**ENDOVASCULAR INTERVENTION IN A CASE OF PRIMARY OSTEOSARCOMA OF THE SKULL BASE IN A MALE CHILD – A CASE REPORT**

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**KEY CLINICAL MESSAGE:**

Skull base osteosarcomas constitute ~ 2% of all brain tumors, with limited treatment strategies. Pre-operative embolization reduces vascularity, resulting in reduced operative time, bleeding and hence better prognosis.

**ABSTRACT:**

**INTRODUCTION:** Osteosarcoma is the most common type of malignancy seen in bones. They mostly affect the metaphysis of long bones. Skull-affected osteosarcoma accounts for 6-10% of cases, mainly affecting the skull's calvaria part.

**CASE REPORT:** A 14-year-old male child presented with symptoms of swelling behind the left pinna, tenderness over the swelling, and right facial deviation. A skull base osteogenic osteosarcoma was identified during diagnostic testing.

**DISCUSSION:** Only 2% of all brain tumors are skull base osteosarcomas, and there aren't many recommended therapy strategies in the literature. Surgery to remove the tumor is a regular practice. But in our case, we employed pre-operative embolization to decrease vascularity, which reduced the amount of bleeding during surgery and cut down on the operating time.

**CONCLUSION:** Embolization before surgery demonstrated a significant benefit due to decreased vascularity, and this can be employed as an effective strategy to reduce intra-operative bleeding and operating time.

**Keywords: Osteosarcoma, Skull Base, Endovascular, Pediatric**

**INTRODUCTION:**

The most prevalent malignant bone tumor is osteosarcoma, which primarily affects children's and adolescents' long bones. However, 6-7% are present in the head and neck. [1] in the pediatric age group, they are primarily in the mandible [2]. The incidence of osteosarcoma is approximately 1:100,000 per year [3], and it can be classified as long-bone osteosarcoma or head and-neck osteosarcoma by location. At the same time, para-osteal sarcomas are rare but tend to reoccur with simple local surgical excision [4]. Unfortunately, the presenting symptom was painful swelling in 51% of cases [5], which does not help differentiate it from benign or malignant. Thus, radical surgery with comprehensive treatment was suggested [6]. The degree of cellular atypia and discernible histologic architecture is used to grade osteosarcomas [2].

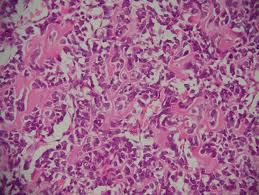
Skull base osteosarcoma as a primary tumor is uncommon, accounting for approximately 1%–2% of all cases [3,7]. Paget's disease and retinoblastoma are predisposing factors [3,8]. It occurs frequently following radiation or chemotherapy for various neoplasms [2]. The clinical characteristics and course of treatment for skull base osteosarcoma have not been well reviewed [6,9]. There is still a debate over the best therapeutic approach and the clinical characteristics of skull-base osteosarcoma. Only 120 cases of primary osteosarcoma of the skull have been documented in the literature since Garland first reported the first case in 1945 [7,10]. The majority of studies investigating its prognosis and treatment have only included case reports and small series due to the rarity of this tumor, particularly in the context of the skull base [9].

**CASE PRESENTATION:**

A 14-year-old male child presented with complaints of facial deviation to the right side for one-month, pulsatile swelling with bruit behind the left pinna for four months, and pain over the swelling for three months. It initially began as a swelling behind the left ear that progressed over time. There was no prior history of comparable swelling, ear discharge, fever, trauma, or chronic suppurative otitis media. On examination, the left pinna is pushed outwards by the swelling on the left occipital and left the mastoid area. The swelling was pulsatile firm in consistency. Its edges were ill-defined and included the temporal and occipital bones. The swelling measured 7\*6 cm in size. Bruit was present over the swelling. The Romberg’s test was positive, the patient swayed to the left while walking, and there was left conductive deafness, left lower motor neuron (LMN) facial palsy, and incoordination on the left side.

Investigations include: Blood test revealed hemoglobin levels of 13 gm/dL, an ESR of 28 mm/hr, and a normal ALP. Eosinophilia was detected in the peripheral smear. The bone marrow test revealed hematopoietic cells that were regularly growing, a slight increase in eosinophilic precursors, and no abnormal cells. A left occipital and temporal bone hyperdense lesion with compression across the left cerebellar hemisphere was identified on CT imaging. This has been hypothesized to be histiocytosis or sarcoma.

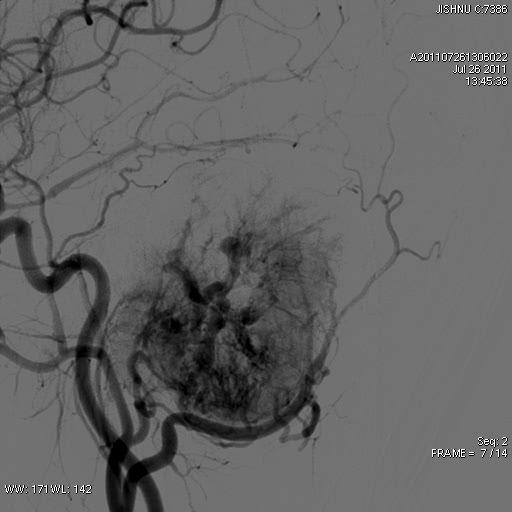
A later MRI scan revealed a moderately sized expansile space-occupying lesion, moderate enhancement noted arising from the left temporal bone and adjacent part of the occipital bone, lobulated intracranial component noted causing marked compression of the left cerebellar hemisphere, intact dura and displaced medially, and moderately sized soft tissue component seen bulging inferiorly. Hemangioendothelioma, intraosseous meningioma, and osteosarcoma are differential diagnoses. The results of the histopathological analysis in (Figure 1) indicated Osteogenic Sarcoma and confirmed the diagnosis.



**Figure 1: Histopathological of the specimen showing Sarcomatous tumor cells with eosinophilic nuclei resembling Osteogenic sarcoma of the skull base**

The treatment involves complete surgical resection of the lesion, with tumor-free margins, chemotherapy, and radiotherapy. To shorten the length of the procedure and reduce vascularity, pre-op embolization was performed. 500–700 microns of PVA dissolved in opac (iodine 300 contrast). To avoid unintentionally embolizing cerebral arteries, it was required to catheterize only specific tumor-supplying branches and then carry out a 4F slip catheter (cook). Embolization of the left occipital artery is seen in Figure 2.

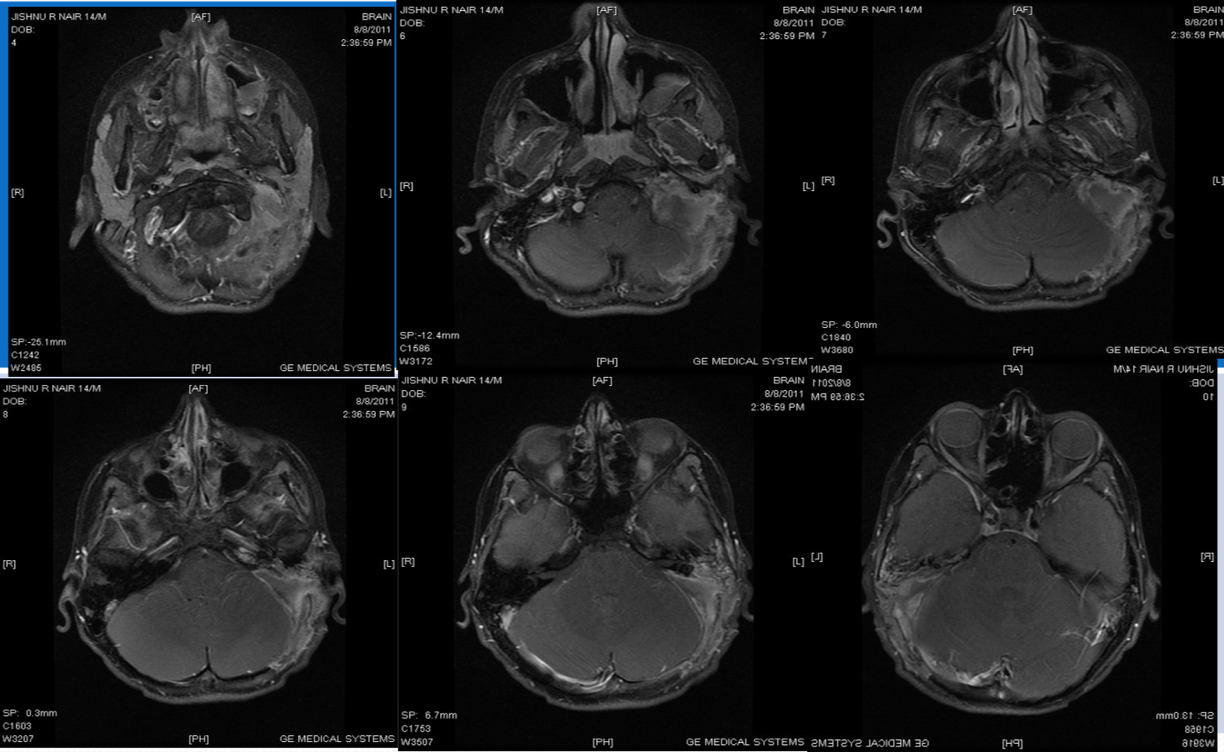
**Figure 2. Embolization of the Left occipital artery branch of the external carotid artery**



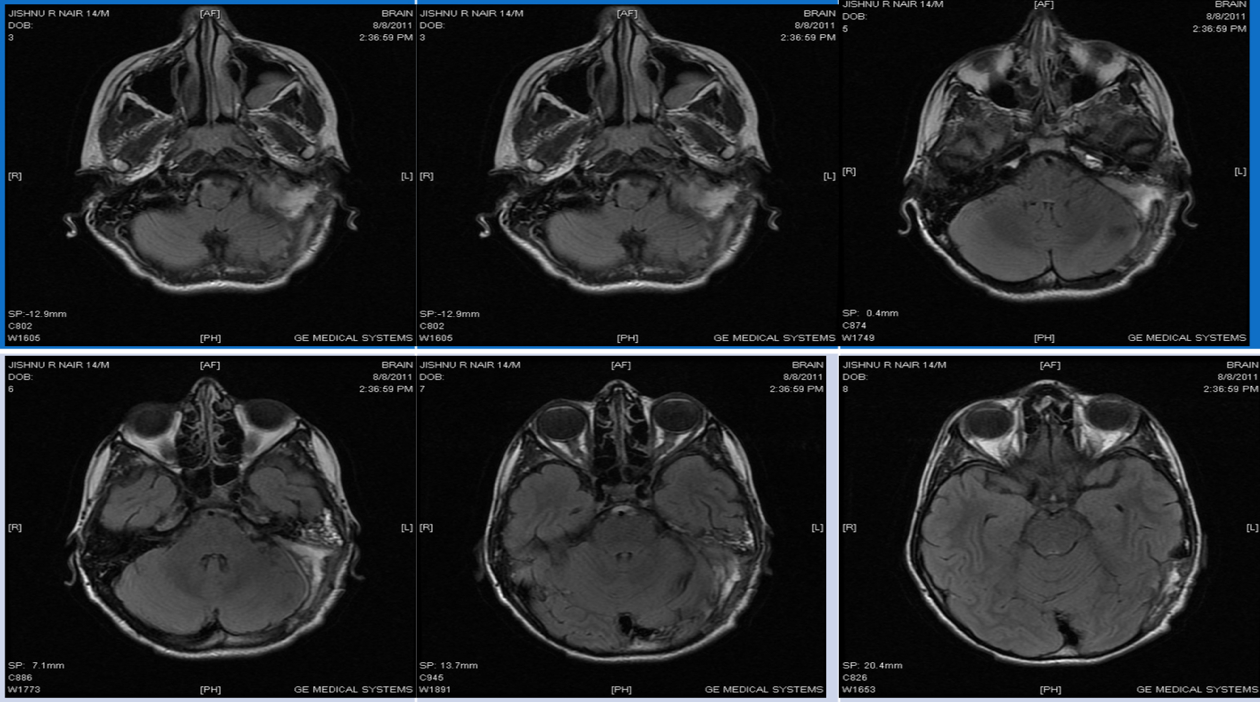
**Figure 3: Shows Pre-embolization alteration**



**Figure 4: Shows Post-Embolization alteration**

Following embolization, he received 1g of methylprednisolone IV and 5 days of antibiotics. Two days later, he underwent Left occipital craniotomy after obtaining written informed consent. Under general anesthesia, the tumor was removed near totally. Figures 5 and 6 depict post-operation MRI T1 and T2 following surgery. The surgery was without any complications, and no post-surgical complications were seen. The patient was asymptomatic and improving.

**Figure 5: Post-surgery T1 weighted MRI**



**Figure 6: Post-surgery T2 weighted MRI**

**DISCUSSION:**

An accurate assessment of the tumor's intracranial extension and the level of ossification and calcification is made possible by a cranial Computed Tomography (CT) scan, which is helpful for diagnosis and follow-up. [6] Magnetic Resonance Imaging (MRI) often reveals vascular irrigation and tissue surrounding the tumor, but it does not detect calcifications as accurately as CT (2,6).  The preferred course of treatment is systemic chemotherapy with cisplatin, methotrexate, and doxorubicin combined with adjuvant radiotherapy or with surgery that involves complete removal of the original lesion, leaving a significant margin of bone tissue tumor-free [10,11]. At the same time, there were attempts to remove the tumor with subtotal resection or gross total resection with free margins confirmed on histology [12].

Primary osteogenic sarcomas of the skull account for only 1.6% of all osteosarcomas and are uncommon. [13]. Osteogenic sarcomas of the skull are exceedingly common in children and the later decade of life when appendicular osseous tumors first appear [13,14]. It manifests as a slowly expanding, painful, or uncomfortable cranial mass. Surgical treatment is the most critical determinant in improving survival [13,15]. However, complete resection of the lesion is seldom achieved. Furthermore, widespread agreement on safety precautions for cranial surgery needs to be widespread. Surgery may be considered initially for minor lesions or if it is possible to produce negative margins. Tumors greater than 5 cm, those with poor histological response to neoadjuvant treatment, partial resection with positive margins, and cerebral invasion are among the prognostic variables contributing to the outcome. [16]

Our patient presented with facial deviation to the right, swelling, and pain behind the left pinna. Upon examination, it was found that the patient swayed. At the same time, walking had left lower motor neuron facial palsy, left conductive deafness, and could not coordinate the left side of the nasal test. There aren't many therapy techniques described in the literature, which is uncommon. The mainstay therapy is surgical excision by craniotomy or adjuvant chemotherapy or radiotherapy. Histological findings and radiological investigations such as CT and MRI scans are used to diagnose definitively. Despite fewer treatment choices, surgical excision is still the most preferred approach [17,18]. Positive surgical margins and high-grade tumors were associated with deleterious effects on survival. [19] Wide surgical margins and complete surgical excision have been associated with improved survival [19,20]. Patients with de novo osteogenic sarcomas were much younger than those with secondary lesions and had a better median survival rate [21]. Non-adjunctive chemotherapy could be started before surgery which could be helpful [22]. In our case, embolization was carried out before surgery to reduce the vascularity and duration of the surgery for a near total resection.

**CONCLUSION:**

Osteogenic osteosarcomas are rarely found in the skull and account for just 2% of skull tumors. There are not many strategic operative methods in the literature. In this instance, preoperative embolization was carried out to facilitate and speed up the surgical removal of the tumor. Since the scarcity of treatment guidelines regarding the Osteosarcoma of the skull, it is our observation that endovascular intervention yielded a better result than the conventional technique because it reduces the bleeding and decreases the complications post-surgery.

**CONFLICTS OF INTEREST:**

None declared.

**AUTHOR CONTRIBUTION:**

All the authors contributed equally in drafting, editing, revising and finalizing the case report.

**ETHICAL APPROVAL:**

The ethical approval was not required for the case report as per the country’s guidelines.

**CONSENT:**

Written informed consent was obtained from the patient to publish this report.

**DATA AVAILABILITY STATEMENT:**

The data that support the findings of this article are available from the corresponding author upon reasonable request.

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