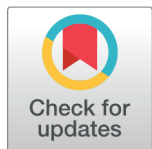


## CASE REPORT



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## Congenital cavernous hemangioma of the skull - A rare case in infants

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### Abstract

Reporting a rare case of Calvarial Cavernous Hemangioma. A 2-months-old-male baby presented with Right Frontal swelling noticed when baby was 20days old rapidly progressive in size. On examination, it was solitary 6\*7cm firm irregular swelling fixed to bone which On CT/MRI showed 6\*5\*4cm mixed density extra axial lesion in right frontal region with bony destruction causing mass effect and mid line shift? neoplastic. Baby had no neurological deficits. CBC showed Pancytopenia. Patient was operated under GA, total excision of the lesion and margin of surrounding eroded bone was performed. Tumor was completely extradural. Histopathology showed Bony trabeculae separated by loose fibrocollagenous tissue enclosing many endothelium lined Vascular spaces. Post operative no recurrence is observed. Cavernous Hemangioma of skull is very rare in infancy and must be included in differential diagnosis of skull lesions . Untreated Cavernous Hemangiomas may show progression. A suspected Neoplastic swelling turned out to be benign lesion. Timely operative intervention prevented complications due to severe anemia because of intralesional bleed.

**Keywords:** Calvaria; Cavernous Hemangioma; Benign

### Introduction

Cavernous hemangiomas (CHs) of the skull are rare tumors arising from the intrinsic vasculature of the bone and are mostly seen in middle age. The incidence of CH in infancy is low comprising 0.7% of all bone neoplasms and the most common site is the vertebral body, followed by the skull. We report a rare case of Cavernous hemangiomas of skull in 2-month-old infant.

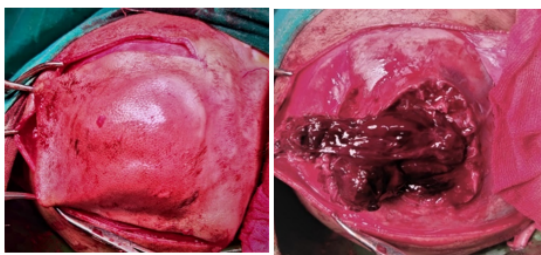
### Case report

A 2-months-old-male baby presented with Right Frontal swelling noticed few days after birth, initially waited for spontaneous resolution as other scalp swellings but it rapidly progressed in size. On examination it was solitary 6cmx7cm firm consistency smooth bosselated immobile swelling in right frontal region.

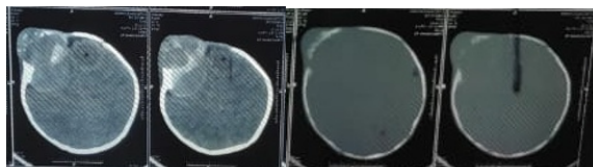
Baby had no developmental delays and no neurological deficits. Radiological investigations showed 6X5X4cm Mixed density Extra axial lesion in Right frontal region with bony destruction causing mass effect and midline shift? malignant lesion. Blood investigations showed decreased hemoglobin and thrombocytopenia.

Patient underwent surgery under general anesthesia. The lesion was completely excised along with margin of surrounding eroded bone. The lesion was extradural with ragged bony edges comprising of clotted blood but no fleshy mass. Underlying dura and brain tissue was unremarkable.

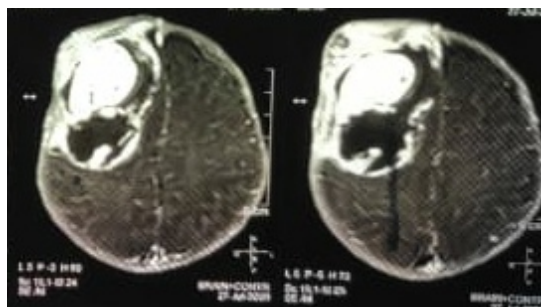
Histopathology revealed Bony trabeculae separated by loose fibrocollagenous tissue enclosing many endothelium lined vascular spaces, consistent with intraosseous cavernous hemangioma.



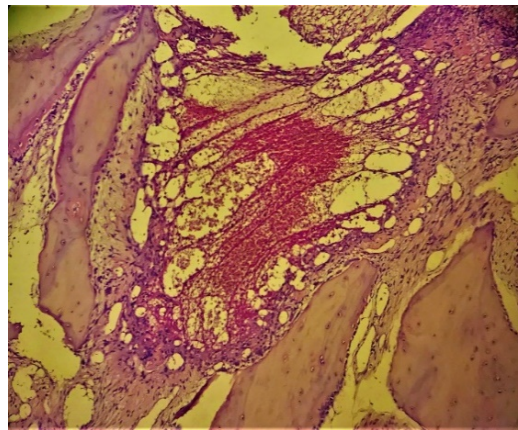
**Fig 1.** Intraoperative images showing clotted blood with ragged bony edges



**Fig 2.** CT images



**Fig 3.** MRI images



**Fig 4.** Histopathological image

## Discussion

Primary Intraosseous cavernous hemangiomas are uncommon neoplasms and comprise 0.7% of all bone tumors. The most commonly involved site is the vertebral column, followed by the skull. Hemangiomas of the calvarial bones account for 0.2% of all bone neoplasms. The parietal and frontal bones are regions of the calvaria most frequently affected by these tumors. The tumors are more common in the female population than in the male population, by a ratio of 2–4 to 1. Although they may be seen in any age group, the peak incidence is during the 2<sup>nd</sup> through 4<sup>th</sup> decades.<sup>(1)</sup> Occurrence of these tumors in the neonatal period is extremely rare.

The bones of the vault of the skull first appear at about Day 30 of gestation. Each parietal bone is ossified from 2 centers, which appear in dense mesenchyme near the tuberosity, one above the other, at about the 7th week in utero. The skull is unilamellar at birth without any diploe. The diploic veins start to develop at around 2 years of age along with the diploe.<sup>(2)</sup>

In the intrauterine period, defective differentiation of primordial vessels, resulting in an abnormal capillary bed, may guide the development of hemangiomas. Calvarial cavernous hemangiomas arise from vessels in the diploic space and are supplied by the branches of the external carotid artery, arising in the skull vault. The middle meningeal and superficial temporal arteries are the main sources of blood supply. Trauma is not thought to be a predisposing factor in the development of these lesions.

The symptoms may be variable depending on the location of the tumor. The most frequent symptoms or signs in calvarial cavernous hemangioma are pain and/or noticeable or palpable bone deformity. Our patient had a variable consistency mass in the right frontal region observed few days after birth.<sup>(2,3)</sup>

In pediatric patients, the differential diagnosis should involve congenital lesions such as encephaloceles and sinus

pericranii; inflammatory lesions such as abscesses; traumatic lesions such as skull fractures, cephalohematomas, and leptomeningeal cysts; neoplastic lesions such as osteoblastoma, desmoplastic fibroma, infantile myofibroma, cranial fasciitis, fibrous dysplasia, lymphoma and metastatic lesions arising from neuroblastoma, rhabdomyosarcoma, fibrosarcoma, angiosarcoma; and other conditions, including epidermoid/dermoid cysts, Langerhans cell histiocytosis, fibrous dysplasia, osteoma, eosinophilic granuloma, aneurysmal bone cyst, and meningioma.<sup>(2)</sup>

The investigation of choice is CT scan which classically depicts lytic expansile and bubbly lesion with a sclerotic rim also known as “sunburst” or “spoke-wheel” appearance. CT also helps in delineating the extent of tumor in to surrounding structures. Magnetic resonance imaging (MRI) signal intensity depends on the amount of venous stasis in the lesion and also on the rate of red bone marrow in to yellow marrow. The lesions enhance on contrast administration intensely on both CT and MRI.<sup>(4,5)</sup>

Histopathology of cavernous hemangioma shows unencapsulated dilated sinusoidal channels that are lined by a single layer of flattened endothelium and are interspersed among bony trabeculae. Preoperative diagnosis of cavernous hemangiomas is often difficult because imaging findings are not specific. Histopathological examination is often necessary to confirm the diagnosis.

The gold standard for the treatment of these tumors is total resection of the affected bone along with a surrounding normal bone edge to ensure that no remnant of the lesion has been left, because any remnant of the tumor may cause recurrence. Although preoperative embolization is recommended to prevent excessive intraoperative bleeding, we did not perform embolization in our case because no definite diagnosis could be established preoperatively. Radiotherapy is known to suppress tumor growth but not

reduce the size of the tumor. Because the resection in our case was total, the patient was not subjected to radiotherapy.<sup>(2,3)</sup>

In the pediatric population, cranial defects often require cranioplasty to avoid progressive soft-tissue depression deformities that may lead to neurological deficits. Allografts and autologous grafts are the options for cranioplasty, and autologous bone grafts should be the first choice in children.<sup>(2)</sup> Allograft materials used for cranioplasty are metallic mesh plates, methylmethacrylate, hydroxyapatite, and high-density porous polyethylene. Cranioplasty was not done in our case since malignancy was not ruled out.

## Conclusions

Although it is extremely rare in the newborn, cavernous hemangioma of the calvaria should be included in the differential diagnosis of skull tumors because surgery provides definitive cure for the disease.

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