Case Report and Literature Review on A Rare Tumor of The Maxilla: Myofibroma

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Case Report

An 11 year-old Hispanic boy presented to USC oral medicine clinic with a large 3.5 x 2.5 cm firm, reddish soft tissue mass distal and palatal to the right maxillary molar. Patient reported the duration of 2 months and recently soreness and bleeding from that area.
Panoramic radiograph (Figure 1) revealed a radiolucent lesion between the right maxillary first and second molars (#3 and #2) with distal displacement of the second and third molars (#2 and 1).

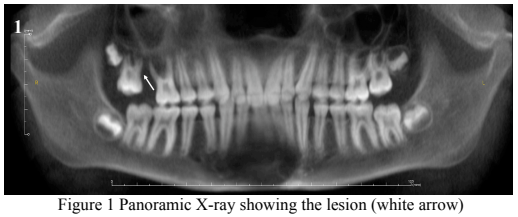


Figure 1 Panoramic X-ray showing the lesion (white arrow)

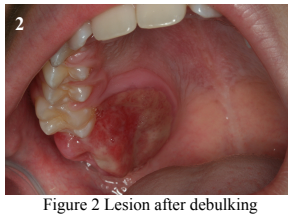


Figure 2 Lesion after debulking

Clinical diagnosis of pyogenic granuloma was made based on clinical features and a debulking biopsy was done by removing the palatal side of the mass (Figure 2). Biopsy specimen was sent to Oral Pathology Consultants at the Ohio State University for histopatholigical diagnosis. The lesion was diagnosed as cellular myofibroma.

Imaging Findings (CT scan)

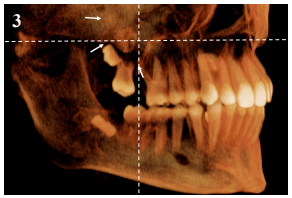


Figure 3 3D reconstruction image

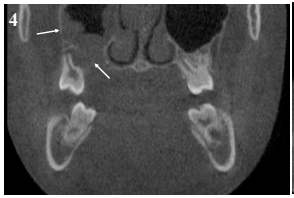


Figure 4 Coronal view of the lesion

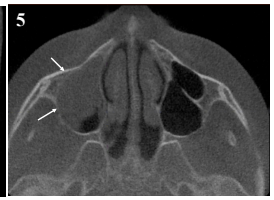
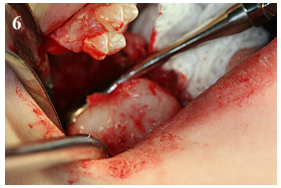
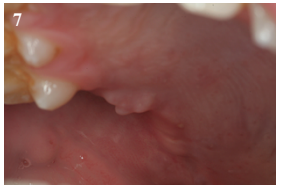


Figure 5 Axial view of the lesion

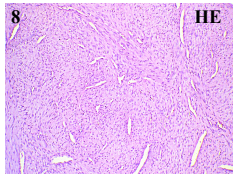
Intra-operative and post-operative findings

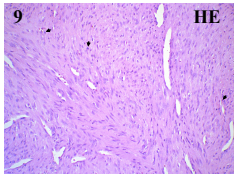


The lesion was attached to the root of #3 but was easily detachable form the surrounding tissues (Figure 6). The surgical area showed good healing 2 weeks after surgery (Figure 7).

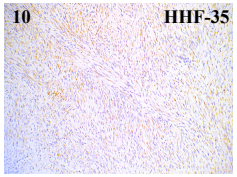


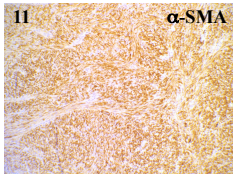
Histopathological findings





The lesion was composed of a uniformly cellular proliferation of spindle cells with elongated ovoid to cigar-shaped nuclei (Figure 8 & 9). The cells were arranged in loose, interlacing fascicles with hemangiopericytoma like areas (Figure 8). A significant degree of pleomorphism and mitoses were also observed (Figure 9, Arrows).





Various immunohistochemical markers were studied to rule out other benign and malignant spindle cell neoplasms. The lesional cells showed positive reaction to HHF-35 (Figure 10) and α-smooth muscle actin (Figure 11) but were negative for S-100, CD-34, Caldesmon, Desmin, CD68 and keratins (Figures not shown).

Literature Review on Myofibroblastic Tumors of Oral Cavity

Myofibroma is a **rare benign mesenchymal tumor of myofibroblastic cells**, primarily occurring in infants and young children. The tumor usually has an aggressive clinical course and can be over-treated as if it were a malignancy. Although most myofibromas are reported in the head and neck region, including oral subsites, myofibroma of the maxilla is very rare. Excellent reviews on oral myofibromas[1,2] and a large case series[3] recorded only **ten cases** of myofibroma in the palate or maxilla from a total of **158 cases** reported **from 1966 to 2005**.
The reported **clinical features** of gnathic myofibromas are: 1. Radiolucent tumor defect associated with developing or erupting teeth 2. Broad, shallow, lytic defects without distinct cortical margins when the tumor occur in the alveolus 3. Central tumor with or without soft tissue mass – partial or poorly developed sclerotic borders around radiolucent center 4. Cortically or periosteally based lesions producing an exophytic soft tissue shadow with a thin radiopaque shell overlying a non-corticated lytic defect in the bone surface.
Treatment: Most commonly accepted treatment is conservative surgery i.e total excision. Local resection of the jaw in large lesion and chemotherapy especially in myofibromatosis were also reported.
Prognosis: Solitary soft tissue and gnathic myofibroma has very good prognosis. Only 10% of the reported cases with difficult surgical access showed recurrence. 50% of patients with myofibromatosis died of other organ involvement.

References

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3) Foss RD, Ellis GL. Myofibromas and myofibromatosis of the oral region: A clinicopathologic analysis of 79 cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 200;89:57-65.