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## **Primary Ameloblastoma of the Temporal Bone**

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## **Abstract**

Ameloblastoma is a locally aggressive tumor derived from odontogenic epithelium. Although benign, its clinical behavior may often times exhibit malignant characteristics. It is marked by slow and persistent growth with infiltration of adjacent tissues. Almost 70% occur in the mandible in patients over 30 years of age. Recurrence of ameloblastoma due to inadequate treatment is frequent. Due to its slow growth, recurrences may present decades after primary surgery. A primary ameloblastoma in an area outside of the mandible, maxilla and infratemporal fossa regions has not been described in detail to date, with only one possible case mentioned in literature. The authors present a case of primary temporal bone ameloblastoma in a 17-year-old male patient. The tumor originated in the left mastoid, infiltrating the lateral semicircular canal, facial nerve and cochlea, adhering to the sigmoid sinus and posterior cranial fossa dura. Although invasion of multiple structures in the infratemporal fossa and the temporal bone leads to variable disease presentation, this case is unique when considering that the first symptom of disease was unilateral recurring sensorineural sudden hearing loss. Surgery required transection of the facial nerve. Histopathology confirmed primary temporal bone ameloblastoma. The difficulties in achieving wide surgical margins, diagnostics and further management are also addressed.

**Keywords:** primary; temporal bone; ameloblastoma; surgery; recurrence

## **Introduction**

Ameloblastoma is a locally aggressive tumor derived from odontogenic epithelium. [1]

Almost 70% occur in the mandible in patients over 30 years of age. Although it is labeled as benign, its clinical behavior may often times exhibit malignant characteristics. It is marked by slow and persistent growth with infiltration of adjacent tissues. Recurrence of ameloblastoma due to inadequate treatment is frequent. [2] Due to its slow growth, recurrences may present decades after primary surgery. A primary ameloblastoma in an area outside of the mandible, maxilla and infratemporal fossa regions has not been described to date, with only one possible case mentioned in literature. [3]

## **Case report**

This case report describes a case of primary temporal bone ameloblastoma in a previously healthy 17-year-old male patient that presented with sudden sensorineural hearing loss on the left side. He had no previous history of dentition disorders or episodes of otitis and otoendoscopy was unremarkable. Following intravenous and oral corticosteroid therapy hearing was restored, but sudden hearing loss recurred a month later on the same side. Computed tomography imaging with contrast enhanced angiography was performed and a multicystic tumor was observed originating in the mastoid portion of the left temporal bone with infiltration of the lateral semicircular canal, cochlea, vestibular aqueduct as well as possible tympanic portion of facial nerve involvement. The sigmoid sinus and posterior cranial fossa dura showed tumor adherence and occipital condyle erosion. No significant contrast imbibition was observed, or posterior cranial dura imbibition, excluding possible

paraganglioma or intraosseal meningioma diagnoses. (Figures 1 and 2) A multidisciplinary tumor board was consulted and surgery was recommended. The mass was reduced through a left-sided mastoidectomy with multiple frozen histopathological section biopsies unable to confirm that the tumor was malignant. Likely facial nerve infiltration was noted and further surgery was prolonged until definitive histopathological diagnosis was available. Definitive histopathology confirmed primary temporal bone ameloblastoma and a second stage surgery in a joint oto-neurosurgical team effort was performed a month after the first stage. (Figure 3) A partial temporal bone resection including an extended mastoidectomy and a canal-wall-up tympanomastoidectomy revealed that the horizontal segment of the facial nerve was infiltrated and was resected until clear frozen histopathological section margins were obtained. The mastoid portion of the tumor was resected with noted adherence to posterior cranial fossa dura and jugular venous bulb and posterior cochlear and lateral semicircular canal bony erosion which was drilled out. Facial nerve decompression, transection and reanastomosis through neurorrhaphy was performed. Six months after surgery, the function of the facial nerve had a IV/VI House-Brackmann left-sided facial paresis score. The patient's postoperative hearing threshold on the left side was normal up to 2 kHz, with deafness above 3 kHz. The immediate postoperative course was uneventful and magnetic resonance imaging after 2 months of follow-up showed residual tumor tissue immediately laterally to the left occipital condyle and posteriorly to the jugular venous bulb, measuring 10x20x15 millimeters. The patient's family was apprehensive of repeated surgery and the neurosurgical team advised an expectative approach, with magnetic resonance imaging after 1 month of follow-up and 7 months of follow-up showing the tumor tissue being stationary in size and position. (Figures 4 and 5)

## **Discussion**

Although ameloblastoma represents 1% of all jaw tumors, found exclusively in the maxillofacial region, with only a single previous reports of a primary temporal bone ameloblastoma published and no information available on the tumors characteristics or patient outcome. [3] Both multicystic and unicystic ameloblastomas exhibit a propensity toward local infiltration and when located in the infratemporal fossa, may spread to the pterygopalatine fossa, parapharyngeal space, orbit, or intracranial space. Multicystic lesions behave more like solid tumors and have a higher recurrence rate than unicystic ameloblastomas. Up to 4.5% of ameloblastomas metastasize, and sometimes present metachronously in the mandible and the maxilla, but there have been no reports of temporal bone metastasis without clear primary tumor localization. There have been speculations on possible implantation of tumor cells in the temporal region during previous surgery in the mandible or infratemporal fossa, but our patient had no previous surgery and the lesion seemed to originate in the mastoid. [2] The possible mechanism of spread in previously surgically treated patients may be incomplete enucleation of the cystic lining in the coronoid process, where unresected fibers of the temporal muscle attachment to the coronoid process act as a bridge for tumor cell implantation in the temporal bone. [4] The invasive borders of the tumor are diffuse, requiring adequate preoperative CT evaluation and resection planning. Margins of 3 centimeters and aggressive resection of solid and multicystic ameloblastomas correlates with a recurrence rate close to zero, due to reduction of residual tumor spread to locally fragmented bone through drilling close to the tumor. [5] It is often difficult to differentiate multicystic from unicystic ameloblastomas, possibly leading to an increased recurrence rate due to insufficient resection margins. [5] Although invasion of multiple structures in the infratemporal fossa and the temporal bone leads to variable disease presentation, this case is unique when considering that

the first symptom of disease was unilateral recurring sensorineural sudden hearing loss.

Treatment of ameloblastoma is usually dependent on its varied biologic behavior, clinical manifestations and radiologic features. [6] In this unique case the proximity and adherence of tumor tissue to vital surrounding structures make achieving these recommendations highly unlikely and optimal future treatment is currently uncertain.

**Disclosure of Interest:**

The authors have no conflict of interest to declare.

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## Figure legends



Figure 1. Preoperative computed tomographic scan showing an infiltrative multicystic ameloblastoma of the left temporal bone (arrow marking tumor position).



Figure 2. Preoperative computed tomographic scan showing an infiltrative multicystic ameloblastoma of the left temporal bone (arrow marking tumor position).

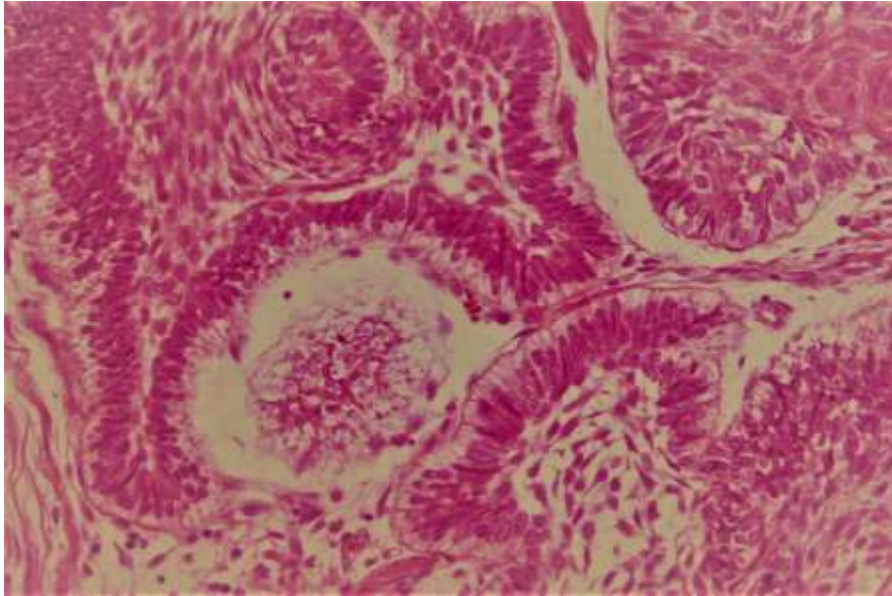


Figure 3. Histopathology showed a follicular growth pattern with tumor islands indicating peripheral and central cystic degeneration (hematoxylin-eosin stain, 200x).



Figure 4. Postoperative T1 FLAIR magnetic resonance image showing residual tumor tissue, surgery-related inflammation and liquid laterally to the left occipital condyle and posteriorly to the jugular venous bulb 1 month after surgery (arrow marking tumor position).

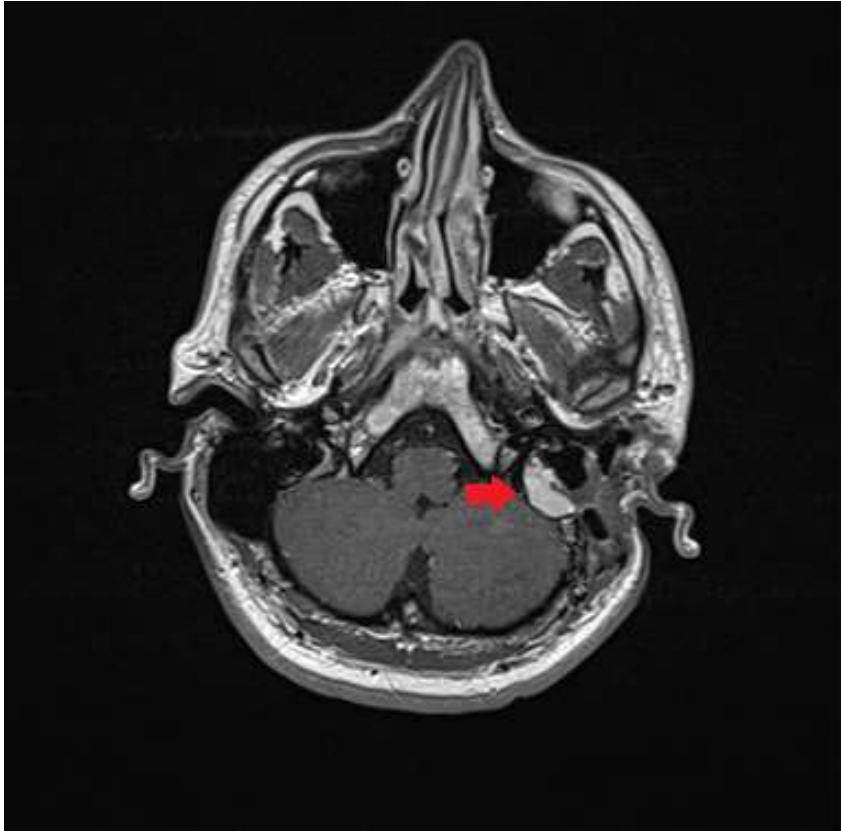


Figure 5. Postoperative T1 FLAIR magnetic resonance image showing residual tumor tissue laterally to the left occipital condyle and posteriorly to the jugular venous bulb 7 months after surgery (arrow marking tumor position).