doi: 10.31674/mjmr.2022.v06i02.003

MJMR

Juvenile Xanthogranuloma: A Case Report

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ABSTRACT

Juvenile Xanthogranuloma (JXG) is a rare condition that can present as early as a neonate or develop later during paediatric age and adulthood. One should be aware of this entity as it can cause extreme anxiety to parents due to the variety of cutaneous features. Our case report details a JXG in an 11-months-old boy with a circumscribed nodule on his scalp, neck, and left arm. The nodule was histologically identified as a JXG following a skin punch biopsy.

Keywords: Juvenile Xanthogranuloma; Nodule, Biopsy

INTRODUCTION

Newborns are affected in Juvenile Xanthogranuloma (JXG) (Mazlan & Aris, 2021). But JXG's true incidence is unknown. It has been reported in just 2371 cases by 122 dermatologists with an average of 12 years of experience, making it a rare condition with less than two cases per dermatologist per year. JXG is commonly characterised by erythematous yellowish papules or nodules that appear on the head and neck. Extracutaneous involvement is quite rare, and the majority of cases involve the eyes (Chang, Frieden & Good, 1996). JXG is the most often encountered non-Langerhans cell histiocytosis in children (Cichewicz *et al.*, 2019). Histiocytes, foam cells, and Touton giant cells serve as histology markers for JXG (Sangüeza *et al*., 1995). The prognosis for JXG is generally favourable; spontaneous regression is the norm. Following regression, patients may experience lingering hyperpigmentation, mild shrinkage, or anetoderma (loss of elastic fibres in the dermis) (Hernandez-Martin *et al.*, 1997). Fatalities are rare, occurring most frequently in newborns with JXG systemic involvement (Höck *et al.*, 2019).

CASE REPORT

An 11-months-old boy presented with a painless nodule over his left arm since he was one month old. The nodules gradually enlarged in size from 2 mm to 30 mm in diameter. The parents also noted multiple nodules over the head and pustules over the neck, gradually enlarging in size since 2-months-old.

The nodules were initially treated as a fungal skin infection with topical cream by the primary care physicians. However, after several unsuccessful attempts, the guardian brought him to the dermatologist for a second opinion.

He was the only child in the family. He was born term at Hospital Tunku Ampuan Afzan (HTAA). He weighed 3.35 kg when he was born. He completed his vaccinations up to 6 months with the pneumococcal vaccine second dose. He had a history of congenital pneumonia after birth. Upon further questioning, none of the family members had the same problem, no family history of malignancy, and no history of atopy. The child was otherwise healthy, active, and thriving. He was not noted to be in pain nor itchy. There was no fever, contact with tuberculosis patients, loss of weight, nor loss of appetite. There was also no history of insect bites and no previous similar presentation.

On physical examination, there are multiple macro nodules over the scalp size of 30 mm x 20 mm (Figure 1); they are well-circumscribed yellowish nodules with erythema at the surrounding margin and hyperpigmented skin. It is firm and non-tender.

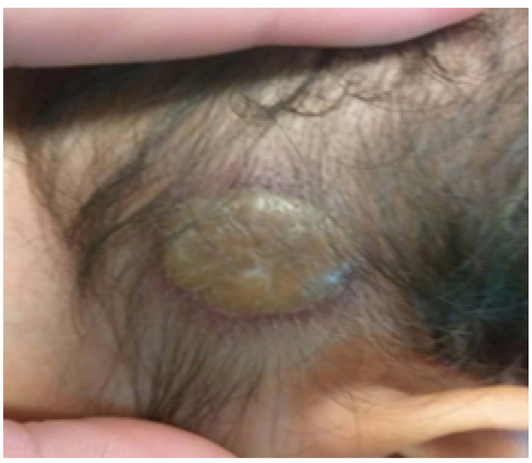

Figure 1: Scalp-tinged orange-brownish macro nodules

Over the left arm, there is a brown nodule with hyperpigmented skin and well-Circumscribed surrounding erythema (refer to Figure 2).

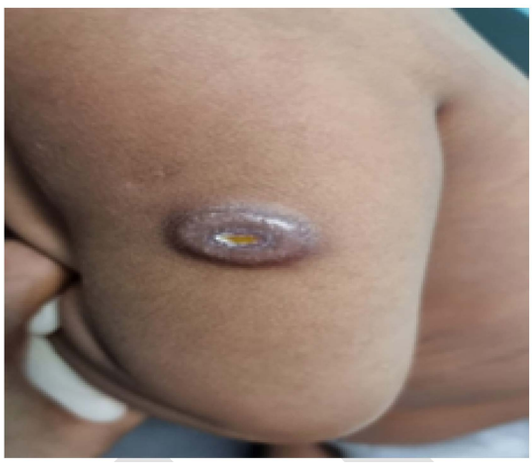

Figure 2: left arm, there is a brown nodule with hyperpigmented skin and well-circumscribed surrounding erythema

On the neck, there are two pustules with a regular margin (Figure 3).

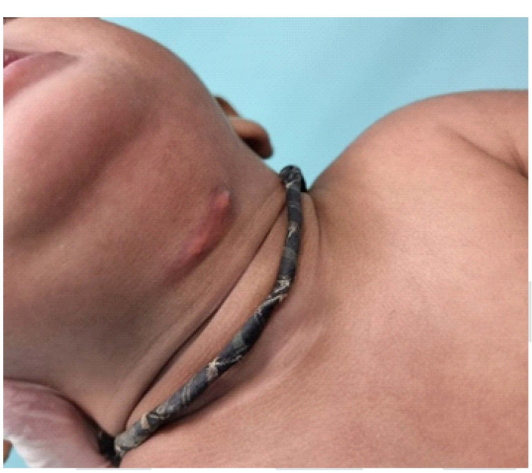

Figure 3: tinged orange-brownish pustules with a regular margin

It is firm and non-tender. There are sub-centimetric lymph nodes at the left inguinal; they are firm and non-tender. Otherwise, there is no hepatosplenomegaly.

Skin punch biopsy of the left arm nodule was done under local anaesthesia. The histopathology examination revealed the proliferation of histiocytes within the dermis in a diffuse pattern, in which some are foamy macrophages (Figure 4).

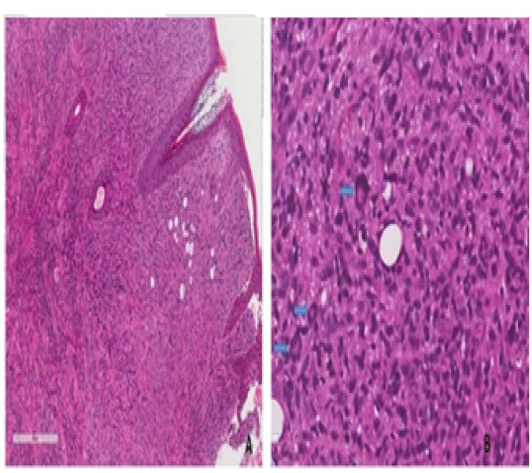

Figure 4: Diffuse dermal proliferation of histiocytes (H&E, 100x magnification (A). Scattered multinucleated giant cells as indicated by the arrow. Foamy histiocytes and eosinophils are also seen.(H&E, 400x magnification) (B).

These histiocytes are CD68 positive. A few eosinophils infiltrated are seen. Scattered multinucleated giant cells are present. There was no malignancy. CD1a and S100 are negative, and the features are consistent with juvenile xanthogranuloma.

DISCUSSION

JXG is primarily a childhood condition, with just 10% of cases occurring in adults. Lesions arise in 64% of patients by the age of 7 months and 85% within a year. In up to one-third of cases, lesions are congenital (Cichewicz *et al.*, 2019) Gianotti *et al*. classified JXG as micronodular (less than 10 mm in diameter, frequently several lesions) or macronodular (10-20 mm), with a broad spectrum of lesions in terms of shape, colour, and number (Gianotti & Caputo, 1985).

Clinical features of JXG are consistent with our patient, as a well-demarcated papule or nodule with a smooth surface is the hallmark of a JXG lesion. It has a typical pink-reddish tint in its early phases, but it gradually becomes a distinguishing yellow, orange, or brownish tone. The lesions are solid in texture, and some contain superficial telangiectasias. JXG is usually asymptomatic, except in some places or with massive lesions. The head and neck are the most commonly affected areas, followed by the trunk and limbs (Dehner, 2003).

In our case, we did a skin punch biopsy to rule out other differential diagnoses. Xanthomatous lesions, mastocytomas, dermatofibromas, Spitz nevus, and Langerhans cell histiocytosis should all be considered upon encounter (Zvulunov, Barak & Metzker, 1995). The histopathologic finding in our case revealed histocyte cells with CD 68 positive and negative for S100 and CD1a, which is consistent with JXG. Histopathologic findings vary according to lesion stage, with early-stage lesions exhibiting a monomorphous infiltrate of lipid-free macrophages that can occupy most of the dermis and later-stage lesions exhibiting an abundance of vacuolated foamy macrophages, Touton-type multinucleated giant cells, particularly in the superficial dermis. JXG lesions are positive for macrophage markers such as CD68, CD163, KiM1P, anti-FXIIIa, vimentin, and anti-CD4 but are often negative for S-100, CD1a, and CD207 (antilangerin), which are unique to Langerhans cells (Sangüeza *et al.*, 2019).

The discovery of a triple relationship between JXG, Neurofibromatosis type 1 (NF-1), and juvenile myelomonocytic leukaemia (JML) (Zvulunov, Barak & Metzker, 1995) sparked great discussion, as subsequent retrospective studies found no increased incidence of JML in individuals with JXG or NF-1 (Cambiaghi, Restano, & Caputo, 2004). Systemic screening is warranted if any extracutaneous manifestation is suspected (Chang, Frieden & Good, 1996). Hence, the patient was referred to a paediatrician and ophthalmologist and awaiting their reviews.

Luckily, the prognosis is excellent with cutaneous JXG with lesions clearing spontaneously and selfheal (Hernandez-Martin *et al.*, 1997). As in this case, even though the diagnosis was missed at the earlier stage, it did not cause any harm to the child. Solitary lesions that hinder function or cause psychological discomfort due to their location (especially among parents) can be surgically removed or treated with various ablative treatments, such as carbon dioxide laser therapy (Cambiaghi, Restano, & Caputo, 2004).

CONCLUSION

This case report highlights the importance of considering other differentials when encountering skin problem which does not improve with medical treatment at primary care and aid in the early detection of JXG. JXG is rare with a multi-faceted presentation of cutaneous features. One should always consider other differentials, hence the need for a biopsy and histopathological correlation. This case report aims to educate health care providers and raise awareness of JXG possible visceral complications and associated medical diseases. Early referral can avoid extreme anxiety, time-consuming, and financial burden to the guardian and patients.

Conflict of Interest

There was no conflict of interest.

ACKNOWLEDGEMENT

No external funding was received for this study.

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