

Ameloblastoma—A Locally Destructive and Invasive Tumour—A case report.



Medical Science

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ABSTRACT

The ameloblastoma is a rare odontogenic tumour . It occurs exclusively in the jaws, with a strong predilection for the posterior region of the mandible. Although benign, the ameloblastoma is a destructive tumour, locally invasive and presents a high rate of recurrence despite adequate surgical removal. A thorough understanding of its clinico-pathological behaviour is essential to avoid recurrence associated with inadequately treated disease. Currently wide resection and immediate reconstruction are the treatment of choice. We present a 45-year-old female patient with an ameloblastoma mandible who was treated with complete resection of a mandibular segment. Reconstruction, carried out during the same surgical procedure, was performed using an free fibular bone graft fixed with titanium plates and screws. The advantages of this procedure include recurrence risk reduction due to segmental resection, reliable mandibular reconstruction and less surgical procedures, allowing full rehabilitation within a shorter period of time.

Case report

45-year-old normotensive, non-diabetic, non-smoker non-tobacco chewer female presented to Gujarat Cancer and Research Institute Ahmedabad with a proliferative lesion in central lower alveolus since last 2 years. The lesion has been progressively increasing in size and was associated with loosening and displacement of teeth. She also complained about difficulty in chewing and closing her mouth.

On examination there was an ulcero-proliferative growth involving lower central segment of mandible up to angle bilaterally. The tumour was involving the floor of mouth anteriorly and lower gingivo-buccal sulcus with loosening and displacement of lower teeth. Multiple neck nodes were enlarged bilaterally. Orthopantomogram was done which showed that growth was confined to lower jaw and was bony in origin. CECT face with 3D reconstruction showed a 51x91x86 mm large lytic expansile mass involving body of mandible, symphysis menti with displacement of teeth with necrotic areas within . Widening of mandible was seen which suggests possibility of primary bone tumour. Incisional biopsy from tumour was suggestive of ameloblastoma.



Wide local excision of the mandible (segmental mandibulectomy), floor of mouth was done. Reconstruction was done with fibular free graft. Post operative recovery was uneventful.

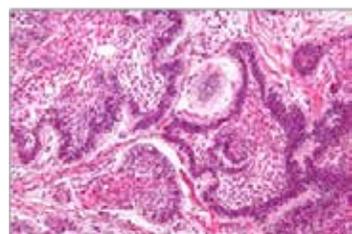


Fig:Micrograph of an ameloblastoma showing the characteristic nuclear palisading and stellate reticulum. H&E stain

Discussion

Ameloblastoma(Adamantinoma) is an uncommon tumour of the mandible and maxilla. It is a locally aggressive tumour that, if neglected, can reach an enormous size and cause severe facial disfigurement and functional impairment. It occurs with equal frequency in both sexes and has its peak incidence in the third to fourth decade of life. Overall, 80% of all ameloblastomas occur in the mandible and 20% in the maxilla.

According to the 2005 classification of tumours of the World Health Organization, the variants of ameloblastomas are the solid/multicystic, the extra osseous ameloblastoma, the desmoplastic and the unicystic type. Multicystic ameloblastoma is the most common variant, making up 85% of all ameloblastomas. It has a propensity to be more aggressive and is associated with a higher recurrence when compared with the other two forms.

Ameloblastomas may be relatively asymptomatic and can be detected incidentally on radiological imaging. When symptoms are present, patients often complain of a slow-growing, painless swelling as in our case report . Less commonly, they may present with pain or paraesthesia loose teeth, or malocclusion. Facial deformity, if present, may range from very mild to severe in delayed presentations. They may also be associated with the presence of unerupted teeth, in particular the mandibular third molar.

They are believed to arise either from embryonic remnants of the epithelial lining of an odontogenic cyst, dental lamina or enamel organ, stratified squamous epithelium of the oral cavity, or displaced epithelial remnants. Its pathogenesis remains unclear .

Radiological investigations provide a useful aid to diagnosis. However, these findings are not pathognomonic and must therefore be confirmed with histological examination. The orthopantomogram (OPG) is a useful first- line investigation and may reveal lucency in the bone of varying size and shape associated with scalloped margins and resorption of the roots of involved teeth . Occasionally it can be a well-demarcated unilocular lesion,

whereas often it presents as multiloculated expansile lucencies with a so called “soap bubble” appearance . Computed tomography (CT) may be useful in the assessment of the extent of the tumour, cortical destruction and extension into the neighbouring soft tissues.

They are rarely considered to exhibit metastatic spread. Metastatic ameloblastoma refers to lesions that metastasize despite having the typical benign histology of ameloblastoma in both the primary and metastatic lesions. Ameloblastic carcinoma, on the other hand, refers to lesions that demonstrate histological features of both ameloblastoma and carcinoma. In contrast to ameloblastoma, ameloblastic carcinoma presents more aggressive clinical features, such as rapid growth, perforation of the cortex, and painful swelling. In most cases, radiographic findings show ill defined radiolucency. However, focal radio opacity may be detected in radiolucent lesions. Ameloblastic carcinoma may metastasize to the regional lymph nodes or lung. In rare cases, brain or multiple bone metastasis have been reported.

Ameloblastoma poses a challenge for all involved not only adequate resection of the tumour but also a functional and aesthetically acceptable reconstruction of the residual defect. Wide resection and immediate reconstruction is currently accepted as the treatment of choice in most cases and are associated with high rate of recurrence if inadequately treated. Treatment choice depends on some factors. Multilocular ameloblastomas have higher recurrence rates than unilocular ones. Age is another important factor when considering treatment options. Ameloblas-

tomas tend to infiltrate bone trabecula of the cancellous bone on the lesions periphery, before a true bone resorption becomes radiologically evident. Therefore, the true tumor margin, often times, goes beyond the apparent clinical or radiographic margin.

Many advocate a safety margin of at least 1cm beyond the tumor radiographic limits. Others advocate segmentary resection or en bloc resection, which allows for total tumor removal and lower recurrence rates. The disadvantage of the segmentary resection is the resulting facial deformity and function loss if not properly rebuilt. In these cases, it is necessary to use grafts of flaps with bone tissue, besides implants and sophisticated surgical techniques with multidisciplinary teams. The reconstruction mode to be employed depends mainly on the defect size. Mandibular segments larger than five centimeters treated with bone grafts tend to have a higher rate of post-operative complications. Such defects must be preferably rebuilt with micro-surgical flaps from the fibula or iliac crest. Another alternative for large defects is osteogenic distraction.

Patient prognosis is difficult to assign, because of the rarity of well documented follow-up information.

Conclusion

We believe that immediate reconstruction after an en bloc resection with safety margins is the best alternative to treat ameloblastomas, since it brings about total disease removal and patient cosmetic and functional rehabilitation in the same surgical procedure

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