Giant Cell Tumor of the Skull Base: A Case Report

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Giant cell tumor of the skull is an extremely rare clinical entity. To our knowledge, this is the first report in Taiwanese literature. In this article, we describe a case of giant cell tumor of the temporosphenoidal region and infratemporal fossa with detailed clinical presentation and radiological studies. Three-dimensional computed tomography was performed preoperatively and provided a useful adjunct to surgical planning. The patient underwent subtotal tumor excision and postoperative radiotherapy. Complete tumor remission was noted in the follow-up study 1 year later.

Key words: Skull neoplasm, giant cell tumor; Skull, temporal bone, sphenoid bone; Computed tomography

CASE REPORT

A 58-year-old male patient had suffered from right facial palsy and tinnitus for about 3 years. However, he paid little attention to these symptoms. He came to our emergency department with the chief complaint of two episodes of vertigo attacks during the past week. He denied any trauma history, and there was no clinical evidence of hyperparathyroidism.

Otologic examination revealed a skin-covered protrusion at the inferior aspect of the right external auditory canal. The audiogram revealed a 25-dB air-bone gap in speech frequency of the right ear. The pertinent neurologic findings were right peripheral facial palsy and sensory deficits over the V1, V2, and V3 distributions of the right trigeminal nerve. Jaw strength was normal. The laboratory data, including serum calcium and alkaline phosphatase, were essentially normal.

High-resolution Computed tomography scans of the temporal bones (with a bone window setting) comprised the initial imaging study at admission. The scans revealed a tumor at the skull base with invasion of the right middle ear as well as destruction of the anteroinferior aspect of the right mastoid, squama of the right temporal bone, and lesser wing of the right sphenoid bone (Fig. 1a). Invasion of the right infratemporal fossa with an intact temporomandibular joint (TMJ) was evident (Fig. 1b). Three-dimensional computed tomography (3-D CT) was then obtained which helped us better understand the spatial relationship of this tumor. It showed a bony defect over the middle cranial fossa (Fig. 1c). Magnetic resonance (MR) imaging was performed thereafter and disclosed a tumor mass involving the right middle cranial fossa and infratemporal fossa. It was mainly isointense to the gray matter on precontrast T1-weighted images (T1WI) (Fig. 2a). After intravenous administration of gadolinium diethylenetriaminepentaacetic acid (Gd-DTPA), this tumor showed strong but inhomogeneous enhancement. The cystic portion of this tumor was hypointense on T1WI, hyperintense on T2-weighted images (T2WI), and septated on postcontrast T1WI (Fig. 2b, 2c). Preoperative angiography was performed. A right external carotid angiogram showed a tumor stained with faint vascularity in the right infratemporal region (Fig. 3).

A right temporal craniotomy was performed. There was a soft,
yellowish-gray, fragile tumor located mainly in the infratemporal fossa. The tumor mass was removed subtotally via a transtempo-marginal approach. Grossly, the tumor had invaded the structures of the petrous ridge, the squama of the temporal bone, the trigeminal nerve, and the pterygoid plate. The overlying dura seemed intact. After operation, the patient was discharged uneventfully with stationary trigeminal and facial neuropathy. He received adjuvant radiotherapy and was followed up regularly.

Histologically, the tumor was composed of a stroma of spindle cells with moderate cellularity and numerous multinucleated giant cells (Fig. 4). Stromal atypia was noted in some parts of the tumor characterized by pleomorphic nuclei and a few mitotic figures. From this microscopic picture, the diagnosis of giant cell tumor was made.

DISCUSSION

Giant cell tumor (GCT) of the bone comprises 3%-7% of primary bone tumors. It has been estimated that 70%-90% of the GCTs are found at the epiphysis of long bones, while the remaining 10%-30% are found in the sacrum, patella, vertebra and skull [1-3, 5]. There is a female predominance, and the age at presentation is usually between 30 and 50 years [5]. However, patients with GCTs of the skull seem to be older than those with GCTs of the long bones [6].

GCT is rarely encountered in the skull. In Dahlin’s series of 407 GCTs, only 3 cases involved the skull [5]. In 1992, Bertoni et al. reviewed some 2046 cases of GCTs and found only 15 cases in the skull bones [6]. The most commonly involved sites of the cranial bones, as published in the English literature, are the sphenoid bones, followed by the temporal bones. Other sites of cranial bone involvement, such as the occipital, frontal, and parietal bones, have been reported [9-11]. It has been suggested that the predominant involvement of these two bones is due to the fact that they arise, as do the long bones, through endochondral bone formation [12, 13]. In contrast, the remainder of the skull is formed from membranous
bone.

The clinical symptoms vary widely as expected depending on tumor size, location, and involvement of intracranial neurovascular structures. Frontal headache, diplopia, and dysfunction of the second through eighth cranial nerves are the usual modes of presentation in sphenoidal involvement [2, 4, 6, 12, 13]. With involvement of the temporal bones, patients usually present with conductive or sensorial hearing loss, vertigo, a mass in the external auditory canal, or even facial palsy [14]. The average duration of symptoms, from the time of onset to admission, is about 6-15 months [12, 13].

Radiographically, an accurate preoperative diagnosis is not readily available because GCT lacks unique radiographic features. On plain skull film, GCT of the skull is a purely lytic lesion as seen at other skeletal sites. On CT scans, the tumor is slightly hyperdense with enhancement after contrast infusion [8]. Cystic change and calcifications within the tumor have been reported, and thus can not be excluded from the diagnosis of GCT [3, 15]. On angiography, GCT can be avascular, moderately vascular, or hypervascular [3, 4, 10, 16, 17]. While CT scans can clearly show bony destruction, MR imaging provides multiplanar images and better definition of the degree of tumor extension. On MRI, GCTs are usually isointense to gray matter on both T1WI and T2WI with homogeneous or inhomogeneous enhancement after infusion of Gd-DTPA on T1WI, depending on the amount of cystic contents [8, 18]. The MRI findings of this case are similar. Once an osteolytic lesion is found in the skull base, the radiological differential diagnosis should include the following: aneurysmal bone cyst (ABC), giant cell reparative granuloma (GCRG), “brown tumor” of hyperparathyroidism, metastasis, chondroblastoma, eosinophilic granuloma, dermoid cyst, and other fibro-osseous lesions [11, 15].

Histologically, GCT shows a predominance of large multinucleated giant cells dispersed in spindle-shaped stromal cells. Unfortunately, multinucleated giant cells may be found in both neoplastic and nonneoplastic bone lesions, rendering a correct pathological diagnosis difficult. The histological differential diagnosis includes GCRG, ABC, chondroblastoma, brown tumor of hyperparathyroidism, osteogenic sarcoma (OGS), malignant fibrous histiosarcoma, and so on [3, 4, 6, 12].

It has been estimated that about 10%-15% of GCTs are malignant [2, 16]. Although usually benign histologically, GCT can be quite aggressive clinically with a tendency for local recurrence, late malignant change, and metastasis [6, 7].

Prognosis based on the clinical and radiological staging is more reliable than that of histological features alone [10]. There is still no standardized method regarding the treatment of GCT of the skull. In most series, regardless of what types of treatment, local recurrence rates are usually between 30% and 40% [3]. Success has been claimed with surgery alone, radiotherapy alone, and a combination of both. Some thought that surgical treatment should be recommended in all operable cases, and radiotherapy should be reserved for those in whom an operation is not possible [3, 9-11, 14]. The role of radiotherapy is still controversial because GCT may be radioresistant and because irradiation increases the risk of late sarcomatous transformation [1, 19]. However, Bell et al. reported that none of their series of 15 patients receiving supervoltage radiotherapy experienced local recurrence or late malignant transformation during a mean follow-up period of 12 years [20]. In addition, Bertoni et al. and Wolfe et al., in their reviews of GCTs of the skull bones, found that the best therapeutic results were those with a combination of surgical excision and radiotherapy [6, 12].

In summary, a case of GCT of the skull base is presented in this article. Detailed imaging studies were undertaken, but we still failed to make a proper diagnosis preoperatively. Pathological examination confirmed the diagnosis.
of GCT. In our case, 3-D CT and MRI were performed preoperatively which offered better planning of tumor resection. Although the tumor was not completely resected because of its location, follow-up MRI study showed complete tumor remission 1 year later after radiotherapy. However, the long-term therapeutic result still needs to be observed.

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顱底巨細胞瘤：病例報告

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顱底巨細胞瘤在臨床上非常罕見，據我們所知，此病例是發表於臺灣文獻之首例。在這篇文章中，我們報告一例顱底巨細胞瘤的臨床症狀及影像表現，並透過三維成像斷層掃瞄
做為術前的輔助工具。此病患接受部分腫瘤切除及術後放射治療，一年後，追蹤磁振掃瞄
檢查，發現腫瘤完全消失。

關鍵詞：腫瘤，巨細胞瘤；顱骨，顱骨，蝶骨；斷層掃瞄；磁振掃瞄