Radiographic Presentation of Fibromyxoma: An Uncommon Entity

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Fibromyxoma is a rare, locally destructive, benign odontogenic mesenchymal tumor which is found exclusively in the jaw region. The maxilla and anterior part of the mandible are rarely affected. It can usually lead to extensive bony destruction with extension into the surrounding structures. The lesion often grows without symptoms and presents as a painless swelling. Histologically and radiologically it is difficult to differentiate this tumor from other odontogenic tumors and may be mistaken occasionally for a malignant pathology. A similar situation happened with our case that was clinically misdiagnosed as malignant lesion initially, but after two repeated biopsies diagnosed as a fibromyxoma.

Keywords: Fibromyxoma, Malignant, Odontogenic, Radiographic, Tumors

INTRODUCTION

Myxomas represents 2.3-17.7% of all odontogenic tumors with fibromyxomas representing a small number of all myxomas.1 A fibromyxoma is an uncommon lesion, accounting for only 1-3% of all cysts and tumors of the jaws. Fibromyxomas are benign, slow-growing mesenchymal tumors that are aggressive, expansile, and locally destructive.2 They usually occur in the second to third decades of life and are rare in children or adults over 50 years of age.3,4

CASE REPORT

A 21-year-old male reported to the Department of Oral Medicine and Radiology with a chief complaint of non-tender swelling in the upper back region of left jaw. The patient first noticed the swelling 8 months back when it was of a smaller size that has gradually increased to present size (Figure 1).

Past dental history included extraction of 27 and 28 due to mobility and after which swelling increased in size. There was no history of discharge of pus, nasal discharge, and breathing difficulties. Past medical history was non-relevant.

Habit history included consumption of betel quid, areca nut, and tobacco chewing since 2 years.

Extra oral examination revealed diffuse extra oral swelling on the left side of the face. Bilateral submandibular lymph nodes were enlarged, tender, and mobile. There was no history of facial and mucosal numbness.

Intraoral examination showed large exophytic growth approximately 5 cm × 4 cm × 4 cm extending from the distal aspect of 25 up to left maxillary tuberosity. The lesion

Figure 1: Proliferative growth with erythematous areas with indentations of lower arch

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showed indentations on the surface from the teeth of the lower arch. On palpation, the lesion was hard in consistency and slightly tender. There was obliteration of the buccal vestibule with Grade I mobility of 25.

Radiological investigation using an intra-oral periapical showed discontinuity of floor of the maxillary sinus (Figure 2). Panoramic radiograph showed the destruction of the floor and posterior wall of maxillary sinus with a faint demarcation of the pterygomaxillary fissure (Figure 3).

Computed tomography imaging of the maxilla showed a well-defined, expansile hypodense lesion arising from the alveolar arch of the left maxilla and extending into the floor of the orbit, maxillary sinus and nasal cavity on the left side. The lesion was seen to erode the anterior, medial and posterolateral wall of the maxillary sinus, hard palate and floor of orbit on the left side (Figures 4 and 5).

Routine hematological and serological investigations of calcium, phosphorus, alkaline phosphatase were found to be normal.

Above features suggested a provisional diagnosis of an aggressive odontogenic tumor. Following this, an incisional biopsy was performed which revealed stratified squamous epithelium with the fibrous connective tissue with mixed inflammatory cell infiltrate, with a diagnosis of fibrous epulis.

However, after excision of the lesion, the histopathological examination showed mucoid stroma that contained spindle or angular cells with abundant collagen fibers which were suggestive of fibromyxoma.

**DISCUSSION**

According to Dutz and Stout, the term myxoma was first used by Virchow in 1863, but the term fibromyxoma was described by Marcove et al. in 1964 who reported extragnathic locations of fibromyxoma. Fibromyxoma has a higher fibrous/myxoid tissue ratio than myxoma and is classified as a specific type of myxoma. Due to the inconsistency in reported cases of fibromyxoma, most of them are largely classified under the term “myxoma,” making the review of the literature difficult.  

Abiose et al. reported that fibromyxomas constituted 20% of tumors of dental origin and 3.73% of all benign and malignant oral tumors which is second in incidence to ameloblastoma. Most of them arise in the second and third decades of life.

**Clinical Feature**

Myxomas/fibromyxomas are usually located intraorally most often in the posterior regions of the mandible (angle

**Figure 2:** Intra-oral periapical with loss of bony trabeculae and outline of maxillary sinus is not traceable

**Figure 3:** Orthopantomogram with faint outline of palate on the left side and outline of maxillary sinus, zygomatic bone, and pterygomaxillary fissure is not traceable

**Figure 4:** (a and b) Computed tomography scan with well-defined radiolucent lesion on the left side of maxilla extending from anteroposteriorly from anterior wall of maxilla up to posterolateral wall of maxillary sinus The lesion has caused erosion of anterior, medial and posterolateral wall of maxillary sinus

**Figure 5:** (a-c) Computed tomography scan with contrast showing hypodense well-defined expansile lesion extending into the floor of the orbit, maxillary sinus and nasal cavity of left side
and ramus) and rarely extraorally. In our case, the lesion was located in posterior maxilla which is not a common location for this pathology.

In their review of fibromyxomas, Farman et al. stated that ratio of mandibular:Maxillary cases were 2:3 and that fibromyxomas were more common in the premolar and molar regions.

Myxomas are mostly asymptomatic, even though some presents with progressive pain in lesions of the maxilla and the maxillary sinus, with subsequent neurologic manifestations. In mandible it produces extensive bone destruction with invasion into the surrounding structures and has a relatively high recurrence rate. Maxilla is less frequently involved by fibromyxoma but behaves more aggressively than that of the mandible as it spreads through the maxillary sinus, as presented in our case.

Radiographical Features
Radiographically, the majority of cases present as multilocular or unilocular radiolucency with or without scalloped borders, and rarely cases can be mistaken for a malignant neoplasm when present with a diffuse and mottled appearance.

The radiological study reveals different appearances, such as “honeycomb,” “soap bubble,” and “tennis racket.” The lesion can be well-defined or diffuse, unilocular or multilocular. Root resorption and displacement may be present. Our case showed unilocular lesion with sclerotic margins.

Pathogenesis
The origin of the tumor is controversial. Since it bears a resemblance to the stellate reticulum, is usually associated with unerupted or missing tooth, and due to occasional presence of odontogenic epithelium some believe it to be of odontogenic ectomesenchymal origin.

Recent studies advocate that myxomas/fibromyxomas arise from the mesenchymatous tissue of the dental follicle, thus being described as odontogenic with fibroblasts playing the major role in cell dispersal. Above explanation fails to explain soft tissue myxomas. They possibly arise from the gingiva and the periodontal ligament.

Some researchers consider it to be an odontogenic fibroma which has undergone myxomatous changes, suggesting that the connective tissue cells are actively secreting intercellular myxoid substance in abundance containing chondroitin sulfate, hyaluronic acid, and variable amounts of collagen fibers. Goldblatt studied myxomas ultrastructurally and concluded that cells in myxoma show many individualities of fibroblasts of the odontogenic apparatus; thus, origin from non-odontogenic mesenchyme cannot be ruled out on the basis of present ultra-structural findings.

Histopathological Features
Myxoma/fibromyxoma shows hypcellularity, stellate spindle-shaped cells within a loose myxoid extracellular matrix with cells presenting with thin, long cytoplasmic extensions that give immature mesenchyme like characteristics to the tissue. The fibromyxoid lesion may show loci of calcification/ossification with a higher amount of collagen fibers and vessels in contrast to a typical myxoma. Actin fibers positive cells are present which suggest that myofibroblasts might play a critical role in cell proliferation in support with the islands of odontogenic epithelium cells and mast cells.

Treatment and Prognosis
The lesion was surgically excised under local anesthesia. Complete resection of the lesion is strongly related to the avoidance of recurrence. The patient should be kept under follow-up for at least 2 years as there is a higher rate of recurrence during this period.

Myxomas/fibromyxomas show a recurrence rate between 25% and 43% which is strongly attributed to the nature of the pathology and presentation of the lesion lacking a sheath, thus making the complete removal of the lesion challenging.

Follow-up
First follow-up was done 8 days after the surgery (Figure 6). It’s been 9 months after the surgery, and there is no evidence of any recurrence.

CONCLUSION
Fibromyxoma of the maxilla is an uncommon clinical entity, but still it is important to keep it in differential diagnosis.
along with other commonly occurring lesions because of its infiltrative and aggressive nature with a high recurrence rate.

REFERENCES


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