Osteosarcoma Arising From the Skull
—Case Report—

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Abstract

A 20-year-old male presented with an osteosarcoma in the right parieto-occipital bone occurring as a painless occipital lump which had rapidly enlarged in the 6 months prior to admission. The neuroimaging appearance resembled intraosseous meningioma. Gross total resection of the tumor was achieved. The final histological diagnosis was osteosarcoma. Osteosarcomas of craniofacial region have a better prognosis than those of the skeletal bones, and distant metastasis is rare. Local recurrence is the most significant factor contributing to poor outcome. Complete excision with negative margins is the key to a better outcome. Adjuvant therapy may be an option in cases of incomplete excision. Advances in target chemotherapy may diminish the significant morbidity associated with these lesions.

Key words: osteosarcoma, skull, calvaria

Introduction

Osteosarcoma is one of the most common bone tumors excluding tumors of hematopoietic origin. Osteosarcoma tends to occur in the long bones of the extremities, whereas only 5% to 6% arise in the craniofacial bones. The vast majority of craniofacial osteosarcomas are located in the zygomatic bone, whereas tumors affecting the skull are rare. We report a case of osteosarcoma of the parieto-occipital bone and describe the radiological features, clinical symptoms, and surgical treatment.

Case Report

A 20-year-old male was admitted with a long history of a painless occipital mass which had rapidly swollen over the previous 6 months. Physical examination found a palpable bony hard mass beneath the scalp, 5 × 5 cm in diameter. His medical history did not indicate any history of Paget's disease, trauma, or irradiation to the head.

Radiography revealed an osteolytic lesion in the occipital bone (Fig. 1). Computed tomography (CT) of the head showed a heterogeneous mass in the parieto-occipital bones, with mild compression of the occipital lobe. Bone windows CT demonstrated a spicular formation of the bone (Fig. 2). Magnetic resonance (MR) imaging of the head revealed a well-circumscribed extra-axial mass, approximately 4 cm in the greatest dimension. T1-weighted MR imaging showed the lesion was composed of two hypoin-
tense areas. T₂-weighted MR imaging disclosed a hyperintense area surrounded by an isointense area. The isointense area was homogeneously enhanced by gadolinium-diethylenetriaminepenta-acetic acid (Fig. 3). Right external carotid angiography showed that the tumor was supplied by a posterior branch of the right middle meningeal artery (Fig. 4). These radiological findings indicated an intraosseous meningioma.

A right parieto-occipital craniotomy exposed the mass just under the scalp and bulging from the parieto-occipital bone, associated with bony destruction, forming an ossicular, tender, and cystic complex. The mass was dissected carefully from the adjacent dura. Gross total removal was performed with a negative margin.

Histological examination of the operative specimen found spindle-shaped cells with cartilaginous differentiation and bone formation. The reticular growth pattern of osteoblasts with associated lace-like strands of osteoid was observed. Epithelial membrane antigen staining was negative. The tumor was located on the surface of the bone rather than

![Fig. 2](image2.png)  
**Fig. 2** Computed tomography scans with contrast medium (left) revealing a densely calcified extra-axial lesion, and a heterogeneously enhanced mass on the bone window setting (right).

![Fig. 3](image3.png)  
**Fig. 3** T₁-weighted magnetic resonance (MR) images disclosing a well-demarcated extra-axial mass appearing as a hypointense area, apparently a cystic lesion, within a round isointense area in the occipital region (left) which was well enhanced by gadolinium-diethylenetriaminepenta-acetic acid (center). T₂-weighted MR image (right) showing the lesion as a hypointense cystic area.

![Fig. 4](image4.png)  
**Fig. 4** Superselective right middle meningeal angiogram, lateral view, showing the tumor mass is enriched with vascularity, and fed by the right middle meningeal artery with the tumor margin.

Neurol Med Chir (Tokyo) 43, February, 2003
Table 1  Summary of reported cases of osteosarcoma affecting the craniofacial region

<table>
<thead>
<tr>
<th>Site</th>
<th>No. of cases (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maxilla</td>
<td>183 (37.5)</td>
</tr>
<tr>
<td>Mandible</td>
<td>225 (46.11)</td>
</tr>
<tr>
<td>Skull</td>
<td>53 (10.86)</td>
</tr>
<tr>
<td>Skull base</td>
<td>9 (1.84)</td>
</tr>
<tr>
<td>Orbit</td>
<td>8 (1.64)</td>
</tr>
<tr>
<td>Sella</td>
<td>3 (0.61)</td>
</tr>
<tr>
<td>Others</td>
<td>7 (1.43)</td>
</tr>
<tr>
<td>Total</td>
<td>488 (100)</td>
</tr>
</tbody>
</table>

Including cases from references 1–10 and the present case.

Discussion

Fifty-three cases of osteosarcomas in the skull have been reported including our present case (Table 1). The occurrence of osteosarcoma in the craniofacial bones peaks in the third decade, whereas that in the skeleton peaks in the second decade. The etiology of osteosarcoma is unknown, but the major risk factors for development of osteosarcoma in craniofacial bones may be similar to those of the long skeletal bones, consisting of exposure to radiation, retinoblastoma, Li-Fraumeni syndrome, and Paget’s disease. Other bone abnormalities, such as fibrous dysplasia, multiple osteochondromatosis, chronic osteomyelitis, myositis ossificans, and trauma, have also been proposed as risk factors.

The general radiographic features are not specific, but may be osteolytic, osteoblastic, or mixed. A spicular pattern of calcification is usually present. The classic “sun ray” or “sun burst” appearance is also seen in 25% to 31% of patients. Osteosarcoma can be classified histologically into three main subtypes: the chondroblastic, osteoblastic, and fibroblastic types. Osteoblastic tumors generally belong to the high-grade tumors (grade 3 or 4), whereas chondroblastic and fibroblastic osteosarcomas are usually either grade 1 or 2. Osteosarcoma was defined as low grade because osteosarcoma is a spindle-cell tumor associated with irregular bone production, low cellularity, fewer than 4 mitoses per 10 high-power fields, and absence of pronounced atypia. However, there is no significant correlation between the histological grading.
of osteosarcoma and the prognosis. The presence of osteoids laid down by malignant stromal cells is regarded as the histological key feature of osteosarcoma.

The recommended treatment has not yet been established, but radical surgery for extirpation including a large margin of normal bone is the most significant factor contributing to a good outcome. Adjuvant therapy, such as irradiation or chemotherapy, can be performed if radical resection cannot be achieved. The role of chemotherapy is less clear for craniofacial tumors. The 10-year survival following resection of large tumors was 69% if the resection was complete, but was only 13% if excision was incomplete.

Local recurrence occurs in 25% to about 50% of cases, usually in the first year after the initial treatment. Distant metastases are seen occasionally. The metastasis usually occurs within the first 2 years. The lung is the major site. The skeleton, the liver, or the neck lymph nodes are predominantly involved. Early pulmonary metastasis usually caused death in patients with long bone osteosarcoma, whereas progressive local recurrence is the most common cause of death in patients with craniofacial osteosarcoma. However, the rate of distant metastasis in low-grade osteosarcoma remains unclear because of the lack of well-organized reports.

The radiological findings of the present case mimicked intraosseous meningioma. The final diagnosis of low-grade osteosarcoma was only established by the histological studies. The highly aggressive nature of the tumor, as evidenced by the high mitotic rate and rapid recurrence despite radical resection, prompts oncologists to perform aggressive chemotherapy and radiation therapy in an attempt to cure the lesion. However, the optimal treatment for the rare low-grade osteosarcomas, such as the current case, has not yet been established. Osteosarcomas in the skull bones presumably have a better prognosis than those in the other skull base or mandibular regions. However, we considered that only surgery for the low-grade osteosarcoma might not be curative. Advances in targeting chemotherapy may reduce the significant morbidity associated with these lesions.

### References


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