Case Report

Odontogenic Myxoma of the Mandible: A Case Report and Review of Literature

Shamimul Hasan¹, and Saurabh Jain²

¹Dept of Oral Medicine and Radiology Faculty of Dentistry Jamia Milia Islamia New Delhi; India
²Dept of Oral and Maxillofacial Surgery Ahmedabad Dental College And Hospital Ahmedabad, Gujarat; India

Article History

Received 14 March 2012
Accepted 10 August 2012

Keywords:
odontogenic tumors,
odontogenic myxomas,
multilocular radiolucency-

Abstract

Odontogenic tumors constitute a heterogeneous group of lesions with diverse clinical manifestations and histopathologic features. Odontogenic myxomas are believed to arise from odontogenic ecto-mesenchyme, and accounts for 3–6% of all odontogenic tumors. The tumor is more common in the mandible, with an age predilection for the 2nd–3rd decade. Radiographically, they appear as initially unilocular radiolucencies, becoming multilocular over time. The radiolucent defect can contain thin, wispy trabeculae of residual bone, which are often arranged at right angles to each other. Large myxomas can also show soap bubble pattern. Treatment modalities vary depending on the tumor size and ranges from local excision, curettage or enucleation- to radical resection. Hereby presenting the case report of a 32 year old male patient diagnosed and treated for odontogenic myxoma of the mandible, along with a detailed review of literature.

Introduction

Myxomas are non encapsulated benign locally aggressive tumors which may occur in both the soft tissue and bone. Macroscopically, a myxoma appears as a mass of characteristically mucoid or slimy material. Microscopically, as first described by Virchow, it consists of stellate cells with long intertwining processes in an amorphous mucoid ground substance (1). The origin of odontogenic myxoma is believed to be the mesenchyme of a developing tooth or the periodontal ligament that has been justified by: Its exclusive occurrence in the tooth bearing areas of the jaws.; frequent occurrence in young individuals.; common association with an unerupted or a developmentally absent tooth.; histologic resemblance to dental mesenchyme; and the occasional presence of sparse amounts of odontogenic epithelium (2).

There appears to be no predilection for either sex in odontogenic myxoma. Approximately 60% of these tumors occur within second and third decades of life. Most studies state that mandible is more commonly affected, particularly in posterior body, angle and ramus. In maxilla, the alveolar and zygomatic processes are the most common sites of occurrence (3). It is usually characterized by a slow painless growing tumor, can cause tooth displacement and mobility, facial asymmetry, delayed eruption of teeth, disturbance of speech and mastication, pain and paraesthesia and oral mucosal ulceration (4). Radiographically, the tumor presents as initially unilocular radioluencies, becoming multilocular over time. Radiographic, characteristics can be described as well-defined corticated, well-defined non-corticated, poorly defined, or diffuse. The internal structure of odontogenic myxomas is described as a unilocular or an expansile - multilocular radiolucency. Multilocular lesions are named "tennis-racket", "soap bubble", or "honeycomb" by some authors (5, 6). Root resorption is rarely seen, though displacement of teeth is relatively common (5).

Despite the fact that odontogenic myxoma shows aggressive local growth, it is believed that it never undergoes malignant transformation or give rise to metastasis (7), but there are reports of presumed benign lesions that then follow an aggressive course with local recurrences (8). Despite the benign nature of these lesions, there is a high rate of local recurrence after curettage alone and in certain cases requires adequate resection (9).

Correspondence to:
Shamimul Hasan
E-mail: shamim0571@gmail
A 32 year old male patient reported to the department of Oral medicine and Radiology, Faculty of dentistry, Jamia Milia Islamia with a slow growing, painless swelling in the left mandibular region since 2 years. History revealed that the patient initially had mobility in mandibular left first and second molar teeth, following which the patient noticed a small swelling which has progressively increased to attain the present size. The past medical history was non-contributory. Extraoral examination revealed a painless bony hard swelling on the left side of face involving the mandibular body and ramus area. The swelling extended superiorly upto the ala-tragus line, inferiorly 2 cms below lower border of the mandible, antero-posteriorly from the symphysis region upto the angle of the mandible. There was marked facial asymmetry, with deviation of the lip to the right side along with obliteration of the nasolabial fold. Single, moveable, nontender left submandibular lymph node was also palpable (Figs.1, 2). Intraoral examination revealed a bony hard swelling extending from mandibular left central incisor to left retromolar region. There was bicortical expansion. On the buccal aspect, the swelling was bony hard with obliteration of the vestibule. On the lingual aspect, there was an overlying soft tissue component. The swelling was firm and yielded on pressure. Indentation marks of the cusps of maxillary teeth on the soft tissue component were seen. Mandibular left second premolar, first molar, second molar and third molar teeth showed displacement, and mandibular left first and second molar teeth were grade 1 mobile (Figs.3, 4). Routine haematological and biochemical laboratory tests were within normal
limits. Haemoglobin was slightly below the normal limits-value of 10.5 gm%. Orthopantomogram (OPG) revealed a multilocular radiolucent lesion extending from mandibular right central incisor, lateral incisor and canine tooth upto the sigmoid notch on the left side. Condyle and coronoid process were not involved. Destruction of left lower border of mandible was evident along with fine radiating bony trabeculae over the lower border. There was lifting of periosteeum along left angle of the mandible. Root resorption with mandibular right central incisor, lateral incisor, canine and mandibular left central and lateral incisor teeth was seen, and OPG showed “tooth floating in air” appearance (Fig.5). The lesion was differentially diagnosed as Odontogenic neoplasm (Ameloblastoma, odontogenic myxoma, keratocystic odontogenic tumor). Malignancy within odontogenic neoplasm (Ameloblastic carcinoma/sarcoma), central giant cell granuloma and vascular neoplasm of bone (Haemangioma). After the informed consent of the patient, incisional biopsy was done and it revealed typical features of odontogenic myxoma, containing loosely arranged stellate or spindle shaped cells within a myxoid matrix (Fig.6). H & E stained section shows parakeratinised stratified squamous epithelium and non cellular underlying connective tissue. Under the histopathologic diagnosis of odontogenic myxoma, a lower cheek flap was raised using Roux lip split incision. Segmental mandibulectomy was done and the tumor mass was resected along with it. The resected specimen (including
right canine up to left condyle) was sent for histopathological analysis (Fig. 8). The histopathology of the excised tumor mass revealed loose myxoid stroma consisting of loosely arranged stellate and spindle-shaped cells, along with round cells. Few of the collagen fibrils were seen that tended to intermesh. Small islands of inactive-appearing odontogenic rests were seen scattered through the myxoid stroma, histopathology suggestive of odontogenic myxoma. (Fig. 9)

Discussion

The German pathologist Rudolph Virchow was probably the first to describe the histologic features of myxofibroma in 1863, although the lesions of jaws were not particularly mentioned. In 1947, Thoma and Goldman first described myxomas of the jaws. Since then, odontogenic myxoma has been a subject of continuous debate (10). Myxomas of the head and neck are rare tumors. Two forms can be identified: (a) facial bone derived, which had been subdivided in the past into true osteogenic myxoma and odontogenic myxoma, and (b) “soft tissue” derived myxoma, derived from the perioral soft tissue, parotid gland, ear and larynx (11).

Odontogenic myxoma is classified as a locally invasive, non-metastasizing benign tumor, which presents 3-6% of all odontogenic tumors of the jaws by World Health Organization (WHO) (5, 12). Though it is a benign neoplasm, it may be infiltrative, aggressive and may recur (13). Previous theories stress that the lesion derives from the neural sheath or is the result of degeneration of fibromas, lipomas and so forth, due to the chronic irritation and the degenerative processes following tissue anoxemia (14). 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 (15). This explanation fails to describe soft tissue myxomas. They probably arise from supporting structures of the teeth like the gingiva and the periodontal ligament (16).

The tumor occurs across an age group that varies from 22.7 to 36.9 years. It is seen rarely in patients under 10 years of age or older than 50 years (17). However, studies by Simon et al. (4) revealed the occurrence of this tumor in a 3-month-old baby, being the youngest and the oldest was 64 year old. The patient in our case was 32 years of age, which is in conformity with what reported in the literature. The mandible appears to be more frequently affected than the maxilla, especially in posterior region. The majority of myxomas are almost always asymptomatic, although some patients present with progressive pain in lesions involving maxilla and maxillary sinus, with eventual neurologic disturbance. Odontogenic myxoma of the jaw has a tendency for extensive bone destruction, invasion into surrounding structures and a relatively high recurrence rate; however, metastasis is rare (18). Odontogenic myxoma of the maxilla is less frequent but behaves more aggressively than that of the mandible, as it spreads through the maxillary sinus (10, 17). When the maxillary sinus is involved, the odontogenic myxomas often fill the entire antrum. In severe cases, nasal obstruction or exophthalmus may be the leading symptoms (19). The patient in the
present case presented with an asymptomatic slowgrowing swelling, involving the body of mandible and ramus.

Barros et al. (7) proposed that the radiologic appearance of the odontogenic myxoma consists of one or two patterns, depending on the evolution of the tumor. The first stage begins with an osteoporotic appearance, with more prominent medullary spaces separated by thin septa of bone. These septa become thinner and more elongated as the tumor infiltrates locally, forming larger areas of osteolysis. During this stage, the lesion acquires its classic radiographic appearance which consists of a multilocular radiolucency with well developed locules, composed of trabeculae tending to intersect at right angles. The bony septa forming the locules usually are straight, thin, elongated, and lacy. According to Eversole-(20) the internal configuration of the bony septa resembled "Lichen planus of the jaw bone". Although some authors have described them as having a soap-bubble or honeycomb appearance, many lesions tend to form more angular locules, resembling the strings of a tennis racket. The second stage consists of the breakout or destructive phase characterized by a loss of internal locules, significant expansion, and perforation of the cortex with invasion into the surrounding soft tissues; in the maxilla, there is extension into the antrum. The early feature of this stage is the appearance of septa beyond the peripheral margin of the lesion, extending at right angles to the margin-thus imparting a hairbrush or sunburst appearance (7).

Orthopantomogram in the present case revealed a multilocular radiolucent lesion involving the left body and ramus of mandible, with extensive destruction of the inferior border, root resorption and "tooth floating in air" appearance. Some odontogenic myxomas may show a mixed radiopaque-radiolucent appearance, which was attributed to the presence of foci of calcification (19). It was suggested that this appearance may be due to residual bone and not to new bone formation, and therefore it was proposed that odontogenic myxomas should be considered in the differential diagnosis of mixed radiolucent radiopaque lesions (19).

Computed tomographic (CT) images of odontogenic myxomas may show any of the following features:

a) Osteolytic expansile lesions with mild enhancement of the solid portion of the mass in the myxoma of the mandible.

b) Bony expansion and thinning of the cortical plates, with strong enhancement of the mass lesion in the anterior maxilla.

c) A soft tissue mass with bone destruction and thinning and strands of fine lace-like density representing ossifications in the maxillary sinus (21).

Magnetic resonance imaging (MRI) revealed a well defined, well-enhanced lesion with homogenous signal intensity on every pulse sequence. The lesion showed intermediate signal intensity on the T1 - T2-weighted images (10). CT and MRI was not performed in the present case, as the patient could not afford the expenses.

Macroscopically, typical cases show an unencapsulated mass; but with well-defined borders. The cut surface is soft, grey white and gelatinous (4). Histologically, it is hypocellular tumor composed of loosely arranged, spindle shaped and stellate cells in a background of abundant myxoid stroma. The cells have round to oval bland nuclei with finely stippled chromatin (22). Atypical nuclei are often encountered, but cellular polymorphism, prominent nucleoli and atypical mitotic figures are not seen (4). The cytoplasm is eosinophilic. Small islands or strands of odontogenic epithelium are rarely observed (23). Immunohistochemistry shows positive staining with vimentine. Desmin, smooth muscle actin, S100, and cytokeratin stains are either negative or equivocal and focally positive (4). The present case showed typical features of odontogenic myxomas- loosely arranged spindle and stellate cells in a myxoid background, together with small islands of odontogenic epithelium.

The tumor is not radiosensitive, and surgery is the treatment of choice (24). The various methods of surgical treatment advocated are simple excision, or enucleation with electrocautery, marginal resection with continuity of bone maintained or resection in extensive lesions or recurrences (25). Since odontogenic myxoma bear a high risk of recurrence, mainly due to its gelatinous aspect and having no capsule, it is necessary that the initial treatment be very efficient. One important aspect that should be taken into account is the possibility that the patients may skip control visits. Thus a possible recurrence will be seen only later on and treatment will be impaired. For all these reasons, resection with broad margins is the most indicated treatment (4). Postoperative preservation of patients with odontogenic myxomas is unclear, especially two in the first 2 years, period of greatest recurrence rate (26). In the present case, segmental mandibulectomy was done and tumor mass was resected. Follow up was made for a period of two years and there was evidence of recurrence.
Conclusion

Odontogenic myxomas are rare benign jaw neoplasms, with a great deal of controversies regarding the histogenesis. The neoplasm should be diagnosed on the basis of radiographic imaging modalities and histopathological examination. Appropriate treatment modalities should be instituted and recurrences should be avoided.

References