Peripheral Odontogenic Fibromyxoma: A Report of a Unique Case
Shiladitya Sil

Abstract
Odontogenic myxoma is a benign, locally aggressive neoplasm that is known to have a high recurrence rate. Fibromyxoma is a variant of myxoma with a higher fibrous–myxoid tissue ratio. The peripheral type of odontogenic fibromyxoma still remains a diagnostic challenge, as very few cases are reported in the literature. One such case is reported here. A 54-year-old male patient reported to our OPD with a diffuse, slowly enlarging growth in the lower right side of the jaw without crossing the midline. Orthopantomogram was done that revealed generalized bone loss with cratering at the lesion site. Incisional biopsy was done along with immune histochemical evaluation for diagnosis confirmation. Surgical excision was done. No recurrence has been reported till now. These lesions clinically mimic any benign peripheral neoplasm. Hence, precise diagnosis is imperative before initiation of management protocols. Wide margin excision and rigid follow-up are recommended to minimize the chance of recurrence.

Keywords: Lichen planus of jaw, Odontogenic fibromyxoma, Odontogenic myxoma, Peripheral odontogenic fibromyxoma.

Introduction
Rudolph Virchow first used the term “myxoma” in 1863.1 The term fibromyxoma was coined by Marcove in 1964 to describe the gnathic variant of odontomes.2

The World Health Organization (WHO) has defined odontogenic myxoma (OM) as a locally invasive neoplasm consisting of rounded and angular cells lying in an abundant mucoid stroma.3 WHO classification of benign odontogenic tumors grouped odontogenic fibromyxoma (OFM) as benign tumors of ecto-mesenchymal origin with or without odontogenic epithelium.4

Myxomas are classified as central and peripheral variants. While peripheral lesions are comparatively less aggressive and encapsulated, the central lesions are generally nonencapsulated tumors with infiltrative capacity into the adjacent medullary bone.5

The mean age of diagnosis of OFM is 30 years. The age range of manifestation is between 1 and 73 years, and rarely OMs have been reported in children below 10 years of age. OFMs have a marginal female predominance with a male to female ratio of 1.5.6

The central lesions are twice as more common in the mandible than compared to maxilla. The peripheral lesions are seen in tongue, nose, cheek, neck muscles, larynx, pharynx, and parotid gland.6

Herein we report a rare case of peripheral odontogenic fibrous myxoma in an adult male patient.

Case Description
A 54-year-old male reported to the OPD with a chief complaint of a growth in the lower right side of the jaw since last 6 months. The lesion was initially small in size, sudden in onset, and with no preceding history of trauma. No relevant family history, medical history, or habit history was elicited. He did not complain of dysphagia or restriction in tongue movement. On inspection, gross facial asymmetry was appreciated due to a diffuse solitary growth measuring 3 × 2 cm involving the right side of the jaw, causing the obliteration of right buccal vestibule, and extending into the floor of the mouth but not crossing the midline. The growth extended anteroposteriorly from 41 to 46 regions. The lesion was associated with an irregular surface without evidence of any discharge or superficial ulceration. Clinically, 43 was involved in the lesion and no other teeth could be appreciated (Fig. 1). On palpation, the...
growth was firm to hard in consistency, nontender, and mobility of the associated teeth could be appreciated. There was no difficulty in occlusion. Generalized tooth mobility was appreciated with poor oral hygiene. A single lymph node was palpable in the right submandibular region which was slightly enlarged and tender.

Orthopantomogram (OPG) revealed generalized horizontal bone loss in the lower jaw with practically no bony structures surrounding the teeth. The right body of the mandible revealed mild cratering at the site of the lesion with 43 and 44. Root resorption was appreciated in 44 (Fig. 2).

Differential diagnosis of ameloblastic fibroma, peripheral ossifying fibroma, peripheral giant cell granuloma, and odontogenic myxoma was made.

Incisional biopsy was planned after obtaining the patient’s consent. Hemotoxylin and eosin–stained sections were examined under 40× magnifications that revealed multiple oval-or spindle-shaped cells and stellate cells arranged haphazardly in a matrix composed of abundant myxoid tissues and few collagen fibrils (Fig. 3). To further confirm the diagnosis of myxoma, immunohistochemical tests were advised for the detection of S-100, smooth muscle antigens, and Vimentin (Fig. 4). All were positive in our case confirming a final diagnosis of peripheral OFM.

Wide margin excision was performed under general anesthesia, and the patient is under follow-up without any evidence of recurrence (Fig. 5).

Discussion

OFM is classified as a specific type of myxoma with a higher fibrous–myxoid tissue ratio than myxoma. Some authors report...
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that fibromyxomas tend to have more prominent collagen fibers than myxoma. While OFM is a relatively rare neoplasm, it accounts for only 3–8% of all odontogenic tumors of the jaw, whereas OM has higher incidence of 2.3–17.7%. As WHO uses the terms myxoma, OM, and OFM interchangeably, there is a gross underreporting of OFM.8

The pathogenesis of OFM remains a matter of considerable debate. It is supposed to be of tooth origin and derived from the mesenchymal portion of tooth germ—most likely of the dental papilla.9 It is not clearly understood what exactly triggers the dental papilla—trauma, systemic diseases may be considered,10 but our case report had no contributing history.

Most of the OFM manifests in the tooth-bearing area, and rarely the lesions can be seen in the condyle and gingiva. The ectopic migration of odontogenic tissues may be responsible for the development of OFM in these sites. Raubenheimer and Noffke reported two cases of peripheral OFM but suggested that these entities were rather rare, and misdiagnosis along with underreporting may be a probable explanation for the same.10

Clinically OFM presents as a slow growing locally aggressive lesion, associated with gross facial asymmetry at an advanced stage. Pain and paresthesia is common in the soft tissue lesions. Pathological displacement of tooth and tooth mobility may be seen, but root resorption is rare. Cortical expansion is usually present at an advanced stage with areas of decortication. OMs are locally aggressive benign lesions that do not exhibit metastasis.11 Similar clinical manifestations were evident in our case report.

WHO classified OM as multiple radiolucent areas of varying size, separated by straight or curved bony septa with poorly defined border.3

The radiographic presentation of OM of the jaw varies from unilocular radiolucency without trabeculations to multilocular mixed lesion with multiple trabeculation that helps in distinguishing OM from malignant tumors arising within the jaw bones because the latter is an aggressive lesion associated with severe bone destruction and is usually devoid of bony septa.12,13

The internal trabecular pattern is characterized by angular or straight bony septa forming square or triangular compartments resembling honeycomb, soap bubble, or tennis racquet. This radiographic feature has been considered the most pathognomic for OM.13,14 Eversole described the internal configuration of the bony septa as lichen planus of the jaw bone.15

Langland and Langlais had described the computed tomographic (CT) findings of OM in three stages-

- Osteolytic expansile lesions with mild enhancement of the solid portion of the mass in the myxoma of the mandible.
- Bony expansion and thinning of cortical plates with strong enhancement of the mass lesion in the anterior maxilla.
- A soft tissue mass with bone destruction and thinning and strands of fine lace-like density representing ossifications in the maxillary sinus.15

Magnetic resonance imaging (MRI) reveals a well-defined, well-enhanced lesion with homogeneous signal intensity on every pulse sequence. OM shows intermediate signal intensity on the T1- and T2-weighted images.16 Unfortunately, CT and MRI were not performed in our case report.

Peripheral lesion of OFM is difficult to diagnose clinically, and histological evaluation is imperative for confirmation. Depending on the site and age, differential diagnosis of peripheral ameloblastoma, peripheral ossifying fibroma, and peripheral giant cell granuloma can be considered.

The central lesions provide a better scope for clinical diagnosis based on radiographic findings. When unilocular and without trabeculae, the tumor closely resembles periapical, lateral, periodontal, and traumatic bone cysts. When multilocular, OM must be distinguished from ameloblastoma and odontogenic keratocyst.17

OM tumor cells are mesenchymal in origin and express S-100, vimentin, and muscle-specific actin. Both were positive in our case. The matrix exhibits different proteins, mostly type-I and type IV, collagen, fibronectin, and proteoglycans.18

In 1976, Goldblat identified two basic types of tumor cells—secretory and non-secretory. The secretory cells resembled fibroblasts and were considered the principal cells contributing to the pathogenesis of OM.19

Simon et al. suggested that radical resection with a margin of 1.5–2 cm of surrounding tissue is the treatment of choice.20 OFMs have a high recurrence rate following excision, about 25–43%. The absence of a capsule and the locally infiltrative growth pattern contributes to the high recurrence rate. The literature suggests that OMs usually recur within the first 2 years following surgical removal. However, recurrence has also been reported over 30 years after the initial surgery.21 Surprisingly, the frequency of recurrence of the OFM of the jaws is higher than that of any other bone for which the jaw lesions have a poorer prognosis.22 Post excision, we had kept the patient under rigid follow-up with no evidence of recurrence.

**Conclusion**

OFMs are locally aggressive benign neoplasms that are known to cause “silent” destruction of the surrounding structures. As OFMs are mostly asymptomatic, their detection depends on secondary manifestations like generalized tooth mobility in an entire quadrant or cortical expansion or facial disfigurement. Histopathological and immune-histochemical diagnosis is mandatory for confirmation of Myxoma. Wide margin excision is the preferred treatment of choice, as the actual margin of the lesion is wider than the radiographic margin.
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