



Case Report

Giant Cell Tumor of the Temporal Bone and Skull Base: A Case Report

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Giant cell tumor (GCT) is a benign tumor that originates from undifferentiated mesenchymal cells of the bone marrow. The cranium as well as temporal bone is a rare location for GCTs. Despite its benign nature, GCT may be locally aggressive and has the potential to recur locally. Furthermore, GCT may give rise to pulmonary metastases (~1%) in addition to causing local bone destruction. Surgical excision is the treatment of choice for patients with GCT. We describe the case of a 56-year-old female who presented with headache and hearing loss with extensive GCT, which originated in the squamous part of the temporal bone and extended into the left mandibular fossa and middle ear. She was treated by total resection of the tumor using left temporal craniotomy approach. In this article, we present a case of temporal bone GCT with its clinical features as well as a review of the related literature.

KEYWORDS: Giant cell, temporal bone, hearing

INTRODUCTION

Giant cell tumor (GCT), which has a benign clinical behavior, arises from undifferentiated mesenchymal cells of the bone marrow. Thus, it is more common in the epiphysis of the long bones. [GCT usually arises from the distal radius and femur and proximal tibia and fibula; it rarely arises from the temporal bone [1]. Despite its benign nature, GCT may be locally aggressive and has the potential to recur locally [2]. Furthermore, GCT may cause local bone destruction and give rise to pulmonary metastases (~1%) [3]. GCTs are seldom located in the cranium and temporal bone and may present with different symptoms depending on their location. Temporal bone GCT causes otalgia, aural fullness, conductive or sensorineural hearing loss, tinnitus, localized swelling in temporal bone or preauricular region, temporomandibular joint dysfunction, and facial paralysis. In this article, a case of temporal bone GCT has been presented with its clinical features.

CASE PRESENTATION

In 2008, a 56-year-old female patient presented with the complaint of headache. A mass with bone destruction in the squamous part of the left temporal bone was found after complete head and neck examination and computed tomography (CT). A biopsy was performed through a craniotomy approach in 2008 at the neurosurgery department. Postoperative histopathologic analysis revealed a benign giant cell lesion. Considering the benign nature of the lesion, the patient was observed through regular follow-ups.

The patient was admitted to the otolaryngology clinic with new-onset headache as well as left-sided hearing loss and tinnitus complaints after 2 years. Informed consent was obtained from the patient. Physical examination showed partial obliteration of the left external auditory canal due to the indentation of the tumor from the inferior wall. The pure tone audiometry (PTA) test showed left-sided 65-dB mixed hearing threshold. CT scan and magnetic resonance (MR) imaging showed an extensive soft-tissue lesion extending to the mandibular fossa with near-total destruction of the temporal bone has shown in Figure 1 and 2. A signal intensity of heterogeneous contrast enhancement was observed in postcontrast T1-weighted MR images, and an intermediate signal intensity was observed in T2-weighted MR images. The patient was taken up for excision of the tumor under general anesthesia. She underwent total tumor excision with craniotomy via middle fossa approach. The tumor was found to be invading the infratem-



Figure 1. MRI shows an extensive soft-tissue lesion extending to the mandibular fossa with near-total destruction of the temporal bone (arrow shows tissue lesion in postcontrast T1-weighted MR images).

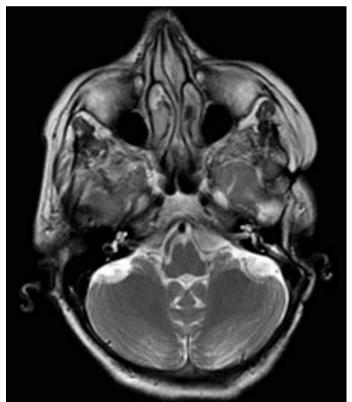


Figure 3. MRI shows that no residual mass or recurrence was detected.

poral fossa, extending anteriorly up to the temporomandibular joint and inferiorly up to the greater wing of the sphenoid bone. A drain was left in the large dead space created by the removal of the tumor. The patient had an uneventful postoperative recovery. Postoperative



Figure 2. MRI shows an extensive soft-tissue lesion extending to the mandibular fossa with near-total destruction of the temporal bone (arrow shows tissue lesion in postcontrast T1-weighted MR images).

histopathology of the tumor revealed a giant cell lesion. Because of its benign nature, the patient was advised postoperative radiological follow-up. A postoperative PTA test showed left-sided total hearing loss. Till the time of writing this article, i.e., 6 years postoperatively, no evidence of tumor recurrence had been detected. Postoperative MR imaging performed in December 2016 showed no residual mass or recurrence has shown in Figure 3 and 4.

DISCUSSION

In the literature, 110 cases of GCTs of the skull have been reviewed. Tumor locations were as follows: temporal bone in 37 patients, sphenoid in 20 patients, occipital in 6 patients, frontal in 2 patients, and temporomandibular joint in 2 patients [4]. GCTs of the skull are most often centered in temporal bone.

GCT comprises almost <5% of benign tumors arising from the bone marrow. Just 2% of all GCTs are seen in cranium [1, 5]. Formation of intramembranous ossification is clarified as infrequent incidence of GCT in craniofacial section [1]. It arises more commonly from the sphenoid bone and to a lesser extent from the temporal bone [1, 6]. GCT originates more often from petromastoid region of the temporal bone [6].

Patients with GCT can present with a variety of symptoms depending on its location in the head and neck region. The associated symptoms are otalgia, headache, localized swelling in temporal or preauricular region, temporomandibular joint disorders, conductive or sensorineural hearing loss, tinnitus, vertigo, fullness of ear, facial weakness, visual field defects, double vision, and loss of vision ^[1, 7]. GCT is more often seen in the fourth or fifth decades of life with a slight female predominance ^[7, 8].

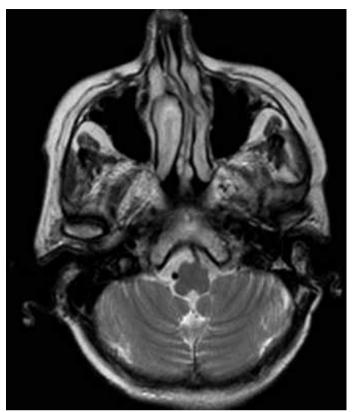


Figure 4. MRI shows that no residual mass or recurrence was detected.

Radiologically, GCTs do not have a specific appearance. Usually, an expanding lytic and destructive lesion is seen in the temporal bone CT. In some cases, CT images show soap bubble appearance; however, soap bubble appearances are also seen in aneurysmal bone cysts ^[8]. These tumors have vascular nature; therefore, they are mostly contrast enhancing. For radiologic differential diagnosis, osteoblastoma, chondrosarcoma, osteolytic metastasis, dermoid cysts, pigmented villonodular synovitis, and other fibro-osseous lesions should be noted ^[1]. MR images generally demonstrate signal isointensity on T1-weighted images and signal hypointensity on both T2- and diffusion-weighted images. GCTs show heterogeneous enhancement after intravenous gadolinium administration ^[3].

Colors of these tumors are largely gray to yellow-brown. They are soft, firm, and friable. Histopathologically, giant cell diagnosis can be inaccurately identified with giant cell granuloma, aneurysmal bone cyst, brown tumor of hyperparathyroidism, fibrous dysplasia, cherubism, and chondroblastoma [1]. Brown tumor is excluded based on normal parathormon and calcium levels.

Histologically, giant cell reparative granuloma (GCG) looks like GCT. GCT contains a large number of nuclei and uncommon mitotic figures; however, giant cell granulomas do not contain as many nuclei. They have clustered giant cells around areas of hemorrhage ^[7]. Giant cell granulomas are associated with reactive processes of traumatic intraosseous hemorrhage. But history of trauma is still in debate ^[7]. Giant cell reparative granulomas are accepted as they have more benign behavior than GCTs. The difference between GCTs and GCGs remains controversial. From GCG to GCT may have a behavior pattern from inflammatory and reparative features to true neoplasms in a spectrum of similar diseases. Some authors

believe GCGs to be the latter type of GCT. Hence, GCGs can be completely cured following surgical curettage. Moreover, GCTs have high recurrence rates up to 40%-60% with a risk of malignant transformation and metastases ^[7].

Surgical resection is the initial choice of treatment. While the local recurrence rate for intralesional surgical approach is 27%, marginal excision recurrence rate is reported to be approximately 8% [9]. Recurrence rates decrease with complete tumor resection. Recurrence seems to be directly related to the extent of resection. However, wide local excision of the tumor is generally inconvenient in the skull base and temporal bone due to the neighboring vital structures and the morbidity associated with surgery. The role of adjuvant radiotherapy in treatment is still controversial. Radiotherapy is recommended for tumors that are not amenable to complete resection, cases with morbidity limiting the use of general anesthesia, and cases with recurrent progressing lesions despite multiple surgeries. Radiotherapy is used as an adjunct therapy. When radiotherapy is used as a single-modality treatment, recurrence rates up to 60%-70% are seen [10]. Radiotherapy in modest doses (35 gray in 15 fractions or equivalent) has been found to be safe and effective [2, ^{10]}. Some authors report that radiotherapy may trigger sarcomatous transformation of GCT [1, 10].

GCT has the potential risk for metastases. GCT (\sim 1%) may give rise to pulmonary metastases in addition to causing local bone destruction ^[2, 3].

CONCLUSION

GCT is an uncommon lesion of the temporal bone. The exact diagnosis can only be made histopathologically. Considering the benign nature of the tumor and the probability of malignant transformation after radiotherapy, surgical removal of the tumor remains the best management option.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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