Case Report

Fibromyxomas of the jaw

Priya Shrish Joshi, Sachin Majati[1], Bhagyalaxmi Hongal[2], Neha Agnihotri[2]

Departments of Oral and Maxillofacial Pathology, [1]Oral and Maxillofacial Surgery, [2]Oral Pathology and Microbiology, Vasantdada Patil Dental College and Hospital, Sangli, Maharashtra, India

ABSTRACT

Myxomas can be found in heart, skin, sub-cutaneous tissue, and centrally in the bone but are rare in the oral cavity. Fibromyxoma (FM), the locally invasive benign tumor of the jaw, though rare in the general population, occurs with excessive frequency among young female patients. The tumor develops from the ecto-mesenchymal portion of the tooth germ and shows an inactive effect of nests of odontogenic epithelium on mesenchymal tissue or as a direct myxomatous change in fibrous tissue; hence, it is also called as odontogenic myxoma. The tumor usually occurs in the posterior mandible and constitutes around 3–6% of all odontogenic tumors. Clinical, radiographic, histopathologic, and immunohistochemical investigations play an important role in the accurate diagnosis of FM. A complete surgical excision with long term follow-up is essential for successful management. This clinical study emphasizes the unusual variations in presentation of FM of the jaws.

Key words: Fibromyxoma, intraosseous, mandible, odontogenic tumor

INTRODUCTION

Fibromyxoma (FM) of the jaws is a primary bone tumor of ecto-mesenchymal origin with or without odontogenic epithelium and comprises not more than 2.3–17.7% of all odontogenic tumors.[1] According to World Health Organization, odontogenic myxoma is defined as a locally invasive neoplasm consisting of rounded and angular cells lying in an abundant mucoid stroma.[2] FMs appear to originate from dental papillae, follicle, 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.[3] FMs usually occur in the second and third decades of life with female predilection. Rare lesions have been described in infants and children. They commonly occur in the premolar-molar region of mandible than the maxilla, are usually unilateral and rarely cross the midline. Myxomas can also be found in various other locations such as intramuscular, cutaneous, or intra-articular.[4]

FM is a locally invasive benign neoplasm. It can be designated either as odontogenic FM, in which the myxomatous element predominates; or odontogenic myxofibroma with a predominance of fibrous tissue.[3] Some studies show variation in clinical and radiographic appearance which makes the diagnosis difficult. Hence, the aim of this paper is to highlight the unusual presentations of FM of the jaws.

Address for correspondence: Dr. Priya Shirish Joshi, Flat No B-3, Avani Appartment, 80 Feet Road, Datta Nagar Vishrambag, Sangli, Maharashtra - 416 415, India.
E-mail: sangeetakov@yahoo.co.in

Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.167863

CASE REPORTS

Case report 1

A 23-year-old female patient reported with a chief complaint of swelling in the upper right cheek region since 2 years. Extraoral examination showed an oval shaped well defined swelling extending from right ala of the nose to the zygomatic arch antero-posteriorly and from the infraorbital margin to the alveolar process supero-inferiorly. The overlying skin was stretched and

Cite this article as: Joshi PS, Majati S, Hongal B, Agnihotri N. Fibromyxomas of the jaw. Clin Cancer Investig J 2015;4:737-40.

surrounding tissue appeared normal. On palpation, the swelling was bony hard in consistency, nontender, nonfluctuant, and noncompressible. Intraoral examination revealed a swelling, obliterating the buccal vestibule and palatal vault; extending from right upper canine to the third molar area. All molars were missing. The computed tomography (CT) scan showed an expansile mass in the right maxilla which completely involved the maxillary sinus, lateral nasal cavity and extended up to the inferior orbital margin [Figure 1a]. Radiographic examination of the excised specimen [Figure 1b] showed a multilocular radiolucency with tennis racket pattern [Figure 1c]. Histopathology of incisional biopsy revealed a loose myxoid stroma with stellate shaped cells. Myxomatous areas were interspersed with the dense bundles of collagen fibers and spindle-shaped fibroblasts [Figure 2a-c] suggestive of odontogenic FM. Immunohistochemical (IHC) analysis for alpha-smooth-muscle actin (α-SMA) was strongly positive [Figure 2d]. Hemimaxillectomy was performed and obturator was fabricated to cover the surgical defect.

Case report 2

A 75-year-old female patient reported to the department complaining of swelling in the upper right palatal region since 6 years. The swelling was initially small and gradually increased to its present size of approximately 5 cm × 3.5 cm. The occlusal radiograph revealed an ill-defined mixed radiolucency in the right maxilla with a displacement of adjacent teeth. Histopathologic picture showed myxomatous areas with fibrous connective stroma and a confirmed diagnosis of FM of the maxilla was given. IHC analysis for α-SMA was strongly positive. The patient was treated with an *en bloc* resection of maxilla and obturator was given.

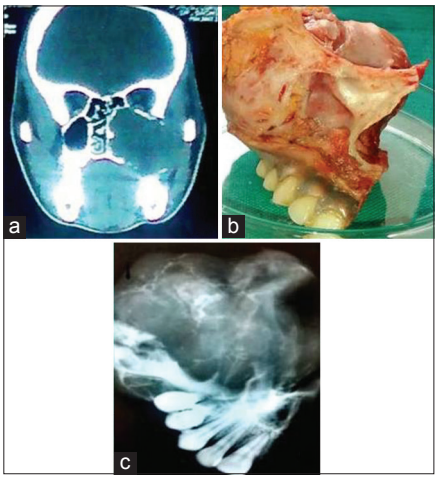

Figure 1: (a) Coronal computed tomography scan showing the destructive expansile lesion in the right maxilla involving the entire maxillary sinus, nasal cavity, and infra-orbital margin. (b) Excised specimen after hemimaxillectomy. (c) Radiograph of the excised specimen showing a multilocular radiolucency with tennis racket pattern

Case report 3

A 50-year-old male patient visited to the department with a chief complaint of swelling in the lower right cheek region extending from midline to the first molar region since 3 years. Intraoral examination revealed an obliteration of vestibular sulcus. Orthopentamograph revealed unilocular radiolucency in the body of the mandible. CT scan showed the expansion of buccal and lingual cortical plate with thinning of the buccal cortex. H and E stained section showed fascicles of collagen fibers arranged in bundles with hyalinization. The fibrous component was admixed with areas of myxomatous tissue showing plump, stellate shaped cells suggestive of odontogenic FM. IHC analysis for α-SMA was strongly positive. Hemi-mandibulectomy was performed.

All patients were on periodic follow-up, no recurrence was noted.

DISCUSSION

FM is a rare locally aggressive intra-osseous, nonmetastasizing tumor of the jaw bones.[5] Rudolf Virchow in 1863 first coined the term myxoma for a group of tumors that had a histologic resemblance to the mucinous substance of the umbilical cord.[1] In 1947, Thoma and Goldman first described myxomas of the jaws.[6] Later in 1964, Marcove *et al*. were the first to report extragnathic fibromyxomas[1].

The tumor develops from the mesenchymal portion of the tooth germ (dental papilla, dental sac).[7] There is an inactive effect of nests of odontogenic epithelium on mesenchymal

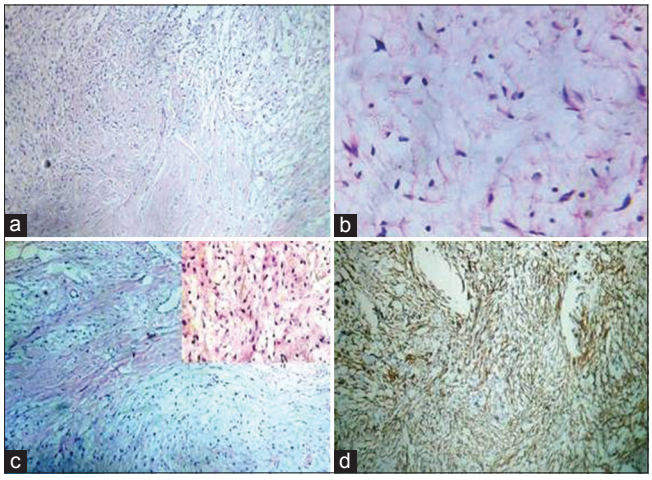

Figure 2: (a) Photomicrograph showing loose myxoid stroma interspersed with dense collagen bundles (H and E, ×10). (b) High power view showing stellate shaped cells in a myxoid stroma (H and E, ×40). (c) Photomicrograph showing fascicles of collagen fibers bundles with hyalinization intermixed with myxomatous areas (H and E, ×10) (Inset) myxoid tissue (H and E, ×40). (d) Strong immune-positivity for alpha-smooth-muscle actin (Immunohistochemical, ×40)

tissue or as a direct myxomatous change in fibrous tissue and hence it is called as odontogenic myxoma.[6]

The prevalence of odontogenic myxoma is 0.04–3.7%. In Asia, Europe and America, relative frequencies between 0.5% and 17.5% have been reported.[3]

FMs mostly occur in second or third decades of life and are predominantly seen in females than males with a ratio of 2:1.[7] But our second case was noted in seventh decades of life which denotes a wide age range variation. FMs are usually located intra-orally in the posterior region of mandible and rarely occur in the maxilla, anterior mandible, or extra-orally.[1] Our first and second case was noted in female patients in posterior maxillary region, but the third case was seen in a male patient and the lesion was extending from midline to the first molar region of mandible. All these features are quite uncommon.

Clinically, myxomas are classified into two forms:

- Facial bone derived which have been subdivided in the past into (a) true osteogenic myxoma and (b) odontogenic myxoma
- Soft tissue derived myxoma (derived from the peri-oral soft tissues, parotid gland, ear, and larynx).

Most of the myxomas are almost always asymptomatic and show an extensive slowly growing swelling with asymmetry of the affected jaw. Lesions are generally painless and ulceration of overlying mucosa may occur.[7] All our cases showed similar presentation.

The cases of FMs in maxilla are more aggressive and have a tendency to involve entire maxillary antrum, the lateral wall of nose or nasal cavity with a displacement of teeth and resorption of tooth roots.[3] Similar features have been reported in two of our cases.

FMs usually present as a unilocular or multilocular radiolucent lesion with well-defined borders, interspersed with fine bony trabeculae giving the appearance of honeycomb, soap bubble, tennis racket, wispy, or spider web. Conventional radiographs present varying radiographic appearances, which are divided into six types: Type I - unilocular; Type II - multilocular (honeycomb, soap bubble, and/or tennis racquet patterns); Type III - involvement of local alveolar bone; Type IV - involvement of the maxillary sinus; Type V - osteolytic destruction, and Type VI - a mixed osteolytic destruction and osteogenesis.[8] In the present case reports, two cases were of Type II while one case was of Type IV appearance. The buccal and lingual cortical plates of mandible may expand occasionally as is described by Kaffe *et al*.[9,10] and our third case showed the same finding.

Histopathological characteristics of the FM include hypocellularity; the presence of stellate or spindle-shaped cells in a loose myxoid extracellular matrix with cells presenting with thin, long cytoplasmic projections, that give the tissue characteristics of immature mesenchyme. The lesions are not encapsulated. Calcification may or may not be present. In our cases, the amount of collagen fibers in the mucoid stroma was more prominent with the absence of odontogenic epithelial rests differentiating it from odontogenic fibroma. Ultrastructural studies of odontogenic myxoma demonstrate basic two types of tumor cells: Secretory and nonsecretory.[3] The secretory cells are considered as principle tumor cells and resemble fibroblast whereas the nonsecretory cells have morphological and functional criteria of a so called myxoblast.

Farman *et al*. have studied the histochemical findings in FMs and have reported 80% hyaluronic acid and 20% chondroidin sulfate in the ground substance. The IHC markers for FM include strong positivity for vimentin, α-SMA, focal S100, and negativity for BCL2, CK19, Ki 67.[3,6] All our cases showed strong immune-positivity for α-SMA.

The differential diagnosis of FMs includes ameloblastoma, odontogenic fibroma, central hemangioma, fibrous dysplasia, odontogenic cyst, aneurysmal bone cyst, central giant cell granuloma, metastatic tumor, well-differentiated liposarcoma, and desmoplastic fibroma. Based on clinical, radiographic, histopathologic, and IHC investigations, all our cases were diagnosed as odontogenic myxoma (FM).

It is stressed that, local excision, curettage, enucleation, radical resection are the treatment options depending on the size and behavior of the tumor. FMs may be infiltrative and aggressive with high recurrence rate and recurrences are generally found in cases which are treated with conservative surgical approach. The recurrence rate of FM ranges from 25% to 43% and have a poor prognosis.[1] To avoid recurrence, all our cases were treated with radical resection and no recurrence was noted on periodic follow-up.

CONCLUSION

Myxomas of head and neck are rare tumors, and they pose a diagnostic and therapeutic challenge. Therefore, the correlation between clinical, radiologic, histopathologic features, and IHC analysis is mandatory to diagnose a lesion. A radical resection with long term follow-up is essential for successful management of the FMs of jaws keeping in mind the high recurrence rate.

Financial support and sponsorship
Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Dietrich EM, Papaemmanouil S, Koloutsos G, Antoniades H, Antoniades K. Odontogenic fibromyxoma of the maxilla: A case report and review of the literature. Case Rep Med 2011;2011:238712.
2. Kramer IR, Pindborg JJ, Shear M. Histological Typing of Odontogenic Tumours. 2nd ed. Berlin: Springer Verlag; 1992. p. 23.
3. Sharma N, Prashant D, Sharma S, Prashant V. Odontogenic myxoma of oral cavity: A case report. Int J Oral Maxillofac Pathol 2013;4:85-9.
4. Weiss SW, Goldblum JR. Enzinger and Weiss's Soft Tissue Tumors. 5th ed. Philadelphia: Mosby Elsevier; 2008. p. 1066-80.
5. Mishra A, Bhatia N, Shukla GK. Fibromyxoma maxilla. Indian J Otolaryngol Head Neck Surg 2004;56:293-5.
6. Gupta I, Keluskar V, Shetti A. Odontogenic fibromyxoma of left maxilla – A case report. J Int Oral Health 2010;2:79-86.
7. Gandhiraj S. Odontogenic fibromyxoma of maxilla, managed conservatively – A rare case report. IOSR J Dent Med Sci 2013;4:15-8.
8. Sharma R, Esht V, Nagrath S, Thapar D. Odontogenic fibromyxoma of maxilla: A rare case report. Int J Dent Med Res 2014;1:42-6.
9. Lu Y, Xuan M, Takata T, Wang C, He Z, Zhou Z, *et al.* Odontogenic tumors. A demographic study of 759 cases in a Chinese population. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1998;86:707-14.
10. Kaffe I, Naor H, Buchner A. Clinical and radiological features of odontogenic myxoma of the jaws. Dentomaxillofac Radiol 1997;26:299-303.