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Giant primary cavernous hemangioma of the skull in an adult A rare calvarial tumor a case report.

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Abstract :
Primary intraosseous cavernous hemangiomas (PICHs) of the cranium are rare benign vascular tumors that account for about 0.2 of all bone tumors and 10 of benign skull tumors. They generally present as osteolytic lesions with honeycomb pattern of calcification. A 24-year-old female presented to us with a large right parietal skull mass that had been present for the last 12 years. Total resection of the lesion was performed. Pathological examination was suggestive of cavernous hemangioma of the skull bone. Cavernous hemangioma should be considered as one of the differential diagnosis in any case of bony swelling of the calvarium so that adequate preoperative planning can be made to minimize blood loss and subsequent morbidity.
Keyword : Adult, cavernous hemangioma, calvarial mass, skull lesion.

Introduction :
Primary intraosseous cavernous hemangiomas (PICHs) of the cranium are rare benign vascular tumors that account for about 0.2% of all bone tumors and 10% of benign skull tumors.(1,2) These typically present as osteolytic lesions in the calvarium with sunburst / honeycomb appearance.(2,4) Total surgical excision is the treatment of choice and the prognosis after complete excision is excellent and recurrence is usually rare.(2,6) We present a rare case of giant cavernous hemangioma of the skull . The clinical presentation, pathology, differential diagnosis and treatment of this rare disorder are discussed.Case report : A 24-year-old female presented to us with history of huge swelling over the right parietal region progressively increasing in size over the past 12 years. There were no significant complaints except for the swelling. On examination, the lesion measured approximately 8 cm ×6cm in diameter in the right

posterior high parietal region. The swelling was firm to hard in consistency, non-tender and immobile. Clinical impression on local examination was that of a hard bony mass arising from the calvarium. Routine hematological examination was normal. General examination : Patient was moderately build and nourished, not anemic,not icteric, Pulse rate - 82, blood pressure- 120/80 Cardiovascular system - s1 s2 normal ,no added sound Respiratory system - Normal vesicular breath sounds heard

Central nervous system examination :

Patient consious, oriented

Higher mental functions normal

Cranial nerves examination normal

Spinomotor system normal.

Investigations :

Blood hemoglobin : 13.4 gms%	Blood sugar : 124 mgs/dl
Total count : P64% L36% E0	Serum creatinine : 0.6 mgs/dl
Differential count : 7200cells/cu.mm	Blood urea : 24 gms/dl
ESR : 5/10mm	
Bleeding time : 3minutes 20 seconds	Serum electrolytes :sodium - 138meq/dl, potassium - 4.6 meq/dl, chloride - 111 meq/dl
Clotting time : 5minutes 15 seconds	
Prothrombin time : 12 seconds	
INR : 0.9	

Plain Computed tomography (CT) of brain with bone window showed a osteolytic mass with sunburst/honeycomb appearance in the right high parietal region measuring 8.4 × 6.2 cm. The involved bone showed complete erosion of both the inner and outer tables. Figure 1- Plain axial CT scan showing a osteolytic mass with sunburst appearance in the right high parietal region involving both the inner and outer table of skull

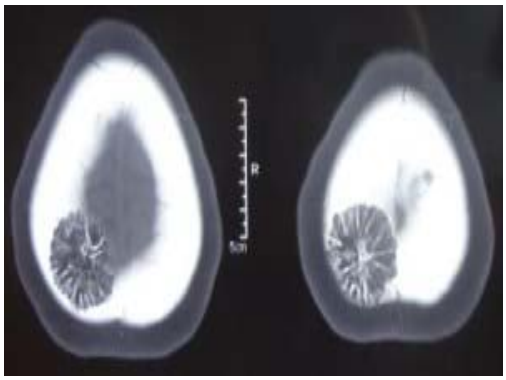
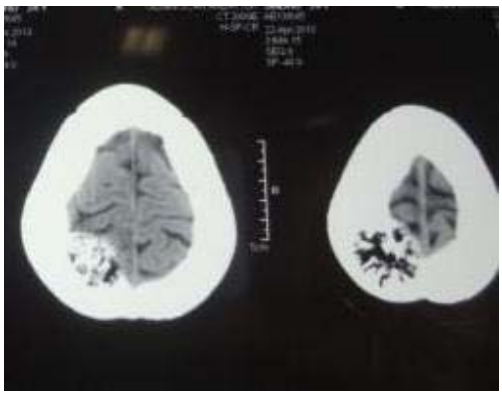


Figure 2 -Axial CT scan showing a osteolytic mass in the right high parietal region with buckling of underlying gray matter



Surgical planning : Since the routine hematological investigations were normal with no neurological deficit and the CT scan showed solitary osteolytic lesion sunburst/honey comb pattern we planned for total surgical excision of the lesion. Patient was operated in supine position under general anesthesia with head turned towards left and fixed in Mayfield head rest. Since the lesion extended upto the midline multiple burr holes were made over the midline and right parieto-occipital craniectomy with total resection of lesion was performed. Intraoperatively lesion was extremely vascular and looked like white coral-filled with vascular soft tissue in honeycomb appearance.After excision of the lesion cranioplasty was done in the same sitting. Patient was extubated on table with no neurological deficit.

Figure 3- Patient positioning for craniectomy

Figure 4 – Multiple burr holes made over midline to avoid sinus injury

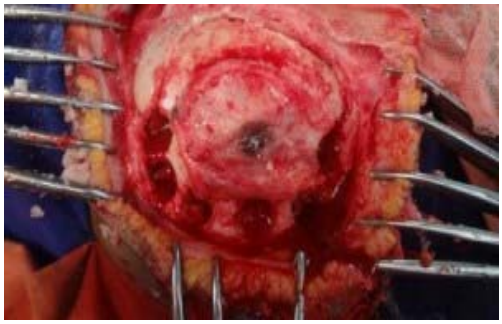
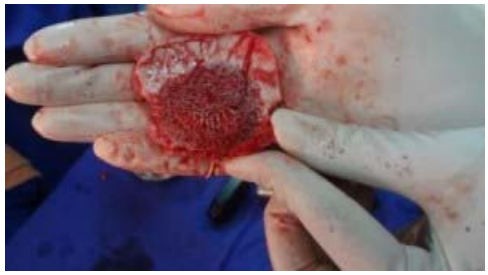


Figure 5 – Lesion with classical honeycomb appearance seen after craniectomy



Histopathology :

The histopathology showed bone bits with mature lamellar bone and bony spicules. The lamellar bone showed concentrically arranged lamellae (collagen) around vascular canal (Haversian canal). Medullary spaces between bony trabeculae showed ectatic thin-walled blood vessels with single layer of flat endothelial cells.

Figure 6 -H and E-stained slide showing bone bits with mature lamellar bone and bony spicules. Medullary spaces between bony trabeculae showing ectatic thin-walled blood vessels with single layer of flat endothelial cells

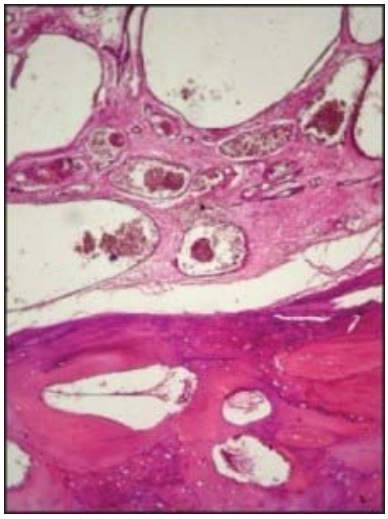
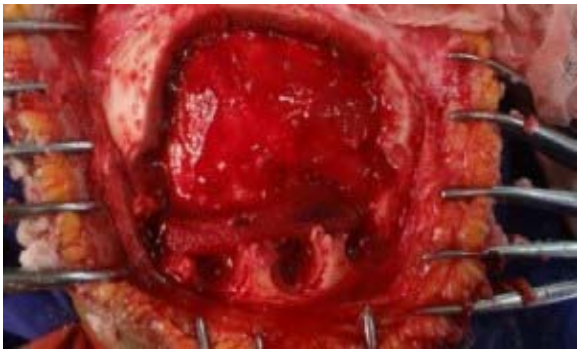
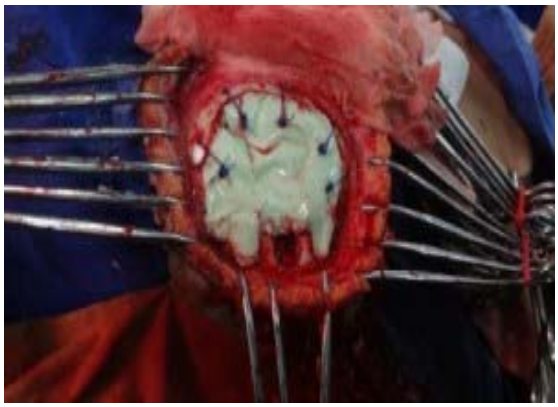


Figure 7 – After craniectomy bone defect showing superior saggital sinus covered with gelfoam Figure 8 – Picture showing cranioplasty with methyl methacrylate





Discussion :

PICHs are rare, benign skeletal tumors most commonly found in the spinal vertebral column. Less commonly, PICHs can involve the bones of the cranium. Overall, they represent 0.7% of all osseous neoplasms.(2) The earliest description in the English literature was in 1845 by Toynbee, who reported a vascular tumor arising in the confines of the parietal bone.(7) Although the origin of PICHs remains obscure, some authors believe that they represent congenital lesions that manifest in adulthood. In a review by Wyke, 70% of cranial PICHs were localized to the calvarium, particularly the parietal and frontal bones.(1) They are predominantly seen in patients in their fourth and fifth decades.(2,8) However, there are a couple of reports of cavernous hemangiomas in neonates or infants which have been misdiagnosed as ossified cephalhematoma.(9,10) Unlike the age predominance, our patient was a young adult (24-year old). The female-to-male ratio ranges from 2:1 to 3:1.(8) Hemangiomas are classified histologically by the predominant type of vascular channel in the lesion: Capillary, cavernous, venous and arteriovenous. In the cavernous variety, the capillaries are widely dilated seperated by fibrous tissue.(5) This variety is the commonest in the calvarium with a preference for the frontal and parietal bone.(2,6) Calcification within the hemangioma is common and may be of three types. The first variant, nonspecific type is either amorphous or, at times, curvilinear. The second type, which is more specific and is the most frequent type of calcification, is the phlebolith. Occasionally, metaplastic ossification may be found in hemangiomas, and this is the third type of calcification.(5) When bone is involved resulting in reactive osteoclastic and osteoblastic remodeling, the characteristic trabeculated, honeycomb appearance is visible on CT.(2,6) Calvarial hemangiomas are slow-growing tumors that are only symptomatic when they become large or compress adjacent neurological structures.(11)They develop in the diploic space constituted by dilated blood vessels with fibrous septa and their vascular supply is frequently from the middle meningeal artery and branches of the external carotid artery.6,8) They are usually solitary but multiple hemangiomas of the skull have been reported with a frequency of 15% of all identified calvarial hemangiomas.(12) Genetics:

Although most hemangiomas of bone are asymptomatic incidental findings, cerebral hemangiomas can

present with seizures, headache, focal neurological deficits, and occasionally hemorrhage. Bone hemangiomas can be expansile lesions, producing local swelling when symptomatic, with associated local pain or fractures.(21) Cerebral cavernous hemangiomas may be hereditary. Some are autosomal dominant, caused by mutations of the KRITI1 gene on the long arm of chromosome 7.(22) This encodes a protein that interacts with the tumor suppressor gene krev-1/rap1a. Occasionally, the mutation gives rise to hyperkeratotic cutaneous venous malformations, as well. Mutations on 7p and 3q are also linked to familial cavernous hemangiomas.(23) Skeletal hemangiomatosis is characteristically not hereditary and is associated with visceral involvement 60 to 70% of the time and lymphangiomatosis.(24) Visceral involvement is mostly of the spleen, liver, kidney, mesentery, chest, and lymph nodes and confers a poorer prognosis. Gorham disease may represent a more severe form of multifocal cavernous hemangiomatosis. It is extremely rare, occurs in children and young adults, and is characterized by resorption of affected bone; hence the name disappearing or phantom bone disease.(24)Since there are no facilities for genetics workup in our hospital we had not done genetic screening.CT is the investigation of choice and calvarial hemangiomas typically appear as a lytic expansile and “bubbly” lesion with a sclerotic rim or a spiculated “sunburst” skull tumor. CT is also helpful in delineating the osseous extension to adjacent skull structures and possible complications such as inner or outer table bone fracture.(8) Nonenhanced CT usually shows a mass isodense with adjacent muscles with intense enhancement after intravenous contrast administration.(5) Other radiological

Author (et al),year	Age group	No.of cases	Radiological presentation
Martinez, 2010	Infant	1	Ossified
Nasrallah, 2009	Paediatric	1	Osteolytic
Gordhan, 2009	Adult	1	Coarse internal trabeculation (sunburst)
Naama, 2008	Adult	3	Osteolytic
Nasser,2007	Adult	2	Osteolytic
Ajja,2005	Adult	1	Osteolytic
Heckl,2002	Adult	4	Honeycomb/sunburst
Khanam,2001	Adult	2	Osteolytic
Suzuki,2001	Adult	1	Osteolytic
Yoshida,1999	Infant	1	Ossified
Pastore,1999	Adult	1	Osteolytic
Cervoni,1995	Adult	3	Osteolytic

investigations include magnetic resonance imaging (MRI) and scintigraphy, which was not done in our case.(2,8,13) A table mentioning various reports and series of solitary intraosseous cavernous hemangiomas of the skull vault in literature is shown below. Table 1- Previously reported cases of solitary primary intraosseous cavernomas of the skull vault in various age groups and their radiological presentation

Differential diagnosis : The differential diagnosis of a solitary circumscribed expansile intradiploic cranial lesion includes an osteoma, aneurysmal bone cyst, giant cell tumor, fibrous dysplasia,hemangioendothelioma,plasmacytoma , Langerhans' cell histiocytosis, sarcoma, meningioma, metastatic disease, Pagets disease and dermoid tumor.(6,13) Rare intraosseous meningiomas, also known as primary extradural meningiomas, represent only 1–2% of all lesions of this group and may be difficult to distinguish from calvarial hemangiomas solely on the basis of location.(8) Management : The best treatment for hemangiomas is total surgical resection of the lesion. (2,6) Embolization before surgery is helpful in preventing excessive bleeding in large tumors.(6,9) Radiation has been attempted to treat hemangiomas, and it has been shown to stop tumor growth; however, it has not been shown to reduce the size of the tumor.(20) Nasrallah *et al have done embolization followed by craniotomy,*preserving the outer table with no recurrence at 3-year follow-up.(14) Cavernous hemangioma of the calvarium is a benign condition of the skull bone which can be completely resected with good results.These lesions usually have typical radiological imaging features, which may, however, show some variations. Cavernous hemangiomas should be considered as a differential diagnosis even in completely ossified calvarial masses and adequate preparation to minimize blood loss such as embolization can be considered if they are adequately investigated considering this possibility.

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