

A rare osteolytic lesion in the mandible: A diagnostic dilemma

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Abstract

Odontogenic myxofibroma is a benign tumour that is rarely encountered in any bone other than jaws. It accounts for only 3–11% of all odontogenic tumours. The pathogenesis of these tumours is still controversial and may arise from myxomatous degeneration or fibrous stroma or from the mesenchymal portion of the tooth germ. Clinically, they are slow-growing and can cause gradual expansion of the cortical plates, loosening, displacement of teeth, and rarely root resorption. Biologically, the tumour is locally aggressive with a high recurrence rate, which warrants extensive surgical treatment. Here, we present a case of central odontogenic myxofibroma in a 45-year-old female resulting in the extensive involvement of the mandible within a month.

Keywords: Fibroma, fibromyxoma, mixed tumour, myxofibroma, myxoma, odontogenic tumour

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INTRODUCTION

Odontogenic myxofibroma (MF) is a seldom benign and locally aggressive neoplasm, which was first described by Virchow in 1863. It is considered as the variant of odontogenic myxoma.^[1] Odontogenic myxoma belongs to a rare group of odontogenic neoplasms involving the jawbones. They originate from the embryonic mesenchymal tissue. The debate on the pathogenesis of these tumours continues to entice pathologists until today. The World Health Organisation (WHO) in 2005 classified odontogenic myxoma as tumours of ectomesenchymal origin with or without the odontogenic epithelium.^[2] However, later in 2017, WHO classified odontogenic myxoma under the category of mesenchymal odontogenic tumours as central and peripheral variants. The peripheral variants are

comparatively less aggressive and encapsulated. Contrarily, central odontogenic myxomas are generally non-encapsulated tumours with infiltrative capacity into the adjacent medullary bone.^[3] Histologically, myxofibroma exhibits delicate fibrous to loose mucoid stroma; this is because of the presence of undifferentiated mesenchymal cells showing fibroblastic differentiation in the myxomatous background.^[2] Here, we are presenting a rare case of odontogenic myxofibroma involving the posterior mandibular region in a middle-aged female without any alarming symptoms.

CASE REPORT

A 45-year-old female reported to the Department of Oral and Maxillofacial Surgery with a chief complaint of

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pain and swelling over the right angle of the mandible for 10 days. The pain was initially dull and intermittent, but for the past 3 days, it was sharpshooting and interfering with the mastication. The swelling was reported to be insidious in onset and progressively increased in size. The patient gave a history of extraction of 43, 46, 47, and 48 four years ago. Her past medical history was non-contributory.

On extra-oral examination, ovoid swelling was present on the lower posterior right side of the face, measuring approximately 3 cm × 2 cm, and the overlying skin appeared normal with no pus discharge. On palpation, it was tender, afebrile, and firm to hard in consistency with an expansion of the buccal cortical plate. A tingling sensation was present on the lower half of the right side of the lips, which was not crossing the mid-line. Right sub-mandibular lymph nodes were tender on palpation. They were soft to firm in consistency on palpation.

On intra-oral examination, a well-defined swelling measuring 2 × 1 cm was palpated along with the buccal cortical plate in the mandibular posterior region with slight lingual expansion. It was extending from the right lower first premolar region to the retromolar pad of the mandible [Figure 1]. The swelling was tender and firm to hard in consistency with the overlying mucosa pink in colour. There was no pus discharge and no bleeding.

On radiographic examination, OPG (orthopantomogram) revealed multi-locular radiolucency with a soap bubble appearance involving the right side of the mandible, extending from the apex of the right lateral incisor to the right sigmoid notch [Figure 2]. There was an additional finding in the OPG, which revealed root-canal-treated 44 and 45 and one impacted 13. 43 was missing. Cone beam computed tomography (CBCT) revealed a well-defined osteolytic lesion, which showed multi-locular homogeneously hypodense radiolucency with a radio-opaque sclerotic margin in the right mandibular arch involving 44 and 45. The radiolucency was extending from the right para-symphyseal region until the sigmoid notch involving the body, angle, and ramus of the mandible [Figures 3-7]. In

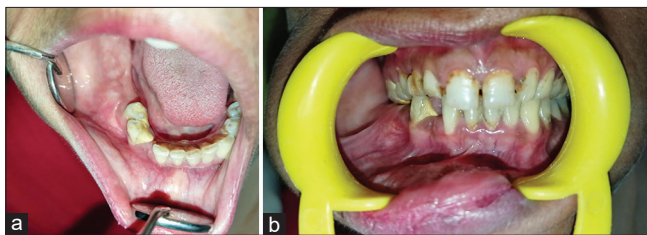


Figure 1: Intra-oral image of the patient showing swelling in the right mandibular buccal vestibule

the coronal view, buccal cortical plate expansion was seen on the right side of the mandible [Figure 4]. Correlating clinically and radiographically, a provisional diagnosis of ameloblastoma was made. Blood investigations were found to be within normal limits.

Fine needle aspiration cytology (FNAC) was performed, but the result was inconclusive. Incisional biopsy was performed and sent for histopathological examination to the Department of Oral Pathology. The Department of Oral Pathology received multiple bits of the soft tissue specimen, measuring 0.9 × 0.8 cm in size, which was soft to firm in consistency. On histopathological examination, haematoxylin and eosin-stained sections revealed fine fibrillar collagen fibres and stellate fibroblasts. At places few foci of odontogenic epithelial rest cells [Figure 8c] were present with clusters of inflammatory cells. Occasional foci of ossification and areas of budding capillaries were also seen [Figure 8a-c]. Based on the histopathology report, a final diagnosis of odontogenic myxofibroma was given.

Because of the aggressive behaviour and destructive nature of the lesion, hemi-mandibulectomy under general anaesthesia of the tumour was performed. One-month follow-up was performed, and the patient was completely asymptomatic.

DISCUSSION

Odontogenic myxofibroma (MF) is a seldom benign intra-osseous neoplasm, which is called 'locally malignant' on account of its exceptionally high local aggressiveness, high recurrence rate, and non-metastasizing nature.^[3] Table 1 highlights the number of cases reported in the literature from the past ten years. On the basis of the data mentioned in Table 1, the mandible is the most common site involved, and marked female predilection can be seen. It is common between the third and the fifth decade, and most of the cases are asymptomatic. On comparing the tabulated data with our present case, we also found similar findings.



Figure 2: OPG reveals large multi-locular lesions in the right mandibular region, extending from the para-symphyseal region to the posterior sigmoid

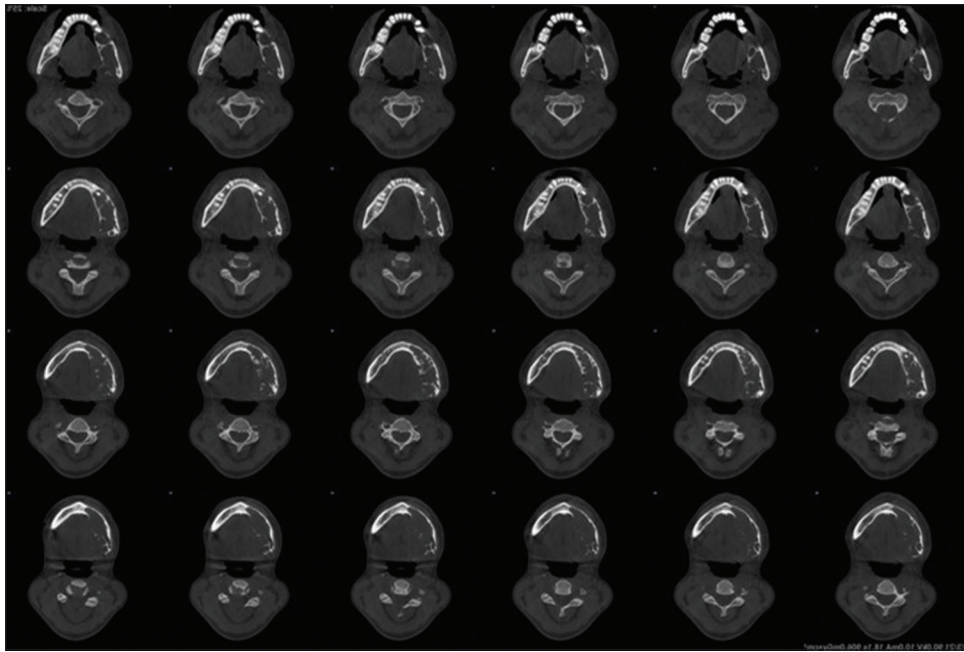


Figure 3: CBCT in the axial plane: A large multi-locular, hypodense region extending from the condylar process to the mandibular right first premolar

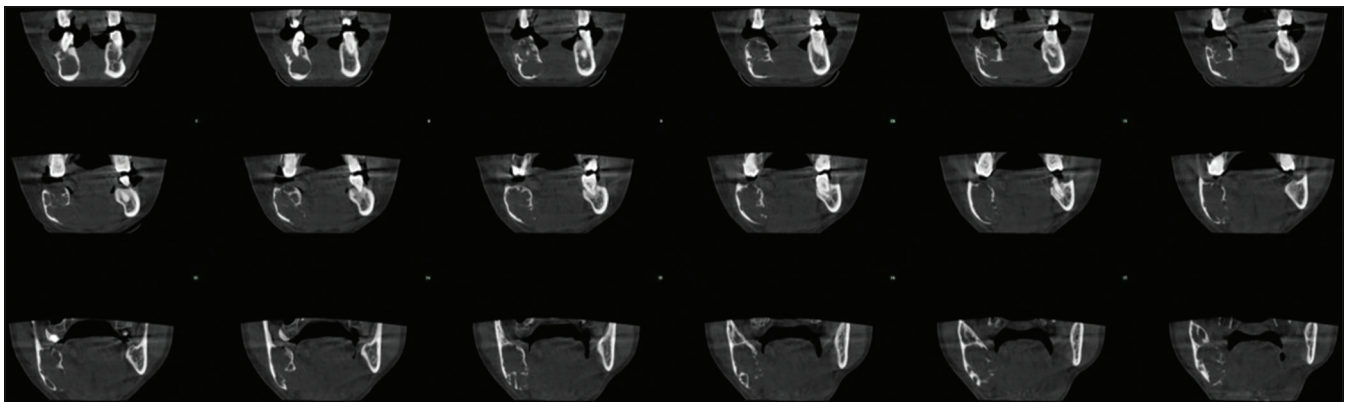


Figure 4: CBCT in the coronal plane: A large multi-locular hypodense region can be seen in the body of the mandible on the right side, with buccal cortical plate expansion

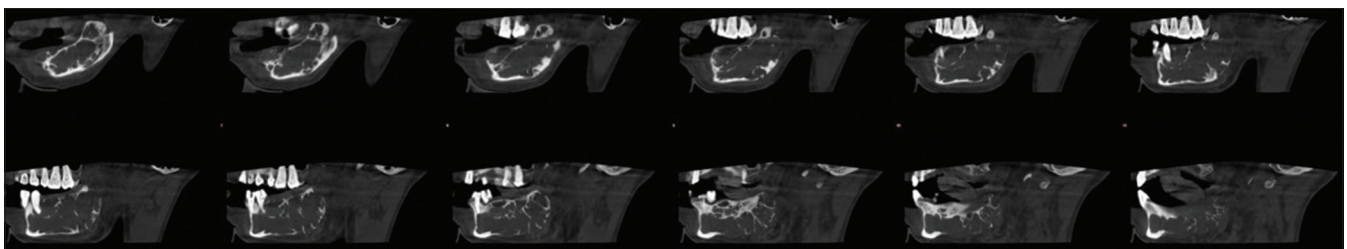


Figure 5: CBCT in the sagittal plane: A large multi-locular hypodense region extending from the right mandibular incisor to the ramus of the mandible

Out of all odontogenic tumours, 2.3% to 17.7% cases of myxoma were reported, among which only a few cases of MF were found. As reported by Meleti M *et al.*^[4] (2015), only 24 specific cases of MF have been reported and described in detail in the English literature since 1950. Approximately 0.05 new cases per million population per year were reported for MF.^[2-4] Most of the cases reported

were diagnosed between the second and fourth decades of life with a peak incidence in the third decade.

The histogenesis of myxoid tumours is still a topic of debate to date. Although Thoma *et al.*^[15] in 1947 theorize that because the myxoma is derived from degeneration of a connective tissue tumour, the myxoma of the jaw

Table 1: Review of literature of odontogenic myxofibroma

Author	Year	Age/Sex	Site Involved	Number of Cases	Clinical Features	Radiological Features	Provisional Diagnosis
Hadidy A <i>et al.</i> ^[20]	2010	15 years/ Male	Mandible	1	Swelling and pain	Multi-locular (honeycomb) radiolucent lesion	TMJ Infection
Infante-Cossio P <i>et al.</i> ^[6]	2011	32 years/ Female	Maxilla	1	Swelling and pain	Multi-locular radiolucent lesion and root resorption	—
Monica Mehendiratta <i>et al.</i> ^[7]	2012	33 years/ Male	Mandible	1	Pain and exophytic growth	Ill-defined unilocular radiolucency	Fibroma
Naresh N <i>et al.</i> ^[8]	2015	63 years/ Female	Mandible	1	Swelling and no pain	OPG showed no bony involvement	—
Sayed F <i>et al.</i> ^[14]	2015	52 years/ Female	Maxilla	1	Swelling and no pain	Large expansile radiolucency without any trabeculations	Ossifying fibroma
Pamolango VT ^[10]	2016	15 years/ Female	Mandible	1	Swelling	Multi-locular radiolucency	Benign odontogenic tumour
SATO Hitoshi <i>et al.</i> ^[9]	2017	8 years/ Male	Mandible	1	Swelling and no pain	Well-defined uni-locular radiolucency	—
Poudel P <i>et al.</i> ^[11]	2016	53 years/ Female	Mandible	1	Swelling and no pain	Multi-locular radiolucency with sclerotic margins	Odontogenic keratocyst
Cankaya Ab <i>et al.</i> ^[15]	2017	39 years/ Female	Maxilla	1	Swelling and no pain	Uni-locular radiolucency and displacement of teeth	—
Avalos N <i>et al.</i> ^[12]	2019	41 years/ Male	Mandible	1	Swelling and no pain	Uni-locular radiolucency and displacement of roots of the involved tooth.	—
Premara JR ^[13]	2020	40 years/ Female	Mandible	1	Swelling and dull pain	A radiolucent lesion with a mixed radiodensity	—
Yilmaz, Yetkin <i>et al.</i> ^[11]	2021	35 years/ Female	Mandible	1	Swelling and pain	Multi-locular radiolucency	—

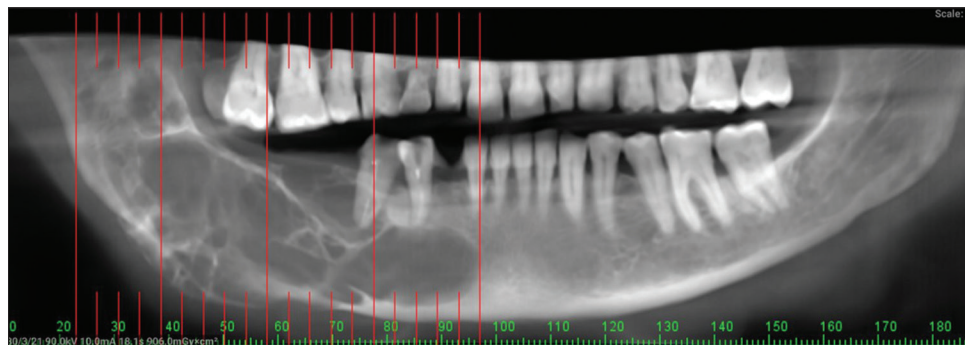


Figure 6: Medium field of view CBCT scan of the mandibular arch reveals a multi-locular hypodense region extending from the right para-symphiseal region until the coronoid process involving the body and ramus of the mandible

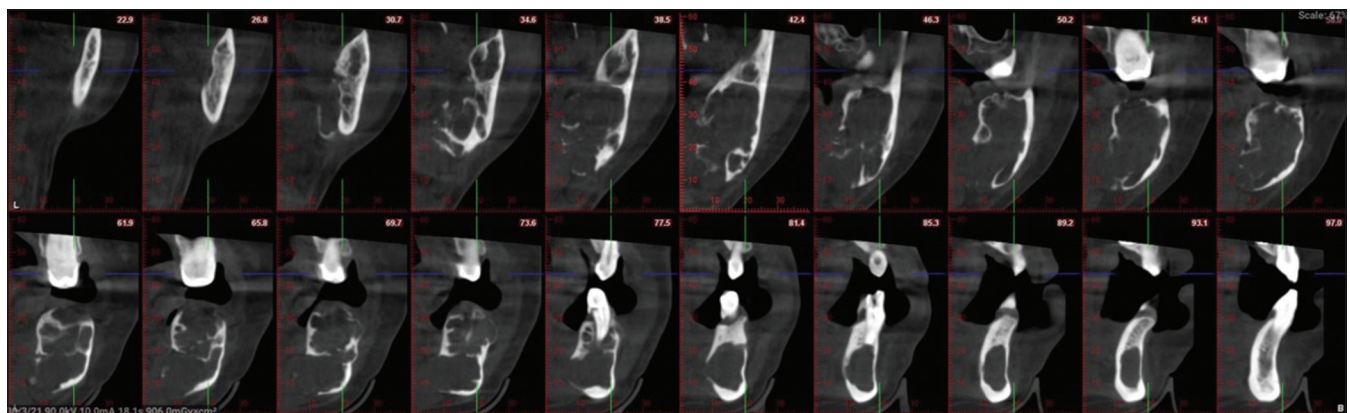


Figure 7: CBCT in the cross-section plane: Multi-radicular radiolucency can be seen extending from the mandibular right lateral incisor to the ramus of the mandible

is, in reality, an odontogenic fibroma that has undergone myxomatous degeneration. Bruce and Royer, however,

believe that the tumour can arise by the proliferation of mesenchymal rests within the alveolar bone. They

postulated that it is possible for embryonic mesenchymal rests located in this area to proliferate and form a myxofibroma.^[15]

Myxomas have two different sites of occurrence: (1) facial bone: myxomas derived from the facial bone are further divided into true osteogenic myxoma and odontogenic myxomas and (2) soft-tissue myxomas of the larynx, parotid, and ear.^[16] Clinically, patients afflicted with a myxofibroma generally comprise a painless and slowly enlarging expansion of the jaw as in our case. However, the pain might be there because of the involvement of neural structures. The premolar and molar regions of the mandible are the most common sites of occurrence for MF. Patients with posteriorly located tumours had a late diagnosis and bigger lesions when compared to those with anteriorly located tumours because disfigurement gets noticed first in the anterior portion. In the present case, the tumour was locally aggressive, involving more than half of the mandible in a very short span of time, which made this case unique.

Radiologically, MF can mimic various lesions of the jaw, which often leads to a mis-diagnosis. They should be differentiated from ameloblastoma, central giant-cell granulomas, and intra-osseous haemangiomas, odontogenic keratocysts, aneurysmal bone cysts,^[17] as mentioned in Table 2. Usually, the compartments of ameloblastoma are round-like rather than the square or triangular spaces of odontogenic fibromas. Compared with MF, the margin and the cortical boundary of ameloblastoma are more

corticated, and ameloblastoma does not invade soft tissues or interrupt the cortical continuity. The differentiating features of MF include the following points: (1) fine and straight septa separating the lesion into triangular, square, or rectangular spaces, which appear as “tennis racket” or “honeycomb” patterns; (2) septa that are frequently scattered to the borders of lesions and appeared perpendicular to the margins; (3) MF that was noted to tend to involve the alveolar process, scallop between the roots, and affect the integrity of the alveolar ridge; and (4) the cortex of MF that appeared normally perforated and the edge of the cortex expanded into the soft tissue.^[17]

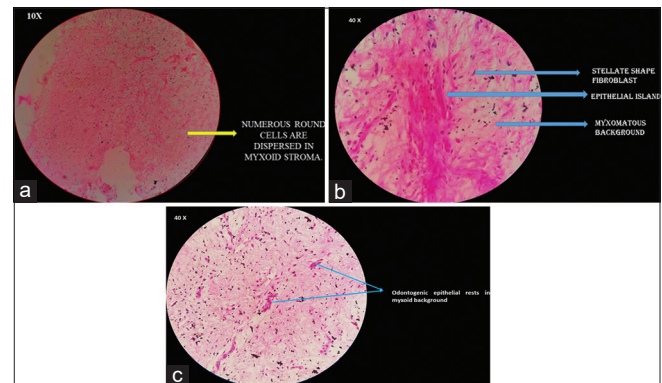


Figure 8: (a) Haematoxylin and eosin-stained tissue section showing fine fibrillar collagen fibres and round fibroblasts with a few inflammatory cells. (b) Haematoxylin and eosin-stained tissue section showing fine collagen fibres, the epithelial island at one focus, and stellate fibroblasts in the myxomatous background. (c) Haematoxylin and eosin-stained tissue section showing odontogenic epithelial rests in the myxoid background at 40 X magnification

Table 2: Differential diagnosis of odontogenic myxoma^[3]

	clinical features	radiological feature	histopathology
ameloblastoma	Painless intra-oral swelling because of expansion of cortical plates. Eggshell crackling/crepitus sound.	Large uni-locular or multi-locular radiolucency (soap bubble appearance)	Odontogenic epithelial islands which consist of tall columnar ameloblast-like cells, with nuclei of reverse polarity. Micro-cysts are present within the epithelial islands.
central giant cell granuloma	Common in the anterior mandible crossing the mid-line.	Small uni-locular lesion, with non-corticated margins.	Numerous multi-nucleated giant cells within the mesenchymal stroma
intra-osseous haemangioma	Rare lesion Intra-oral swelling and numbness in the posterior region of the jaw.	Multi-locular radiolucent defect, with small loculations (honeycomb appearance). Large malformations may show a “sunburst appearance”.	Proliferation of numerous plump endothelial cells. Well-defined vascular spaces are evident.
odontogenic keratocyst	Swelling Expansion of cortical plates. Parathesia of the lower lip.	Small solitary Uni-locular to large multi-locular radiolucency. Septa present are coarse and curved.	Corrugated para-keratinized/ortho-keratinized luminal epithelium and prominent basal layer.
aneurysmal bone cyst	Rare lesion An asymptomatic expansile lesion in the condyle and coronoid. Pathologic fracture. Perforation of the cortex.	Uni-locular radiolucency, radiopaque, or mixed. Root resorption is common.	Multiple sinusoidal blood-filled spaces. Multi-nucleated giant cells are present. Osteoid formation.

Histopathologically, the MF consists of a huge amount of inter-cellular substance rich in acid mucopolysaccharides and made up of fibroblasts and myofibroblasts dispersed in loose myxomatous connective tissues. A few areas may show mild pleomorphism that does not correlate with the rate of recurrence of these tumours.^[16] The amount of collagen and mucoid material determines whether it can be called myxofibroma (MF) or fibromyxoma.^[15]

Odontogenic myxomas express three varieties of cells as follows: spindle cells, stellate cells, and hyaline cells. Stellate cells are actively positive for transferrin, alpha 1-AT, S-100 protein, and vimentin. Hyaline cells combine with alpha 1-ACT and alpha 1-AT.^[18]

Treatment options may vary from a conservative approach to enucleation of the lesion or curettage of the cavity to radical surgery. Recurrence is usually observed during the first 2 years after the first treatment. MF shows a recurrence rate between 25% and 43%.^[19,20]

CONCLUSION

MF is a benign, locally aggressive, painless, slow-growing tumour with a high recurrence rate. Based on clinical and radiological features, they are often mis-interpreted. Therefore, it is highly recommended that the patient must undergo radiographic investigation and biopsy to improve the outcome of treatment. To avoid recurrence, in-depth histopathology knowledge about the tumour of myxomatous origin is recommended.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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