

CASE REPORT

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Temporal Bone Chondroblastoma: a Rare Entity

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ABSTRACT

Introduction: Chondroblastoma is an uncommon benign, locally destructive tumor that usually arises from epiphyses of the long bones. Temporal bone chondroblastoma is an extremely rare occurrence. Chondroblastoma arise from immature cartilage cells and it may display certain malignant features by invading surrounding structures and metastasizing to adjacent sites. **Aim:** To present a case of extradural temporal bone chondroblastoma and discuss the clinical presentation, radiographic findings, histology and particularly the surgical management of the case. **Case report:** We report a case of a 31-year-old man who presented with a painless left temporal swelling and left sided hearing loss for four months. Computed tomography (CT) scan revealed an aggressive mass involving the left preauricular region with temporal mastoid bone erosion. Magnetic resonance imaging (MRI) showed an extra-axial left temporal mastoid mass pushing the left temporal lobe superiorly. The patient underwent complete excision of the temporal bone tumor. The final histopathological diagnosis was in keeping with chondroblastoma. **Conclusion:** Temporal bone chondroblastoma is rare but an aggressive condition. Complete tumor resection via an appropriate approach that enables adequate exposure will lead to a favorable outcome.

Keywords: Chondroblastoma, Temporal bone, Surgical excision, Management.

1. INTRODUCTION

Chondroblastoma occurs in the epiphysis of long bones and represents approximately 1% of all primary bone tumors (1). It was first reported by Codman in 1931, and the term was corrected to chondroblastoma of the bone by Jaffe and Lichtenstein in 1942 (2). Chondroblastoma involving the temporal bone is extremely rare (3). Approximately 81 such cases of temporal bone chondroblastoma have been reported (4). Although, pathologically, chondroblastoma is a benign tumor, it may invade and metastasize to adjacent structures (5). Radiation therapy may be effective in suppressing tumor growth but is only recommended for inoperable cases as it can induce sarcoma development (1, 6). Surgical curettage showed a recurrence rate of 55% among temporal bone chondroblastoma patients (7). Therefore, complete surgical resection is the gold standard of treatment (8).

2. AIM

We present a case of extradural temporal bone chondroblastoma that was completely resected. We will discuss the clinical presentation, radiographic findings, histology and particularly the surgical management of this case of temporal bone chondroblastoma.

3. CASE REPORT

A 31-year-old male presented to our department with a four-month history of left temporal painless swelling and left hearing loss. Upon clinical examination, there was a diffuse, firm, non-tender, non-erythematous and immobile mass in the left temporal region (Figure 1). Otoloscopic examination showed a left external auditory canal mass occluding the lumen of the external ear canal. Pure tone audiogram showed left moderate to severe conductive hearing loss. Neurological and other systemic examinations were unremarkable.

The computed tomography (CT) scan demonstrated an aggressive mass in the left preauricular region with intracranial and left external auditory canal extension causing destruction of left mastoid air cells (Figure 2a). Brain magnetic resonance imaging (MRI) revealed a lobulated soft tissue mass of the left temporal bone measuring 5.6cm x 3.7cm x 3.8cm with left temporalis fas-

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Figure 1 Preoperative image showing left temporal mass A) Posterior view B) Anterior view

cia and temporalis muscle extension. It was an extra-axial mass extended into the left temporal lobe without dura involvement. The tumor was hypointense on T_1 - and T_2 -weighted images and showed heterogeneous enhancement on post-intravenous gadolinium enhanced images (Figure 2b). A tissue biopsy was obtained from the left preauricular mass showing the presence of multinucleated osteoclastic giant cells and positive on S100 staining, in keeping with chondroblastoma.

The patient underwent excision of the tumor via middle cranial fossa approach. This was performed through a C-shaped incision or “question mark” incision via the preauricular skin crease towards the retromandibular region. Intraoperatively, a vascular solid cystic tumor had destroyed temporal bone, particularly the mastoid part and the squamous part with extension into the middle cranial fossa (Figure 3a). The tumor had invaded the temporalis muscle and was seen attached to the dura mater. A neurosurgeon resected the middle cranial fossa component and found the tumor to be fully extra-dural. The tumor was excised completely together with the infiltrated temporalis fascia and muscle; dura and facial nerve was identified and preserved. The eroded temporal bone was resected to normal bone by drilling with a diamond burr. Accordingly, the middle cranial fossa defect with exposed dura resulted from the tumor resection and tumor erosion was reconstructed with a temporalis muscle flap (Figure 3b).

The postoperative course was uneventful without any complications. The patient was discharged three days later. Histopathological analysis of the resected tumor was consistent with chondroblastoma. To date, there is no evidence of tumor recurrence in this patient.

4. DISCUSSION

Chondroblastoma of the temporal bone typically occurs in older males, between 30-40 years of age; conversely, patients with tumors that arise from the epiphyseal cartilage of the long bones are generally between 20-30 years of age (3). The most common subsite with involvement of temporal bone chondroblastoma is the squamous part, often originating from the cartilage (9).

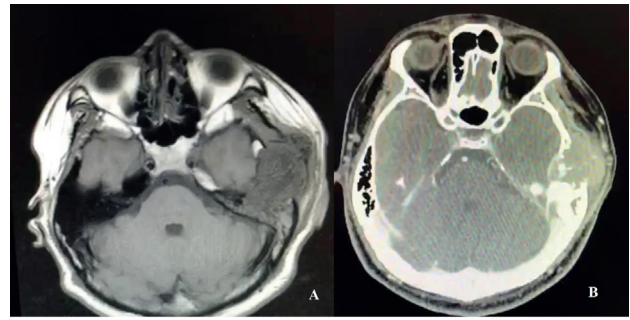


Figure 2. Radiological scan A) MRI temporal-axial view T1 weighted B) CT scan temporal-axial view. Soft tissue tumor over the left temporal region causing compression of the temporal lobe and local bony erosion.

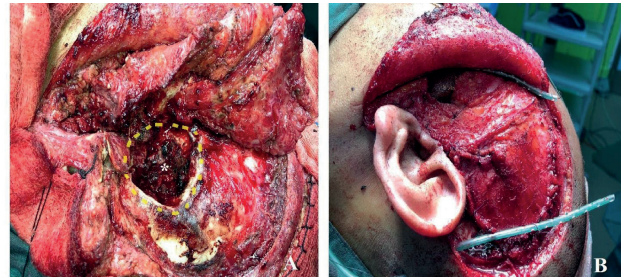


Figure 3. Intraoperative image showing A) Tumor site post excision B) Reconstruction of defect with Temporal Flap (Asterisks : Exposed dura ; Dotted yellow lines : Bony defect secondary to tumor invasion)

Although it is a benign lesion, it may show aggressive features including soft tissue infiltration, pulmonary metastasis and local recurrence (10).

Reid et al. described the most common presenting symptoms as hearing loss (49%), cranial nerve involvement (43.2%), facial swelling (22.2%) and otalgia (19.8%) (4). The duration of presenting symptoms is usually more than one year (3). Our patient was a 31-year-old male with a four-month history of left temporal swelling and hearing loss, which fulfills the classical criteria of temporal bone chondroblastoma.

Common histopathological findings of temporal bone chondroblastoma include polygonal cells, focal “chicken-wire” calcifications and osteolytic-like giant cells (11). Differential diagnosis of temporal bone chondroblastoma includes cholesteatoma, giant cell tumor, chondrosarcoma and aneurysmal bone cyst. Histopathological examination is used to differentiate chondroblastoma from those pathologies, except giant cell tumor which would require further immunohistochemistry staining to differentiate the two (8). Focal positive immunohistochemical staining for S100 protein is exclusively found in chondroblastoma (11). CT scans of temporal bone chondroblastoma typically show an expansile mass with local bony destruction (11). Radiological investigation is essential to look for bony involvement and to aid in surgical planning for complete resection. Intracranial involvement seen on CT imaging will alert the surgeon that neurosurgical co-management may be needed intraoperatively (4). MRI is the best tool to delineate intracranial soft tissue involvement, particularly differentiating dura mater from tumor. Dural invasion often demonstrates thickening and strong enhancement of

the involved region on MR images (8). T1 images usually show low intensity and low to high intensity on T2 images with heterogeneous enhancement of the mass on gadolinium contrast-weighted images (12).

Various treatment modalities have been reported previously, including total en bloc excision, curettage, irradiation and a combination of irradiation and surgical excision (11). Complete surgical resection is the treatment of choice for temporal bone chondroblastoma patients. Other treatments, such as irradiation, may be effective in suppressing tumor growth, but can induce sarcomatous changes from chondroblastoma to chondrosarcoma. Therefore, radiotherapy is restricted to inoperable or recurrent or after incomplete surgical excision (3, 7). There is no role for chemotherapy in treating temporal bone chondroblastoma (4). Preoperative planning of the surgical approach is decided according to the extension and location of the tumor (8). Adequate surgical field exposure is important when the tumor is adjacent to vital structures such as the brain, cranial nerves, carotid artery or jugular vein (11). Aggressive attempts to remove the tumor completely without adequate exposure will be hazardous (7).

In most cases, the tumor can usually be separated free from the adhering dura by careful dissection. However, dural invasion and infiltration would require excision and appropriate reconstruction of the dura. In the case of Temporomandibular joint (TMJ) invasion, excision of the glenoid fossa or removal of a mandibular condyle is needed to achieve complete resection of the tumor (8). Successful complete excision of a temporal bone chondroblastoma via the skull base approach has been reported. The middle cranial fossa approach in combination with a total mastoidectomy via a retroauricular incision is preferred by most Ear, Nose and Throat (ENT) surgeons with multiple sources citing a low recurrence rate during follow-up of one to four years. On the other hand, neurosurgeons prefer the middle fossa approach, which has shown promising outcomes without tumor recurrence for up to four years (8). Kurokawa et al. concluded that the middle fossa approach provides poorer exposure compared to the trans zygomatic approach with resection of the mandibular condyle to expose the tumor; thus, the latter allows superior exposure and total excision done under direct visualization (7).

In our case, the middle fossa approach was adequate to fully expose the tumor and allow excision to be done under direct visualization of the tumor. The tumor was successfully separated from dura and the TMJ was preserved. In most cases and as reflected in our patient, complete resection of the tumor is effective and sufficient as a single modality treatment. Such an approach is often favorable with low recurrence rate (8). Radiation therapy has been recommended only in selected cases of incomplete tumor removal, as it may induce sarcomatous changes from chondroblastoma to chondrosarcoma. Overall recurrence after total tumor removal is low, i.e. approximately 20%, compared to surgical curettage, with a recurrence rate of about 55% (7, 12). Higher re-

currence rates are seen in cases with chondroblastoma containing areas of aneurysm bone cyst (11).

5. CONCLUSION

Temporal bone chondroblastoma is rare but an aggressive condition. Complete tumor resection via an appropriate approach that enables adequate exposure will lead to a favorable outcome. Despite a low recurrence rate, long-term clinical follow-up is still needed to rule out any possible recurrence in the future.

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