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CASE REPORT

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Uncommon Presentation of a Giant Cell Tumor of the Skull in a Pediatric Patient: A Case Report

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ABSTRACT

Giant cell tumours (GCTs) of the bone rarely affect the cranium and are even more unlikely to occur in preschool children. Due to its local aggressiveness and high recurrence rates, the entity is classified as an intermediate malignant tumour. We report on a unique cranial GCT case in a 5-year-old female who presented with a 8 month history of progressive swelling of the left side of calvaria and right lower limb monoparesis. A cranial computerized tomography (CT) revealed a large contrast-enhancing osteolytic bony mass of the left parieto-occipital region with intracranial extension. Extensive systemic workup for malignancy yielded negative results. The patient underwent a gross total resection of the tumour via a left parieto-occipital craniectomy, followed by titanium mesh cranioplasty. Histology was consistent with a bone GCT, including positive CD68 and vimentin immunostaining, lack of expression of p63, and Ki67 less than 2%. At 2 months follow-up she is neurologically intact. This report of an unusual clinical presentation and successful management of a giant pediatric cranial GCT contributes valuable insights to the existing literature on the topic.

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Introduction

Giant cell tumour (GCTs) of the bone are benign but locally aggressive rare neoplasms that primarily occur in the epiphysis of long bones [1]. The skull is a rare site of occurrence accounting for <1% of cases, most commonly in the sphenoid and temporal bones [1]. The tumor typically occurs in the third to fourth decades of life and is rarely seen in patients under 20 years of age. Pediatric and adult GCTs share the same characteristics [2,3]. The tumour radiographically appears as an osteolytic lesion and is histologically defined by numerous multinucleated giant cells that are diffusely distributed among a background of mononuclear histiocytic cells and giant cell tumour stroma cells [4]. Positive immunohistochemical stains include H3G34W, p63, SATB2, and CD68 [5,6]. Gross total resection is the most commonly recommended treatment choice [7]. Considering the rarity of intracranial GCTs in the pediatric age, we report a unique case and describe its clinical manifestation, radiological features, histological and immunohistochemical findings, and surgical management. Thereby we aim to contribute valuable insights to the existing literature on pediatric cranial GCTs.

Patient Information

A 5-year-old female presented to the Lagos State University Teaching Hospital with the complaint of progressive bony swelling on the left side of the scalp and right lower limb weakness persisting for 8 months.

Clinical Findings

A 7 x 6 x 4 cm painless left parieto-occipital bony mass and right lower limb monoparesis of power grade 3, were found on examination. Other medical history and neurological examination were unremarkable. A contrasted cranial computerized tomography (CT) scan revealed a distinct 80 x 72 x 42 mm osteolytic bony mass of the left parieto-occipital region, with an intracranial extension, displaying avid enhancement. Some mass effect was visible (Figure 1). Considering the medical history and physical examination findings, the radiology report listed solitary fibrous tumour as the most probable primary diagnosis. A comprehensive workup to rule out metastatic disease yielded

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negative results.

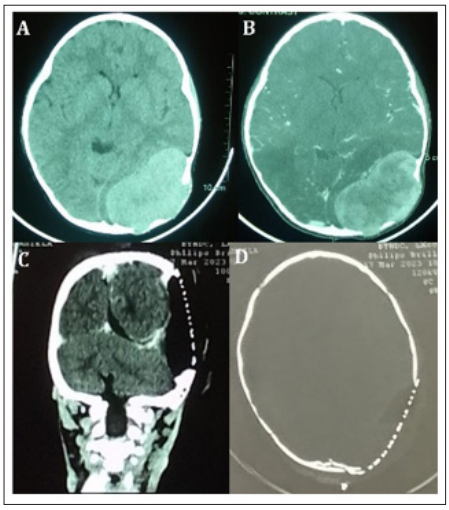

Figure 1: Preoperative and Postoperative Computerized Tomography Findings

A: An extensive hyperdense expansive process of the left

parietooccipital region, destructive to the bone and extending noticeably into the intracranial

space.

B: The underlying brain parenchyma is displaced and edematous. Contrast-enhanced

image shows contrast uptake.

C: Postoperative coronal section shows complete tumor removal and no damage to the underlying parenchyma and axial section

D: normal bone structure at the resection margin and osteosynthetic material used in reconstruction.

Timeline

Timeline of treatment course is summarized in Figure 2.

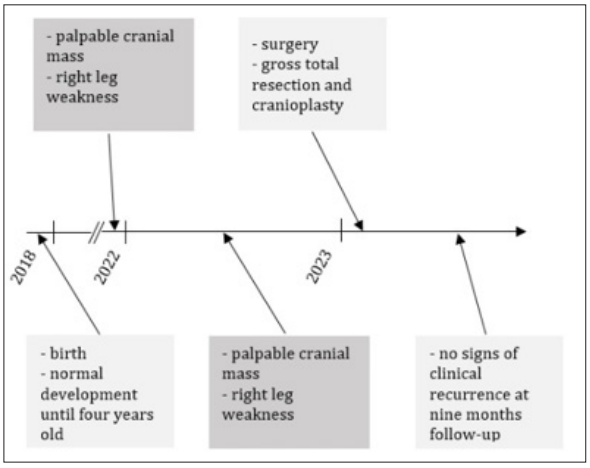

Figure 2: Timeline of Events

Therapeutic Intervention

The patient was scheduled for a procedure under general anesthesia aimed at total removal of the lesion via a left parieto-occipital craniectomy, followed by primary cranioplasty with titanium mesh. Intraoperatively, the left parietal and temporal bones appeared paper thin, with areas of complete bone erosion and a visible underlying tumour. The tumour was a well circumscribed extradural grayish-yellow mass (Figure 3), with heterogeneous soft-to-firm consistency. There was no dural infiltration. Gross total resection of the tumour was achieved (Figure 3). The resulting parieto-occipital bone defect was reconstructed using a titanium mesh (Figure 3). The course of surgery was uneventful, therefore the patient was extubated in the operative theater and placed in a room at the neurosurgical ward. Postoperative recovery was uneventful and she was discharged 8 days after the surgery.

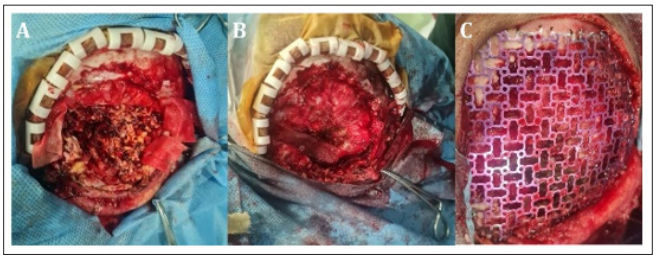

Figure 3: Intraoperative Findings

A: Features typical of a giant cell tumor - red-brownish cut color, nodular, circumscribed, friable, destructive to the bone, without sclerosis of the bony edge or periosteal reaction.

B: The underlying dura is not invaded.

C: Postoperative bony defect was reconstructed using a titanium mesh.

Histopathological examination of the tumour sections showed a dense fibro-collagenous tissue with multiple osteoclast-like giant cells within the lesion. The stroma was composed of spindle and round to oval mononuclear cells, including macrophage-like and primitive mesenchymal cells with a poorly defined cytoplasm and few mitotic activities. There was mild infiltration by lymphocytes, plasma cells, and hemosiderin histiocytes. Multiple vascular channels were seen with focal fibrosis and degenerate bone. These features were indicative of a bony GCT (Figure 4). Immunostaining with CD68 and vimentin was positive, and there was no p63 expression (Figure 4). The Ki67 proliferative rate was less than 2%.

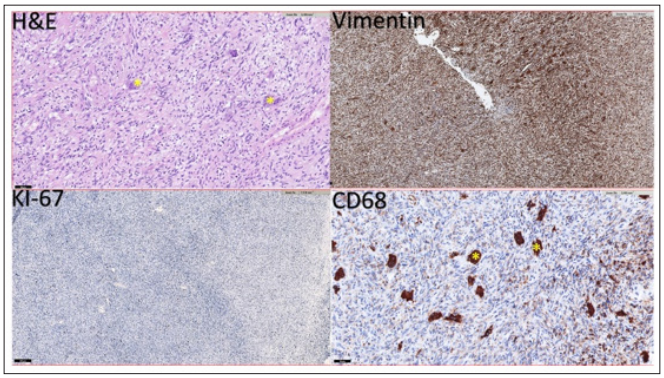

Figure 4: Histological/Immunohistochemistry Findings

Histology shows a highly cellular lesion populated by giant multinucleated cells (asterisk), Vimentin and CD68 positive staining, and low (<2%) Ki-67 index.

Follow-up and Outcomes

The patient regained full motor strength at four weeks post-surgery. Currently, nine months post-surgery, she remains in good overall conditions, with no clinical or radiological evidence of recurrence.

Discussion

Giant cell tumors (GCTs), also known as osteoclastoma, are remarkably rare in children, and in the rare instances of pediatric occurrence they affect the epiphysis of long bones, that is bones that form via endochondral ossification [1]. The skull is a rare site of occurrence, accounting for <1% of cases. Common GCT sites in the skull are the temporal and sphenoid bones, which develop through endochondral ossification [8]. In this communication, we reported a parietal bone GCT, which is a remarkably rare site of occurrence in the skull. Even though bone GCTs are considered benign lesions, they can be locally aggressive invading the surrounding tissue1. Further, a malignant variant has also been reported, with metastasis to the lungs [9].

In our case, histological criteria, including Ki67 levels, as well as intraoperative findings, were indicative of a benign GCT. Since gross total resection (GTR) was achieved, we consider the surgery to be curative. Previously, GTR has been established as the mainstay of GCT treatment. In case GTR is not attainable, adjuvant radiotherapy (RT) and or chemotherapy (CT) have been proposed as adjuncts [10]. However, CT and RT are both burdened by adverse effects that lower the quality of life in children. Weng et al. have recommended adjuvant RT to patients with malignant pathology and a Ki-67 index ≥10% regardless of the extent of resection, residual tumours regardless of the Ki-67 index, and recurrent bone GCT [11]. Although RT is associated with longer overall survival, it also causes delayed morbidity, such as endocrine dysfunction, secondary malignancy, and neurocognitive decline [12]. Advancements in photon-based radiotherapy have minimized these effects [13]. Bocanegra-Becerra et al. reported no recurrence over the time of follow-up in the majority of patients that had either GTR or subtotal resection (STR) + RT in a review of the management of GCT of the occipital bone [10]. Our patient had a GTR with a follow-up of nine months so far, with no clinical or radiological evidence of recurrence. Different markers, such as H3G34W, p63, SATB2 and CD68 have been found to be positive in GCTs. Among these, H3G34W is considered a reliable marker [14-16]. In our case, we were able to obtain immunohistochemical staining for P63, CD68 and Ki68. H3G34W staining was unavailable in our institution at the time of this report.

Informed Consent

The parent of the patient consented to reporting this case.

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