Radiologic Features with Pathologic Correlation of an Unusual Large Intraosseous Skull Cavernous Hemangioma

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Patient: Female, 58
Final Diagnosis: Intraosseous skull hemangioma
Symptoms: Palpable, painless hard mass in forehead • no headaches, altered mental status, seizures, or focal neurological deficit
Medication: —
Clinical Procedure: Cerebral angiogram and embolization of feeders • afterwards, she underwent a bifrontal craniectomy and cranioplasty
Specialty: Radiology
Objective: Rare disease
Background: Intraosseous cavernous skull hemangiomas are rare benign vascular tumors that are usually found incidentally on imaging, with an asymptomatic and slow-growing course. We present a case in which the patient had a mass on her forehead for many years, which began to grow rapidly after head trauma. Imaging characteristics play a crucial role in the diagnosis and description of this disease, and in differentiating it from other more common calvarial lesions that may present with a similar clinical picture. Here, we report an unusual presentation of a large skull hemangioma and discuss the different radiologic imaging findings and pathologic correlations.
Case Report: A 58-year-old female with history of a lump on her forehead since childhood, which began to grow rapidly after experiencing a closed-head injury. Due to its large size, she went on to seek further management. Radiologic images revealed a frontal skull lesion suggestive of an intraosseous hemangioma. She underwent embolization of the tumor, and 2 days later underwent bilateral frontal craniectomy and cranioplasty. Histopathologic findings confirmed this diagnosis.
Conclusions: Intraosseous skull hemangioma may be confidently diagnosed and differentiated from other skull lesions by its imaging characteristics. An accurate diagnosis is essential to selecting correct management and avoiding complications.

MeSH Keywords: Bone Neoplasms • Hemangioma, Cavernous • Skull

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Background

Intraosseous skull hemangiomas are rare benign tumors of endothelial origin that account for 0.2% of all bone tumors and 10% of the benign tumors of the skull [1]. Intraosseous hemangiomas are usually found in the vertebral column, but are rarely seen in the cranium. The frontal bone is the most commonly involved, followed by the parietal bone, temporal bone, and, less frequently, the occipital bone [2]. They are mostly encountered in adults in the 4th to 5th decades of life, the ratio being 3:2 more common in females than in males. To date, there is no documented racial variation in the frequency of skull hemangiomas [3,4].

The etiology of a skull hemangioma remains unclear. Some are thought to be congenital, while others authors propose trauma as a causative factor [2,5,6]. These benign tumors develop from the vessels in the diploic space that arises from branches of the external carotid, usually being the middle meningeal and superficial temporal arteries [7]. They can mimic other more common skull lesions and therefore can be difficult to diagnose preoperatively. We present a rare case of a large skull hemangioma that started growing rapidly after a traumatic event in a 58-year-old female patient. Our goal is to emphasize the imaging characteristics of this tumor and to demonstrate its pathological correlation. The clinical presentation, differential diagnosis, and treatment of this disorder are also discussed.

Case Report

A 58-year-old woman presented with a history of a small bump on her middle forehead since childhood, which began to grow rapidly for the past year after she received a closed-head injury from a ladder that fell on her head. She denied symptoms such as headaches, seizures, altered mental status, or any focal neurological deficit. Other medical history and review of systems were insignificant. Physical examination revealed a palpable, painless, hard mass. The overlying skin was mobile and normal in appearance.

Plain skull radiographs showed an expansile lytic lesion along the frontal skull, in which the original impression of the mass by a radiologist was a meningioma (Figure 1). Subsequent CT and MRI studies without contrast were performed. The CT scan revealed a large, well-demarcated, trabeculated mass with a sunburst pattern arising from the frontal skull (Figure 2A, 2B). The brain MR showed the mass to be mostly T2 hyperintense and T1 isointense, with interspersed hyperintense components (Figure 3A, 3B). Given the characteristic sunburst pattern, the preferential diagnosis was a hemangioma.

Neuroendovascular Services decided to perform a cerebral angiogram, which revealed a vascular mass with feeders from the left middle meningeal artery, and went on to embolize the tumor with coils (Figure 4A, 4B). Two days later, Neurosurgery Services operated on the patient under general anesthesia, and in a supine position via a bifrontal subperiosteal approach, the mass was exposed. A reddish-brown, highly vascular lesion, with cavernous blood-filled spaces between the bony trabeculae was seen at the bifrontal region. The mass expanded externally and intracranially, and the inner surface of the mass had significantly adhered to the dura mater. A bifrontal craniectomy with complete resection of the tumor was performed, including a margin of surrounding uninvolved bone. Subsequent cranioplasty with a polymer implant was done to cover the 9×10 cm skull defect (Figure 5A, 5B). The gross specimen showed a protruding, well-delineated lesion from the bone (Figure 6A–6C). Histological examination concluded that, in effect, this was a cavernous hemangioma (Figure 7). Post-operative CT revealed complete resection of the tumor with expected post-surgical changes (Figure 8). The patient was discharged 2 days later without complications.

She was evaluated in the outpatient clinic 5 weeks later. She did not present any symptoms such as focal neurological deficits, seizures, altered mental status, or headaches at this time. After 5 months, she was re-evaluated and has remained asymptomatic with optimal healing of the surgical site, demonstrating a well-restored contour of the forehead.

Discussion

Intraosseous hemangiomas are classified histopathologically according to the vascular network as venous, arteriovenous, cavernous, or capillary type [3]. The cavernous hemangioma is composed of large, thin-walled vessels and sinusoids lined...
Figure 2. Sagittal (A) and axial (B) bone window CT images show a sharply marginated, expansile lesion arising from the frontal skull with a sunburst pattern of trabecular thickening radiating from a common center. Note the associated expansion of the inner and outer tables of the frontal skull.

Figure 3. (A) Sagittal T2WI MR without contrast demonstrates a large expansile lesion arising from the frontal skull with mostly hyperintense components, mainly the result of slow flow or venous stasis with a contribution from fat. The internal hypointensities represent the thickened trabeculae. Note how the mass is resulting in compression upon the frontal lobe and in deformity of the overlying scalp. (B) Sagittal T1WI MR without contrast shows lesion to be mostly isointense, which is less bright than the overlying fatty scalp. Hyperintense components are likely caused by fatty tissues and/or hemorrhage.
with a single layer of endothelium [8]. On gross inspection, the tumor tends to be well-demarcated, fleshy, vascular, and unencapsulated with cystic red cavities.

Intraosseous skull hemangiomas are usually discovered incidentally on imaging given its asymptomatic course in the majority of the cases [3]. In this case, however, further management was sought due to esthetic concerns given its large size. There are several radiologic features that help radiologists arrive at the correct diagnosis. Skull x-rays usually demonstrate a lytic, expansile lesion with a honeycomb or sunburst pattern [4,9].

Head CT confirms the findings of plain film because of its excellent characterization of trabecular and cortical details [7]; it will present an expansile bone lesion with thin borders and a characteristic sunburst pattern of trabecular thickening radiating from a common center. Tumor expansion may result in erosion of the internal and external plates. Homogeneous enhancement is seen upon intravenous contrast administration [3].

Brain MRI characteristics largely depend on the proportion of fat and vascularity the lesion has. In T1-weighted images, intermediate signal intensity is predominantly seen, with scattered foci

Figure 4. (A) Left external carotid angiogram (ECA), anteroposterior view, with arterial phase shows the tumor shadow (red arrow) fed by the left middle meningeal artery (yellow arrow). (B) Left ECA status post embolization with coils (yellow arrows). Note the lack of vascularity reaching the tumor shadow (red arrow).

Figure 5. (A, B) Intraoperative photographs showing that the vascular mass has a reddish color and is well delineated from the bone.
of hyper- and hypo-intense T1 signal. Low signal intensity on T1-weighted images usually indicates a decreased amount of fatty tissue or a greater amount of vascularity. On T2-weighted images, a heterogeneous hyperintense mass is seen. This appearance is due to cavernous or cystic vascular spaces containing stagnant blood. Thickened trabeculae demonstrate low signal intensity in all sequences. Additionally, hemangiomas avidly enhance after gadolinium contrast administration [10].

Utilization of preoperative angiography and selective embolization has been a controversial topic in the literature. Some authors state that vascular control is not necessary if an adequate margin of normal bone is resected [11]. However, many authors recommend preoperative embolization, especially of larger vascular lesions, to reduce intraoperative blood loss [4,8,12–14]. Angiography confirms the hypervascularity of the lesion, demonstrating a delayed blush, with distinguishable

Figure 6. Photographs of gross specimen. (A) In the center there is a dark blue, slightly spongy protruding lesion (yellow arrow) with a rim of normal, uninvolved bone around the circumference of the lesion (red arrow). (B) Note the expansile effect of the tumor in the frontal skull, with the tumor protruding at the inferior aspect (yellow arrow). (C) Cross-section reveals a red, bloody, spongy interior with some friable areas.

Figure 7. Histology demonstrates endothelium-lined sinusoidal cavities filled with erythrocytes.

Figure 8. Head CT without contrast (3D reconstruction), anterosuperior projection, status post bifrontal cranioplasty.
feeding arteries but no draining veins [8]. The most common arterial feeders are branches of the middle meningeal, superficial temporal, and posterior occipital arteries [15]. In our case, we felt that preoperative angiography and embolization of the feeding arteries before undergoing surgery limited intraoperative bleeding and helped maintain a clean field for resection.

In some series, nearly 50% of intraosseous hemangiomas occur in the craniofacial bones, predominantly in the calvarium. The spine is involved in approximately 20% of cases. However, if autopsy data is taken into account, the vertebral bodies are the most common site of this lesion. Less frequently, hemangiomas involving the major long tubular bones of the lower extremities and ribs have been documented. Hemangiomas involving long tubular bones are usually larger than those of the skull and spine, since they usually become symptomatic when they grow large [16].

The differential diagnosis includes any solitary expansile intradiploic cranial lesion with normal overlying skin. As most scalp lumps are of benign origin, meningioma is usually considered because it is one of the most frequent entities. Another diagnostic consideration is metastasis because most malignant scalp masses are of metastatic origin. Other differential diagnoses that must be considered are: lymphoma, osteoma, aneurysmal bone cyst, giant cell tumor, Langerhans cell histiocytosis, sarcoma, fibrous dysplasia, and dermoid tumor [3]. Nevertheless, with careful examination of the lesion’s imaging characteristics, we can arrive at an accurate diagnosis.

The characteristic appearance of honeycomb or sunburst pattern of white spicules radiating from the center of a radiolucent round or oval defect is an important imaging feature that can distinguish a hemangioma from other lesions. It is crucial to recognize these features, as this will aid physicians in selecting the best management for the patient and to avoid complications. The correlation with histologic findings showing multiple thin-walled vascular spaces lined by endothelial cells and enclosed by loose connective tissue and bony trabeculations is important to arrive at a definitive diagnosis [17].

Skull hemangiomas rarely require treatment, and indications include mass effect, hemorrhage control, and esthetic improvement. As was seen with our case, the rapid increase in size was probably due to repeated hemorrhage, especially given the history of trauma. If it is indicated, the treatment of choice is total resection with an adequate normal bone margin to reduce the risk of bleeding. Cases of recurrence after complete resection have not been described [18]. Prior to surgery, embolization of the tumor may be performed to reduce the intraoperative blood loss [13]. When surgical removal is not possible, radiotherapy is another alternative, but radiotherapy only prevents the tumor from growing and it cannot eradicate the lesion [19].

**Conclusions**

Intraosseous skull hemangiomas are rare benign tumors that can mimic other more common calvarial tumors. Although histopathological confirmation after surgical resection is the definitive method for diagnosis, in most cases the diagnosis can be made with certainty by its imaging findings. Imaging characteristics play a crucial role in diagnosing this entity and can prevent complications such as severe hemorrhage from routine biopsy and curettage.

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