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Case Report

Odontogenic myxoma - A report of two cases

V.R. Chandrababu Pamidi[1], Sridhar Reddy Erugula[2,*], Divya Jahagirdar[3], Gude Venkata Naga Sai Pratap, Vinay Jahagirdar

[1]*GSL Dental College, Rajahmundry, Andhra Pradesh, India*
[2]*Dept. of Oral Pathology and Microbiology, MNR Dental College and Hospital, Sangareddy, Telangana, India*
[3]*Dept. of Oral Pathology, Government Dental College and Hospital, Hyderabad, Telangana, India*

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ABSTRACT

Odontogenic myxoma is a rare intraosseous neoplasm that has the potential for extensive destruction of the jaws. It is thought to be derived from mesenchymal portion of tooth germ. Odontogenic myxoma mainly affects the mandible, with a peak incidence in the second to fourth decades of life and have predilection for the female sex. In this article, we report two cases of odontogenic myxoma in 31-year-old and 35 year old male patients that involved right maxilla and right mandible respectively.

1. Introduction

The term myxoma is derived from greek word 'muxa' which means mucous. Myxoma is a benign primitive connective tissue tumour which is gelatinous in nature. It mainly affects the heart but it can occur in other locations also.[1] When it occurs in jaws, it is referred to as odontogenic myxoma. They account f or 3-6% of odontogenic tumors with incidence rate of 0.07 per million.[2] Odontogenic myxoma (OM) occur in both tooth bearing and non-tooth bearing areas of jaws; they are usually aggressive in nature and often undergo distant metastases. They commonly affect young adults between 25-30yrs with slight female predilection. It a slow growing tumour but in some instances the growth is rapid due to accumulation of ground substance in tumor.[3] In this article, we report two cases of odontogenic myxoma; one in maxilla and one in mandible.

2. Case Report 1

A 31 year old man presented with chief complaint of swelling involving right side of face since 2 years. The swelling was initially small it gradually increased to present size. There was no history of pain or tenderness. Past medical and family histories were uneventful. On extra oral examination, diffuse swelling was seen extending superiorly 1.5cm away from inferior orbital rim, inferiorly till lower border of mandible and medially 2.5 cm away from corner of lip, laterally 0.5 cm away from angle of mandible. On palpation, the swelling was firm, non-fluctuant and no local rise of temperature. Intraoral examination revealed diffuse bosselated swelling over the right posterior alveolar region from maxillary 1st premolar to maxillary tuberosity with obliteration of labial vestibule (Figure 1). Computed tomography was done and CT-axial showed a diffuse radiodense mass extending from alveolus on right side to the infra - orbital rim obliterating the right maxillary antrum and conchae completely. 3D CT showed bony erosion perforating the cortical plates in right maxillary premolar-

* *Corresponding author.*
E-mail address: drerugulasridharreddy@yahoo.com (S. R. Erugula).

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molar area (Figure 2). Fine needle aspiration cytology was negative. Differential diagnosis included odontogenic tumours and malignant neoplasm involving maxillary post erioralveolar ridge extending till sinus. Hemi maxillectomy of right maxilla was done and specimen was sent for histopathological examination. The biopsy specimen was in multiple pieces, creamy whitish to brown in colour and soft in consistency. Histological examination of excised specimen showed tumour mass without encapsulation. It showed spindle cells and stellate cells in an abundant myxomatous and fibrillary stroma interspersed with thin blood vessels (Figure 3). Diagnosis of odontogenic myxoma was given.

3. Case Report 2

A 35 year old male patient presented with a chief complaint of swelling in the right lower back tooth region since one and half year. Swelling was gradual in onset, there was no associated pain. On extra oral examination, milddiffuse swelling about 1.5 x 2.5cm was observed extending superiorly 2.5cm away from zygomatic arch, inferiorly lower border of mandible, med ially 3cm away from corner of lip and laterally 1cm away from anterior border of ramus (Figure 4). On palpation, it was firm in consistency and non-fluctuant. Intra oral examination revealed diffuse swelling involving edentulous alveolar ridge IRT 45, 46 regions. Radiological investigation was done and Orthopantomogram (OPG) showed circumscribed unilocular radiolucency IRT 45, 46 regions. Based on the clinical and radiograph ic features differential diagnosis included residual cyst, ameloblastoma and central giant cell granuloma. Excision of the lesion was done and specimen was sent for histopathological examination. On gross examination, the lesion appeared white in colour with soft gelatinous texture. The haematoxylin and eosin stained soft tissue sections showed connective tissue stroma with loosely arranged collagen bundles intermixed with sparsely arranged spindle shaped and stellate shaped fibroblasts with long cytoplasmic extensions in a myxomatous background. Odontogenic epithelial rests were observed in few areas.

Fig. 1: **(a)** Extra oral showing swelling on the right side of face; **(b)** Intra oral photograph showing buccal and palatal cortical expansion

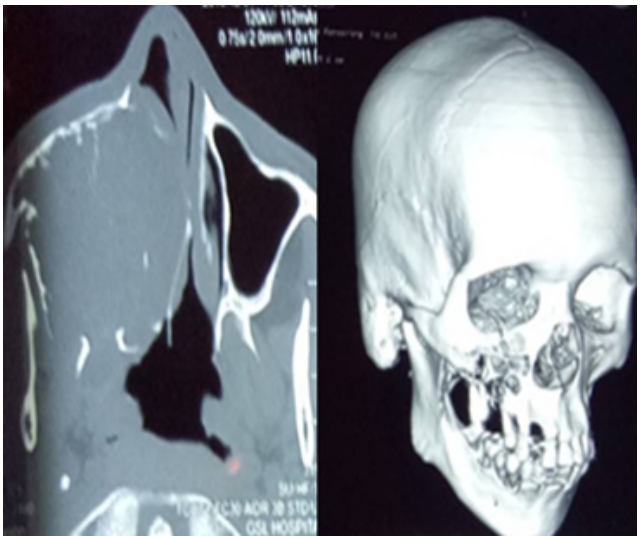

Fig. 2: Computed tomography (CT) and 3d bone CT images showing perforation of cortical plates.

4. Discussion

Odontogenic myxoma is a rare intraosseous slow growing neoplasm which follows aggressive course. The exact etio-pathogenesis of OM is still obscure. There are several theories proposed for the origin of OM which include: (1) origin from primitive odontogenic ectomesenchyme, (2) undifferentiated mesenchyme tissue of periodontal origin, (3) it may be due to myxomatous change of an odontogenic fibroma, (4) fibroblastic - histiocytic origin, (5) myofbroblasic origin.[4,5] Resemblance to pulp ectomesenchyme and o ccurrence in to oth bearing areas support that it s origin from mesenchyme of developing tooth whereas ability of producing collagen (myxofibroma) supports its origin from fibroblasts.[4] Ultrastructural studies reveal the presence of fibroblast like cells (myxoblasts) which have capacity to synthesize mucopoly-saccharides. Protein kinase A regulatory subunit type 1A PRKAR1A mutations are reported and rarely mutations of Gs alpha gene are noted in sporadic cases.[6] The role of epithelial strands in OM still remain unknown, some authors state that these strands induce formation of myxoma cells whereas other consider them as residual rests.

The incidence of OM is equal in both sexes however some studies showed slight female predilection. The tumour

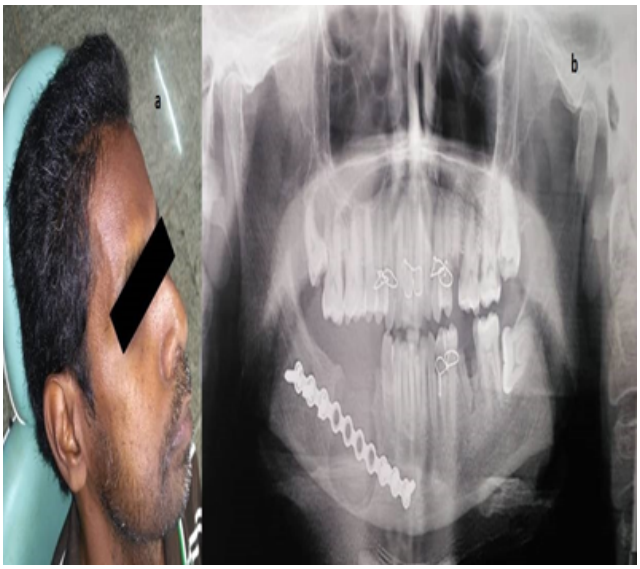

Fig. 3: **(a)** Extra oral photograph showing mild swelling on the right side; **(b)** Orthopantomograph (OPG) showing unilocular radiolucency.

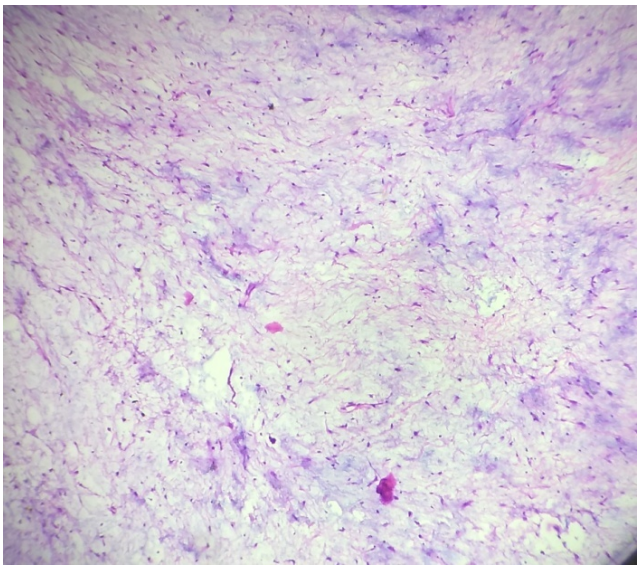

Fig. 4: Photomicrograph of H&E stained sections showing sparsely arranged stellate s haped mesenchymal cells within myxoid stroma (10 X View).

usually affects mandible compared to the maxilla and the most common site of occurrence is premolar-molar region. Peripheral odontogenic myxoma in the gingiva is reported by Raubenheimer et al.[7] According to Lo Muzio et al posterior lesions tend to be larger when compared to other areas.[8] It can cause cortical expansion and displacement of teeth with root resorption is reported in some cases. In the first case we noticed cortical expansion and displacement of teeth. Maxillary lesions tend to infiltrate maxillary sinus and sometimes can cause exophthalmos. Radiologically small lesions (<4cm) may exhibit unilocular radiolucency and larger lesions multilocular radiolucencies. In this case report, OPG revealed unilocular radiolucency in mandibular case and multilocular radiolucency in maxillary case. The multilocular radiolucencies arise due to formation of locules by the intersection of trabeculae at right angles giving a varied radiological appearance which include soap bubble, honeycomb, spider web or tennis racket appearances.[9] Because of the lacy pattern, Eversole called OM as ' Lichen planus of jaw bone.[10] Sometimes mixed radiolucency and radio opacity is seen due to focal calcifications due to residual bone.

Computed tomographic (CT) may reveal an osteolytic expansile lesion with thinning of cortical plates. In the maxillary tumours expansion is to an extent that they tend to fill the maxillary sinus.[11] A well-defined expansile multiloculated mass can be seen on magnetic resonance imaging (MRI).[12] CT and MRI offer greater advantage over conventional radiographs as accuracy of margins and extent of tumours are important in order to avoid incomplete surgical removal.

Gross examination of OM reveals white gelatinous slimy material covered by a layer of bone giving tender coconut appearance or it may appear as a white/gray to yellow solid mass. The tumours will have a poorly defined non - capsulated peripheral margin. Solid tumour usually exhibit multi -nodularity and bone will be frequently found in cut sections.[13]

On histological examination, the tumour is monotonous and may exhibit odontogenic epithelial islands. Connective tissue stroma ranges from loose to fibrous to hyalinised tissue. It is hypocellular or acellular in nature with minimal vascularity. When fibrillar component is more it is called as odontogenic fibromyxoma[14] Stellate cells have long cytoplasmic process which is often anastomozed and with small hyperchromatic nuclei. Care must be taken to differentiate it from myxoneurofibroma, myxoliposarcoma and myxochondrosarcoma. Ultra chemical studies showed the presence of hyaluronic acid, chondroitin sulphate and orosomucoid protein.[15] The stromal expression of matrix metalloproteinases 2 & 9 explain invasive nature of the tumour.[16] Because of gelatinous nature of OM, it infiltrate through thin layers of tissue planes and hides in trabeculae suggesting that the high recurrence rates is due to inadequate removal of lesion rather than inherent biological behaviour. Hence most authors prefer block resection rather than conservative therapy, in first case maxilla is completely resected and in the second case complete excision of tumour along with the borders.[9] Periodical clinical and radiographical monitoring is required as it has high recurrence rates and follow up over atleast two years is mandatory. The present cases are followed up for two years and no recurrence was observed.

5. Conclusion

The confirmatory diagnosis of OM is by histopathological examination. Local invasiveness nature of OM and varied radiographical appearances may result in misdiagnosis which may result in recurrence of tumour.

6. Source of Funding

None.

7. Conflict of Interest

None.

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Author biography

V.R. Chandrababu Pamidi Reader

Sridhar Reddy Erugula Senior Lecturer

Divya Jahagirdar Intern

Gude Venkata Naga Sai Pratap Intern

Vinay Jahagirdar Medical Student

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