

Asymptomatic solitary nodule in hard palate

AK Douieb,¹ MD, Bayoumi Eassa,² MD

¹Consultant, Department of Dermatology, HPLM, Larache, Morocco

²Department of Dermatology, Farwaniya Hospital, Kuwait

A- 31 year old Indian male patient presented with painless lesion in his oral cavity. The condition was first noted at 9th year of age. The lesion was very small and stationary for about 19 years and after that it had slowly enlarged one year ago. There was no history of trauma or a history of surgery during childhood. Also, there was no congenital anomaly at birth. This patient had a history of diabetes for 5 year and hypertension for 2 years. There was no past history of the condition. In addition, family history was negative of similar condition.

Oral examination revealed a circumscribed nodule, 9mm in diameter and located in the middle of hard palate. It was whitish to yellowish in color, On palpation, it was a firm nodule and not tender. The surface of the lesion was soft. In addition, the nodule was fixed and not mobile (Fig. 1). Examination of lymph nodes of head and neck showed no significant findings.



Fig. 1 whitish to yellowish colored nodule in the middle of Hard Palate.

What is the clinical diagnosis?

- Oral fibroma
- Adenoma of salivary gland
- Xanthoma
- Neuroma
- Schwannoma

A shave excisional biopsy was done under local anesthesia by a dental surgeon. Microscopic examination of the lesion revealed dome shaped lesion with a dense advential infiltrate of benign polygonal to spindle-shaped fibroblastic cells, mixed with numerous foamy histiocytes including multiple Touton cells with foamy, vacuolated cytoplasm (Fig. 2). There were also scattered lymphocytes. The immunohistochemical study demonstrated that the histiocytic cells were positive for CD68, but negative for CD1a, S-100 and Langerin.

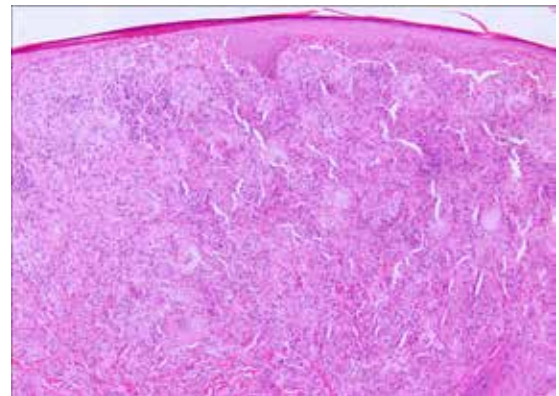


Fig. 2 A dense advential infiltrate formed of benign polygonal to spindle-shaped fibroblastic cells, mixed with numerous foamy histiocytes including multiple Touton cells with foamy cytoplasm in addition to lymphocytes.

Correspondence: Dr. AK Douieb, Consultant, Department of Dermatology, HPLM, Larache, Morocco, Email: douieb@menara.ma

DIAGNOSIS

Xanthogranuloma (XG)

DISCUSSION

Xanthogranuloma (XG) a member of the non-Langerhans cell group of histiocytic proliferative disorders and it is a relatively uncommon benign cutaneous fibrohistiocytic lesion.^{1,2} In spite of the head, neck and trunk are the most common sites for JXG, it can appear anywhere on the body, including the groin, scrotum, penis, clitoris, eyelid, toenail, palms, soles and lips. Extracutaneous involvement is usually restricted to the eye, specifically the iris, but may also occur in bone, (Erdheim-Chester disease), lung and liver.³ It is present at birth in 5% to 17% of the cases but it mainly arises within the first year of life.⁴ Our case of XG was localized in hard palate and this case can be considered as a rare site for XG.

Histiocytic diseases are currently identified by their component cells. In the right clinical context, lesional cells that are CD1a+/Langerin+/S100+ can be identified as Langerhans cell histiocytosis (LCH) without looking for ultrastructural Birbeck granules. A standard nonautomated avidin-biotin peroxidase complex method without antigen retrieval was used in the immunohistochemical analysis to explore the immunoreactivity to those antibodies. The non-LCH are a diverse group of disorders defined by the accumulation of histiocytes that do not meet phenotypic criteria for the diagnosis of Langerhans cells. Although some may have a hemophagocytic component, the definition excludes primary and secondary forms of hemophagocytic lymphohistiocytosis.⁵ XG is a benign cutaneous fibrohistiocytic lesion and it is a type of granulomatous process, at times accompanied by lipid deposits. Adamson,²

who first described this lesion in 1905, defined single or multiple cutaneous nodules in infancy as congenital xanthoma multiplex. The lesions were designated nevoxanthoendothelioma by Mc Donagh in 1912,⁶ who considered this disorder to be derived from endothelial cells. However, widespread recognition of an entity resembling XG occurred in 1954.⁷ A solitary cutaneous lesion is the most common presentation but it may occur as a soft tissue lesion with or without organ involvement.¹ JXG has been documented in many visceral locations such as lung, bone, testis, gastrointestinal tract, heart, eye and oral cavity.⁸⁻¹⁰ It can manifest as a multisystem disease.^{1,11} The lesions of XG are common in the young child. Median age of onset is two years, but lesions may be present at birth. Lesions vary in size, but children younger than six months of age tend to present with multiple lesions with predominance in the head and neck. The male preponderance is much higher (12:1) in young infants with multiple skin lesions.¹

Neurofibromatosis type 1 and juvenile chronic myelogenous leukemia may be associated with XG. Ocular JXG occurs in the very young child, with 92% of patients being younger than two years of age, and may occur without concomitant skin involvement.¹²⁻¹⁵ Eye involvement is usually but not always unilateral, and commonly presents with an asymptomatic iris tumour, a red eye with signs of uveitis, unilateral glaucoma, spontaneous hyphema or heterochromia iridis.¹⁶

Differential diagnosis of XG including xanthoma, neurolemmoma, molluscum contagiosum (pearly, dome-shaped papule with central umbilication), hemangioma and neurofibroma (firm lesion, associated café-au-lait spots).¹⁷

Microscopic examination from suspected case

is used to diagnose the presence of a non-LCH, but differentiation between the different subtypes is based mostly on immunohistochemistry and the clinical setting. Histological features of the non-LCH shows well circumscribed nodules with dense infiltrates of histiocytes. Those that involve the skin usually infiltrate the dermis. Giant multinucleate cells are variable in number and there is also a variable degree of predominantly perivascular and perilesional inflammatory cells. Touton giant cells (seen in 85% of cases of JXG, in a recent series),¹ are characterized by a wreath of nuclei around a homogeneous eosinophilic cytoplasmic centre, while the periphery shows prominent xanthomatization. Electron microscopy has revealed a variety of nonspecific organelles including dense bodies, worm-like bodies and popcorn bodies.¹⁶ The cells of histiocytes and giant cells are monocyte-macrophage in origin. They label strongly with macrophage markers such as CD 68 and HAM. On the other hand, S-100 protein immunoreactivity, which is a marker for the diagnosis of LCH, is typically absent.^{18,19}

In most of cases, XG is most often a self-limiting disease that often spontaneously regress.¹ Lesions may resolve completely or may leave a residual atrophic or hyperpigmented scar. The pathogenesis is unknown, and the initiating stimuli may be one of many infections or physical factors.^{13,14} Conservative management of these tumours has been advocated.¹² Despite the likelihood of spontaneous regression, it is often decided to excise the lesion(s) for esthetic or diagnostic reasons, as was the case for our patient. Excision of the lesion is an adequate treatment and recurrence is uncommon, although it has been documented.¹⁸⁻²⁰ Our patient was treated surgical excision without recurrence of 3 years follow up.

REFERENCES

1. Dehner LP. Juvenile xanthogranuloma in the first decades of life. A clinicopathologic study of 174 cases with cutaneous and extracutaneous manifestations. *Am J Surg Pathol.* 2003; 5:579-93.
2. Jung T, Emmert S, Günzl HJ, Neumann C, Rüniger TM. Congenital manifestations of juvenile xanthogranuloma (large nodular form) *Hautarzt.* 2000; 51:423-26. doi: 10.1007/s001050051145.
3. Cypel TKS, Zuker RM MD. Juvenile xanthogranuloma: case report and review of the literature. *Can J Plast Surg.* 2008; 16 (3):175-77. Autumn.
4. Hernandez-Martin A, Baselga E, Drolet BA, Esterly NB. Juvenile xanthogranuloma. *J Am Acad Dermatol.* 1997; 36:335-67. Chang MW. Update on juvenile xanthogranuloma: unusual cutaneous and systemic variants. *Semin Cutan Med Surg.* 1999; 18:195-205. doi: 10.1016/S1085-5629 (99)80017-0.
5. Curtis T, Wheeler DT, Cibis GW, Law SK, Rowsey JJ, Brown LL, Roy H. Juvenile xanthogranuloma. [<http://emedicine.medscape.com/article/1209681-overview>].
6. Sangüeza OP, Salmon JK, White CR Jr, Beckstead JH. Juvenile xanthogranuloma: a clinical, histopathologic and immunohistochemical study. *J. Cutan Pathol.* 1995; 22 (4):327-35. doi: 10.1111/j.1600-0560.1995.tb01415.
7. Liang S, Liu YH, Fang K. Juvenile xanthogranuloma with ocular involvement. *Pediatr Dermatol.* 2009; 26 (2):232-34. doi: 10.1111/j.1525-1470.2009.00893.
8. Karcioglu ZA, Mullaney PB. Diagnosis and management of iris juvenile xanthogranuloma. *J. Pediatr Ophthalmol Strabismus.* 1997; 34 (1):44-51.
9. Adamson HG. Society intelligence: the dermatological society of London. *Br J Dermatol.* 1905; 17:222.
10. Helwig EB, Hackney VC. Juvenile xanthogranuloma (nevooxanthoendothelioma) *Am J Pathol.* 1954; 30:625-26.
11. Püttgen KB. Juvenile xanthogranuloma (JXG) [<http://www.uptodate.com/contents/juvenile-xanthogranuloma>].
12. Sonoda T, Hashimoto H, Enjoji M. Juvenile xanthogranuloma. Clinicopathologic analysis and immunohistochemical study of 57 patients. *Cancer.* 1985; 56:2280-86. doi: 10.1002/1097.
13. Zelger B, Cerio R, Orchard G, Wilson-Jones E. Juvenile and adult xanthogranuloma. A histological and

- immunohistochemical comparison. *Am J Surg Pathol.* 1994; 18:126-35. doi: 10.1097/00000478-199402000-00002.
14. Tomaszewski MM, Lipton GP. Unusual expression of S-100 protein in histiocytic neoplasms. *J Cutan Pathol.* 1998; 25:129-35. doi: 10.1111/j.1600-0560.1998.tb01704.x.
 15. Wee SH, Kim HS, Chang SN, et al. Generalized eruptive histiocytoma: A pediatric case. *Pediatric Dermatol.* 2000; 17:453-55.
 16. Wee SH, Kim HS, Chang SN, et al. Generalized eruptive histiocytoma: A pediatric case. *Pediatric Dermatol.* 2000; 17:453-55.
 17. Kesavan TM, Sreedevi PK. Juvenile xanthogranuloma. *Indian Pediatr.* 2005; 42:950.
 18. Sonoda T, Hashimoto H, Enjoji M. Juvenile xanthogranuloma. Clinicopathologic analysis and immunohistochemical study of 57 patients. *Cancer.* 1985; 56:2280-86.
 19. Zelger B, Cerio R, Orchard G, Wilson-Jones E. Juvenile and adult xanthogranuloma. A histological and immunohistochemical comparison. *Am J Surg Pathol.* 1994; 18:126-35.
 20. Cohen BA, Hood A. Xanthogranuloma: Report on clinical and histologic findings in 64 patients. *Pediatr Dermatol.* 1989; 6:262-66.