CASE REPORT

Central odontogenic fibromyxoma of mandible: an aggressive odontogenic pathology

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SUMMARY

Myxoma of the jaws which was first described by Thoma and Goldman in 1947 is a rare neoplasm and its rate of prevalence and incidence is not available. Myxoma term, according to 1992 WHO classification, is used along with odontogenic myxoma (OM) and myxofibroma. There are two forms of myxomas or fibromyxomas that are recognised in head and neck region: one is derived from the facial skeleton and the other is derived from the soft tissue. Most of the OM are located intraosseously, but peripheral ones are also recognised. OM behaves differently from myxomatous tumours of long bones, which recur more often and may transform into malignancy. A majority of these lesions occur between 2nd and 4th decade. In the pathogenesis of OM, dental papilla, dental follicle and periodontal ligament tissues have been implicated as possible 'germ centres'. This case describes an uncommon finding of central odontogenic fibromyxoma, throwing light on its epidemiology, clinical, histopathology, molecular and treatment aspects.

BACKGROUND

Central odontogenic fibromyxoma is a non-existing aggressive pathology which extends beyond the radiological limits of the main tumour and it is infiltrative in nature. A handful of cases are given in the literature. Here, we would like to share its out-and-out features with the oral and maxillofacial fraternity through this journal.

CASE PRESENTATION

A male patient aged 15 years reported with swelling on the right lower and middle third region of face extending until the midline of 1-year duration. The swelling was associated with mild, dull non-radiating pain which was relieved on occasional medications. There was a history of being hit by a basketball on the right side of the jaw about 1½ years. On extraoral examination (figures 1 and 2), a single diffuse swelling was found, measuring ~6×5 cm extending from the midline of the face to the right ramus of the mandible. The swelling was soft, non-fluctuant and non-tender on palpation.

Intraoral examination revealed (figure 3) a well-localised swelling extending from 42 to 48 teeth which was obliterating buccal and lingual vestibule. Correlating with clinical findings, a provisional diagnosis of ameloblastoma was given and after ruling out FNAC findings, clinical differential diagnosis as mentioned below was given:

- ▶ giant cell lesion,
- ▶ central odontogenic fibroma.

CT scan investigation (figures 4–7) revealed a large cystic osteolytic lesion causing breach in the cortex. Findings were suggestive of neoplastic aetiology. Incisional biopsy was advised to ascertain the nature of pathology, which revealed myxoma.

All preoperative investigations were made and the patient was deemed fit for surgery. The patient was operated under general anaesthesia and segmental resection of the right side of the mandible was performed followed by free fibula graft.

The pathological segmental resected specimen obtained (figure 8) was soft gelatinous in consistency and grey-white to tan-yellow in colour with irregular borders.

Histopathology of the specimen (figure 9) revealed connective tissue stroma composed of haphazardly arranged, stellate, spindle-shaped and round cells in abundant loose myxoid stroma that contained numerous collagen fibrils. Figure 10 reveals small islands of inactive-appearing odontogenic epithelial rest scattered in the myxoid ground substance. Correlating with clinical and radiological findings, final diagnosis of central odontogenic fibromyxoma was made. Apart from this, special staining with Alcian blue showed strong positivity (figure 11) of myxoid stroma, and immunohistochemistry with BCL-2 (figure 12) showed mild to moderate staining of the stroma and odontogenic islands.

During the postoperative period of 1, 3 and 6 months and 1 year, follow-ups were uneventful.

INVESTIGATIONS

- ▶ FNAC to rule out cystic/vascular lesion.
- ▶ CT sections (axial, coronal and sagittal).
- ▶ Haematological examination (haemoglobin: 17 g%, BT: 2 min, CT 5 min and random blood sugar 130 mg/dL).
- ▶ For better definition and extension of intrabony and soft tissue, it was judged by different 3D reconstructions.
- ▶ To ascertain the exact nature of pathology, histopathological examination was advised, which revealed characteristic features of central odontogenic fibromyxoma.
- ▶ Special staining with Alcian blue revealed positive staining of myxoid stroma, which is very much present in myxoma tumours.
- ▶ As the given pathology was aggressive involving the soft tissue along with eroding the entire right section of the mandible, immunohistochemistry with BCL2 marker revealed positivity in the stroma and odontogenic islands.

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Figure 1 Extraoral swelling revealing huge swelling present on the right side of the mandible extending until the left portion of the chin.

Figure 2 Profile view revealing swelling present on the right lower and middle third of face.

Figure 3 Intraoral swelling revealing a well-localised swelling extending from 42 to 48 teeth which was obliterating buccal and lingual vestibule.

DIFFERENTIAL DIAGNOSIS

A working diagnosis of ameloblastoma was made because:

- Unicystic ameloblastoma, a variety of ameloblastoma, is commonly seen in the second decade of life as suspected in our case.

Clinical differential diagnosis: After non-productive fine needle aspiration biopsy, irrespective of unerupted wisdom tooth, cystic lesions were ruled out and giant cell lesion, central odontogenic fibroma as a clinical differential diagnosis was given.

Radiological differential diagnoses as mentioned below were given:

- Conventional ameloblastoma commonly gives multilocular soap-bubble appearance as seen in this case.
- Central giant cell granuloma presents as multilocular radiolucency.
- Central haemangioma gives multilocular radiolucency.
- Central odontogenic fibroma presents as multilocular radiolucency.
- Malignancy shows breach in the cortex of bone and infiltration in the surrounding soft tissue as seen in the present case.
- Odontogenic myxoma (OM) commonly gives multilocular radiolucency as seen in the present case.

Histological differential diagnosis: An enlarged dental follicle with myxoid changes or the dental papilla closely resembles OM. Peripheral odontoblastic bordering of cells present in enlarged follicle is in contrast to odontogenic myxoma tumours.

Myxomatous degeneration is seen particularly in fast-growing tumours like fibrosarcomas, chondrosarcomas and liposarcomas as seen in the present case.

After correlating with history, clinical, radiological findings, grossing and confirming histopathologically, central odontogenic fibromyxoma as a final diagnosis was given.

TREATMENT

Decision of treatment was based on the radiographical and clinical findings, which revealed complete intrabony nature with significant soft tissue counterpart. Radiological findings showed expansile soap bubble lytic septated cystic lesion with breach in the adjoining cortex. As there was significant infiltration in the adjoining structures, incisional biopsy confirmed myxoma and ruled out malignancy. Surgical intervention in the form of segmental resection with 1–2 cm safe margin was performed on the right side of the mandible which resulted in the complete resolution of the pathology. Success of the treatment was assessed by

Figure 4 Sagittal CT sections revealing multilocular lesion showing bony and soft tissue invasion.

the complete resolution of the lesion. Postoperatively during the follow-up period of 1, 3 and 6 months showed no recurrence and the patient was asymptomatic.

OUTCOME AND FOLLOW-UP

The patient is undergoing regular follow-up after uneventful recovery and removal of sutures. The patient showed good healing without any scar (figure 13). So far, there is no recurrence.

DISCUSSION

OM was first described in the year 1947 by Thoma and Goldman as rare benign tumour of the tooth-bearing areas of

the jaw bones.¹ The myxoma (fibromyxoma), which is a benign neoplasm of uncertain histogenesis, has a characteristic histological appearance and often behaves in a locally aggressive, infiltrating fashion.² Under the WHO classification of odontogenic neoplasms, myxoma comes under 'odontogenic ectomesenchyme with or without included odontogenic epithelium'.³

The prevalence of OM is generally between 0.04% and 3.7%.⁴ In Asia, Europe and the USA, relative frequencies between 0.5% and 17.7% have been reported.⁵

Age at the time of diagnosis ranged from 1 to 73 years, with the mean age of 30 years; 75% of cases were detected between the second and fourth decade commonly affecting the mandible compared with the maxilla.⁶ It very rarely occurs in the paediatric age group,⁷ but in the present case, it appeared in a boy aged

Figure 5 Axial CT sections revealing multilocular lesion causing breach in the cortex of bone and soft tissue infiltration. Pathology is extending from the canine to third molar region.

Figure 6 Coronal CT sections revealing multilocular lesion showing bony and soft tissue invasion.

15 years affecting the mandibular molar region. According to Kaffe *et al*'s⁸ study, female to male ratio of OM was 1.5:1. Female preponderance among gender is the most common feature reported in several studies.^{9–11}

Radiologically, OM has a multilocular trabecular pattern typifies as 'honey comb', 'soap bubble', 'tennis racket', wispy and spiderweb in appearance.¹² The present case showed soap-bubble appearance with breach in the cortex of bone and infiltration in the surrounding soft tissue.

Most OM are asymptomatic, although some patients present with progressive pain in lesions involving the maxilla and maxillary sinus, with eventual neurological disturbances.¹³

The WHO 1992 classification and the present authors describe OM clinically, histologically as invasive neoplasm consisting of rounded and angular cells lying in abundant mucoid stroma.¹⁴ In the given case, all the characteristic features of OM and numerous loose collagen fibres were present which diagnosed it as central odontogenic fibromyxoma.

Figure 8 Pathological resected specimen measuring ~9.5×6.5 cm, greyish white to tan yellow in colour, gelatinous soft in consistency with irregular borders.

There are many epicentres which are implicated for the pathogenesis of OM like dental papilla, dental follicle and periodontal tissues as given by Adekeye *et al*. The given epicentres for pathogenesis are still a dilemma for origin and yet to be proved because some studies signify that OM stroma has no resemblance to dental follicle and it may be a dual fibroblast-histiocytic origin.¹⁵ This duality arises because of the hypothetical origin of fibroblasts to produce mucopolysaccharides which mimic myxoid tumours histopathologically.¹⁶ This theory is still uncertain whether a familiar situation of 'chicken and egg' for histogenesis of these tumours. Table 1 depicts various molecular markers and their implications in OM.

Owing to high recurrence rate after conservative treatment and considerable debate on its appropriate treatment, complete surgical resection is the standard approach for the management of this tumour.^{18 19} An extensive search of the literature on the management of these tumours showed 45.5% of mandibular cases were treated surgically by enucleation, curettage or en bloc

Figure 7 Three-dimensional reconstruction revealing intrabony and soft tissue component on the right side of the mandible.

Figure 9 H&E-stained section 10× photomicrograph revealed connective tissue stroma composed of haphazardly arranged, stellate, spindle-shaped and round cells in abundant loose myxoid stroma that contained numerous collagen fibrils.

Figure 10 H&E-stained section 40× photomicrograph revealed small islands of inactive-appearing odontogenic epithelial rest scattered in the myxoid ground substance.

Figure 11 Alcian blue special staining revealed strong positivity of myxoid stroma.

Figure 12 Immunohistochemistry with BCL-2 marker revealed mild to moderate positivity in odontogenic islands and surrounding stroma.

Figure 13 Postoperative photograph revealing good healing.

Figure 14 Postoperative OPG revealing free fibula graft reinforced with plating.

resection.^{1 9 18–20} Liquid nitrogen therapy eliminates the neoplastic cells by devitalising the bone and hence is considered to be a better alternative to radical resections. The healthy vital bone is formed as due to the presence of inorganic substances in the vicinity of the healthy tissue.²¹ These tumours are radioresistant and hence radiotherapy is never a choice of treatment.²²

Maxillary lesions are also surgically enucleated and preferably intraoral Caldwell-Luc approach is recommended along with a prosthetic maxillary obturator for rehabilitation.¹¹ The rate of recurrences is attributed to the ability of the tumour to infiltrate into the surrounding bone and the approach of treatment should be radical rather than conservative. Incomplete removal

Table 1 Immunohistochemical markers for odontogenic myxoma¹⁷

IHC markers	Implication and expression
MIX-1	Positivity due to mesenchymal odontogenic origin
VIMENTIN	Positive in mesenchymal cells
S-100	Positive
ACTIN	Positive
Collagen land VI, tenascin, fibronectin, proteoglycans	Positive and expressed in matrix component
AGNOR	Positive
Ki-67	Low proliferation index
MMP-2 and BCL-2	Positive

of the tumour is responsible for recurrence more than the biological behaviour. The rate of recurrence was reported to be 25% when the tumour is treated conservatively.²³ Boffano *et al*²⁴ suggested that lesions of size >3 cm are considered for radical resections and bloc resections and tumours of lesser diameter are better treated by enucleation or curettage. There should always be a follow-up period of 2 years postsurgically which represents the maximum activity of the tumour recurrence and a period of 5 years recommended for surgical healing with periodic check-up and review.^{9 10 20 22 25} In the present case, segmental resection of the right side of the mandible followed by free fibula graft reinforced with plating was carried out and no recurrence was observed during 1-year follow-up.

Patient's perspective

I came to this hospital with my father because of huge swelling in my right side of jaw. After getting operated, I am completely alright. I am in constant touch with my doctor.

Learning points

- Patients with odontogenic myxoma owing to gelatinous nature of tumour and absence of capsule have a strong propensity of recurrence, so long-term follow-up for early recurrence is always warranted.
- Being an aggressive pathology, prognosis should always be assessed by molecular IHC markers.
- There is scarcity of odontogenic fibromyxoma cases. Life of clinician becomes difficult because of its behaviour, treatment planning and complicated prognosis.

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