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What is This?
Case Report

A Large Juvenile Xanthogranuloma within the Tongue

Fred M. Baik, MD1, Nicole K. Andeen, MD2, Stephen C. Schmechel, MD, PhD2, and Neal D. Futran, MD, DMD1

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Juvenile xanthogranuloma (JXG) is a benign, self-limiting disorder that typically involves the cutaneous regions of the head and neck. Infrequently, the oral cavity is involved, which presents unique diagnostic and treatment considerations. We present the first descriptive case report of a large, entirely submucosal JXG of the tongue. Withdrawal from review was granted by the Institutional Review Board of the University of Washington.

Case Report

A 13-year-old female was referred to our clinic with a chief complaint of tongue soreness. Three months prior to presentation, she noted mild pain in her tongue with swallowing. While her parents believed her speech to be “thicker” in character, she denied difficulty breathing and had been maintaining her weight. Her past medical history was unremarkable. Examination of the oral cavity demonstrated fullness of the tongue without mucosal abnormalities. An irregular, submucosal mass was palpable from ventral and dorsal aspects of the tongue. The base of tongue was clear, and her vocal cords were mobile. Magnetic resonance imaging (MRI) of the head and neck revealed a large, encapsulated midline tongue mass measuring 4.4 × 4.1 × 3.8 cm (Figure 1). The patient had normal thyroid tissue present.

We proceeded with operative biopsy of the mass. Histology demonstrated a monomorphic population of plump to spindled histiocytic cells with admixed eosinophils, clusters of Touton giant cells (Figure 2, arrows), and rare cells with intracytoplasmic microvesicular lipid; approximately 4 mitoses per 10 high-powered fields were present. Due to the symptomatic effects of the growing mass, complete excision of the mass via a sublingual approach was performed. The patient’s airway remained patent during her hospitalization, and she was discharged after tolerating a soft diet. At 6-month follow-up, the patient had normal tongue motion and no evidence of recurrence.

Discussion

Juvenile xanthogranuloma is a rare, benign histiocytic disorder of the non-Langerhans cell histiocytosis type. Other names attributed to this disease include nevoxanthoendothelioma and congenital xanthoma multiplex.

Classic, cutaneous JXG affects 1% to 2% of healthy infants and young children with a slight male preponderance.1 The head and neck region is involved in two-thirds of cases.2 These lesions are firm and solitary but can cluster together, are typically subcentimeter in size, and appear yellow, brown, and/or red in color. Juvenile xanthogranuloma is a clinical and histologic diagnosis; routine blood tests are not recommended. It appears to be nonhereditary. However, the finding of JXG in

Figure 1. T1 sagittal magnetic resonance imaging demonstrating a well-encapsulated midline tongue mass.

1Department of Otolaryngology–Head and Neck Surgery, University of Washington, Seattle, Washington, USA
2Department of Pathology, University of Washington, Seattle, Washington, USA

Corresponding Author:
Fred M. Baik, MD, Department of Otolaryngology–Head and Neck Surgery, University of Washington, 1959 NE Pacific St, Box 356515, Seattle, WA 98195, USA.
Email: fbaik@uw.edu
a child with neurofibromatosis is associated with a 20- to 32-fold increase in the development of juvenile chronic myelogenous leukemia.1

Rarely, JXG involves extracutaneous, or systemic, sites. The eye and testis are the more frequent sites affected, although JXG has been found to involve all major organs, the temporal bone, paranasal sinuses, salivary glands, oropharynx, and larynx.2 Juvenile xanthogranuloma of the oral cavity is exceedingly rare, and there are only 6 reported cases of JXG of the tongue.2-5 To our knowledge, this is the first descriptive case report of a large, entirely submucosal JXG of the tongue.

Histologically, JXG is characterized by a diffuse population of phagocytic cells with a great variability in the amount of admixed eosinophils, Touton giant cells (arrows), and rare cells with intracytoplasmic microvesicular lipid. Given the slightly older age of our patient and the location of disease, extranodal sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) could also be considered; however, the lack of alternating lymphoid aggregates and the presence of Touton giant cells and eosinophils made this much less likely.

Treatment for classic, cutaneous JXG is usually not required, since these lesions spontaneously involute with time. In contrast, spontaneous regression is not characteristic of extracutaneous JXG. For JXG of the tongue and oral cavity, excisional biopsy is recommended for functional and diagnostic reasons. In the case presented, we suspect the submucosal nature of the tumor permitted growth to large dimensions prior to detection.

Author Contributions
Fred M. Baik, data gathering, surgery, literature review, manuscript drafting; Nicole K. Andeen, pathology consultation, manuscript review and approval; Stephen C. Schmechel, pathology consultation, manuscript review and approval; Neal D. Futran, surgery, manuscript review, final approval.

Disclosures
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