

Odontogenic Myxoma: Clinico-radiographic Presentation – A Case Report

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Abstract: Odontogenic myxoma (OM) is a rare tumour arising in the jaws. It is considered to be derived from ectomesenchymal portion of tooth germ. They are slow growing, non-metastasizing, often asymptomatic with local aggressiveness due to its infiltrative nature and hence high recurrence rate, with a high incidence of occurrence in the mandible. Most frequently occurs in second to third decade of life, seldom occurs beyond these age groups. According to the World Health Organization (WHO), the odontogenic myxoma is classified as an odontogenic tumor of ectomesenchymal origin. The radiographic appearance of OM is not specific and the diagnosis is frequently unexpected following surgical removal of the lesion. We present a case of odontogenic myxoma in an Eighty-year old male patient, which had acquired large dimensions and involved the right maxilla including maxillary sinus of right side resulting in gross facial deformity within a span of two months.

Keywords: Myxoma, maxillary sinus, odontogenic tumors.

I. Introduction

Myxomas are benign, slow growing and locally aggressive mesenchymal origin neoplasm. [1] It can be found in various parts of body like skin, subcutaneous tissues and heart. Myxomas of head and neck region are rare. [1, 2] Two forms of myxoma have been identified. Those derived from facial bones can be either osteogenic myxoma or odontogenic myxoma (OM). Another form is derived from soft tissue like perioral soft tissue, parotid gland, ear or larynx. [2, 3] OM represents an uncommon benign neoplasm constituting of about 3-6% of all odontogenic tumors. [2-5] They are benign tumors, non-metastasizing, non-encapsulated, derived from the primitive mesenchymal tissues of dental follicle, dental papilla or periodontal ligament. The evidence for its odontogenic origin arises from its almost exclusive location in the tooth-bearing areas of the jaws, its occasional association with missing or unerupted teeth and the presence of odontogenic epithelium. [6] World Health Organization defined odontogenic myxoma as a benign, locally invasive neoplasm characterized by rounded and angular cells lying in an abundant mucoid stroma, that replaces the cancellous bone expanding the cortex. [7] There are only few reports on the relative frequency and incidence of OM in the available literature. However, in Asia, Europe and America relative frequencies between 0.5% and 17.7% have been reported. Clinically, odontogenic myxoma is a benign painless, invasive, slowly enlarging mass causing marked asymmetry of the face. [8] It commonly involves the mandibular premolar and molar regions. Females have higher predilection than males. It usually occurs in second and third decades of life and causes expansion of bony cortices, displacement and loosening of teeth. [9] Radiographically, its appearance ranges from unilocular pericoronal radiolucency with variable trabecular pattern giving rise to soap bubble, tennis racket, or honey comb appearance. The “sunray” or “sunburst” appearance has also been reported in the literature. Root resorption is rarely seen, though displacement of teeth is relatively common. [10]

Histopathologically, OM is defined as non-encapsulated benign odontogenic tumor of mesenchymal origin that is locally invasive and consists of rounded and angular cells that lie in abundant mucoid stroma. It is composed of round and spindle-shaped cells lying in a rich myxoid stroma [7]. The tumor is rich in extracellular matrix [ECM] represented by type I collagen, fibronectin, tenascin, chondroitin sulphate and especially rich in hyaluronic acid [11,12,13]. Small islands of apparently inactive odontogenic epithelial rests may be scattered through the myxoid substance [14,15]. There is a microscopic similarity between odontogenic myxoma and the dental papilla [16]. According to Kim et al [17] myxomas do not have the epithelial lining found in many dental follicles.

The treatment plan varies from curettage to radical excision. Complete surgical excision can be difficult as the lesion is not encapsulated and because the myxomatous tissue infiltrates adjacent bone tissues.

II. Case report

An 80-year-old male patient presented with a complaint of pain and swelling over the right side of the cheek area for the past two months. Patient was relatively well two months back but then noticed a pain and swelling in the upper right back tooth region of jaw, for which he visited a local dentist and drainage was done from the swelling. After six months of drainage, pain and swelling developed again. Pain was radiating in nature which causes discomfort to patient while eating.

Extraoral examination revealed a diffuse, ill-defined, firm swelling of size 5 cm × 4 cm involving the right cheek extending superior-inferiorly from infraorbital rim to the zygomatic process of maxilla. (Fig. 1) Medially the lesion was found to extend to the lateral wall of the nose with obliteration of the nasolabial groove and laterally to about 1 cm in front of tragus; the skin over the lesion appeared normal and pinchable. The swelling was tender and afebrile.

Intraorally well defined around 3 cm in size, oval shaped swelling was present extending from upper right second premolar extending till maxillary tuberosity, thereby obliterating the right buccal vestibule. (Fig. 2) Swelling was soft, non-fluctuant, tender and without any discharge. Effect on the dentition cannot be appreciated as the patient was completely edentulous. Medical history was not significant.

The patient underwent panoramic radiographic investigations that suggested radiolucent area around 3×4 cm in size over the maxillary alveolar process obscuring the maxillary sinus. The internal structure of the lesion has mixed radiolucent and radiopaque appearance. (Fig.3)

Coronal and axial sections of cone beam computed tomography suggested obliteration of the right maxillary sinus and none of the wall of maxilla sinus was appreciable. Also deviation of nasal septum towards left side can be appreciated.

Based on the clinico-radiographic findings provisional diagnosis of benign odontogenic tumor was given. Differential diagnosis includes fibro-osseous lesion, salivary gland tumor and carcinoma of maxillary sinus.

Incisional biopsy was done from the lesion in which two bits of soft tissue specimen which was creamish white in colour, measuring about 4.5×2.5 cm, having jelly like material was taken. (Fig. 5) H&E stained section showed highly cellular stroma, which are loosely arranged, showing spindle shape and stellate cells. The tumor cells are interspersed with tiny capillaries and occasional strands of collagen. (Fig. 6) Based on the histologic picture the final diagnosis of odontogenic myxoma was given. Patient is planned for resection of the lesion and is under follow-up.

III. Discussion

Myxoid tumors of soft tissue represent a heterogeneous group of lesions that exhibit significant differences in biologic behaviour ranging from harmless to malignant neoplasm. [18] Rudolf Virchow in 1863 coined the term myxofibroma, for a group of tumors that had histologic resemblance to mucinous substance of the umbilical cord. [2-5] OM was first mentioned in the literature by Thoma and Goldman in 1947. [21] In 1948, Stout redefined the histologic criteria for myxomas as true neoplasm that do not metastasize and exclude the presence of recognizable cellular components of other mesenchymal tissues, especially, chondroblasts, lipoblasts and rhabdomyoblasts. [2,3]

We have reported a case of odontogenic myxoma involving the maxilla and the maxillary sinus. Its slow but aggressive growth rate makes it reach large sizes and, when involving the maxilla, it can invade the entire maxillary sinus, as was the case here.

The tumor occurs across an age group that varies from 22.7 to 36.9 years. [22,23] It is rarely seen in patients younger than 10 years and older than 50 years of age, but in the present case the patient is 80 years old and this age group is quite unusual according to literature. According to Farman *et al.*, suggested mean age of maxillary OM for men was 29.2 years and 35.3 years for women while mandibular OM in men occur at mean age of 25.8 years and 29.3 years for women. [11] Gunahan *et al.* and Regaziet *al.* reported a distinct predominance in females (64–95%) and a predilection for the mandible. [12,13] Keszler *et al.* noted that 8% of OMs occurred in children of <16 years. [14,15] Posterior Mandible is more frequently affected than the maxilla. [3-6] According to Reichart and Philipsen, mandibular myxomas accounted for 66.4%, with 33.6% in the maxilla. Whereas 65.1% of mandibular cases located in molar-premolar region, 73.8% were located in same areas of the maxilla. [2, 3, 16] In the present case the patient is male and the area affected is maxilla.

OM is commonly described as a slow growing tumor and generally is symptomless. There are also scattered reports of rapid enlargement [19, 20]. Bony perforation, with subsequent invasion into the soft tissues, which is reported to be common in large OMs, was not observed in this patient. Paraesthesia, mobility of teeth and ulceration were among the other complaints that patients usually presented with, but were not present in our case.

Odontogenic myxoma is a benign painless, invasive, slowly enlarging mass causing marked asymmetry of the face. [3] It commonly involves the mandibular premolar and molar regions and females have higher

predilection than males in contrast to our case.[21] The tumor is often associated with congenitally missing or unerupted tooth. It grows slowly and may or may not cause pain. OM of the maxilla may be asymptomatic or on examination, it may present as heaviness, swelling of cheek or palate, malocclusion or loosening of teeth.[3] Eventually it causes expansion and may grow quite large if left untreated. It may also invade the maxillary sinus, often fills the entire antrum and cause exophthalmos. [21] Recurrence rate is high as 25% have been reported. This high recurrence rate may be explained by the lack of encapsulation of the tumor, its poorly defined boundaries, and the extension of nests or pockets of myxoid (jelly-like) tumor into trabecular spaces where they are difficult to detect and remove surgically.

Myxomas in the maxilla usually involve the alveolar process in the premolar and molar regions and the zygomatic process of maxilla rather than anterior maxilla. The periphery of the lesion is usually well defined and often has a corticated margin. However, the outline of some lesions, especially in the maxilla is poorly defined. On conventional radiograph the tumor presents as a unilocular or multilocular radiolucent lesion with fine, bony trabeculae within its interior structure, expressing as “honeycombed”, “soap bubble”, or “tennis racket” appearance. Unilocular appearance may be seen more commonly in children and in the anterior part of the jaws. Dental displacement and loosening of teeth is a relatively common finding. The lesion also frequently scallops between the roots of adjacent teeth and in rare cases the roots may show resorption. The “sunray” or “sunburst” appearance has also been reported in the literature.

CT findings in odontogenic myxoma are varied and may reveal osteolytic expansile lesions with mild enhancement of the solid portion of the mass. Some cases may show bony expansion and thinning of cortical plates with strong enhancement of the mass lesion. A soft tissue mass with bone destruction and fine lace-like density representing ossifications in the maxillary sinus may be seen in some cases. [24] Radiological diagnosis of odontogenic myxoma is difficult because of overlapping features with other benign and malignant bone lesions. Some odontogenic myxomas may show a mixed radiopaque-radiolucent appearance which is ascribed to the presence of foci of calcification. Thus diagnosis should be considered in mixed radiolucent-radiopaque lesions [8, 19]. Differential diagnoses include ameloblastoma, central giant cell granuloma, intraosseous haemangioma, aneurysmal bone cyst, glandular odontogenic cyst, cherubism, metastatic tumor and, in cases of unilocular lesions, simple cysts and odontogenic keratocyst. In older patients, the possibility of a malignancy should not be ruled out.

On gross examination of the specimen, the gelatinous, loose structure of the myxoma becomes obvious [25]. Microscopically, the myxoma is made up of loosely arranged spindle-shaped and stellate cells. The intercellular substance is mucoid. The tumor is usually interspersed with a variable number of tiny capillaries and occasionally strands of collagen [26]. In case of fibromyxoma, the amount of collagen in the mucoid stroma is more prominent. Remnants of odontogenic epithelium have occasionally been noted, sometimes being surrounded by a narrow zone of hyalinization. The microscopic features in this case were compatible with that of odontogenic myxoma, which is comprised of spindle or stellate cells in a mucoid material. Since it is an infiltrative, aggressive disease, with a high recurrence rate, treatment should aim at complete surgical excision of the mass along with excision of bony margins.

Surgical excision is the recommended treatment ranging from conservative curettage [27, 28] to radical excision [29]. However, owing to its potential for local infiltration, simple enucleation and curettage alone have been associated with a high recurrence rate of 10 to 33% [8, 30]. Thus radical excision with burring of the cavity borders with a drill should be performed with maximum preservation of surrounding structures.

IV. Conclusion

Clinical and radiological aspects of maxillary odontogenic myxomas are not conclusive, it is necessary to have a histopathological exam for the final diagnosis. Because of its high rate of recurrence, especially due to its gelatinous and mucous aspect, surgical treatment through bone resection is the most indicated treatment modality, and the patient must be followed up closely for years.

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Fig. 1 Extraoral view showing swelling on right maxilla with obliteration of nasolabial fold



Fig. 2 Intraoral view showing swelling at posterior maxilla of 14-18 region



Fig. 3 Orthopantomogram showing poorly defined mixed radiolucent and radiopaque area at right posterior maxilla

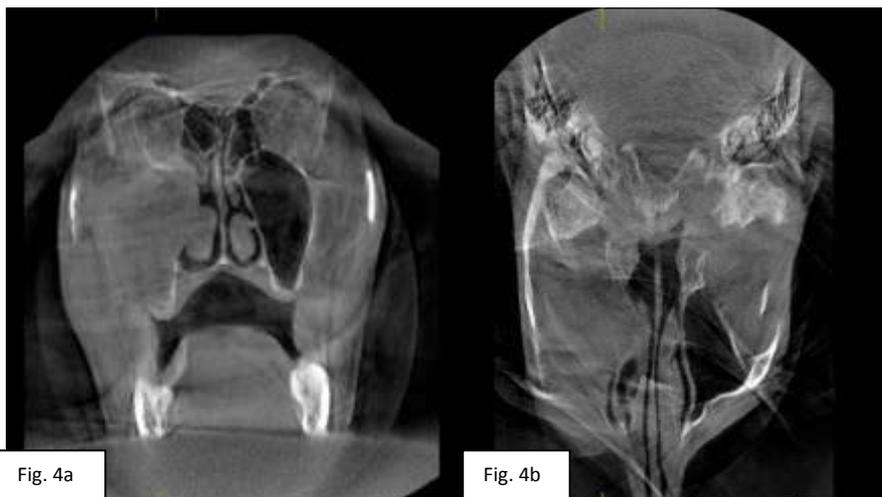


Fig. 4 (a,b) Coronal section and axial section of CBCT showing complete obliteration of maxillary sinus & deviated nasal septum.

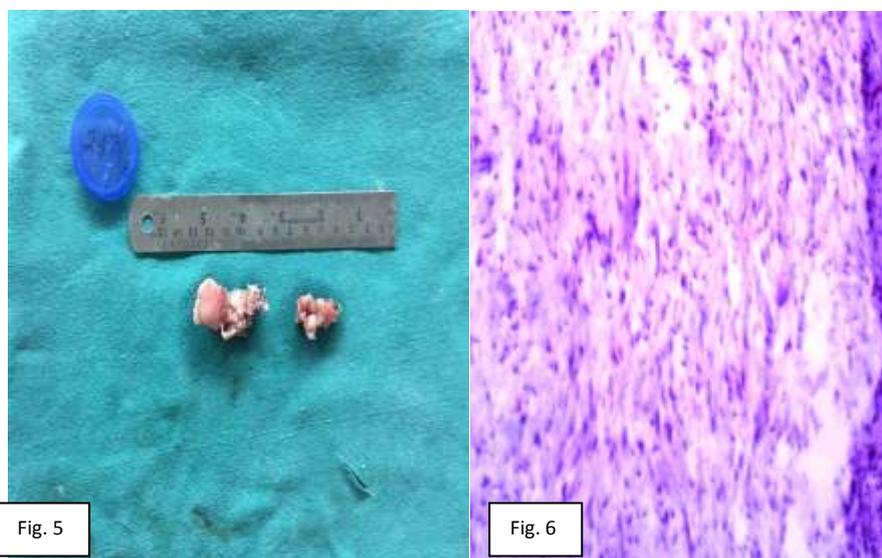


Fig. 5 Soft tissue specimen, creamish white in colour, measuring about 4.5×2.5 c, having jelly like material

Fig. 6 H&E stained section (40X) showed highly cellular stroma, which is loosely arranged, showing spindle shape and stellate cells.