

Juvenile xanthogranuloma of the nose

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ABSTRACT

Juvenile xanthogranuloma (JXG) is a relatively uncommon, benign histiocytic proliferative disorder. One should be aware of this entity in order to make a diagnosis with the possibility of visceral involvement as well as associated medical conditions related to it. The present case report describes a case of JXG in a two-year old boy who presented with a circumscribed papule located over the vestibule of the right nostril. The lesion was histologically diagnosed as a JXG after surgical excision of the mass.

Keywords: Benign neoplasm, excision, juvenile xanthogranuloma, vestibule

INTRODUCTION

Juvenile xanthogranuloma (JXG) is a member of the non-Langerhans cell group of histiocytic proliferative disorders and is a relatively uncommon benign cutaneous fibrohistiocytic lesion.¹ Adamson, who referred to 'congenital xanthoma multiplex' as a single or occasionally multiple cutaneous lesions that occurred in infants and showed spontaneous involution, first described JXG in 1905. The lesions were designated 'nevoxantho-endothelioma' by McDonagh in 1912, who considered them to be derived from endothelial cells. However, since Helwig and Hackey in

1954 showed the fibrohistiocytic origin of the tumour on the basis of clinicopathologic evaluation, the term 'juvenile xanthogranuloma' has been commonly used.² It is characterised by firm, rubbery eruptions in the head, neck and upper trunk. It is initially reddish papular or nodular, which later turns yellow and finally to brown flattened plaques or macules. Large lesions may ulcerate.³

We report the case report of a two-year-old boy with JXG localised to the vestibule of nose.

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CASE REPORT

A two-year-old boy presented with a soft tissue mass over the right nostril, which was first noticed as a small area of redness two

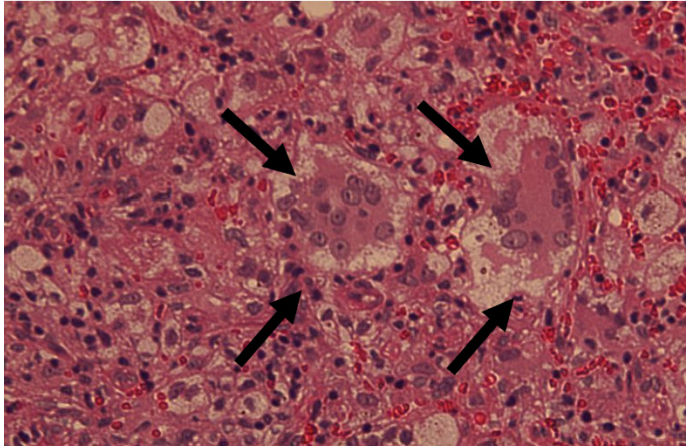


Fig.1: Granulomatous infiltrate with presence of multinucleated giant cells (Touton cells) indicated by arrows, (H&E stain, x100).

months earlier. The mass continued to grow and became quite prominent. There was no bleeding or crusting noted over the mass. The patient was otherwise healthy.

Physical examination revealed the mass to be a well-circumscribed papule that was located over the vestibule of the right nostril. It measured approximately 5mm in diameter. It was not attached to the right inferior turbinate. On palpation, it was firm and non-tender. There was no ulceration of the overlying skin.

The patient underwent an excisional biopsy under general anaesthesia without any complications. Histological examination revealed the presence of inflammatory cells mainly of neutrophils, haemosiderin laden macrophages and Touton giant cells, some with lipidised cytoplasm. There was no evidence of malignancy or granuloma seen. Immunohistochemical analysis of the lesions was positive for CD68. These clinical and histological findings were consistent with the diagnosis of JXG.

Post-operatively, the operated site

epithelialised and healed without any complications. There was no recurrence observed after six months of follow up.

DISCUSSION

JXG is a common form of non-Langerhans cell histiocytosis that predominantly affects infants and children. It is a benign cutaneous fibrohistiocytic lesion and a type of granulomatous process, at times accompanied by lipid deposition.¹

Solitary or multiple yellowish papules or nodules on the head, neck and trunk characterise its usual presentation. It may also occur as a soft tissue lesion with or without organ involvement. Although the skin of the head, neck and trunk are among the most common sites for JXG, intranasal lesion is very rare. To our knowledge only two other cases have been reported in English medical literature.⁴⁻⁵

JXG has been documented in many visceral locations such as the brain, intestine, liver, heart, kidney, appendix and lung.⁶ It can manifest, as a multi-system disease and the intramuscular lesion tends to be larger

than cutaneous ones. ¹ Reported medical conditions associated with the lesion are neurofibromatosis, urticaria pigmentosa, Niemann-Pick disease and myelogenous leukaemia. Therefore, a careful assessment of the lesion with appropriate investigation is essential to determine the definite treatment and outcome.

JXG is a disease often observed in early childhood. Median age of onset is two years, but lesions may be present at birth. ¹ It has been reported that five to 17% of cases arise within the first year of life. ⁷ However they are not confined to childhood and may be found in patients of any age.

The pathogenesis of JXG is unknown. The initiating stimuli may be associated with infection or physical factors. ⁵ The clinical course tends to be benign and lesion spontaneously regresses over a period of months to years. ⁸ JXG may resolve completely or leave behind an area of residual hyperpigmentation or atrophy. ⁹

Histologically JXG shows well-circumscribed nodules with a dense infiltrate of histiocytes. Those that involve the skin usually infiltrate the dermis. A wreath of nuclei around a homogenous eosinophilic cytoplasmic centre characterises Touton giant cell, seen in 85% of cases, while the periphery shows prominent xanthomatisation. ¹ The presences of Touton giant cell is typical of JXG, but is not specific and may be absent. ⁹ Fibrosis may also be observed, especially in older regressing lesions. Immunohistochemical analysis is therefore important and lesions are positive for CD68 and HAM56 and negative for S-100 and CD1a. The latter are mark-

ers for the diagnosis of Langerhans cell histiocytosis. ²

Excision of the solitary lesion should be considered to establish a histological diagnosis. However, in view of spontaneous regression, a conservative approach has been advocated. ³ Excision of the lesion is an adequate treatment and recurrence is uncommon. ¹ Despite the likelihood of spontaneous regression, it is often decided to excise the lesion for esthetic or diagnostic reason, as was the case for our patient. Another important factor is the concern of parents in which a malignancy cannot be excluded.

In conclusion, JXG can be readily recognised when it presents as the usual yellow cutaneous nodule. Although primarily a dermatological entity, one should be familiar with this entity, and should be considered in the differential diagnosis of benign soft tissue tumour of the nose.

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Brunei Darussalam — Healthcare in Pictures



Picture of the right showing Hjh Hapsah Bte Hj Manudin (Previous Acting Principal of Nursing, Ministry of Health) with two young patients, Nasib and Alex (patient with clubbed foot) taken outside of the Paediatric ward of the Brunei General Hospital (1972). Picture on the left showing Hjh Hapsah with three paediatric patients.
