CASE REPORT

Ameloblastoma: an aggressive lesion of the mandible

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SUMMARY

Ameloblastoma is a benign locally invasive epithelial odontogenic tumour comprising 1% of all tumours and cysts arising in the jaws. It is commonly found in the third and fourth decade in the molar ramus region of the mandible. Among all types of ameloblastoma, multicystic ameloblastoma is believed to be locally aggressive lesion that has the tendency for recurrence. In this report we present a large multicystic ameloblastoma in the left body-ramus region of the mandible in a 55-year-old woman. This large lesion was diagnosed with the help of CT and was successfully managed by hemimandibulectomy with simultaneous reconstruction using iliac crest bone.

BACKGROUND

Ameloblastoma is the most common benign odontogenic tumour of the jaws that constitutes about 1% of all cysts and tumours of the jaws.^{1,2} It is generally a painless, slow growing, locally aggressive tumour causing expansion of the cortical bone, perforation of the lingual or the buccal cortical plate and infiltration of the soft tissues. It has peak incidence in third and fourth decade of life but can be found in any age group with equal gender predilection (1:1).¹⁻⁵ The relative frequency of mandible to maxilla is reported to be varying from 80–20% to 99–1%. In the mandible majority of ameloblastomas are found in the molar ramus region.^{1,3}

In a conventional radiograph, ameloblastoma can present as either unilocular or multilocular corticated radiolucency; the bony septae results in a honey comb or soap bubble appearance, or tennis racket pattern. In some places, cortical plates are spared and expanded where as in other region they are destroyed; root resorption is a common finding.⁶ Buccal and lingual cortical plate expansion is more common in ameloblastoma than in other tumours.⁷

Conventional radiograph is sufficient for small mandibular lesions but maxillary lesions and extensive lesions require CT and MRI to establish the extent of the lesion.⁷

The challenge in managing ameloblastoma is in achieving complete excision and reconstruction of the defect when the tumour is large.¹ Ameloblastoma is treated by enucleation, curettage or surgical excision depending on size and type of the lesion. The rate of recurrence ranges from 17.7% for en bloc resection to 34.7% for conservative therapy. Wide resections with a safety margin of healthy bone to prevent local recurrence were preferred.²

This report gives a comprehensive knowledge regarding the epidemiology, classification, occurrence, behaviour, diagnosis and treatment outcome of ameloblastoma along with the presentation of the case.

CASE PRESENTATION

A 55-year-old woman reported with a swelling (figure 1) on the left side of the face since 2 years and pain while chewing food since 3 months. The swelling was insidious in onset and gradually increased to the present size. There was no history of trauma or toothache or decrease in the size of the swelling or any discharge from the swelling. The patient was experiencing pain while chewing hard food. The patient also had altered sensation over the left cheek region. She was a known pan chewer. On examination, there was a solitary ill-defined diffuse swelling over the left middle and lower third of the face (figure 1) measuring about 5×8 cm extending superoinferiorly from the left pretragal region to the lower border of the mandible and mediolaterally 1 cm from the left corner of the mouth to the left lateral border of the mandible. The surface was smooth and the skin overlying the swelling was stretched and was of normal colour with no secondary changes to be found. It was non-tender and hard to the palpating fingers.

An intraoral examination revealed an ill-defined solitary swelling (figure 2) in the left lower posterior buccal vestibule extending anterioposteriorly from 34 to the retromolar region and mediolaterally 1.5 cm from the buccal surface of the molars

Figure 1 Extraoral photograph of the patient, revealing a diffuse swelling over the left side of the face.

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Figure 2 Intraoral photograph revealing a diffuse swelling from tooth 34 to retromolar region.

to 1 cm lingual to molars with smooth surface and mucosa overlying was stretched and similar to adjacent mucosal colour with no secondary changes to be found. It was non-tender and hard in consistency with buccal and lingual cortical plate expansion. There were clinically missing teeth (37 and 38). Considering the clinical findings, a tentative diagnosis of benign tumour of the left side of lower jaw was made. Ameloblastoma was thought as first in the list of differential diagnosis as it is the most commonly occurring tumour in the mandibular molar ramus region in this age group. Second, odontogenic myxoma was considered, which has similar site of occurrence. An incisional biopsy was made and the specimen was subjected to histopathological examination.

INVESTIGATIONS

The patient was subjected to radiographic and routine haematological examination. The haematological findings were not significant.

COMPUTED TOMOGRAPHY

The axial CT of the jaw (figures 3 and 4) revealed a very large well-defined radiolucent expansile lesion in the left body and

Figure 3 Plain axial CT revealing an expansile radiolucent lesion in the left body ramus region of the mandible causing expansion of the ramus.

Figure 4 Axial CT with contrast revealing a multilocular radiolucent lesion involving the left body and ramus of mandible.

ramus of the mandible with multilocular appearance causing expansion of the body and ramus.

CHEST RADIOGRAPH

The screening chest radiograph (figure 5) did not show any evidence of metastasis.

HISTOPATHOLOGY

The histopathological examination of the biopsy specimen revealed (figure 6) the presence of follicles that consist of a core of loosely arranged angular cells resembling the stellate reticulum of an enamel organ. A single layer of tall columnar ameloblast-like cells had been found surrounding the central core. The nuclei of these cells were located at the opposite pole to the basement membrane (reversed polarity) with few follicles revealing, microcyst formation. These findings were strongly suggestive of follicular ameloblastoma.

RADIOLOGICAL DIFFERENTIAL DIAGNOSIS

Odontogenic keratocyst, odontogenic myxoma, central giant cell granuloma and Brown's tumour of hyperparathyroidism.

Figure 5 Plain chest radiograph showing no evidence of metastasis.

Figure 6 Photomicrograph revealing, follicles which consists of a core of loosely arranged angular cells resembling the stellate reticulum of an enamel organ (A). A single layer of tall columnar ameloblast like cells seen surrounding this central core. The nuclei of these cells are located at the opposite pole to the basement membrane (reversed polarity) (B) with few follicles showing microcyst formation (C).

TREATMENT

As the lesion was very extensive, a hemimandibulectomy (figure 7) was performed along with reconstruction using iliac crest bone (figure 8).

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful. The patient's aesthetics and function was restored. The patient was followed up for 6 months with no evidence of complication or recurrence (figure 9). Currently the patient is under biannual follow-up.

DISCUSSION

Ameloblastoma is a benign epithelial odontogenic tumour often aggressive and destructive with the capacity to erode bone and invade adjacent structures.⁴ Ameloblastoma of the lower jaw can progress to variable sizes (1–16 cm) and cause facial asymmetry, displacement of teeth, malocclusion and pathological fractures.¹ In the present case also the patient's clinical examination revealed a large hard swelling in the ascending ramus and molar region of the mandible which had caused the facial asymmetry, loose teeth and expansion of the buccal as well as the lingual cortical plate.

Aetiology

Ameloblastoma arises from epithelial cellular elements and dental tissues in their various phases of development.¹

Figure 7 Surgically excised specimen (hemimandible).

Figure 8 Intraoperative photograph showing placement of bone graft which is secured to the other half of the mandible.

Epidemiology and prevalence

Although ameloblastomas occur with equal frequency in both genders they are found slightly in higher frequency in women than men⁴ as seen in our case.

In the ameloblastoma of the mandible, the majority (70%) are located in the molar ramus region and 10–15% are found in association with an unerupted tooth.⁴ In the present case also ameloblastoma was found in the molar ramus region and was not associated with any unerupted tooth.

Classification

Ameloblastoma are classified as either extraosseous (peripheral) or intraosseous. Peripheral ameloblastomas presents as either sessile or pedunculated, slow growing mass that is confined to the gingiva or alveolar mucosa, without involving the underlying bone.⁶ Intraosseous ameloblastomas of the jaws are further

Figure 9 Follow-up photograph of the patient after 6 months.

classified as unicystic, desmoplastic, mixed cystic and solid types.⁷ The mixed cystic and solid forms demonstrate more aggressive behaviour and are known for recurrence.⁶ The histopathological variants are the follicular and plexiform types, followed by the acanthomatous and granular cell types.⁴ Uncommon variants are desmoplastic, basal cell, clear cell ameloblastoma, keratoameloblastoma and papiliferous ameloblastoma.⁸ Among all these variants, plexiform pattern is less aggressive with low recurrence.⁹

Clinical presentation

Ameloblastoma presents as a painless, slow growing hard mass¹ and in our case also, it was painless and hard swelling which took about 2 years before the patient had developed symptoms. Other clinical presentations of the disease were pain or anaesthesia of the affected area. In the present case also the patient had paraesthesia over the left cheek region.

Becelli *et al*¹⁰ observed that, in mandibular ameloblastoma half of the patients presented with swelling of the affected region (38.3%), paraesthesia of the innervated region of the mandibular nerve (13.3%) and altered occlusion of teeth in 10% of cases.

Radiographic features

Radiographically, ameloblastoma appear as radiolucent lesion that may have either unilocular or multilocular appearance with or without cortical plate expansion.¹

Treatment

For reporting ameloblastomas, it seems acceptable to group the treatment regimens into three modalities being conservative that includes enucleation and curettage, marsupialisation and radical surgery which includes resection with or without continuity defect.¹¹

For solid-multicystic ameloblastoma of the mandible, a resection of the jaw should be approximately 1.5–2 cm beyond the radiological limit, in order to ensure that all the 'microcysts' and 'daughter cysts' are removed.³ In our case also marginal clearance of 2 cm was achieved as histopathology of specimen revealed it to be follicular variety.

There are different methods of mandibular reconstruction of large defect with microvascular surgery using donor site from fibula, iliac crest, scapula and radial forearm.¹² In our case as well tumour was treated with hemimandibulectomy as it was quite an extensive lesion involving body as well as the ramus of the mandible. Hemimandibulectomy, simultaneously with reconstruction using iliac crest bone reduces the morbidity while retaining the aesthetics of the patient.

Outcome and recurrence

The chance of recurrence seems to be more dependent on the method of surgical treatment.¹¹ In general, annual follow-up for at least 10 years is recommended. Few authors have recommended annual follow-up for 5 years and thereafter once in every 2 years till 25 years.¹¹ Our patient is also under follow-up with no evidence of recurrence.

To conclude, ameloblastoma is the most commonly occurring odontogenic tumour in the mandibular body ramus region in the middle age group, yet other differential diagnosis such as

odontogenic keratocyst, odontogenic myxoma, central giant cell granuloma has to be ruled out with the help of advanced radio-diagnostic tools. The best treatment of ameloblastoma is aggressive en bloc resection with simultaneous reconstruction.

Learning points

- ▶ Ameloblastoma should always be considered first in the list of differential diagnosis for any swelling in the mandibular posterior region in the middle age group.
- ▶ Conventional radiographs are sufficient for small lesions but large lesions require advanced imaging such as CT for better surgical management of the lesion.
- ▶ Multilocular/solid ameloblastoma has the highest recurrence among all types of ameloblastoma. Hence wide resection of the lesion with a safety margin of healthy bone will prevent recurrence of such lesion.
- ▶ Resection with simultaneous reconstruction using various grafts will restore aesthetics of the patient as well as function of the jaw.

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Competing interests None.

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