# Intraoral juvenile xanthogranuloma

## A case report and literature review

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Juvenile xanthogranuloma rarely occurs in the oral cavity and has received little attention. A case of histologically documented juvenile xanthogranuloma of the oral cavity is described. This is the first intraoral case reported in the Oriental race and in the vestibule. Pertinent literature regarding intraoral lesions of this condition is also reviewed. **(ORAL SURG ORAL MED ORAL PATHOL ORAL RADIOL ENDOD 1996;81:450-3)**

Historically, juvenile xanthogranuloma (JXG) has been described as a benign, self-limited, regressing, fibrohistiocytic lesion of infancy and childhood. The condition was originally termed *congenital xanthoma multiplex* in 1905 by Adamson[1] who observed multiple cutaneous cephalic, nuchal, and truncal lesions in a 2½-year-old boy. The term *juvenile xanthogranuloma* was proposed by Helwig and Hackney[2] in 1954. Serum lipid studies are characteristically normal.[3-5] It is usually a cutaneous lesion most commonly located on the head, neck, and upper trunk, but it may involve extracutaneous sites, especially the periorbital tissues.[6] A few cases of JXG with lung,[7] testis,[2] or pericardium[8] involvement have been reported. Oral lesions of JXG are exceedingly uncommon if the number of cases reported in the literature is any indication of frequency of occurrence. To the best of our knowledge, only nine histologically documented intraoral cases have been previously reported in the English-language literature.[9-14] The purpose of this article is to present an additional histologically-documented example of this uncommon disorder originating in the vestibule, the first intraoral lesion described in this location. Pertinent literature regarding the previously published intraoral JXGs is also reviewed.

## CASE REPORT

A 14-year-old Chinese boy complained of a solitary, raised, nontender, soft, yellowish, smooth-surfaced lesion that had been present for more than 1 year. The lesion, which was located in the vestibule adjacent to the left mandibular cuspid, measured approximately 1 cm by 1 cm in diameter (Fig. 1). Radiographic examination showed no abnormalities. The clinical impression was a lipoma. No cutaneous lesions were present, and serum lipid levels were within normal limits. The lesion was excised and submitted for light and electron microscopic examination. The biopsy site healed without complications, and after about 5 years' follow-up, there has been no recurrence.

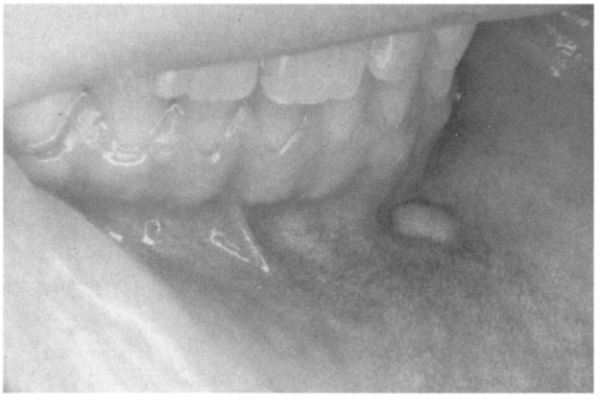

Fig. 1. Solitary, raised, yellowish, smooth-surfaced lesion on vestibule adjacent to left mandibular cuspid.

### Histopathologic findings

Hematoxylin-eosin–stained sections revealed unremarkable surface oral epithelium and underlying fibrous connective tissue that contained an infiltrate of histiocytes and lymphocytes within a fibroblastic stroma (Fig. 2). Only a few eosinophils were present. Touton giant cells with a wreath-like configuration of the nuclei were noted in the histiocytic infiltrate (Figs. 3 and 4). On electron microscopic examination, the Touton giant cells contained numerous irregularly shaped nuclei with cytoplasmic lipid. No Langerhans' (Birbeck) granules, which are characteristic of Langerhans' cell histiocytosis, could be found. Periodic acid-Schiff and methenamine silver stains did not reveal fungal organisms, and foreign material was not present. A histopathologic diagnosis of JXG was made.

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## DISCUSSION

The occurrence of JXG in the oral cavity is exceedingly uncommon and has received little attention. The mean age of the reported cases of intraoral JXG is 8 years (range, 9 months to 16 years) and although too few cases have accumulated to draw definite conclusions, a male to female ratio of 7:3 is noted. No visceral lesions have been found to occur simultaneously with the reported intraoral lesions. On the other hand, only one intraoral lesion has appeared simultaneously with cutaneous disorder.[11] The size of the lesion was described in eight cases[11-14] and ranged from 0.3 cm to 2 cm in greatest diameter. Usually the intraoral JXG was described as a symptomless, smooth-surfaced lesion, but an easily bleeding surface was reported in one case[11] and surfaces with ulceration were noted in three others.[12] Twenty percent of cutaneous lesions are present at birth but most develop between 6 and 24 months of age and affect both sexes equally.[2] However, only one previously reported case of intraoral JXG occurred from birth.[14]

Several significant clinical features of the present case may be noted. This is the first report of intraoral JXG occurring in the Oriental race. Seven previously reported intraoral cases,[10-13] excluding those of Kjaerheim and Stokke[9] and Patel et al.[14] who did not state the patients' race, have been found in Caucasians. Cohen et al.[12] found that intraoral lesions seemed to occur more frequently in the midline of the palate, gingiva, and lateral border of the tongue. The present case developed in the vestibule. The lesions of JXG may vary in color from reddish or reddish-brown in the early lesion to yellowish in older lesions.[15] Four previously published intraoral cases,[9-14] as well as the present lesion, were yellowish or yellowish-brown in color. These may represent more mature lesions as most of them had been present for more than 1 year at the time of biopsy. Clinically, the oral JXG may be misdiagnosed as abscess,[9] irritation due to foreign body,[10] pyogenic granuloma,[12] mucoepidermoid carcinoma,[12] and fibro-epithelial polyp.[14] In addition, the lesion may mimic lipoma, granular cell tumor, lymphoid aggregate, or verruciform xanthoma clinically.

JXG should be completely excised and the biopsy site examined for proper healing because up to 20% of cases have been reported to recur. No recurrence has been noted for the present case after a follow-up period of about 5 years.

The histologic and cytologic diversity in JXG have been discussed by Marrogi et al.[15] and Zelger.[16] Microscopically, JXG must be differentiated from Langerhans' cell histiocytosis. In the present case, only a few eosinophils were found but there were many Touton giant cells, which are typical for JXG and absent in Langerhans' cell histiocytosis.[17] Furthermore, no Langerhans' (Birbeck) granules could be found with electron microscopy. Therefore these pathologic findings favor the histologic diagnosis of JXG rather than Langerhans' cell histiocytosis.

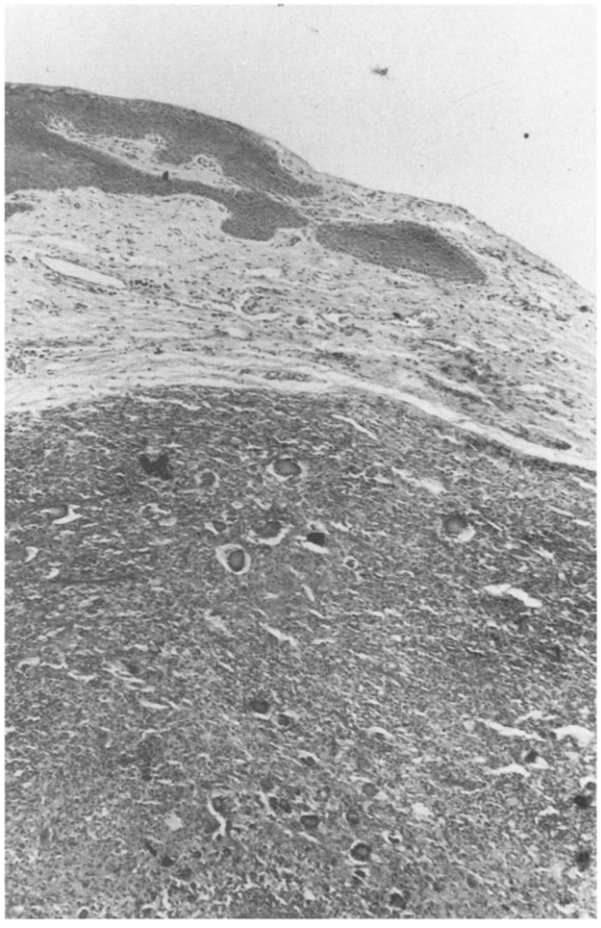

Fig. 2. Low-power view of JXG exhibits an infiltrate of histiocytes and lymphocytes with a fibroblastic stroma. Many Touton giant cells were present within the infiltrate of the histiocytes. (Hematoxylin-eosin stain; original magnification ×40.)

The histogenesis of JXG remains to be clarified. Whether it represents a true neoplasm or a reactive process is still not completely understood. JXG has been considered to perhaps represent a reactive virally-induced lesion associated with cytomegalovirus infection.[18] In addition, the involutional behavior of some JXGs might at least in some cases suggest the reactive hypothesis.[19]

In conclusion, this is the first reported oral JXG lesion to occur in the Oriental race and in the vestibule. It provides an additional histologically documented

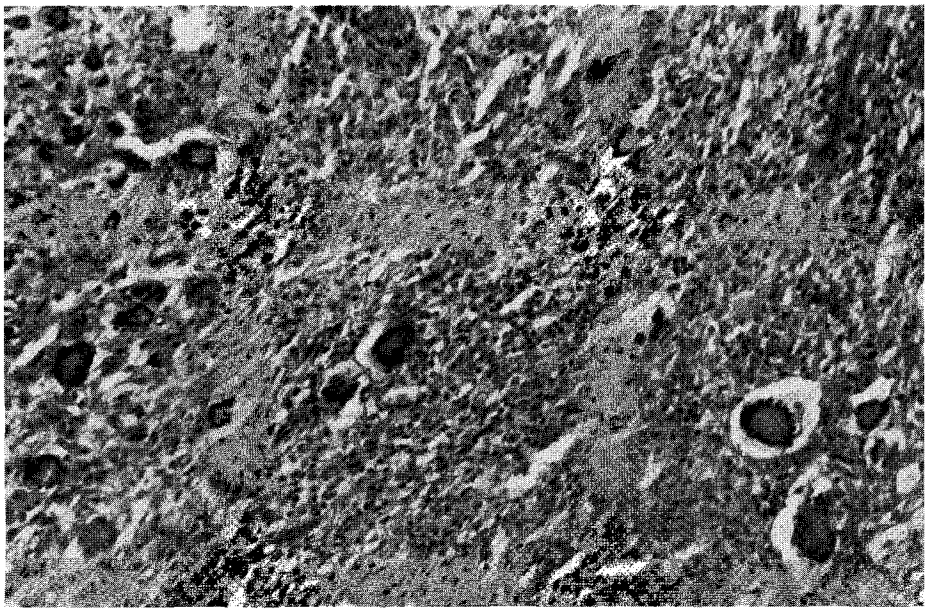

Fig. 3. Medium-power view demonstrates Touton giant cells located within the histiocytic and lymphocytic infiltrates. (Hematoxylin-eosin stain; original magnification ×100.)

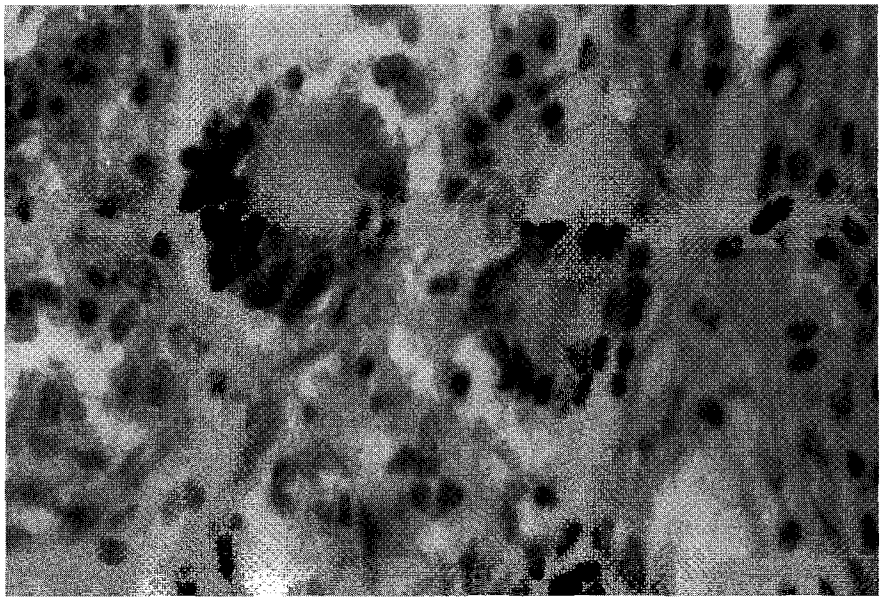

Fig. 4. High-power view of Touton giant cells with wreath-like configuration. (Hematoxylin-eosin stain; original magnification ×400.)

case of this uncommon benign disorder of the oral cavity.

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