Case Report

An Interesting Case Report of Odontogenic Myxoma

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Abstract

Odontogenic myxoma is an uncommon benign tumor that affects the mandible, among individuals with a mean age of 25 -35 years and a predilection for the female gender. We describe a case of central odontogenic myxoma in a 10-year-old patient who presented with a swelling on the right side of the mandible. We have emphasized the importance of radiological examination that demonstrates its classic 'tennis racket" pattern along with clinical and histopathological comparison in arriving at a diagnosis. A review of current existing updated literature has been done to differentiate with other lesions that show overlapping characteristics. Since odontogenic myxoma has a high rate of recurrence, the treatment depends upon the age of the patient and the size of the lesion. It is commonly treated by curettage and resection. A regular follow-up is pertinent to rule out recurrence.

Keywords: *Odontogenic Tumors, Odontogenic Myxoma, Bony Lesions, Multilocular Radiolucencies*

Introduction

Odontogenic myxoma is a locally aggressive benign neoplasm that affects the bones of the jaws. It was first described in 1947, by Goldman and Thoma. **[**1**]** Regezi and colleagues have reported that odontogenic myxomas account for 3-6% of all odontogenic tumors. **[**2**]** Eversole described it as a 'lichen planus of the jaw bone" due to its internal configuration. **[**3**]**

Its origin is believed to be from odontogenic mesenchymal elements of the dental papilla. Histologically, it presents as loosely arranged rounded cells in the mucoid stroma. **[**3**]** The lesion is rare in children younger than 10 years, with the mean age being 25 -35 years and a higher prevalence among woman as compared to men (3:2) The lesions usually demonstrate as a painless swelling which shows buccal and lingual cortical plate expansion when it occurs in the mandible. The lesions are commonly treated by curettage and resection. **[**3**]**

The radiological appearance of this lesion is a tendency to occur in tooth-bearing areas, molar region followed by the premolar area. Barros and colleagues (1969) described its radiologic appearance in two stages: the first stage is an osteoporotic appearance, with the medullary spaces divided by thin septa of the bone, these septa become thinner and elongated as the tumor infiltrates locally. The classic radiographic appearance of this lesion is a multilocular radiolucency with bony septa being straight, thin, elongated, and lacy. Other authors have recognized the angular locules resembling strings of a tennis racket. The second stage is a destructive phase, with significant expansion and perforation of the surrounding area. Root resorption and tooth displacement may occur. **[**2**]**

World Health Organization (WHO) classified odontogenic myxomas as benign mesenchymal odontogenic tumors. There are central and peripheral variants with the central being aggressive and non-encapsulated. **[**4**]** When dense collagenous stroma is evident the term-fibro myxoma/myxofibroma can be used interchangeably. **[**4**]** Due to this dual pattern, the odontogenic myxoma is believed to be a continuum of odontogenic fibroma. **[**5**]**

The present report highlights a case of central odontogenic myxoma in a 10-year-old patient who presented with a swelling on the right side of the mandible.

Case Description

A 10-year-old patient reported to the clinic with a complaint of swelling on the lower right back tooth region for 1 week. The patient's parent gives a history of the appearance of swelling a week ago before which the patient was normal. The patient does not give any history of trauma, hot fomentation, or

application of topical medicine. The swelling was initially small in size and has gradually progressed to the current size. The swelling is not associated with any pain or discharge. No other associated symptoms were reported. The patient's past medical/dental history, drug history, and family history were non-contributory. On clinical examination revealed facial asymmetry: the swelling measured 2 x 3cm, extending from the right corner of the mouth to 2cm away from it and superoinferiorly from lower lip to 3cm inferior to it. **[**Figure 1**]** On palpation, the lesion was non-tender, firm to hard with smooth margins. Buccal cortical plate expansion was evident with no local rise in temperature.

Figure 1: Evidence of facial asymmetry due to presence of swelling on right lower third region of the face (As indicated by arrow)

On soft tissue examination, vestibular obliteration was present in #83, #84 region. **[**Figure 2**]** On hard tissue examination, deep dental caries was present in relation to 85, with dental caries in relation to #75 and #84. A provisional diagnosis of dentigerous cyst secondary to impacted #44 was made. A differential diagnosis of ameloblastoma, odontogenic keratocyst, and ameloblastic fibroma was considered. The patient was initially subjected to radiographic investigations and routine hematological investigations. The orthopantomography and intraoral periapical radiograph showed a well-defined multilocular radiolucency was present at the periapex of #84, #85 extending superoinferiorly from alveolar crest up to lower border of the mandible and mediolaterally from the distal aspect of #42 to mesial aspect of #46. Diffuse radiolucency present in the occlusal aspect of the crown in relation to #85 and disto-occlusal aspect of #84 involving enamel, dentin, and pulp suggestive of dental caries. Tooth buds of #45, #44, #43 were inferiorly displaced and #42 was mesially displaced. Root resorption was present in relation to #84, #85, and #83. Straight septations at right angles to each other were present (tennis racket appearance). The lower border of the mandible showed thinning of the cortex. **[**Figure 3,4**]**

Figure 2: Intraoral examination shows the evidence of buccal cortical expansion in the right lower vestibule (As indicated by arrow)

Figure 3: Intraoral periapical radiograph of the region shows characteristic criss-crossing septae at right angles (tennis racket pattern)

Figure 4: Orthopantomogram. Shows a multilocular, radiolucent lesion with fine septae in the right body of mandible

To ascertain the clear extent of the lesion and its effect on the surrounding structures, a 3D imaging modality of CBCT was advised. CBCT revealed a solitary well-defined radiolucency in the right mandible measuring 25.18 mm x 44.47 mm, the lesion extends from the distal aspect of 41 to the mesial aspect of 46. The internal structure appears completely radiolucent with multiple septae giving a multilocular appearance. The mandibular canal cannot be traced medially to 46. The axial section reveals expansion of the buccal and lingual cortical plate with thinning of the buccal cortical plate, lesion measures 20.38 mm buccolingually in the right premolar region. The coronal section revealed multiple thin wispy septae which are straight and crisscrossed. **[**Figure 5a, 5b, 5c**]** A radiographic diagnosis of odontogenic myxoma in the right mandible was given along with a differential diagnosis of ameloblastoma.

Figure 5 (A, B) : CBCT view (Coronal Section); CBCT view (Axial Section)

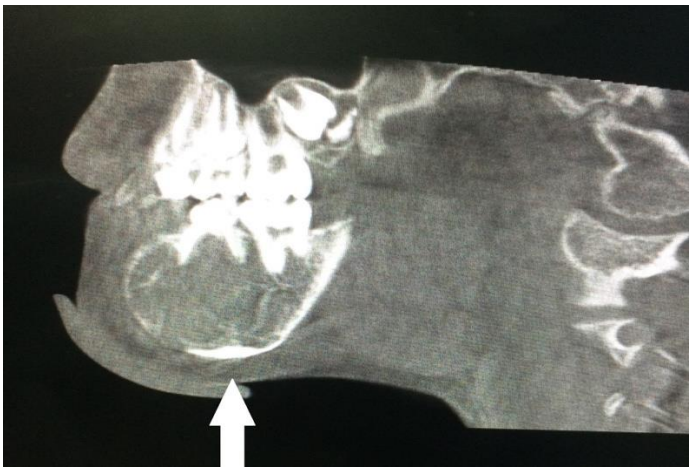

Figure 5C: CBCT view (Coronal Section)

All hematological investigations were found to be within normal limits. Incisional biopsy was performed and the gross appearance for the specimen was cream-white in color, soft to firm inconsistency. Upon staining cut section with H & E, it revealed loosely arranged, spindle-shaped stellate cells with myxoid stroma, and the stroma is not representing odontogenic mesenchyme. The lesion is interspersed with fine capillaries. **[**Figure 6a, 6b**]** The findings were suggestive of a diagnosis of odontogenic myxoma. En bloc resection was done for the removal of the tumor. The patient's 6 months follow-up clinical examination was uneventful with no evidence of recurrence.

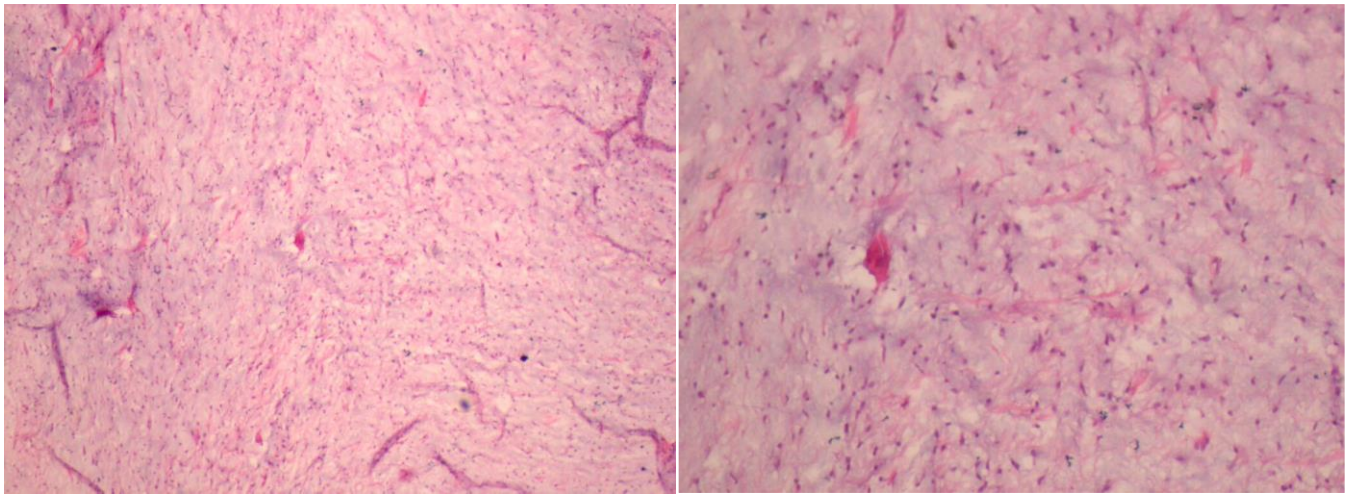

Figure 6A: Spindle- or stellate-shaped mesenchymal cells within loose myxoid stroma. Odontogenic epithelial islands. (As indicated by arrow) (H&E, ×10) **6B:** Spindle- or stellate-shaped mesenchymal cells within loose myxoid stroma. (H&E, ×40)

Discussion

Odontogenic myxomas, also termed odontogenic fibro myoma or myxofibroma, are rare benign tumors. They occur mainly in the hard, bony tissues of the face and a small minority occur in paranasal sinuses and soft tissues. They are locally invasive. **[**6, 7, 8**]**

Since the myxomas of the bones were found almost exclusively in the jawbones, it led to studies that presumed the origin is the mesenchymal portion of the tooth germ **[**8**]**. The incidence of odontogenic myxoma ranges from 0.5% to 19% among the lesions of the jaw. **[**9, 10**]** Regezi reported an incidence or 3% of 706 odontogenic tumors. **[**2**]** The rarity of this lesion must be a contributing factor to this wide variation in incidence.

According to Simon et al, the peak incidence of its occurrence is in the third decade of life with a marked bias towards females as compared to males 1:2. **[**9**]** Whereas Shafer et al, stated an absence of sex predilection. We have used the term "unusual" to describe the present report of the occurrence of odontogenic myxoma in a 10-year-old boy because these lesions have mostly been recorded in adult women. **[**8**]**

Odontogenic myxomas are more common in the mandible **[**7**]** and are typically associated with a tooth such as a premolar or molar. **[**11**]** According to Speight, it is most often found in the mandibular premolar area **[**6**]**. Despite being of benign nature, they become locally invasive by breaking through the borders and infiltrating the surrounding normal bone. **[**12**]** The expression of matrix metalloproteinase 2 and 9 which degrade the extracellular matrix (ECM) are responsible for the invasive nature. **[**13**]**

Clinically they appear as usually a slow-growing mass with symptoms appearing later, primarily because of the mass effect. Pain, paraesthesia, ulceration, and tooth mobility are some of the symptoms associated with it, although none of these symptoms were found in the present case. A few cases of myxomas crossing the midline were usually found in the mandible. **[**10**]** In the present case, the lesion was present in the region of the body of the mandible with displaced unerupted teeth.

On gross examination, odontogenic myxoma appears as greyish white, with a gelatinous cut surface. The tumor may have a true capsule or be encapsulated. In the present case, the capsule was absent. **[**11**]**

Histologically, the tumor demonstrates a mucoid-rich ECM, with scattered cells, connective tissue fibers, bony trabeculae, irregular calcifications, scant blood vessels, and sparse capillaries. Stellate or spindle-shaped cells with small hyperchromatic muclei are present. **[**12**]** The lesions tend to imitate dental pulp microscopically. **[**2**]** Our case showed all of these classical features.

The radiographic description of this lesion has varied between; always radiolucent, usually radiolucent, or mixed radiolucent-radiopaque. **[**14**]** The margins are classified as corticated or non-corticated. **[**15**]** It can appear as a unilocular or multilocular entity on radiographs with the multiloculated variant larger than the unilocular one. **[**14, 16**]** Other varieties have also been described as mottled, soap-bubble **[**17**]**, tennis racquet **[**15**]** or honeycombed **[**8**]**. The current case showed a multilocular appearance with a tennis racket pattern. Zhang et al. classified the radiological appearance of odontogenic myxoma into six groups. **[**18**]** The tumor tends to displace adjacent teeth, but root resorption has been rarely reported. **[**8, 12**]** In our case, the radiological appearance was of a multilocular, corticated type with tennis racket septation. Evidence of both tooth displacement and resorption was seen.

Arriving at an accurate diagnosis through routine radiographs alone is a difficult feat as many features of odontogenic myxomas tend to overlap with other tumors. Some tumors that can be suggested based on radiological appearance include odontogenic keratocyst, radicular cyst, ameloblastoma, central giant cell granuloma, aneurysmal bone cyst, intraosseous haemangioma, etc. **[**8, 19, 20**]** Some differentiating features have been tabulated in Table 1. Pertaining to the multilocular appearance in the current case, ameloblastoma was considered a differential diagnosis. A final diagnosis can only be arrived at on the basis of clinical features, radiographic presentation, and histopathology. CT, CBCT, MRI, and immunohistochemistry can aid in a proper diagnosis. **[**21**]**

Table 1: Differential diagnosis of odontogenic myxomas **[**8, 19, and 20**]**

		Odontogenic Myxoma	**Odontogenic Keratocyst**	**Amelobl astoma**	**Ameloblastic Fibroma**	**Aneurysmal Bone Cyst**
Age group		3rd Decade	2nd – 4th decade	3rd – 4th decade	1st-2nd decade	1st-2nd decade
Site		Mandible > Maxilla	Posterior mandible	Mandible Rarely maxilla	Mandible > Maxilla	Mandible Rarely maxilla
Sex		Female > Male	Slight male predilection	Not significan t	Male>Female	Not significant
Radiologi cal Appearan ce	**Margin s**	Ill defined	Smooth, well defined, mediolateral expansion	Smooth, scalloped , corticate d	Well defined, sclerotic margin	Smooth, well defined, Buccal and lingual cortical expansion
	Locula tion	May or may not be present	Multilocular	Uni/Mult ilocular	Uni/Multilocu lar	Uni/Multilocular
	Interna l Struct ure	Trabaculae/S eptae rarely seen. If present, very fine.	Not described	Trabacul ae/Septa e present	Trabaculae present	Rare

	Effect on surrounding Structures	Root resorption: Absent Tooth Displacement: Common	Rare	Root resorption: Common Tooth Displacement: Common	Associated with impacted teeth	Rare
Histopathology		Collagen/reticulin fibres with single hyperchromatic nuclei. Stellate shaped cells present interspersed with fine capillaries	Cystic cavity with cheesy material. Thin, fibrous capsule, lined by stratified squamous epithelium. Palisading basal layer with satellite cysts	Cystic changes and squamous metaplasia seen with presence of varying amount of fibres	Spindle or angular cells mimicking dental papilla with fibrous connective tissue	Blood filled cavities with multinucleated giant cells
Recurrence		More than 25%	30%	50-90%	Rare	8-60%

The infiltrative nature of the lesion makes complete extirpation of the tumor difficult. Surgery remains the treatment of choice, with the site and size of the tumor determining the treatment protocol. The various surgery types could be wide excision, enucleation, and curettage, resection, or surgeries involving resection of adjacent tissues. **[**22**]**

Odontogenic myxoma has a notoriously high recurrence rate of up to 25% after curettage **[**6**]**. A minimum follow-up period of 5 years without recurrence is recommended by some researchers before performing reconstructive surgeries. **[**23**]**

Conclusion

Odontogenic myxoma is an atypical entity among head and neck lesions. It shadows various other tumors and cysts, in its clinical, radiological and histopathological appearance. A rigorous step-by-step diagnostic process with thorough investigations can rule out potentially malignant lesions. As there is no consensus on the treatment to be followed, in the current case of a young growing patient, conservative surgery with follow-up was appropriated. Broader research with patients in the younger age groups can propel the informative evidence we currently have, towards making a quicker refined diagnosis.

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