Received: 25 February 2024 | Revised: 16 April 2024 | Accepted: 1 May 2024

DOI: 10.1002/ccr3.8912

CASE REPORT

Clinical Case Reports Open Access WILEY

Juvenile xanthogranuloma manifesting in the forehead: A case report

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Key Clinical Message

A 3 -year-old boy presented with a forehead nodular mass, which was excised and confirmed histologically as Juvenile Xanthogranulomma (JXG). It affects children with a predilection for the head and neck region. A relatively rare, benign, histiocytic proliferative cutaneous disorder with a potential for malignancy. A prompt and wide resection is recommended.

KEYWORDS

children, head and neck, juvenile xanthogranuloma

1 | INTRODUCTION

Juvenile Xanthogranuloma (JXG) is a relatively uncommon, benign, histiocystic proliferative cutaneous disorder that typicaly affects children, with the head and neck being the most common sites.[1] The lesion is a non-Langerhans cell histocytosis whose exact prevalence remains unknown.[2] Cutaneous lesions are usually asymptomatic and most of them spontaneously involute over a course of several years.[3]

JXG is a proliferative disorder of histiocystic cells of the dermal dendrocyte phenotype and is diagnosed clinically and confirmed after biopsy for histological analysis.[3] We present a case of JXG prominently manifesting in the forehead of a 3-year-old boy.

2 | CASE REPORT

2.1 | Case history/examination

A 3-year-old boy first presented with a forehead firm, nontender mass that had evolved over about 1 month. During a follow up appointment after 1 week, ulceration of the mass (Figure 1) was noted for which his parents confirmed traumatic injury.

3 | METHODS

A haemogram indicated a full range of normal parameters with hemoglobin at 11.9 g/dL. A differential diagnosis of Langerhans cell histiocytosis, Xanthomatous lesions, dermatofibromas and the Spitz nevus was made pending the definitive one.

3.1 | Treatment and Investigation

The treatment plan was excision under general anesthesia. The lesion was excised through an elliptical incision and closure accomplished utilizing a one layer monocryl suturing. The tissue was sent for histopathology evaluation. The patient has been follow up for 6 months and the recovery has been uneventful.

4 | RESULTS

Histological analysis showed a delimited completely excised lesion of proliferated histiocystes admixed with lymphocytes, foamy microphages and touton type giant cells.

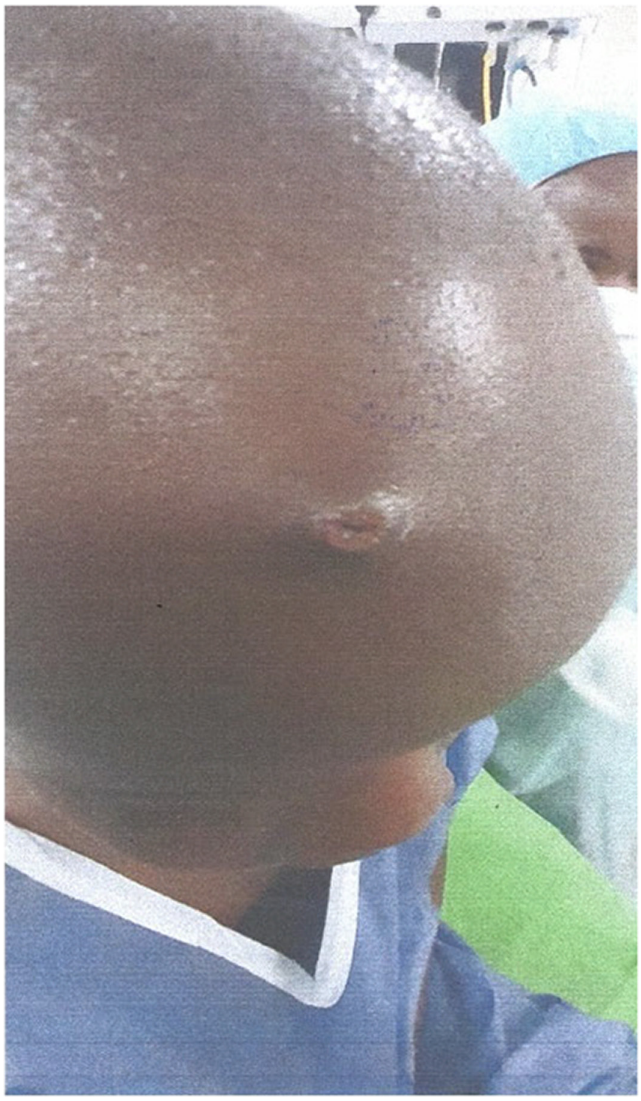

FIGURE 1 Clinical Photo. A traumatically ulcerated forehead nodular lesion.

The features were consistent with a benign histiocytic lesion the JXG (Figure 2A,B).

5 | DISCUSSION (TREATMENT AND FOLLOW-UP)

JXG is a rare non-Langerhans cell histiocytic disorder which was first described in 1905 by Adawson as a congenital xanthoma multiplex that belongs to a group of histiocytic lesions with a macrophage heterogenous phonotype and variable factor XIIIa and fascin reactivity.[4] One study outlined the diagnostic criteria for JXG, as typical Touton cells resette shaped nuclei amid eosinophilic or vacuolar cytoplasm, this was in keeping with the patients histopathology.[5,6]

The true incidence of JXG may be underestimated since many lesions, especially those which are solitary and small (in up to 90% of the patients), may go unrecognized.[3] However, lesions manifesting in exposed areas as the present case are noted early and should raise concern for prompt intervention. Furthermore, traumatic ulceration may commonly occur during play.

Meticulous histopathological analysis should be performed in order to rule out concurrent systemic malignant processes. The etiology of JXG is unknown although some authors postulate the existence of an alteration of the macrophage response to a nonspecific stimulus such as trauma and viral infection, a hypothesis that still lacks evidence.[7] This lesion which has predilection for the eyes is rarely linked to systemic manifestations.[8] It has been recommended that enlarging soft tissue masses in children in whom a malignancy cannot be excluded by physical or radiological examination be totally excised where feasible, when no functional compromise from surgery is anticipated.[9] As for the systemic Langerhan cell histiocytosis the treatment protocol is chemotherapy and for the Central nervous system, the management may not have an effect on the patients mortality.[10] While in the present case aesthetic compromise was notable, the incision design was intended to extirpate the entire lesion. A month

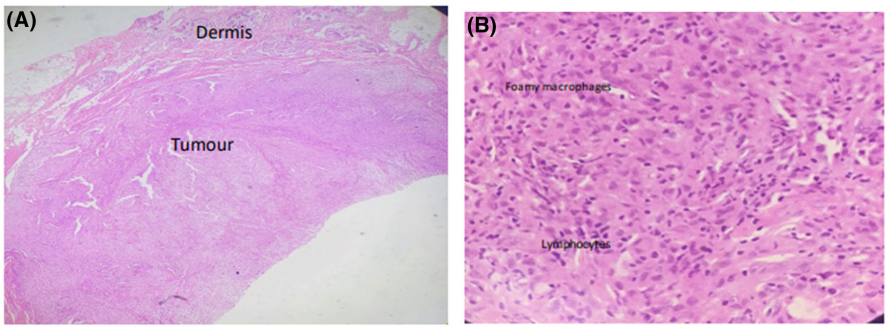

FIGURE 2 (A) (×4 magnification). Histological Illustration of a well dermacated lesion delimited completely excised lesion. (B) (×400 magnification) of proliferated histiocytes admixed with lymphocytes foamy macrophages and touton type giant cells.

after follow up of the surgical intervention the linear scar had healed well with no evidence of hypertrophy. The patient remains under review for over at least 6 months.

AUTHOR CONTRIBUTIONS

Fawzia Butt: Data curation; writing – review and editing. **M. L. Chindia:** Conceptualization; supervision. **M. Mung'ania:** Data curation. **Richard Owino:** Methodology.

ACKNOWLEDGMENTS

We greatly appreciate Claris Kamula for skillfully preparing the manuscript.

FUNDING INFORMATION

There was no funding available for this report.

CONFLICT OF INTEREST STATEMENT

None.

DATA AVAILABILTY STATEMENT

Data supporting this case report will be available once this article has a accession number.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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How to cite this article: Chindia ML, Butt FMA, Mung'ania M, Owino RO. Juvenile xanthogranuloma manifesting in the forehead: A case report. *Clin Case Rep*. 2024;12:e8912. doi:10.1002/ccr3.8912