An Uncommon Mesenchymal Tumor of Maxilla

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ABSTRACT

Myxoma of jaw is a very rare benign mesenchymal tumor, which is slow growing, painless, and exhibits aggressive behavior locally. It has a prevalence rate of about 0.04 to 3.7%. It frequently occurs in the 3rd and 4th decades of life, with more female predilection in the ratio of 1:1.6. The mandible is more commonly involved than the maxilla predominantly in the premolar and molar regions, associated with swelling, pain, and loosening of teeth. The radiographic features are variable; it may appear as unilocular or multilocular radiolucency with “honey comb” or “tennis racket” pattern with cortical expansion and tooth displacement. Corrective management of myxoma involves a radical primary resection despite significant functional and esthetic sequelae due to its locally aggressive behavior and high recurrence rate. Here, we present a rare case of myxoma involving the maxilla of a 50-year-old female patient.

Keywords: Aggressive behavior, Benign, Mesenchymal tumor, Myxoma.

CASE REPORT

A 50-year-old female patient visited the Department of Oral Medicine and Radiology with a chief complaint of painless swelling in her upper left jaw since 1 month. Patient history revealed the swelling was initially small in size, which gradually increased to its present size. There was no history of pain, pus discharge, bleeding, and trauma. Patient also gave history of intermittent bleeding with mucopurulent discharge from left nose, loss of smell, watery discharge of left eye along with headache and heaviness of head.

Postoperative healing was uneventful. Recent medical history unveils that the patient is a known case of hypertension since 2 weeks and is under medication (tab Rancil 5 mg). Family history was noncontributory. On general physical examination, patient was moderately built and nourished.

On extraoral examination, a tear drop-shaped erythematous mass measuring about 0.3 × 0.3 cm was present irt left nostril (Fig. 1). Palpation of nasal mass was not possible due to inaccessibility. Solitary left submandibular lymph node was palpable measuring about 0.5 × 0.5 cm, round in shape, soft in consistency, nontender, and mobile.

Intraorally, a diffuse swelling was present over the left anterior and posterior hard palate measuring about 5 × 3 cm, extending anteroposteriorly from 23 region to mesial aspect of 27. Mediolaterally from 0.6 cm away from midpalatal raphe to attached gingiva irt 23, 24 region and approximately 1 to 1.5 cm away from midpalatal raphe to attached gingiva irt 25, 26 region, mucosa overlying the lesion appeared normal. Mandible appears to be more frequently affected than the maxilla. Myxomas of head and neck can be identified in two forms: (1) Facial bone derived, which has subdivided into true osteogenic myxoma and odontogenic myxoma and (2) soft tissue myxoma derived from perioral soft tissue, parotid gland, ear, and larynx. True myxomas tend to infiltrate the surrounding structures and often recur after excision.

INTRODUCTION

Myxoma is a tumor of primitive indifferent mesenchyme, closely mimicking the structure of mucoid connective tissue of umbilical cord. The term myxoma was first coined by Virchow in 1863. Stout (1948) defined myxoma as true mesenchymal tumor, which neither metastasizes nor includes chondroblasts, lipoblasts, myoblasts, or recognizable tissues. Myxoma rarely occurs in head and neck region, with an incidence of approximately 0.07 new cases per million people per year at a prevalence rate of 0.04 to 3.7%. It can occur in the heart, skin, subcutaneous tissue, and centrally in the bone, but myxomas of jaws are encountered rarely. Mandible appears to be

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gingival recession, and periodontal pocket present in 25, 26, 27, and 16. Hard tissue examination revealed displacement of teeth from 25 to 27 region, clinically missing tooth in 45, grade I mobility in 25, 31, 41, grade III mobility in 26, 27, root stump in 46, supra and subgingival calculus, and extrinsic stains. Based on history and clinical examination, a provisional diagnosis of recurrent case of mucocele of left maxillary sinus, chronic generalized periodontitis, and nasal polyp in left nose was given. Differential diagnosis of benign tumor arising from left maxillary sinus and malignancy arising from nose or maxillary sinus was given.

Radiographic investigations included intraoral peri-apical radiograph (IOPAR), occlusal radiograph, paranasal sinus (PNS) view, orthopantomograph. The IOPAR in 25, 26, 27 revealed diffuse radiolucency seen on periapical area of mesial root in 26, 27 with 2 to 3 mm horizontal bone loss below the cementoenamel junction (Fig. 3). Occlusal radiograph revealed buccally displaced in 25 and 26 with no expansion of bone (Fig. 4). Orthopantomogram revealed retained root stump in 36, generalized horizontal bone loss, clinically missing tooth in 45, radiopacity seen on mesial root of 37 suggestive of idiopathic osteosclerosis. A well-defined radiolucency was present measuring about $4 \times 1.5$ cm extending mediolaterally from distal aspect of 23 to 27 region and superoinferiorly extending from crest of alveolar ridge to the hard palate (Fig. 5). Hazziness was present over the left maxillary sinus and floor was also not appreciated. Paranasal sinus view that revealed haziness over left maxillary sinus as well as medial wall was not appreciated, which is strongly suggestive of maxillary sinus tumor involving the oral cavity (Fig. 6).

Patient was subjected to multislice computed tomography (CT) with IV contrast to perceive the exact extension of the lesion. The report revealed slight deviation of nose toward right side, well-defined, poorly enhancing lesion diffusely involving whole left maxillary sinus with expansion of its walls and remodeling of it extending laterally into cheek and inferiorly into oral cavity with...
break in hard palate. Inferolaterally, it extended into cheek, superiorly till floor of orbit with focal breech in orbital floor, and involved left ethmoid air cells superiorly (Fig. 7). Posteriorly, it extended into posterior nare extending to right side. Medially, the nasal septum was pushed. Mucosal thickening of right maxillary sinus was noted, which is strongly suggestive of malignancy arising from maxillary sinus invading into oral cavity (Fig. 8).

Hematological investigations were within normal limit except alanine transaminase (serum glutamic pyruvic transaminase) enzyme level, which was elevated up to 153 U/L (normal range 7 to 49 U/L).

Histopathological examination of incisional biopsy from nasal polyp revealed tissue fragments devoid of lining epithelium with an encapsulated tumor composed of loose cellular proliferation of spindle to stellate cells arranged on fibromyxoid stroma, cells with round
hyperchromatic nucleus along with moderate amount of eosinophilic cytoplasm, and no evidence of pleomorphism and mitotic figures. Proliferative blood vessels surrounded by stellate cells and periphery of tumor was covered by hemorrhage and mixed inflammatory cells, which confirmed the diagnosis of fibromyxoma (Fig. 9).

Patient was operated under general anesthesia and hemimaxillectomy was performed and obturator was placed (Fig. 10). The excised specimen was subjected to histopathologic and immunohistochemistry (IHC) analysis. The report revealed typical features of myxoma, loosely arranged stellate cells with elongated cytoplasmic process and abundant myxoid stroma, thin-walled capillaries, edema, and inflammatory cells in the periphery. There was no evidence of increase in mitosis (Fig. 11). The IHC reports revealed negative for S-100 protein marker, which confirmed nonneural origin (Fig. 12).

DISCUSSION

Myxoma is defined as “a tumor composed of sometimes spindle-shaped cells set in a myxoid stroma containing mucopolysaccharide, through which courses very delicate reticulin fibers in various directions and is locally invasive”. Virchow (1863) coined
the term myxoma. Myxoma of the jaws was identified by Thoma (1954) who described two types of myxoma in orofacial region: (i) Odontogenic myxoma and (ii) osteogenic myxoma, the former he regarded as benign, while the latter as malignant. Myxomas can occur in heart, skin, subcutaneous tissue, and centrally in the bone, but myxomas of the jaws are encountered rarely. It accounts for only 1 to 3% of all tumors of the jaws, with an incidence rate of 0.07% new cases per million people per year.6

Myxomas of jaws may be odontogenic in origin (HmTison, 1973). As bony myxomas almost exclusively occur only in the jaws, it is also suggested that they arise from the mesenchyme of the tooth germ (Pindborg, 1965).7 Myxomas are prevalent in the 3rd and 4th decade, rarely occur in children or adult over 50 years of age, with female predilection in ratio of 1:16.4 The mandible appears to be more frequently affected than the maxilla. According to Reichart and Philipsen,8 mandibular myxomas accounted for 66.4%, with 33.3% in maxilla. Our case presented at the age of 50 and maxilla was involved. The maxilla and anterior region of the mandible are rarely affected.2 When found in the maxilla, it usually behaves more aggressively than that of the mandible involving the zygoma, maxillary sinus, and even the orbits as presented in our case. Hence, our case has unique clinical presentation. They are slow growing, invariably isolated tumors. Sometimes, pain and loosening of teeth may be observed. However, in the present case, the lesion was located in premolar and molar regions of the maxilla along with displacement and mobility of teeth. The exact etiology of myxoma is unknown. Depending on their fibrous tissue contents, the consistency may vary from soft to firm. Occasionally, lesion may be infiltrated by fat, acquiring a yellowish tinge resembling a lipomatous tumor.9

According to the different amounts of myxoid component and fibrous tissue, degree of cell polarization, the tumor can show various radiological appearances.10 Myxomas present a variety of radiographic features. They can present as unilocular or multilocular radiolucencies containing delicate or sclerotic trabeculations, with a “honey comb,” “soap bubble,” or “tennis racket” appearance. Lesions greater than 4 cm tend to be multilocular and smaller lesions tend to be unilocular. The radiolucency may have clearly defined border or poorly defined and diffuse.11 In our case, the lesion appeared as unilocular, measuring about 4 × 1.5 cm with well-defined border with no trabeculation. Conventional radiographs have certain limitations such that exact location and extent of lesion cannot be appreciated. Hence, three-dimensional imaging modalities, such as CT and magnetic resonance imaging are mandatory to ascertain the exact extension and location of lesion.

Radiographic differential diagnosis of unilocular lesion without trabeculae includes periapical, lateral, periodontal, and traumatic bone cyst, whereas ameloblastoma, intraosseous hemangioma, aneurysmal bone cyst, and central giant cell granuloma were considered in multilocular lesion.10,11 A biopsy is, therefore, necessary to ascertain an accurate diagnosis.

Microscopically, myxoma is made up of spindle-shaped and stellate cells, with long fibrillar processes that tend to intermesh. The loose tissue is hypocellular, mucoid intercellular substance with no evidence of significant mitotic activity. The tumor is interspersed with thin capillaries and collagen. Similarly, our ultrastructural findings showed tumor with myxoid stroma, thin-walled capillaries, and stellate cells with elongated cytoplasmic processes with no evidence of increase in mitosis.12 This tumor has an extremely rare malignant version called myxosarcoma.13 Neoplastic cells of myxoma were positively stained for transferrin, ferritin, alpha 1 antitrypsin, S-100 protein, vimentin, and actin. It is a tumor of dual fibroblastic histocytic origin, and cells may be of myofibroblastic origin.14 However, in our present case, IHC examination revealed negative for S-100 protein, suggestive of nonneural origin of the lesion.

Myxomas of the jaw have a recurrence rate of 25 to 43%, which is strongly related to the nature of the lesion; without a sheath (encapsulated) and its potential for secretion of bioactive enzymes, such as hyaluronic acid and acid phosphatase, it contributes to locally infiltrative nature,15 thus making the complete removal difficult, thereby, leading to poor prognosis. As the tumor is not radiosensitive, surgery is the only treatment of choice.16 Corrective management of myxoma involves a radical primary resection despite significant functional and esthetic sequelae. Enucleation or curettage is inadequate treatment as chances of recurrence with aggressive behavior are high. Recurrence can be minimized with extensive partial or total resection procedures; aggressive treatment modality is indicated in the maxilla due to proximity of vital structures.10 Since maxilla was involved in our case, hemimaxillectomy was performed.

CONCLUSION

Myxoma of maxilla is a rare entity with varied clinical and radiographic appearance. Correlation among clinical, radiological, and histologic features is mandatory to arrive at an accurate diagnosis of lesion. Here, we present a very rare case of myxoma involving the maxilla. Complete removal of the tumor, leaving no remnants attached to the soft tissue or bone, should be considered. Whichever surgical approach is chosen, the patient should be
observed over a long term, because of the well-known potential of myxomas to recur. The case showed no evidence of recurrence till date.

REFERENCES