CASE REPORT

Odontogenic myxoma in a 52-year-old woman

Hari Ram,¹ Gagan Mehta,¹ Manoj Kumar,² Parveen Lone³¹Department of Oral and Maxillofacial Surgery, King George's Medical University, Lucknow, Uttar Pradesh, India²Department of Radiodiagnosis, Chhatrapati Shahuji Maharaj Medical University, Lucknow, Uttar Pradesh, India³Government Dental College, Jammu, Jammu & Kashmir, India**Correspondence to**
Dr Hari Ram,
hariram81@yahoo.com

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SUMMARY

Odontogenic myxoma (OM) is a rare benign but locally aggressive tumour of the jaws. It is usually seen in the second to third decade of life. Women are more frequently affected than men and it has more predilections for the mandible rather than the maxilla. OM presents as an asymptomatic swelling in most of the cases. Owing to the non-capsulated and aggressive nature of OM, a high rate of recurrence has been reported. Here we present a case of OM in a 52-year-old woman managed by segmental mandibulectomy. Sign of recurrence was seen after 18 months of follow-up.

BACKGROUND

Odontogenic myxoma (OM) is a benign but locally aggressive and non-metastasising neoplasm of the jaws.¹ The term myxoma was used by Virchow in 1871 to describe tumours with histological features similar to the mucinous tissue of the umbilical cord.² Odontogenic origin of OM is supported by its exclusive occurrence in close proximity to tooth bearing areas of the jaw and histological similarity to the stellate reticulum of a developing tooth.³ Most frequently, OM occurs in the second to third decade of life. Children below the age of 10 years and adults over 50 years of age are rarely affected.^{4 5} OM is more common in women, male-to-female ratio is 1:2.⁶ There is a strong predilection for the mandible as compared with maxilla, ratio being 2.5:1.⁶ OM presents as an asymptomatic progressive swelling without pain and paresthesia in most of the cases. The treatment of choice is surgical excision.⁷

CASE PRESENTATION

A 52-year-old woman presented to the department of oral and maxillofacial surgery with symptoms of progressively increasing swelling over the left side of the face. On systemic examination, the patient was found to be healthy. History revealed development of an asymptomatic swelling in lower jaw since 1 year. It gradually caused loosening of ipsilateral molar teeth. She got her molars extracted 2 months back, after which there was a sudden increase in swelling involving the left body of mandible.

Extraoral examination revealed a swelling of about 5×4 cm over the left body of mandible extending anteroposteriorly from the left corner of mouth to 2 cm anterior to the angle of mandible and superoinferiorly from the level of tragus to the lower border of mandible. Overlying skin was normal in colour and texture. On palpation it was firm and hard in consistency (figure 1).

On intraoral examination a bilobed swelling was seen measuring about 4×4.5 cm extending from

Figure 1 Extraoral view showing swelling at the left side of the face.

buccal mucosa to lateral border of tongue and canine to retromolar triangle. Few capillary growths were seen on the lingual lobe of swelling. Swelling was firm in consistency and non-tender. The mucosa overlying the area of the lesion was of the same colour and texture as the surrounding mucosa (figure 2).

Figure 2 Intraoral view showing bilobed growth on the mandible.

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Figure 3 Histopathology of the lesion.

INVESTIGATIONS

Incisional biopsy was performed and histopathological examination revealed sheets of loose stellate cells with long branching cytoplasmic processes. The stroma was myxoid comprising of cells varying from slender, spindle shaped to plump and oval cells. Scattered occasional island and cords of odontogenic epithelium were also evident in the stroma ([figure 3](#)). Orthopantomogram (OPG) showed destruction of bone on the left side of mandible crossing the midline ([figure 4](#)).

CT scan of the face revealed an expansile destructive soft tissue attenuation measuring about 4.3×4.5×5.0 cm in size with multiple calcifications involving predominantly the left half of mandible. Anteriorly, the lesion was seen involving the subcutaneous planes with extension up to the mentum. Posteriorly, the lesion was extending up to the angle of mandible with the involvement of the lower part of ramus of mandible. Medially, the lesion was seen involving the mylohyoid muscle and abutting the genioglossus muscle pushing it medially. The lesion was seen involving an inferior alveolar process in the region of premolars and molars ([figures 5 and 6](#)).

DIFFERENTIAL DIAGNOSIS

When a clinician encounters a typical multilocular radiolucency, the lesion like:

- ▶ ameloblastoma,
- ▶ odontogenic fibroma,
- ▶ central haemangioma,
- ▶ central giant cell granuloma,
- ▶ chondromyxoid fibroma,
- ▶ rhabdomyosarcoma, should be included in the differential diagnosis.

Figure 4 Orthopantomogram showing bone destruction at the left side.

The honey-comb variant of myxoma usually shows fine trabeculations within small lobules, which are not present in ameloblastoma.

The giant cell granuloma occurs most commonly in the anterior region of the jaws, whereas myxoma is seen most frequently in the ramus and the premolar–molar area of the mandible.

In case of central haemangioma a pumping tooth (a tooth that can be pushed apically and then rebounds to its original position) or localised gingival bleeding around a tooth is usually encountered which is not seen in myxoma. Also easy aspiration of a large quantity of blood is seen in central haemangioma but not in myxoma.

TREATMENT

Segmental resection of mandible was performed, extending from the right canine to the left half of ramus with clear margins of approximately 1.5 cm on each side of the lesion ([figure 7](#)). Following the resection, a 2.4 mm reconstruction plate was adjusted and fixed with screws on each side of the defect ([figures 8 and 9](#)). Histopathology of excised specimen was again consistent with OM.

OUTCOME AND FOLLOW-UP

The patient was under regular follow-up and signs of recurrence were seen after 18 months of surgery. The patient was planned for second surgery but she did not turn up ([figure 10](#)).

DISCUSSION

There has been a lot of controversy about the origin of myxomatous neoplasms. Myxoma of the jaw is considered a tumour of odontogenic origin.⁸ Histological resemblance to dental mesenchyme, especially the dental papilla and the presence of a sparse amount of odontogenic epithelium and association with an unerupted tooth or a developmentally absent tooth supports its odontogenic origin.⁹ Stout¹⁰ redefined histological features of OM and defined them as a true mesenchymal tumour.

OM is a locally invasive and non-encapsulated tumour which does not metastasise and exhibits slow and asymptomatic expansion,¹¹ but perforation of the cortical borders of the affected jaw may be noted in few cases.¹² OM occurs in individuals of second to third decade of life.^{4 5} According to Kaffe *et al*¹³ the mean age of the patients at the time of diagnosis was 29.6 years. The mandible is more commonly affected than the maxilla in a ratio of 66.4:33.6%. Mostly it is seen in the molar and premolar regions.¹⁴ Women are affected more than men.⁶

OM of the mandible often causes bony expansion that leads to loosening of teeth, root resorption, malocclusion, ill-fitting dentures and facial deformity. In advanced cases there may be pain, paresthesia and anaesthesia over the affected region. Maxillary lesions cause bony expansion or extension into the maxillary sinus.¹³ In advanced cases, nasal obstruction or exophthalmos may be seen.¹³ Aetiology of myxoma is unknown. Myxomas are usually slow-growing neoplasms; local trauma like tooth extraction, blunt injury, complication of a Le Fort I osteotomy during secondary cleft palate repair has been shown to increase their rate of growth.²

Tumour is seen as a radiolucent unilocular or multilocular lesion in the radiograph, described as a honeycomb, soap bubble or tennis racket lesion^{15 16} that often displaces teeth or sometimes resorbs roots of teeth.¹³ Occasionally, myxomas may exhibit features of an ameloblastoma or a central giant cell granuloma.¹⁷

Histologically, myxoma consists of loosely arranged spindle shaped and stellate cells with ovoid hyperchromatic nuclei and

Figure 5 CT showing the coronal view of the lesion.

Figure 6 CT showing the axial view of the lesion.

Figure 7 Excised specimen.

Figure 8 Intraoperative photograph.

scant cytoplasm, many of which have long fibrillar processes that tend to intermesh. The ground substance in OM is mucinous in nature with tiny capillaries and occasional presence of collagen strands.^{2 17 18} Binucleated cells or atypical nuclei are frequently seen. Pleomorphism, prominent nucleoli, mitotic figures and multinucleation are rarely seen.^{6 17}

Different treatment modalities like curettage, curettage with peripheral ostectomy, segmental resection, block resection or en bloc resections have been described in the literature. Treatment of OM depends on site, size and biological behaviour of the lesion. Conservative management of myxomas by lesion excision and curettage with liquid nitrogen cryotherapy have also been reported.¹¹

Reconstruction of bony defects less than 5 cm can be done with buccal fat pad⁷ in maxilla or using corticocancellous iliac crest bone graft in case of mandible.¹⁹ Larger defects (more than 5 cm) usually require primary prosthetic reconstruction followed by a final obturator.²⁰ Mandibular lesions can be reconstructed with a reconstruction plate followed by vascularised fibular free flap,²¹ iliac crest graft,²² costochondral graft or scapular osteocutaneous free flap.²³ Owing to the high recurrence rates of OM delayed reconstruction is the treatment of choice.¹ We performed segmental mandibulectomy with clear margins of 1.5 cm and reconstruction was performed with reconstruction plate.

Figure 9 Postoperative orthopantomogram showing reconstruction with plate.

The main cause of recurrence is the incomplete removal of the tumour, therefore, the lesion should be excised with adequate disease free margins. Recurrence rate of 10–33% has been reported when the tumour boundaries are poorly defined,^{12 16} whereas simple enucleation and curettage alone can have recurrence rates up to 25%.^{24 25} Owing to the high rate of recurrence we excised with safe margins of 1.5 cm but signs of recurrence were seen after 18 months of surgery.

Though OM shows aggressive local growth but malignant transformation or metastasis has not been reported.²⁵ We report this case of OM which is a comparatively rare entity and was seen in a patient of above 50 years which is unusual for this age group.

Figure 10 Postoperative photograph of the patient.

Learning points

- ▶ Odontogenic myxoma (OM) occurs most commonly between the second and third decade and less common in the first decade and beyond the third decade. In our case it was in a 52-year-old woman.
- ▶ While diagnosing OM, the most frequently occurring multilocular lesions including ameloblastoma, central haemangioma and giant cell granuloma must be considered and should be ruled out.
- ▶ Although benign but OM is a non-encapsulated and locally aggressive lesion and a high rate of recurrence has been reported. So a long-term follow-up is necessary.

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