Unique Image Characteristics of an Occipital Primary Chondroblastic Osteosarcoma: A Rare Case Report and a Brief Literature Review

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Abstract

Keywords
► chondroblastic osteosarcoma
► occipital bone
► skull base
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► magnetic resonance imaging

Primary osteosarcomas of the skull and skull base are rare and comprise < 2% of all skull tumors. In head and neck osteosarcomas, the chondroblastic subtype occurs most frequently, which has an exceedingly poor outcome, but its image characteristic remains unknown. Herein, we report a case in the right occipital bone of the skull base and the unique characteristics of image. Pathologic examination of the surgical specimens led to the diagnosis of chondroblastic osteosarcomas. We believe those image characteristics can improve the understanding of skull chondroblastic osteosarcoma and the preoperative diagnosis.

Introduction

Osteosarcoma (OS) develops most frequently in the extremities, and it is the most common histologic form of the primary bone cancers.¹ ² Head and neck OSs are rare, comprising only 6 to 10% of all OSs.³ ⁴ They typically present in the third or fourth decade of life and comprise only 1% of all pediatric head and neck malignancies. The most common craniofacial sites affected by OSs are the mandible and maxilla, followed by the calvaria and then the skull base.⁴ ⁶ On cytology, OS can be divided into several pathologic types, including the pleomorphic, epithelioid, chondroblastic, small cell, mixed, and osteoclast-like giant cell types.⁶ In head and neck OSs, the chondroblastic type occurs most frequently.⁷

Skull base OSs can be challenging to resect and an aggressive surgical approach can result in poor cosmetic outcome.⁸ Imaging plays a crucial role in the diagnosis of each subtype of OS and ultimately in patients’ survival because the diagnosis is based on a combination of histopathologic and imaging features. The therapeutic options and prognoses for different types of OS differ from each other, so correct diagnosis is essential.⁹ ¹⁰ Magnetic resonance imaging (MRI) or computed tomographic (CT) scan should be used to assess the extent of the primary tumor.¹¹

In this case report, we describe a pediatric patient of occipital OS of the chondroblastic type. The chondroblastic type of OS has an exceedingly poor outcome.¹² However, the detailed imaging description of such cases have not been reported in the previous literatures. We present the CT, MRI, and enhanced MRI features of this case, followed by a brief review of the related cases reported in the previous literatures

Case Report

A 9-year-old boy was admitted to our hospital with a major complaint of a growing mass on his head. Physical examination found a firm and tough mass on the right occipital that showed no tenderness upon palpation. CT scan showed the right occipital bone to be irregularly thickened with fluffy and cloudy calcification, with a mass deriving from the
internal occipital protuberance extending toward the basilar part of the occipital bone, invading the neighboring jugular foramen, the sublingual neural tube, and the mamillary process. On MRI, the lesion was 4.5- × 5.5- × 6.5-cm in size with calcifications areas of hypointensity in T1- and heterogeneous in T2-weighted series. Contrast MRI showed peripheral and septal enhancement in the interior side of the tumor (Fig. 1). Significant mass effect was present, distorting the cerebellar hemisphere, pons, and the forth ventricle, which led to hydrocephalus, and the oppression of the sigmoid sinus and the transverse sinus. Histopathology examination reported lace-like osteoid material abutting the neoplastic cells (Fig. 2), corresponding to the features of chondroblastic OS, and occipital bone chondroblastic OS was the final definitive diagnosis. A subtotal resection of the tumor was performed followed by radiation therapy. The patient died after half a year of local recurrence.

Discussion
Craniofacial OSs are rare. They typically present in the third or fourth decade of life, account for fewer than 5% of OSs in children, and comprise only 1% of all pediatric head and neck malignancies. The most common craniofacial sites are the mandible and maxilla, followed by the calvaria and then the skull base. Our case in the right occipital bone of skull base is a very rare location. A search of the English language literature revealed 22 cranial OSs previously reported in children (Table 1): 12 calvarial tumors and 10 tumors of the skull base. The mean age of the pediatric patients with cranial OS was 12.2 years old in this table. The patient in our case suffered at a younger age. On cytology, OS can be divided into pleomorphic, epithelioid, chondroblastic, small cell,
mixed, and osteoclast-like giant cell types. Our case is a chondroblastic subtype, which occurs most frequently in head and neck OSs.

The etiology of OS is unknown, but the major risk factors for development of OS 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. The skull is a favored site for OS arising out of 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 presenting symptoms varied with the location of the tumors. The maxillary or cranial lesions usually produced no pain, which was in accordance with our case; however, mandibular tumors frequently presented with focal painful swelling. Other common presenting symptoms include headache, cranial nerve palsies, exophthalmos, and visual impairment due to different location of the tumor. CT best demonstrates tumor mineralization, especially when minimal, and it is usually able to demonstrate tumor extension into the soft tissues. Hemorrhage, necrosis, and unmineralized, chondroblastic, or fibroblastic components of the tumor will appear as areas of low attenuation on CT if present. Unlike any other conventional OSs, we see fluffy calcification in our case, and we believe it is a characteristic of OS. The osteoblastic subtype is most common with nearly 90% containing variable amounts of cloudlike opacities.

### Table 1

| Author and Year | Age at diagnosis | Location | Extent of resection | Adjuvant therapy | Follow-up | Outcome |
|-----------------|------------------|----------|---------------------|------------------|-----------|---------|
| Garland, 1945   | 17, M            | Occipital| NR                  | RT               | NR        | NR      |
| Reddy et al, 1973 | 8, F            | Occipital| Biopsy              | RT               | NR        | Dead, progressive disease |
| Goodman and McMaster, 1976 | 15, F         | Parietal-occipital | NR              | Chemotherapy and RT | 6         | Alive, disease free |
| Wang et al, 1981 | 17, M            | Frontal-occipital | NR               | RT               | 6         | Dead, progressive disease |
| Benson et al, 1984 | 11, M          | Frontal  | NR                  | Chemotherapy     | 12        | Alive, disease status |
| Sundaresan et al, 1985 | 11, M          | Parietal | STR                | STR              | 36        | Alive, progressive disease |
|                   | 13, F            | Skull base|                  |                  | 66        | Alive, disease free |
|                   | 11, M            | Parietal  | STR                | Chemotherapy     | 9         | Dead, progressive disease |
|                   | 15, F            | Frontal   | GTR                | Chemotherapy     | 144       | Alive, disease free |
| Mark et al, 1991  | 14, M            | Anterior skull base | NR           | Chemotherapy and RT | 12        | Dead, progressive disease |
| Shramek et al, 1992 | 8, M            | Parietal-occipital | GTR            | Chemotherapy and RT | 16        | Alive, progressive disease |
| Salvati et al., 1993 | 11, M         | Frontal-occipital | STR              | RT               | 9         | Dead, progressive disease |
| Chander et al, 2003 | 15, F           | Frontal   | GTR                | Chemotherapy     | NR        | NR      |
| Ellison et al, 1996 | 11, F           | Skull base| STR                | Chemotherapy     | NR        | NR      |
| Gadwal et al, 2001 | 9, M            | Sphenoid | NR                 | RT               | 9         | Dead, progressive disease |
|                   | 1, M             | Sphenoid  | NR                 | RT               | NR        | NR      |
| Chennupati et al, 2008 | 14, F         | Skull base| Biopsy            | Chemotherapy     | 12        | Alive, progressive disease |
| Kirby et al, 2011  | 16, M            | Parietal  | GTR                | Chemotherapy     | 5         | Alive, disease free |
| Oakley et al, 2011 | 15, M            | Anterior skull base | GTR            | Chemotherapy     | NR        | NR      |
| Ohno et al, 2011  | 14, F            | Anterior skull base | STR            | Chemotherapy     | 26        | Dead, progressive disease |
| Meel et al, 2012  | 10, M            | Sphenoid  | Biopsy             | Chemotherapy     | 18        | Alive, disease free |
| Caroline et al, 2014 | 14, M          | Parietal  | GTR                | Chemotherapy     | 16        | Alive, disease free |
|                   | 12, M            | Skull base| GTR               | Chemotherapy     | 12        | Alive, disease free |
| He et al, 2016    | 9, M             | Occipital | STR                | Chemotherapy     | 6         | Dead, progressive disease |

Abbreviations: GTR, gross total resection; NR, not reported; RT, radiation therapy; STR, subtotal resection.
in accordance with the current literature. Areas that demonstrate either a heterogeneous enhancement pattern or lack enhancement are the preferred sites for biopsy because they are more likely to contain both chondroid and osteoid elements that are necessary for the correct diagnosis.\textsuperscript{21,22} Chondrosarcomas show similar image characteristic, but they occur in an older age with a mean age of 57 years old. DWI can also help identify chondroblastic OS. Chondroblastic OSs also have significantly higher minimum and maximum apparent diffusion coefficient (ADC) values compared with other conventional OS subtypes, but they have a lower minimum ADC and similar maximum ADC value compared with chondrosarcoma.\textsuperscript{23}

Skull base OSs can be challenging to resect, and an aggressive surgical approach can result in poor cosmetic outcome. Thus, skull base tumors have a poorer prognosis than mandibular or maxillary tumors.\textsuperscript{3} Complete surgical excision is the mainstay of treatment of the primary tumor. Local recurrence is the main reason of treatment failure and a high mortality in head and neck OSs. Positive margins and a high excision is the mainstay of treatment of the primary tumor. Therapeutic regimens.

This subtype has some particular image characteristic, which helps surgeons identify before surgery and set early therapeutic regimens.

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