

Case Report

Giant unicystic ameloblastoma of maxilla- successful endoscopic management

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ABSTRACT

This 32-year-old male patient presented with complaints of restricted mouth opening and gross facial asymmetry owing to the massive jaw swelling on the right side. After a thorough examination, a diagnosis of unicystic ameloblastoma of maxilla. Although, the standard of care surgical approach was through Weber-Fergusson incision, a concerted attempt to excise the tumour with the minimally invasive endoscopic route was made successfully. The patient was symptom free now with complete resolution of swelling on a regular follow up post operatively. This case report highlighted the modern day protocol of organ preservation concepts in surgical management of a rare tumour in otolaryngology practice.

Keywords: Ameloblastoma, Maxilla, Denker's, Endonasal approach, Unicystic, Facial asymmetry, Infra temporal fossa

INTRODUCTION

Ameloblastoma is a locally aggressive benign tumor affecting the mandible and less commonly the maxilla. It arises from either remnant odontogenic epithelium within the bone or from odontogenic cysts or from the epithelium of the sinonasal tract. Ameloblastomas constitute the second most common neoplasm of odontogenic origin. As per literature, solid ameloblastomas rarely affects maxillary bone.¹⁻³ Maxillary ameloblastomas secondarily invade through the nasal and paranasal sinuses.

Solid ameloblastomas exhibit a persistent and slow growth. They are non-encapsulated and infiltrate into marrow spaces with pseudopods. This leads to frequent recurrence of the tumour following surgical removal. Long term follow up is therefore imperative because this lesion has been shown to recur even up to 30 years following primary treatment.⁴

The aim of this uncommon case report was to highlight a case of unicystic solid ameloblastoma arising from the maxilla, which was infrequently encountered in clinical otolaryngology practice.

CASE REPORT

A 32 year old male presented with a gradually progressive right sided facial swelling in the last one year. Clinical examination revealed an approximately 6×5 cms, diffuse, firm, non-tender swelling over right side of face with skin over the swelling being normal with normal sensations preserved along with grade I trismus. The swelling was palpable intraorally in the right upper gingivo-labial sulcus. Ocular and dental examination did not reveal any clinical abnormality (Figure 1).

A diagnostic nasal endoscopy showed a fullness of the lateral wall of the right nasal cavity.



Figure 1: Preoperative picture of the swelling over right maxillary region.

A CT scan of the paranasal sinuses with 3D reconstruction demonstrated an expansile lytic lesion measuring 5 x 4.3 x 5.4 cm centred within the right maxillary sinus, with the bony origin from the anterolateral wall and the floor of right maxilla. The right maxillary sinus was found to be thinned out due to bony remodelling and dehiscence anterolaterally with extension of the mass into the soft tissues of premaxillary region. There was involvement of superior alveolar process due to its extension inferiorly with resorption and blunting of roots of premolar and molar teeth. Superior, posterior and medial walls of the maxillary sinus were thinned out with extension into the infratemporal fossa (Fig.2).

A biopsy of the lesion was done through a sublabial approach. Histopathological examination showed odontogenic epithelial islands composed of peripheral palisading columnar cells with keratin and stellate reticulum like cells. These findings were suggestive of an ameloblastoma.

A modified Denker's endoscopic extended trans nasal surgical approach with an additional sublabial access to the infratemporal fossa was performed to excise the tumour completely along with wide margin of clearance(Fig 2).Total ethmoidectomy was performed. Fovea ethmoidalis was found to be free of the tumor. Since the mass had destroyed the lateral nasal wall, subsequent excision of the necrotic bone was done . Coblation system was used for careful delineation of the mass from the infratemporal fossa. Blood loss was minimal during the procedure. There was no destruction of the posterior wall of the maxillary sinus or erosion of teeth along floor of maxillary sinus. Orbital floor and floor of maxillary sinus were preserved. Surgical challenge was mainly the access to reach disease free margins all round the lesion. However , with the use of

high speed skull base drill system and angled endoscopes, excision was complete. Intra operative blood loss was minimal too.

Histopathologic examination of the excised tissue specimen confirmed the diagnosis of ameloblastoma (Fig 3).

Post-operative period was uneventful. Clinical examination confirmed intact vision, normal extraocular movements, and normal sensation over the cheek and near normal facial symmetry (Fig 4).

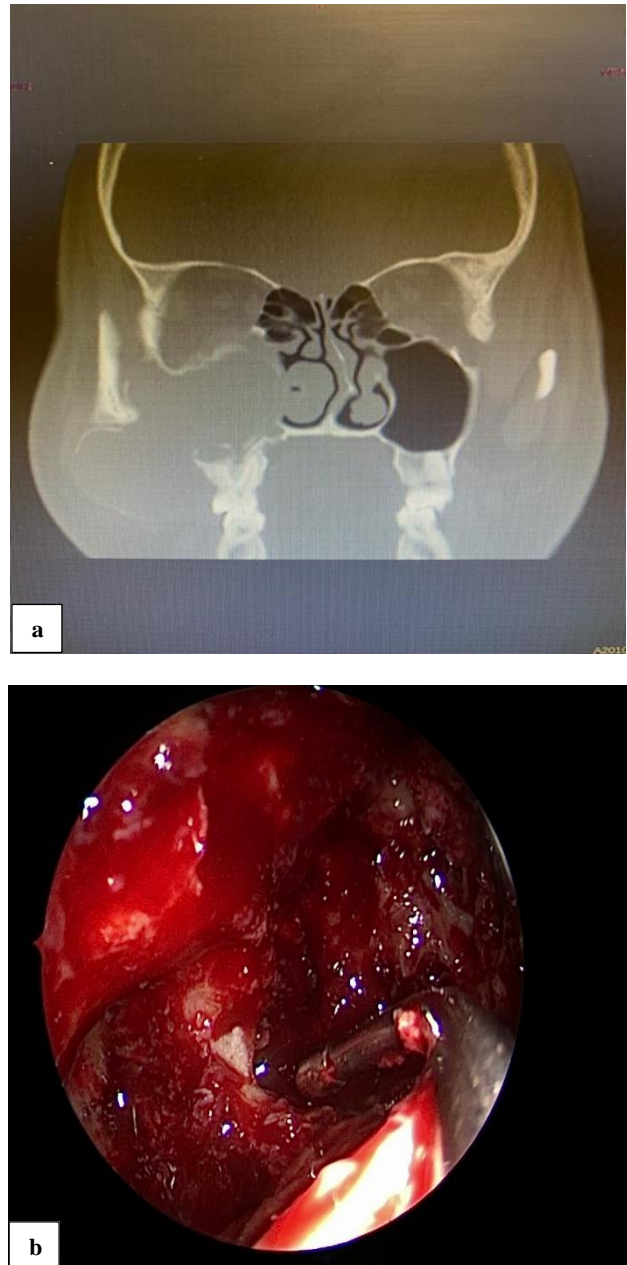


Figure 2: (a) Coronal computed tomogram showing the lesion within the right maxilla antrum; (b) intra operative picture showing dissection from right infratemporal fossa.

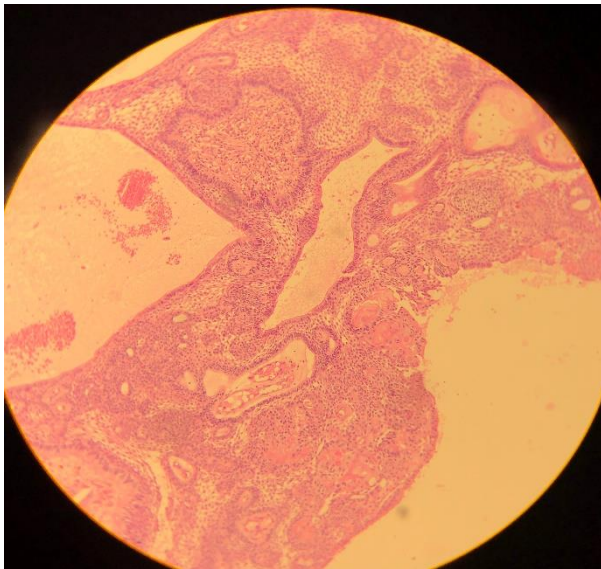


Figure 3: HPE demonstrating odontogenic epithelial islands (ameloblastoma).



Figure 4: Pre and Postoperative views.

Review CT scan was done 3 months postoperatively and this showed no evidence of recurrent or residual lesion. The patient is being reviewed periodically. On last review which was 3 months after surgery, the patient continued to be free of recurrence and symptoms.

DISCUSSION

Sinonasal ameloblastomas are rare tumors comprising less than 1% of all sinonasal tract tumors.⁵

Ameloblastomas of maxilla presented approximately 15 to 25 years later than those occurring in the mandible.^{5,6} Ameloblastomas were present during third or fourth

decade of life. Sinonasal ameloblastomas have been suggested to originate from the pluripotent cells of the basal layer of the sinonasal epithelium.⁷ Chronic inflammation may lead to neoplastic transformation of the retained odontogenic epithelium and hence to the formation of an ameloblastoma.⁸

Maxillary ameloblastomas presented as a painless, slow growing mass. Other clinical features included nasal obstruction, facial enlargement, swellings of the cheek, gingiva or hard palate.⁹ Extension into the paranasal sinuses, orbit, nasopharynx or skull base occurred due to the lack of thick cortical bone. This can cause delay in the diagnosis of the disease.¹⁰ Abundant vascularity of the maxilla helped in the dissemination of the disease. Patients with unilocular ameloblastoma had a better prognosis than those with multilocular disease.¹¹ Patients presenting with pain over the swelling may be due to secondary infection.¹²

The differential diagnosis included radicular cyst, odontogenic keratocyst, adenomatoid odontogenic tumor, ameloblastic fibroma, craniopharyngioma, odontogenic myxoma and glandular odontogenic cyst. Histopathological confirmation was required for the definite diagnosis. MRI and CT scans were routinely done for all sinonasal lesions.

In 2005, WHO classified ameloblastomas into solid/multicystic (conventional); extra-osseous/peripheral; desmoplastic and unicystic.¹³ The solid/multicystic ameloblastoma can histopathologically be divided into a follicular and a plexiform type. The plexiform pattern was the most common pattern in sinonasal ameloblastomas. Follicular type can be further subdivided into a spindle cell type, an acanthomatous type, a granular type and a basal cell type.¹³ The plexiform type contained basal cells arranged in anastomosing strands of epithelium with peripheral columnar cells demonstrating reverse polarity of their nuclei. Hyperchromasia of the nuclei and basal cytoplasmic vacuolization were commonly identified features. Loosely arranged, angulated cells with a stellate reticulum-like component were present between the epithelial strands. This component was less conspicuous in the plexiform type.⁵

A number of treatment modalities were available like wide excision, curettage, enucleation, cryotherapy, laser surgery, radiotherapy and chemotherapy.⁹ Enucleation of the lesion can lead to recurrence, possibly because of insufficient removal. Ideally 1-3 cms of surrounding healthy bone was to be resected as advocated by most surgeons. Regular follow up was required once a year for the first five years and in cases of maxillary involvement, a follow up period of at least 10 years was recommended.⁷

Endoscopic management in recent times had resulted in less radical surgery, decreased morbidity and better tumor

control. In this case a combined endoscopic and external approach resulted in complete excision due to better visualisation.

CONCLUSION

Timely assessment with help of imaging studies and biopsy help in early diagnosis and treatment planning of ameloblastomas. Histopathological diagnosis is needed for confirmation of diagnosis. Regular follow up is needed to rule out recurrence, rarity of maxillary ameloblastoma in modern medicine, relatively young age group affliction, radical extended endonasal surgery as standard of care and no specific aetiological factors.

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