CASE REPORT

Rare case of adult Xanthogranuloma: A case report

Fatima Hasan, Najia Ahmed

Abstract

Xanthogranulomas are the most common form of non-Langerhans cell histiocytosis. They are benign, asymptomatic, and self-healing, affecting mostly infants, children, and very rarely adults. They present clinically as erythematous to yellow-brown papules. In children they can be single to multiple but in adults they are solitary. We present the case of a 23-year-old Pakistani man with an erythematous to yellow-brown papule on his neck that persisted for 1.5 years. The results of an excision biopsy showed histopathological features of histiocytes, tuoton giant cells and necrobiosis corresponding to xanthogranuloma. We emphasize the importance of considering xanthogranuloma in skin coloured nodules.

Keywords: Xanthoma, Non-Langerhan cell histiocytosis, Skin tumour.

DOI: https://doi.org/10.47391/JPMA.5314

Submission completion date: 08-01-2022

Acceptance date: 09-06-2022

Introduction

Xanthogranulomas (XG) are the most common form of non-Langerhans cell histiocytosis.[1] They are benign, asymptomatic, and self-healing, affecting mostly infants, children, and rarely adults.[2] Histiocytosis is categorised into three groups as Langerhans cell histiocytosis (LCH), non-Langerhans cell histiocytosis, and malignant histiocytosis. Within these groups are multiple disease subtypes, and xanthogranuloma falls within the non-LCH group.[3]

Usually, xanthogranuloma is termed as Juvenile xanthogranuloma (JXG). Although around 10% of cases manifest in adulthood, 81% of cutaneous JXG cases manifest as a solitary lesion. The solitary form is more common in cases of adult onset.

The pathogenesis of XG is unknown. Although aetiology has been linked to various physical and infectious factors, most investigators believe that it is caused by a reactive response to an unknown stimulus.[4] In both the juvenile and adult variants of xanthogranuloma, patients present with cutaneous, discrete, well-demarcated, reddish to yellow, dome-shaped papules and nodules that often are asymptomatic.[5] Rarely systemic involvement can occur with deep soft tissues being the most common site involved followed by liver, spleen, lungs and central nervous system. Due to self-healing nature of the cutaneous XG, systemic treatment is controversial. However spontaneous regression is not common in adult xanthogranuloma. Treatment options for solitary lesions include surgical excision, retinoids and carbon dioxide laser. It is a benign condition with a good prognosis and is not typically invasive.[3]

We present a case of JXG in a 23 year old male, being very rare in adults.The importance of considering XG in differential diagnosis of erythematous to skin coloured papules/nodules is emphasised.

Case Report

A 23-year-old Pakistani man presented on 21st April, 2021 at PNS Shifa Hospital Karachi, with an asymptomatic, gradually enlarging papule on his neck, as seen in Figure 1. The papule had persisted for 1.5 years. He had no other skin lesion. He gave no history of headache, seizures or photophobia. On physical examination, the patient had a solitary, well-demarcated, reddish, dome-shaped, and flat-topped, 0.5- to 1-cm papule on front of his neck with surface telangiectasia. Scaliness and central depression were absent. The lesion was firm in consistency, and no other similar lesion was found elsewhere. Rest of the

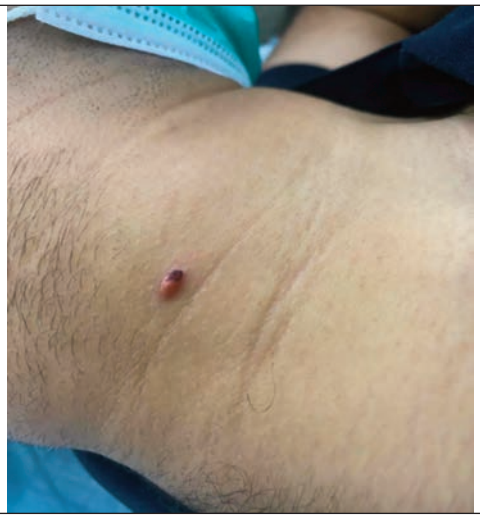

Figure-1: Skin coloured- well demarcated solitary nodule on neck. Crusting on the top corner developed after an attempt to extract it as milia.

[1,5,6]Department of Dermatology, PNS Shifa Hospital, Karachi, Pakistan.

Correspondence: Fatima Hasan. e-mail: fatima17hasan@gmail.com

ORCID ID. 0000-0003-1984-9656

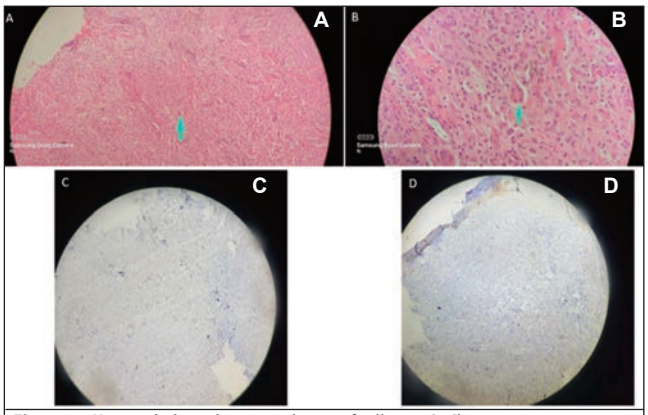

Figure-2: Histopathology showing splaying of collagen. B. Characteristic tuoton giant cell. C. Immunostaining showing CD1A negativity in dermis. D. Immunostaining showing S- 100 negativity.

cutaneous examination was unremarkable. Ocular examination by ophthalmologist was unremarkable.

Laboratory investigations, including complete blood count, liver and renal function test, were within normal range. Serum lipid profile and fasting blood sugar were normal as well. Ultrasound abdomen done to rule out hepatomegaly or splenomegaly was normal.

The papule was excised and sent for histopathological examination. It revealed dense infiltrate of small histiocytes in the dermis, Tuoton giant cell and necrobiosis, as seen in Figure 2B. The cells stained positive for S-100 and negative for CD1a, as seen in Figure 2C & 2D. Margin clearance was achieved histopathologically.

The papule was excised completely under local anaesthesia of 0.1% lignocaine with adrenaline using strict aseptic measures. The wound was closed with silk 3-0. The patient was counselled regarding the benign nature of the lesion. Stiches were removed after 1 week.

Discussion

Xanthogranuloma, a non-Langerhan cell histiocytosis clinically manifests as cutaneous lesions predominantly affecting the head and neck but less commonly the extremities.

In our case there was a single papulonodular lesion in front of the neck in accordance with majority of the cases in literature. However there have been cases of multiple cutaneous lesions as reported by Tian et all.[6]

Extracutaneous involvement only occurs in 4% children and 5-10% overall. Ferreira et all[7] reported absence of extracutaneous involvement and so did it show in our case.

Furthermore literature review has shown that histopathology has been highly useful to confirm the diagnosis. The common finding observed have been Tuoton giant cell and histiocytes with lipid laden macrophages.[8] These findings may be absent in early stages of the disease, making the diagnosis difficult. In our case, histopathology showed similar changes.

Vahabi-Amlashi et al reported little improvement with systemic retinoids[9] in purely cutaneous lesions. In our patient there was no systemic involvement so there was no rationale for systemic treatment. Complete surgical excision was done for diagnostic, therapeutic and aesthetic purpose.

Although recurrence has been documented in studies after excision, our patient had no recurrence until 6 months of follow-up.

Conclusion

This was a rare case of adult-onset xanthogranuloma that shows that this diagnosis should be considered when dealing with skin coloured nodules in adults.

Informed Consent: We have not disclosed patient's identity or personal information. Permission was taken before including his picture in the article.

Disclaimer: None.

Conflict of Interest: None.

Funding Sources: None.

References

1- Tan LC, Tan KB, Aw CW. Unusual presentation of adult xanthogranuloma: a case report. Singapore Med J 2014;55:e25-7. doi: 10.11622/smedj.2013207.

2- Achar A, Naskar B, Mondal PC, Pal M. Multiple generalized xanthogranuloma in adult: case report and treatment. Indian J Dermatol 2011;56:197-9. doi: 10.4103/0019-5154.80416.

3- Bastianpillai J, Haloob N, Panchappa SA, Marais J. The Rare Case of an Adult-Onset Xanthogranuloma of the Paranasal Sinuses: A Histological Dilemma. Case Rep Pathol 2020;2020:e2847821. doi: 10.1155/2020/2847821.

4- Ederle A, Kim KH, Gardner JM. Eruptive xanthogranuloma in a healthy adult male. J Cutan Pathol 2017;44:385-7. doi: 10.1111/cup.12877.

5- Asarch A, Thiele JJ, Ashby-Richardson H, Norden PS. Cutaneous disseminated xanthogranuloma in an adult: case report and review of the literature. Cutis 2009;83:243-9

6- Tian X, He S, Li R, Wang J. Multiple Adult Xanthogranuloma: A Rare Case Report with Characteristics. Asp Biomed Clin Case Rep 2021;4:88-92. doi: 10.36502/2021/ASJBCCR.6235

7- Ferreira BR, Cardoso JC, Reis JP, Tellechea O. Multiple adult-onset xanthogranuloma, an uncommon diagnosis. An Bras Dermatol 2017;92:294-5. doi: 10.1590/abd1806-4841.20176216.

8- Martínez-García M, Silvestre-Torner N, Aguilar-Martínez A, Burgos-Lázaro F. Multiple Xanthogranulomas in an Adult Patient with Myelodysplastic Syndrome. Case Rep Dermatol 2020; 2020:e8826715. doi: 10.1155/2020/8826715

9- Vahabi-Amlashi S, Hoseininezhad M, Tafazzoli Z. Juvenile Xanthogranuloma: Case Report and Literature Review. Int Med Case Rep J 2020;13:65-9. doi: 10.2147/IMCRJ.S240115.