A case of unicystic ameloblastoma of maxillary sinus: A rare case report

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Ameloblastoma is a slowly growing, locally invasive tumor that primarily originates from enamel tissue and odontogenic epithelium. On the brain magnetic resonance imaging of a 34-year-old female patient, a 3cm-sized cystic mass was found in the right maxillary sinus. A computed tomography of the paranasal sinuses with contrast enhancement showed irregular internal contrast enhancement occupying the space in the right maxillary sinus. A right endoscopic sinus surgery (ESS) showed a cystic mass and extensive tissue sampling was conducted. Histopathological examination confirmed the diagnosis of ameloblastoma. Concurrent chemoradiotherapy (CCRTx) was administered by oncology and radiation oncology departments. Three years follow up showed no recurrence.

Keywords: Unicystic ameloblastoma, endoscopy, maxillary sinus, surgery.

INTRODUCTION

Ameloblastoma is an undifferentiated tumor that primarily originates from enamel tissue and odontogenic epithelium. It grows slowly and invades the surrounding tissues locally. According to WHO classification of head and neck tumors in 2017, ameloblastomas are odontogenic epithelial tumors, classified unicystic, extraosseous/peripheral, and metastasizing ameloblastoma.

Unicystic ameloblastoma (UA) presents as a cystic lesion in the jaw, as observed clinically, and radiologically. Histologically, it is characterized by a typical cystic cavity lined by epithelial cells and showing luminal or mural tumor growth. UA accounts for 6% of ameloblastoma cases, with 50% occurring in individuals in their teens. It is more commonly found in the mandible followed by maxilla. UA in the maxillary sinus is very rare. To my knowledge, there was no case report of UA treated by endoscopic sinus surgery in the English literature. This case of UA was diagnosed incidentally and was treated with endoscopic sinus surgery.

CASE PRESENTATION

A 34-year-old female presented to the outpatient department of neurology with headaches, leading to the performance of a Magnetic Resonance Imaging (MRI) brain scan. Abnormal findings in the sinuses were discovered, and she was referred to the otolaryngology department. MRI revealed a cystic mass occupying a 3cm-sized space in the right maxillary sinus, along with findings suggestive of sinusitis. The T2-weighted images showed a peripheral rim with hypointense signal intensity with strong enhancement in the outer wall and internal septation. No specific findings were observed during nasal endoscopy examination.

Subsequently, a Computed Tomography (CT) scan of the paranasal sinuses with contrast enhancement (CE) was performed, revealing irregular internal contrast enhancement occupying space in the right maxillary sinus and a low-density lesion with posterolateral bone erosion (Fig. 1). There were suspicions of a organized hematoma or tumor, leading to the performance of right endoscopic sinus surgery (ESS). During the surgery, a cystic mass was observed using right MMA, and extensive tissue sampling was conducted. The mass was...
meticulously removed using tools such as microdebriders and giraffe forceps, leaving no visible remnants (Fig. 2).

![Fig. 2: Intraoperative endoscopic findings. A. right maxillary sinus cystic mass B. no residual gross lesion after removal.](image)

Histopathological examination revealed angular epithelial islands within a dense collagenous stroma, characterized by peripheral palisading columnar cells at the basal layer, and reverse polarized stellate cells, leading to the diagnosis of ameloblastoma. Consultation with the departments of oncology and radiation oncology resulted in the administration of concurrent chemoradiotherapy (CCRTx) with cisplatin (CDDP). The patient has been under follow-up observation for three years without any evidence of recurrence (Fig. 3).

![Fig. 3: Postoperative 3 year follow up showed no recurrence. A. endoscopic finding. B. enhanced paranasal sinus CT axial view.](image)

DISCUSSION

Ameloblastomas usually present with slow-growing, painless swelling in the mandible or maxilla. Occasionally, it is incidentally discovered during imaging examinations conducted for other reasons. In this case, the patient had no symptoms such as facial swelling, but the ameloblastoma was incidentally discovered during a brain MRI scan conducted due to headaches.

The unicystic variant of ameloblastoma is primarily found in children and is believed to originate from the dental follicle of an unerupted tooth or from a previously existing dentigerous cyst. The most commonly observed location is in the area of the third molar. In this case, despite the patient being an adult female, it is determined that the origin was in the area of the right maxillary third molar.

Ameloblastoma typically presents as a distinct-bordered, uni- or multicystic radiolucent expansile lesion on CT scans. UA on the other hand, often appears clinically or radiographically as an odontogenic cyst, and it is primarily diagnosed through histopathological examination. CT or MRI scans may reveal unilocular radiolucency with a thinly corticated appearance associated with jaw expansion and an unerupted tooth. In this case, there were suspicions of organizing hemATOMA or tumor. However, following ESS and excisional biopsy, a definitive diagnosis was made.

Histologically, ameloblastoma is composed of two cell types. The peripheral cells resemble ameloblasts and basal cells, while the central cells resemble the stellate reticulum and suprabasal epithelial cells. The diagnosis of UA can be challenging as soft tissue involvement is generally absent, and the cyst wall is usually membranous. Nuclear palisading may be only minimally observed, and stellate reticulum can be overlapped by inflammation in many cases. UA exhibits two histological variations: luminal and mural. Luminal refers to the presence of ameloblastoma epithelium within the cyst wall, while mural indicates infiltration of tumor islands into the fibrous wall, similar to conventional ameloblastoma. Ackermann et al, classified UA as Luminal, Intraluminal/plexiform and Mural UA. In this case, the presence of papillomatous plexiform ameloblastic tissue growing within the cyst indicates an intraluminal/plexiform UA, classified as group II.

The principle of surgical treatment for ameloblastomas is en bloc resection. Due to its high recurrence rate, it is recommended to maintain a margin of 1-1.5 cm for classic (solid/multicystic) ameloblastoma. However, extensive resection may raise concerns regarding aesthetics, functional impairment, and psychological issues, which may lead to considering conservative surgeries such as marsupialization, enucleation, or curettage. Nevertheless, recent meta-analyses have reported a high recurrence rate of 40% with conservative surgery. For UA, conservative surgeries such as extensive
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resection, excision, marsupialization, chemical cauterization, curettage, radiation therapy, and a combination of surgery and radiation therapy can also be considered.\(^1\) UA, being a less aggressive form of ameloblastoma, can be successfully removed through simple enucleation or other conservative surgeries.\(^8\) In this case, although margin clearance was not achieved during the sinus endoscopic surgery, all visible tumors were successfully removed. Chemotherapy is not considered effective for localized ameloblastoma, but it may be the only treatment option in cases of metastasis. Ameloblastoma is believed to be sensitive to platinum-based chemotherapeutic agents.\(^9\)

Similar to chemotherapy, radiation therapy also has a limited role in the treatment of ameloblastoma. It can be used in cases where there is residual disease microscopically or macroscopically after surgery, when the patient is not a suitable candidate for surgery, or when re-resection is not possible.\(^10\) In this case, a cystic mass in the right maxillary sinus was identified during ESS, and extensive tissue examination and excision were performed. Although the removal was done in a piecemeal manner, it was confirmed that no residual tissue was left behind after the surgery. However, due to the lack of secured margins, the presence of residual tissue was suspected both under the microscope and grossly. Therefore, in collaboration with the medical oncology and radiation oncology departments, platinum-based concurrent chemoradiotherapy was administered. As of 3 years post-surgery, there has been no recurrence during follow-up.

In summary, UA in the maxillary sinus is very rare. When there is a unilocular radiolucency in the jaw, it is important to approach it with caution as UA exhibits clinical and radiographic similarities to odontogenic cysts and tumors. It is recommended to perform resection with a sufficient margin whenever possible. Long-term follow-up is necessary.

REFERENCES