Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Pediatric Radiology

Occipital Chondroblastic Osteosarcoma in a Pediatric Patient: Imaging Features of a Rare and Aggressive Tumor

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DOI: 10.36347/sjmcr.2022.v10i11.009 | **Received:** 17.09.2022 | **Accepted:** 28.10.2022 | **Published:** 09.11.2022

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Abstract **Case Report**

Primary osteosarcoma of the skull base is a very rare entity, representing less than 2% of all skull tumors. The chondroblastic subtype is the most frequent in this particular localization, the outcome remains poor. Imaging features are not well illustrated in literature. We report the case of a pediatric patient presenting a primary osteosarcoma of the occipital bone revealed by an occipital lump and signs of cranial hypertension. Pathology examination confirmed the diagnosis of chondroblastic osteosarcoma.

Keywords: Osteosarcoma, occipital bone, computed tomography, magnetic resonance imaging.

INTRODUCTION

Osteosarcoma is the primary bone cancer; it mostly develops in the extremities. Skull base localization is very rare representing only 6 to 10% of all Osteosarcoma. The most common craniofacial localizations affected are the mandible and maxilla, followed by the calvaria and finally the skull base. Osteosarcoma can be divided into several pathologic subtypes: the pleomorphic, epithelioid, chondroblastic, small cell, mixed, and osteoclast-like giant cell types. Distant metastasis is generally rare, however local recurrence is frequent and it's a contributing factor in the poor outcome of these tumors. The guarantee for the best outcome is complete excision with negative margins. Adjuvant therapy is an option in cases with incomplete excision. Imaging techniques (CT scan and MRI) are essential in the diagnosis process; they also provide information regarding the extension of the tumor [1-3].

In this case report, we describe the case of a pediatric patient with occipital chondroblastic Osteosarcoma. We discuss the imaging features of this tumor on CT scan and MRI, followed by a brief review of the literature.

CASE REPORT

A 10-year-old girl was admitted for a mass on the occipital region of the skull growing for over a year. The mass has been increasing in size gradually. She also presented signs of intracranial hypertension. Her past medical history was unremarkable. Physical examination found a firm mass of the right occipital region beneath the scalp, with no tenderness upon palpation. Neurological examination showed neither sensory nor cranial nerve deficit.

CT scan showed a lytic lesion causing destruction of the left occipital bone as well as a heterogeneous invasive mass in the parieto-occipital bones invading the occipital parenchyma, the neighboring structures and the soft tissue with multiples calcification areas (Figure 1).

On magnetic resonance imaging (MRI), it was a well-circumscribed extra axial mass measuring 8x11x11 cm, it showed heterogeneous signal on T1-weighted and in T2-weighted images with restricted diffusion. Postcontrast the mass exhibited intense and heterogeneous enhancement. Mass effect was present, distorting the occipital lobes, the right ventricule and splenium of the corpus callosum. We also note invasion of the cerebellar tentorium and significant mass effect on the cerebellum hemispheres, which led to tonsillar herniation. The mass seems to invade the superior sagittal sinus, the confluence of sinus, transverse sinus and the vein of Galen (Figure 2). The extension to soft tissue was prominent.

The mass was partially excised. Histological examination showed lace-like osteoid material abutting the neoplastic cells suggestive of chondroblastic osteosarcoma. The patient received adjuvant chemotherapy and died few months after.

Figure 1a

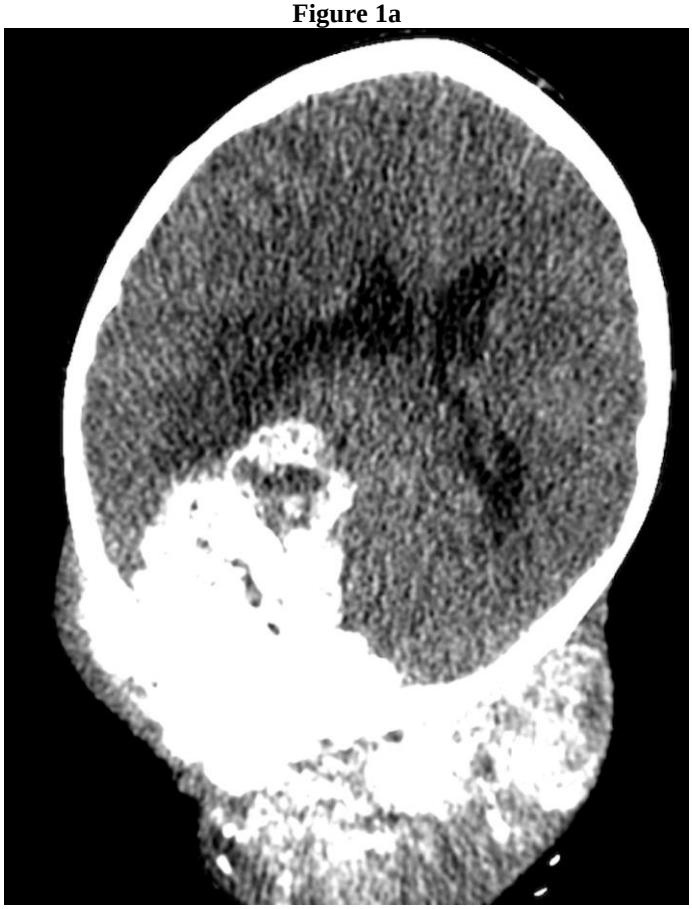

Figure 1b

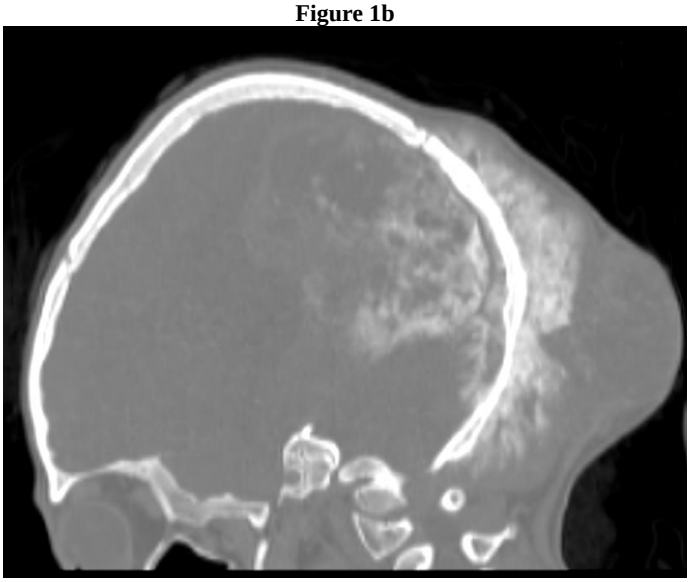

Figure 1: Axial section in the parenchymal window (a) and sagittal section in the bone window (b) showing a voluminous osteolytic process of the occipital scale, poorly limited, with irregular contours, heterogeneous, with calcifications.

Figure 2 a

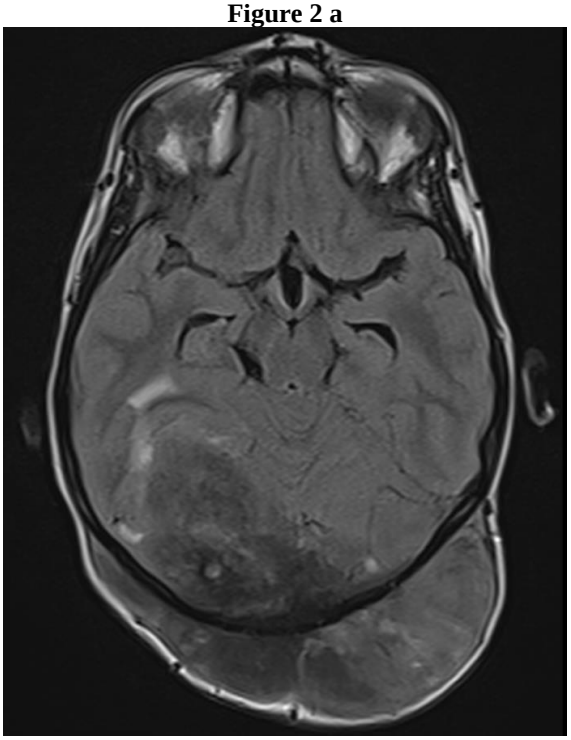

Figure 2b

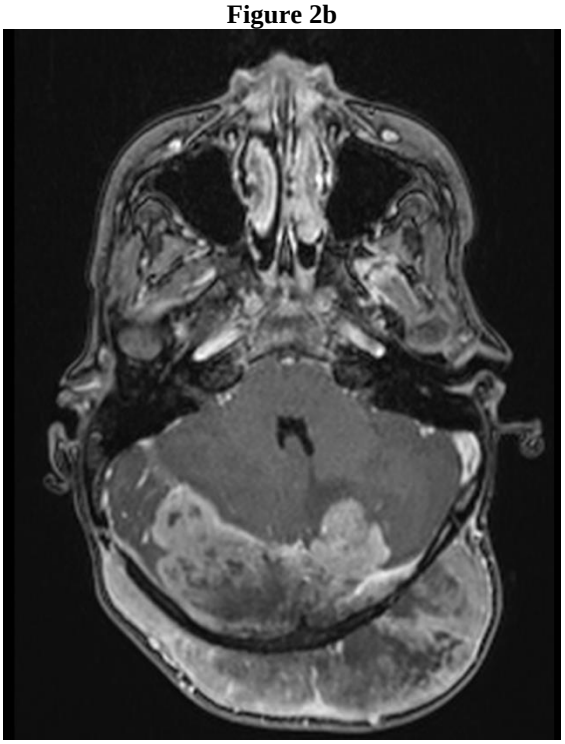

Figure 2: Axial section in T2 FLAIR sequence (a) and in T1 axial section after injection of Gadolinuim (b) showing a voluminous tumor process of the occipital scale with endo and exocranial development, poorly limited, with irregular contours, heterogeneous signal, seat of areas in T2 signal void

DISCUSSION

Primary osteosarcoma of the skull is rare, only few cases have been reported. They usually present in the third or fourth decade of life, occurrence in the pediatric population is less common. Causing factors may include radiation exposure, Paget's disease, Li-Fraumeni syndrome, and retinoblastoma. However the etiology remains uncertain [3]. The revealing symptoms are variable depending on the location of the tumor; patients usually report headache, cranial nerve palsies, visual impairment and signs of cranial hypertension. Similar to osteosarcoma of the extremities the mass

tends to have a slow growth [4]. However unlike osteosarcoma of the extremities; distant metastases are quite rare compared to local recurrence [5, 6].

CT scan using bone windows in the key to diagnosis, it shows bone growth with lytic regions as well as periosteal remodeling [4, 7, 8]. It also demonstrates tumor extension into soft tissue. The presence of the necrosis, hemorrhaging and unmineralized, chondroblastic or fibroblastic components should be noted. A similar case was published by the journal of neurological surgery reports, noted the presence of fluffy calcifications and suggested it was characteristic of this tumor [9]. In our case, the tumor displays multiple fluffy calcifications.

MRI is especially used to assess soft tissue involvement. The mass usually presents isointense signal on T1-weighted, hypointense on T2-weighted with well-defined margins [10]. In our case the mass showed heterogeneous enhancement post contrast. Literature suggests no enhancement or heterogeneous enhancement in most areas of the tumor [9]. MRI can also help deciding the site of the biopsy; usually areas that present heterogeneous enhancement or no enhancement are the preferred sites because they usually contain both chondroid and osteoid elements [11]. Diffusion weighted images can be helpful to identify chondroblastic Osteosarcoma. These tumors have significantly higher minimum and maximum apparent diffusion coefficient (ADC) values compared with other conventional osteosarcoma subtypes, but they have a lower minimum ADC and similar maximum ADC value compared with Chondrosarcoma [12]. The aggressiveness is assessed by its tendency to extend beyond the bone margins and invade the surrounding soft tissues [3].

Skull base osteosarcoma can be aggressive and difficult to resect, therefore it has a poor prognosis compared to other localizations that are more accessible. In our case the tumor could not be entirely removed and patient underwent chemotherapy and died few months after.

CONCLUSION

Skull base chondroblastic osteosarcoma is a rare aggressive cancer with poor prognosis. Determining the different imaging features will help identify and assess the extension of the tumor and thus suggest early and appropriate treatment plans.

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