Case Report

Giant Cell Tumor of Infratemporal Fossa and Mandibular Condyle: A Case Report

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INTRODUCTION

The giant cell tumor (GCT) is a rare primary bone neoplasm and constitutes approximately 5% of all primary bone neoplasms [1]. GCT mainly arises from the epiphyses of long bones, and it generally has a slight female dominance in the second to the fourth decade of life [2]. GCT is a benign neoplasm but often exhibits aggressive behavior, which leads to a high rate of local recurrence or malignant transformation. Complete excision of tumor is the established treatment of choice [3].

Approximately 2% of all GCTs occur in the head and neck regions, and most involve the maxilla and mandible. GCTs are only rarely encountered in the skull; in the few reported cases, tumors arose mostly in the sphenoid and occasionally in the temporal bone. The clinical features of GCT arising from the temporal bone depend upon the site of origin in the temporal bone, and trismus is an unusual presentation [2, 4]. Here, we report the case of a huge GCT of the petrous and squamous portions of the temporal bone, which extended into the left mandibular ramus and middle ear and include a review of the literature.

CASE PRESENTATION

A 31-year-old male presented with a progressive difficulty in opening his mouth that persisted for 6 months. There was no history of trauma or infection in the left ear, and dental examination did not show any findings of a specific disease. During otoscopic examination, a yellowish-brown mass was observed behind the posterior half of the intact left tympanic membrane, with no evidence of hearing loss by pure tone audiometry (Figure 1). Temporal bone computed tomography (CT) revealed a mass with a size of 4.1x6.9x5.2 cm³ expanding into the squamous and petrous portions of the left temporal bone and extending into the mandibular ramus inferolaterally and into the middle ear posteriorly (Figure 2). Brain magnetic resonance imaging (MRI) demonstrated that the lesion was a predominantly low-signal intensity region on T1-weighted images, an intermediate signal intensity region on gadolinium-enhanced T1-weighted images (Figure 3). To make a correct diagnosis and to determine the therapeutic option, an open biopsy was performed at the site of the squamous portion of the temporal bone. A subsequent histopathological examination revealed that the lesion was composed of multinucleated giant cells admixed with mononuclear stromal cells. The stromal cells were round- to-oval polygonal or elongated shaped; therefore, the lesion was diagnosed as a GCT (Figure 4). No evidence of distant metastasis was provided by positron emission tomography-CT (PET-CT).

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A left temporal craniectomy was performed using an extended pterional approach for complete surgical resection with tissues of the infratemporal fossa, glenoid fossa, and mandibular ramus (Figure 5a). In addition, complete canal wall down mastoidectomy was performed to remove any tumor invading the middle ear. The mass was mainly located in the attic area, surrounded the ossicles, and bulged through the squamous portion of the temporal bone; the round and oval windows and ossicles were intact. Surgery was performed with the cooperation of the Departments of Oral and Maxillofacial Surgery, Neurosurgery, and Otolaryngology. Using this radical resection with marginal drilling and electrocauterization, we were able to remove all tumor tissues and achieve a clear resection margin. All cranial nerves and inner ear structures were preserved, and the skull base was reconstructed using the pericranial flap and temporalis muscle (Figure 5b). No further treatment, including radiotherapy, was administered. During the last follow-up, 9 months after the operation, the patient showed no evidence of recurrent disease (Figure 6).

**DISCUSSION**

GCT is a rarely encountered bone tumor that occurs at a frequency of approximately one per million per annum. It originates from the connective tissue of the bone marrow and mainly involves the epiphyses of long bones, most commonly the lower femur. Although rare, GCT can occur in the bones of the skull, usually in the sphenoid, the temporal bone, or the ethmoidal bones. It usually presents with

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**Figure 1.** Otoscopic finding. A yellowish-brown mass was observed behind the posterior half of the intact left tympanic membrane.

**Figure 2.** a, b. Computed tomographic images with bone window algorithm (a). The axial image demonstrates a large lytic lesion on the right with erosion into the left temporal bone (b). The coronal image demonstrates the same left-sided lesion with significant bone remodeling.

**Figure 3.** a-c. Preoperative magnetic resonance imaging (MRI) scans showing a well-defined mass with a size of 4.1×6.9×5.2 cm in the left temporal bone that exhibited low signal intensity on axial T1-weighted images (a), heterogeneous contrast enhancement (b), and intermediate signal intensity on axial T2-weighted images (c). The mass had not infiltrated into the surrounding tissues.
a slight female dominance in the second to the fourth decade of life \cite{3,5}. There are no clinical symptoms specific to GCT. Patients usually present with pain and limited range of motion caused by the proximity of tumor to the related area. The clinical presentation of GCT of the temporal bone depends upon its site of origin. Patients typically complain of pain behind the ear on the lesion side, hearing difficulty, swelling in the concerned region, and facial paralysis \cite{6}. Our patient presented with progressive trismus, which is an unusual presentation.

On conducting radiographic studies, it was found that GCT is typically a nonspecific, relatively well-circumscribed, expansile, osteolytic mass lesion without abundant matrix calcifications on plain films or CT scans. However, plain radiography cannot usually be distinguished from other radiolucent lesions. GCT of the skull tends to be more aggressive and presents as a purely osteolytic lesion. CT scanning is useful for assessing the extension of the bony erosion and demonstrates prominent bony trabeculation and loculation areas \cite{2,7,8}. MRI can be useful in evaluating the extension into the intramedullary and soft tissues. Few reports issued on the MRI characteristics of skull GCT describe a nonspecific, heterogeneously

![Figure 4. Pathological findings. Round-to-oval polygonal or elongated mononuclear cells were evenly mixed with numerous multinucleated giant cells. H-E stain; ×200](image)

![Figure 5. a, b. Operative findings. The tumor in the temporomandibular joint was completely removed (a). The surgical defect was reconstructed with a temporal rotational flap (b)](image)

![Figure 6. a, b. Postoperative magnetic resonance imaging (MRI) scans. Axial T1-weighted images (a) and axial T2-weighted images (b). MRI scans demonstrate the absence of a well-defined mass in the left temporal bone](image)
enhancing mass of variable, frequently intermediate signal intensity on T1- and T2-weighted images [7, 8]. However, GCT of the skull does not demonstrate any unique radiographic features, and a definitive diagnosis requires a comprehensive consideration of clinical features and often histopathological confirmation. The differential diagnosis on radiological images includes aneurysmal bone cyst, chondroblastoma, dermoid cyst, chondrosarcoma, giant cell reparative granuloma, and pigmented villonodular synovitis [2, 8]. In the described case, temporal bone CT showed an expansile osteolytic lesion in the temporal bone, and MRI revealed a heterogeneously enhancing mass on gadolinium-enhanced T1-weighted images.

Histological examination is essential to diagnose GCT; the tumor cells are composed of homogeneously dispersed, large, multinucleated giant cells distributed uniformly amid a vascularized background network of round, oval, or spindle-shaped stromal cells [9]. GCT is regarded as benign. However, it is locally aggressive and may even metastasize to the lung. The treatment of choice for GCT is surgical excision. The recurrence rate depends upon the adequacy of treatment; regardless of the site of presentation, marginal resection or curettage is intimately associated with a high recurrence rate (40%–60%). Wide radical excision is the preferred treatment and has a low recurrence rate (7%). With regard to local recurrence rates and prognosis of GCT, they correlate with the extent of resection. However, GCT are not always completely removed. It depends on the location of GCT; therefore, some authors have reported their experience with adjuvant therapy [3, 4]. In most cases, radiation therapy is reserved for inoperable or non-radically operated cases. However, radiation therapy increases the possibility of sarcomatous transformation soon after the completion of therapy. Interestingly, the likelihood of malignant transformation following radiation therapy significantly decreased after orthovoltage irradiation was replaced by megavoltage irradiation [5–6]. Therefore, radiation therapy is recommended for GCT when the lesion cannot be completely removed or for those patients who cannot undergo surgical resection because of comorbid factors. Chemotherapy is also indicated for cases that remain inadequately controlled after surgery and irradiation [10]. In our case, adjunctive therapy was not administered because the tumor was considered to have been completely removed by surgery.

Several rational approaches for surgical excision have been suggested for GCT in different regions of the temporal bone [9, 10]. The surgical approach depends on the position and extent of the tumor. In the present study, the patient had a tumor located in the petrous and squamous regions and in the middle ear that extended into the mandibular ramus. We selected left temporal craniectomy via an extended pterional approach and canal wall down mastoidectomy to easily obtain access to the mandibular ramus and to allow better visualization of structures of the skull base in the surgical field.

This paper presents a case of GCT of the temporal bone with an unusual presenting symptom, namely trismus. The patient was treated by radical tumor excision using a combined surgical approach. In summary, GCT is a rare primary bone disease. Therefore, the possibility of GCT should be considered in the differential diagnosis of a bony lesion of the craniofacial region until a final diagnosis is made.

Informed Consent: Informed consent was not obtained as this was a case report without recognizable photographs of patient.


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REFERENCES