CASE REPORT

Giant cell tumour of the posterior cranial fossa: a case report

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ABSTRACT. Giant cell tumours (GCTs) of the cranium preferentially affect the sphenoid and temporal bones. We report a 19-year-old patient with GCT involving the left occipital bone and petromastoid portion of the temporal bone. CT and MRI revealed a predominantly expansive soft-tissue mass of the posterior cranial fossa. The patient underwent surgery. Permanent histopathological sections and immunostains revealed the presence of a GCT. The radiological features and method of surgical intervention of this rare lesion are discussed.

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The giant cell tumour (GCT) is considered to be a locally aggressive benign tumour. GCT of bone is an uncommon primary bone neoplasm, which typically occurs at the epiphyses of long bones and rarely manifests in the skull. GCTs of the cranium represent only 1% of all GCTs and preferentially affect the sphenoid and temporal bones [1–6]. Primary GCTs of the posterior cranial fossa are exceedingly rare. Here, we report a case of GCT in the posterior cranial fossa and review the literature regarding its radiological features and the method of surgical intervention used for treatment.

Case report

A 19-year-old female reported with a recurrent paroxysmal headache localised at the occipital region which she had experienced for several years. Since the symptom was consistently relieved by rest she did not seek or receive medical examination or treatment. 10 days prior to admission at our hospital, the usual headache symptom gradually aggravated and recurrent vomiting developed. Physical examination revealed no significant mass or tenderness in the head and routine neurological examination and laboratory examinations revealed no abnormalities.

CT scan showed an expansile, destructive soft-tissue mass across the occipitomastoid suture involving mostly the left occipital bone and petromastoid portion of the temporal bone. The mass appeared to have originated from the diploic space; it had expanded into the inner table and destroyed the outer table (Figure 1a,b). MRI showed the tumour tissue was isointense on T_1 weighted imaging (WI) and slightly hyperintense on T_2 flair, relative to the white matter. Within the tumour, the

hypointensity on T_1 WI and hyperintensity on T_2 flair indicated necrosis (Figure 2a,b). At the axial and sagittal planes of the gadolinium-enhanced MRI, the tumour tissue was markedly enhanced (Figure 3a,b). The tumour did not invade the dura mater or brain parenchyma.

During surgery, the colour of the lesion in the skull was normal. The mass was completely excised by *en bloc* removal. The inner table of the skull had evidently expanded but the underlying dura was not involved. Histopathological examination revealed large, multinucleated giant cells that were uniformly dispersed among the oval or spindle-shaped stromal cells (Figure 4). Immunohistochemical staining indicated the presence of Kp-1 and vimentin, and CD34-positive immunoreactivity was detected. The histological diagnosis was GCT.

Post-operative radiotherapy was not administered. The patient was re-checked by MRI every 3 months for 1 year after surgery and there was no evidence of recurrence.

Discussion

GCTs account for approximately 4–9.5% of all skeletal tumours and 18–23% of benign tumours [7]. This tumour type develops by endochondral ossification and most GCTs (70–90%) occur at the epiphyses of long bones. Only 1% of GCTs present in the skull [1, 8], with the most common cranial sites being the sphenoid and temporal bones. The fact that the sphenoid bone and petromastoid portions of the temporal bone arise from endochondral ossification in the skull can explain why GCTs in the skull are mostly found in the sphenoid and temporal bones [3, 5, 7]. To date, only a few cases of GCT have been reported to have derived from the occipital, frontal and parietal bones. In these cases, the bulk of the tumours have involved the occipital bone; a minor portion has been associated with the petromastoid portion of the temporal bone. The site and condition involving both endochondral and intramembranous ossification are exceedingly rare. In the English language literature, only

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Figure 1. (a) CT scan of the bone window at the occipital bone slice shows the mass originating from the diploic space expanded into the inner table (thick arrow), with prominent bony trabeculation (thin arrow) and destroyed outer table. (b) CT scan of the bone window at the temporal bone slice shows the mass expanded beyond the inner table (small arrow) across the occipitomastoid suture (large arrow).

four case reports of GCT of the occipital bone have been published [6, 9–11] but no case report describing GCT involving endochondral and intramembranous ossification exists. Thus, GCT occurrence in intramembranous ossification remains largely unexplained. Many hypotheses have been postulated including the presence of aberrant cells and the occurrence of metaplasia at local primitive connective tissue [6].

Histopathological sections of these types of tumours have demonstrated a uniform distribution of multinucleated giant cells in a background of bland, oval- to short-spindled stromal cells. Recent experiments have characterised GCT as consisting of three cell types: osteoclast-like multinucleated giant cells, round mononuclear cells resembling monocytes and spindle-shaped, fibroblast-like stromal cells [12]. These studies further

suggested that the stromal spindle cell is responsible for tumour proliferation and the giant cells and monocytes are the reactive components of the tumour [12, 13].

Compared with GCT of long bones, GCT of the skull lacks the characteristic signs of expansion and the “soap bubble” appearance [14]. Moreover, GCT in the skull tends to be aggressive. Radiologically, this form of GCT usually manifests as apparent cortical destruction of soft tissue. On CT scan, the appearance of reliquus bone flap within the tumour is difficult to differentiate from calcification in chondroitic bone tumours in the skull base. MRI can be used to assess intramedullary and soft-tissue extension. On MRI scan, the tumour’s signal is irregular owing to the onset of necrosis, haemorrhage and cyst formation. In this case, the characteristics of CT and MRI are similar to those in the long bone, which

Figure 2. The tumour tissue is indicated as isointense on T_1 weighted image (a) and as slightly hyperintense on T_2 flair (b), relative to the white matter. The expansile inner table of the skull is hypointensity on T_1 weighted image and T_2 flair (white arrow). The region of necrosis is hypointensity on T_1 weighted image and hyperintensity on T_2 flair (black arrow).

Figure 3. The tumour tissue is enhanced markedly by gadolinium-enhanced T_1 weighted MRI at the axial plane (a) and sagittal plane (b).

show extensive expansion, necrosis and cyst formation within the tumour and obvious enhancement post-contrast. It is proposed that these characteristics may be related to the tumour site. There is expansile space for GCT in the posterior cranial fossa relative to the skull base; thus, by the time a patient presents with obvious symptoms the volume of the tumour is relatively large. In addition, there is no overlapped structure in the posterior cranial fossa and the signs of the lesion are shown clearly.

Surgery and radiation is the general treatment strategy applied to GCT in long bones because of the high recurrence rate after surgery and the unresectable nature of the tumour. The management of GCT in the skull is dependent upon the tumour location. Because there are many important blood vessels and nerves surrounding the skull base, the fact that tumours of GCT are mainly located at sphenoid and temporal bone leads to total surgical excision being dangerous and unfeasible. It is therefore postulated that the treatment of surgical excision and radiation is a rational method for GCT at

the skull base. A series of cases reported by Bertoni et al [7] used treatment strategies involving surgical resection and radiotherapy. The effectiveness of treatment was satisfactory after follow-up. However, the GCTs had originated from other sites of the skull as well as the cranium, making total surgical excision possible. It is unclear as to whether the radiotherapy was ultimately necessary. Two cases involving the cranial vault, documented by Coumbaras et al [15] and Ulu et al [16], employed no post-operative radiotherapy and no recurrence was observed during the follow-up period.

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Figure 4. Haematoxylin and eosin staining of the tumour (original magnification $\times 20$) reveals large, multinucleated, giant cells uniformly dispersed among the oval- or spindle-shaped stromal cells.

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