
CASE REPORT**Odontogenic Fibromyxoma of Maxilla: A Case Report***CD Mounesh Kumar^{1*}, KV Suresh², Praveen Reddy B³, KV Umashankar³**¹Department of Oral & Maxillofacial Surgery, ²Department of Oral Medicine and Radiology, School of Dental Sciences, Krishna Institute of Medical Sciences, Deemed University, Karad -415539, (Maharashtra), India; ³Department of Oral & maxillofacial surgery, College of Dental Sciences, Davangere - 577001, (Karnataka), India***Abstract:**

Odontogenic fibromyxoma (OM) is a rare locally invasive, non metastasizing benign neoplasm found exclusively in the jaws. It commonly occurs in the second and third decade, and the mandible is involved more commonly than the maxilla. The lesion often grows without symptoms and presents as a painless swelling. The radiographic features are variable so the diagnosis is not easy in many cases. It poses a diagnostic and therapeutic challenge due to its morphology and biological behaviour. A case of OM of the maxilla with unusual radiographic and histological features is described in a 27-year-old female.

Keywords: Myxoma, Fibromyxoma Odontogenic tumor, Multilocular radiolucency

Introduction:

Fibromyxomas of head and neck are rare noncapsulated benign neoplasms. The WHO defines a myxoma as a locally invasive neoplasm consisting of rounded and angular cells that lie in the abundant mucoid stroma. The term "odontogenic myxoma" is often applied when the tumor occurs in the jaws to reflect its odontogenic origin [1]. OM is considered to be the second commonest odontogenic tumor in many countries, however only 0.5 to 17.7 % of

them have been reported in Asia, Europe and America. It comprises around 3-6% of all odontogenic tumors [2].

Case Report:

A 27-year-old female patient reported to the department, complaining of an asymptomatic unilateral swelling in the right maxillary posterior region since one year. The swelling was initially small and gradually increased to its present size of approximately 5 x 3.5 cm. An extra oral examination revealed a diffuse, bony hard, painless, immobile swelling on the right side of face, obliterating the naso-labial fold. (Fig. 1) Buccal and palatal cortices were expanded, and there was no history of paresthesia. Intraorally swelling was extending from distal porting of the right central incisor to mesial side of second right molar with obliteration of buccal vestibule. (Fig. 2) Lateral incisor, canine and second molars were palatally displaced. First premolar was missing. Associated teeth showed Grade II mobility. Skin over the swelling showed scarring and right lower eyelid was pushed upwards. Routine hematological and biochemical investigations were normal.

The panoramic radiograph showed a well-defined radiopaque lesion involving right maxillary antrum. Teeth in the affected region showed displacement. First premolar was horizontally

impacted. (Fig. 3) The CT image showed an expansile mass in the right maxilla, which completely obliterated the maxillary sinus. Incisional biopsy was performed, histopathological examination of specimen showed a loose myxoid stroma with interspersed dense collagen bundles. Numerous stellate to plump spindle shaped cells in the stroma. Cells were active with granular cytoplasm, dense diffuse collection of fibrils and fibroblasts and small inconspicuous strands of odontogenic epithelium (Fig. 4). It was diagnosed as odontogenic fibromyxoma, and the lesion was surgically excised with en bloc resection of the maxilla and obturator was fabricated to cover the surgical defect. (Fig. 5) The course was uneventful after the surgical removal of the tumor. On gross examination, Specimen measured around 6.5x6 cms. in dimension, whitish in color, firm to hard in consistency and was roughly wedge shaped.

Fig. 1: Extra Oral Photograph showing Asymptomatic Swelling in Right Maxilla



Fig. 2: Intraoral Photograph showing Diffuse Swelling in Right Maxilla.



Fig. 3: OPG showing Radiopaque Lesion in Right Maxilla

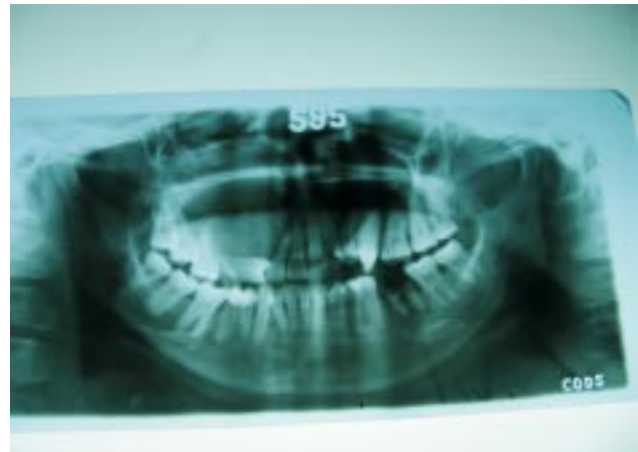


Fig. 4: Histopathology Photograph showing Loose Myxoid Stroma with Interspersed Dense Collagen Bundles and Numerous Stellate to Plump Spindle Shaped Cells in the Stroma

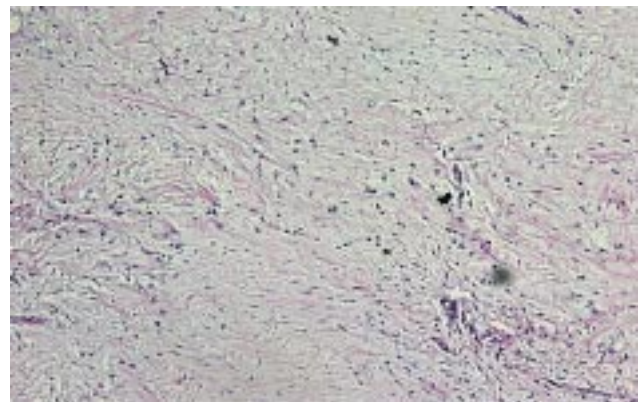


Fig. 5: Intraoperative photograph showing the resection of tumor

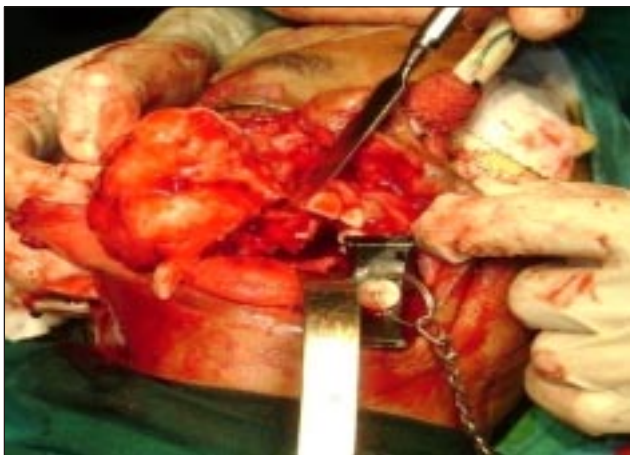


Fig. 6: Postoperative Photograph



Discussion:

Odontogenic myxomas (OM) are rare tumors derived from embryonic mesenchymal elements of dental anlage [2, 3]. World Health Organization has classified OM as a benign tumor of ectomesenchymal origin with or without odontogenic epithelium [4]. It appears to originate from the dental papilla, follicle or periodontal ligament [5, 6]. Virchow in 1863

coined the term myxoma for a group of tumors that had histologic resemblance to the mucinous substance of the umbilical cord. In 1947, Thoma and Goldman first described myxomas of the jaws.

OM is a locally invasive benign neoplasm. The invasiveness is attributed to the biological nature of the tumour. The OM exhibits abundant extracellular production of ground substance and thin fibrils by the delicate spindle-shaped cells. These undifferentiated mesenchymal cells are capable of fibroblastic differentiation also [7-9]. Depending upon the pattern of differentiation, the histological nature of the tumour varies. It may have complete myxomatous tissue or varying proportions of myxomatous and fibrous tissue. In the latter case it can be designated either as odontogenic fibromyxoma, in which the myxomatous element predominates; or odontogenic myxofibroma, with predominance of fibrous tissue [10, 11].

The tumour occurs across an age group that varies from 22.7 to 36.9 years. It is rarely seen in patients younger than 10 years of age or older than 50 [13]. Our case has presented at the age of 27 years, which is in conformity with that reported in the literature. The mandible appears to be more frequently affected than the maxilla, especially the posterior region. In our case posterior region of the maxilla is involved. The majority of myxomas are almost always asymptomatic, although some patients present with progressive pain in lesions involving maxilla and maxillary sinus.

OM of the maxilla is less frequent but behaves more aggressively than that of the mandible, as it spreads through the maxillary sinus [11, 13]. Displacement and mobility of teeth are rela-

tively common. It may be associated with unerupted teeth. Cortical expansion can occur and large lesions can cause perforation [5, 7, 12, 13]. These features have been in accordance with present case.

Radiographically, the tumour presents as a unilocular or multilocular radiolucent lesion with well-defined borders with fine, bony trabeculae expressing 'honeycomb', 'soap bubble', 'tennis racket', wispy or 'spider web' appearance. Unilocular appearance may be seen more commonly in children and in the anterior part of the jaws [6, 8]. Displacement of teeth is a relatively common finding, root resorption is rarely seen and the tumour is often scalloped between the roots [4, 5, 7]. Premolars and molars regions are most commonly affected. In the present case OPG has showed well defined radiopacity with displacement of associated teeth and impacted first premolar.

The differential diagnosis based upon the radiographic, clinical and surgical findings includes ameloblastoma, odontogenic myxoma, central giant cell granuloma, ameloblastic fibroodontoma and odontogenic fibroma central hemangioma and odontogenic keratocyst [15, 16].

The tumor is not radiosensitive, and surgery is the treatment of choice [2, 12]. The lack of a capsule and infiltrative growth pattern is responsible for high rate of recurrence when conservative enucleation and curettage are performed. Radical resection including a margin of 1.5–2 cm healthy bone has been the mode of treatment for all cases.

The clinical presentation of present case has suggested an infiltrative tumour of the maxilla. Hence, surgical excision with segmental

maxillectomy has been performed. Prognosis of myxomas of the jaw is generally good. Recurrence typically occurs during the first 2 years after removal although recurrence has been described over 30 years after original surgery [17].

Conclusion:

Myxomas of head and neck are rare tumors. It poses a diagnostic and therapeutic challenge hence correlation of clinical, radiological and histopathological features are essential when trying to diagnose lesions which lack the characteristic appearance. A complete surgical excision along with proper long term follow up is essential keeping in mind the high recurrence rate, for the successful management of the myxomatous tumours.

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