reported which are associated with morbidity. The male to female ratio of cutaneous JXG is about 1.4:1 in children, while in adults no sex predilection exists [9]. The confirmation of clinical diagnosis can be made by skin biopsy.

Characteristic histologic findings in juvenile xanthogranuloma are: dense dermal histiocytic infiltrate and Touton Giant cells which are multinucleated, with homogeneous eosinophilic cytoplasmic center and xanthomatization in the periphery [6]. These giant multinucleated cells may vary in number. Simultaneously, perivascular or perilesional inflammatory cells may be present at times. The ultrastructural studies of lesions has showed non specific organelles ranging from worm-like bodies and popcorn bodies [10]. Immunohistochemistry has an important role in the differential diagnosis between Langerhans cell histiocytosis (LCH) and JXG. The histiocytes and giant cells present in JXG are of monocyte-macrophage in origin. They therefore, show strong immunoreactivity to CD68 & HAM. S-100 protein immunoreactivity, which is a marker for the diagnosis of LCH, is typically absent [11, 12]. In most cases with JXG, S-100 protein was non reactive, but scattered cells may show weak cytoplasmic reactivity. However, in histological examinations showing S-100 positivity, the diagnosis of JXG should not be ruled out and clinical correlation is always advised [1].

Differential diagnoses are Langerhans cell histiocytosis, pyogenic granuloma, Spitz nevus, urticaria pigmentosa, xanthomas and molluscum contagiosum. Juvenile xanthogranuloma may be associated with neurofibromatosis type I and myelomonocytic leukemia. Spontaneous regression usually occurs within 1-3 years and recurrence is rare. Large size nodules or cosmetic reasons are the main indication for surgical removal of nodules. Chemoradiotherapy, immunosuppression (steroids, cyclosporine, methotrexate) and surgery have been used in systemic cases [13].





Legends :

**Fig 1:** H & E section showed subepithelial round to polygonal large cells with vesicular nucleus and prominent eosinophilic nuclei lying in diffuse sheets, cord and trabeculae in an inflammatory background. These cells are immunoreactive for CD68, Vimentin and negative for p40, S100, CD19, Cd1a. Ki67 index was less than 1%.

### Conclusion

JXG is a self-limiting disorder seen in young children and usually requires no treatment. Regression of tumor is seen over a period of 6 months to 3 years. It usually presents after minor trauma. Extracutaneous involvement is rare. Juvenile xanthogranuloma should be considered in differential diagnosis of cutaneous nodule in young children as wide local excision is the only treatment required in these case.

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