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Congenital cavernous hemangioma of the calvaria

Case report

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The authors present the case of a 6-month-old infant with a cavernous hemangioma of the parietal bone and discuss the radiological, operative, and pathological features and differential diagnosis of these extremely rare lesions in infants. Only 1 case of an infant with a calvarial cavernous hemangioma without intracranial invasion has previously been reported, and that case involved a 4 month old.

Although a cavernous hemangioma of the calvaria is extremely rare in the newborn, this condition should be included in the differential diagnosis of calvarial lesions. During surgical treatment of calvarial cavernous hemangiomas, utmost attention should be paid to avoid blood loss, which could be fatal in infants.
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Key Words • **calvarium** • **congenital tumor** • **hemangioma**

Intraosseous hemangiomas are rare, slow-growing, benign tumors of the blood vessels. They comprise 0.7% of all bone neoplasms; the most common site is the vertebral column, followed by the skull.[9,14] Hemangiomas are histologically classified as cavernous and capillary. Most of the calvarial hemangiomas are of the cavernous type, whereas vertebral hemangiomas are most frequently of the capillary type.[10,11] Intraosseous cavernous hemangiomas can be found in any location in the skull but most frequently seen in parietal and frontal bones, respectively and may occur at any age but patients usually present for treatment in the 2nd to 4th decades.[6,7,9,14,20] Only 1 case of an infant with a calvarial cavernous hemangioma without intracranial invasion has previously been reported; the patient in that case was 4 months old at presentation.[27] The authors present the case of a 6-month-old infant with a cavernous hemangioma of the parietal bone and discuss the radiological, operative, and pathological features and differential diagnosis of these extremely rare lesions in infants.

Case Report

History and Presentation. This 6-month-old male infant presented with a right parietal skull lesion that had been present since birth. He had been delivered vaginally, and no traumatic intervention had been reported during his delivery. The patient's parents described the lesion as a soft mass at the time of birth. The physicians interpreted that lesion to be a caput succedaneum, which might be expected to resolve, but, in the course of time, the mass became solid and enlarged. Therefore, the patient was brought to our department for further investigation.

Examination. Physical examination revealed no abnormality except a right parietal lesion. The lesion was hard in consistency and 5 cm in diameter. The patient's head circumference was within the normal range. The findings on neurological examination were completely normal.

Posteroanterior cranium radiographs showed an asymmetric enlargement of the calvarial bones on the right side. The outer table was prominent with an associated area of nonspecific increased radiopacity (Fig. 1). Computed tomography showed a soft-tissue–density mass that expanded in the diploic space on the right side. Both the inner and outer tables were increased in thickness, and there were some defective areas on the cortex of the inner table (Fig. 2). On T1-weighted MR images (Fig. 3) there was an isointense expansile mass, which bulged externally, was limited to the diploic space, and enhanced after contrast administration (Fig. 4). The lesion was heterogeneously hyperintense on T2-weighted images (Fig. 5).

This article contains some figures that are displayed in color online but in black and white in the print edition.

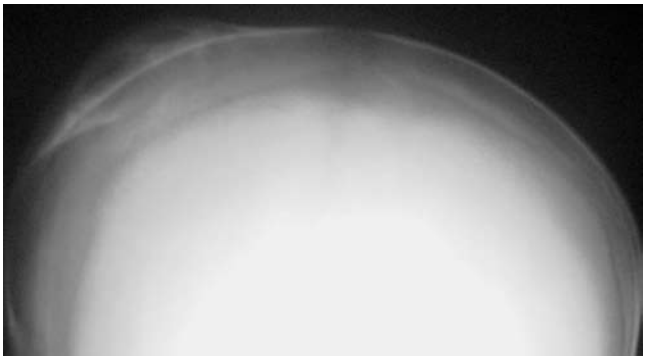


FIG. 1. Radiograph showing asymmetrical enlargement of the right parietal bone.

Operation. The patient underwent a right parietal craniectomy with excision of the parietal lesion and a margin of surrounding uninvolved bone. The lesion tended to bleed considerably, and the craniectomy was completed with utmost attention paid to avoiding blood loss. No involvement of the underlying dura mater was observed. On gross examination, the right parietal lesion was well delineated and reddish brown in color and appeared to originate from the diploe. There was erosion of the inner table of the skull, but no erosion of the outer table. After total excision of the tumor, cranioplasty was performed with high-density porous polyethylene (Medpore, Porex Industries). Postoperative CT showed complete removal of the lesion.

Histological Findings. Histological examination revealed thin-walled large dilated vessels filled with erythrocytes between the bony trabeculae (Fig. 6), consistent with an intraosseous cavernous hemangioma. In the center of the lesion, an organizing hematoma containing hemosiderin-laden macrophages was seen.

Discussion

Intraosseous 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.[22,24,27] Intraosseous calvarial cavernous hemangiomas may be unifocal or multifocal, but the vast majority of reported cases are unifocal. The parietal and frontal bones are regions of the calvaria most frequently affected by these tumors. The tumors are

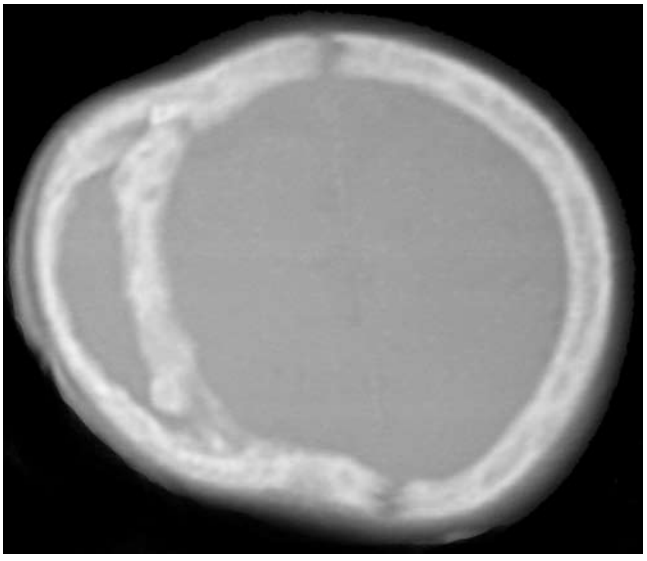


FIG. 2. Computed tomography demonstrating increased thickness in both tables and some defective areas on the cortex of the inner table.

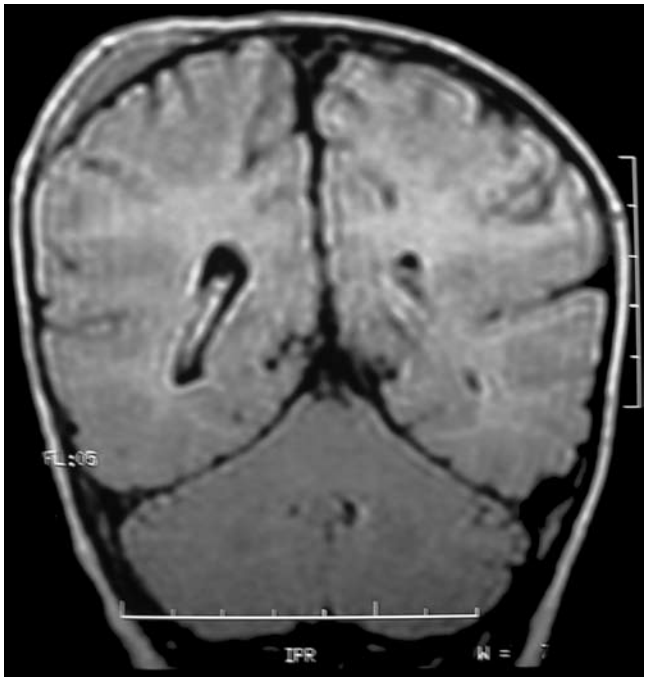


FIG. 3. Coronal T1-weighted MR image revealing an isointense expansile mass that bulges externally and is limited to the diploe.



FIG. 4. Coronal T1-weighted MR image demonstrating enhancement of the lesion after administration of contrast medium.

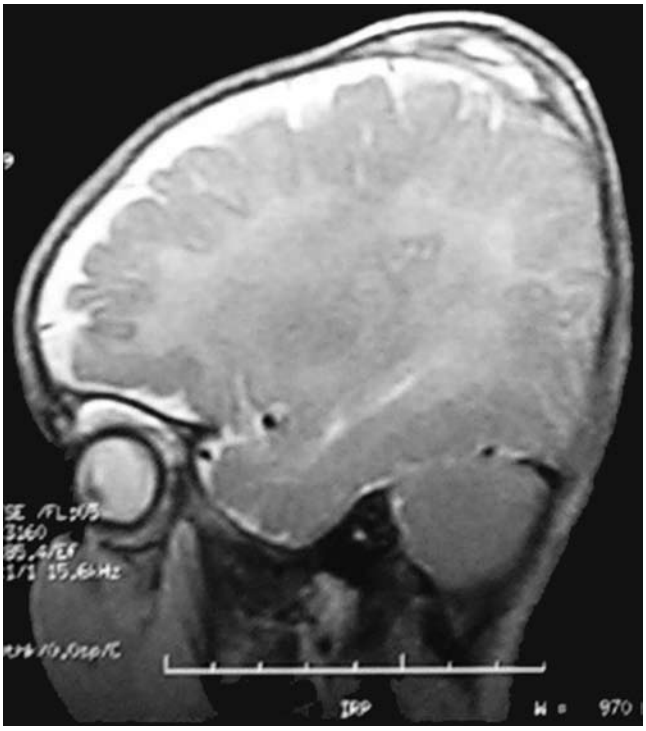


Fig. 5. Sagittal T2-weighted MR image demonstrating a heterogeneously hyperintense lesion.

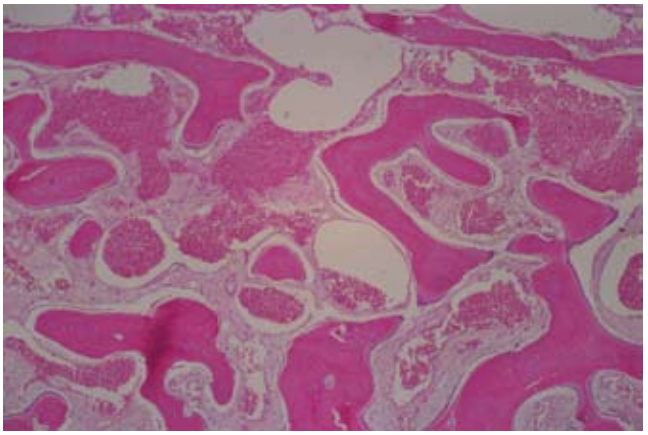


Fig. 6. Photomicrograph of a section of the lesion showing thin-walled cavernous vascular spaces between the bony trabeculae. H & E. Original magnification × 100.

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 2nd through 4th decades.[19,24,27] Occurrence of these tumors in the neonatal period is extremely rare. To date, there is only 1 report of a purely calvarial cavernous hemangioma in a neonate—the case described by Yoshida et al.[27] Honda et al.[9] and Koulouris and Rao[13] reported on newborns with cavernous hemangiomas affecting the calvaria but not limited to it. Honda et al. described a newborn presenting with a temporal bone cavernous hemangioma with intracerebral extension, and Koulouris and Rao reported a case of multiple calvarial hemangiomas affecting not only the calvaria but also the zygoma and maxilla with the involvements of the skull base, frontal and petrous temporal bones, and squamous temporal bone. Our case is the second report of a cavernous hemangioma affecting the neonatal calvaria that had been present since the birth of the patient.

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.[25] In the intrauterine period, defective differentiation of primordial vessels, resulting in an abnormal capillary bed, may guide the development of hemangiomas.[17,27] 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.[10,11] Trauma is not thought to be a predisposing factor in the development of these lesions.[19,24,27]

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.[11] Our patient had a soft mass in the right parietal region at birth, which became solid over time.

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.[2,5]

Although preoperative radiological studies usually provide insufficient information for the precise diagnosis of calvarial cavernous hemangiomas, imaging modalities for the diagnosis include radiography, CT, and MR imaging. Most often the imaging findings are nonspecific and can mimic those of calcified/ossified cephalohematoma. Thus, it is usually not possible to arrive at an exact diagnosis preoperatively, and the nature of the lesions can only be confirmed with histological diagnosis. Microscopically, the lesion in our case was markedly different from calcified cephalohematoma. In contrast to simple calcified cephalohematoma, it showed endothelial-lined cavernous spaces between bony trabeculae (Fig. 6).

The typical sunburst appearance of a calvarial cavernous hemangioma on imaging is due to osteoblastic remodeling with trabecular bone following osteoclastic activity of the tumor.[14,20] This characteristic imaging feature may not be seen in every case, as it was not evident in ours. Furthermore, in our case there was no outer table erosion but there was inner table erosion, although outer table erosion is more common in these lesions.[13] Intensity of the hemangiomas is variable on MR imaging.[1,11,26] Hyperintensity on

T1-weighted images obtained after contrast administration, the only images that were strongly suggestive of calvarial cavernous hemangioma in our case, is thought to be a differentiating feature of these lesions. Although they are not definitive in discriminating calvarial cavernous hemangiomas from the other lesions, CT and MR imaging are also valuable diagnostic studies for determining the presence of intracranial extension.

In cephalohematoma there is collection of blood beneath the periosteum. The periosteum adheres to the bone at its margin and can be elevated over its entire surface. When the absorption of the hematoma does not occur rapidly, subperiosteal osteogenesis begins at the attached margin of the periosteum all around the circumference of the bone. If absorption of the clot does not proceed rapidly, the newly formed bone at the edge grows subperiosteally over the dome of the bulge until at one stage only a small area at the apex of the lesion lacks an osseous covering. Over time this space is also filled in, and the hematoma becomes completely roofed over, and in a few weeks it seems, in palpation, as firm as the rest of the skull.[4] Thus, in calcified/ossified cephalohematoma the original calvarial bone forms the inner wall and the newly ossified bone the outer wall, and the inner wall remains intact. The outer table may be intact or defective depending on the stage of ossification. In our case, there were defective areas at the inner table which were thought to be not consistent with cephalohematoma.

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,[10,11] because any remnant of the tumor may cause recurrence. Although preoperative embolization is recommended to prevent excessive intraoperative bleeding,[11,24] we did not performed 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.[8,11,24] Because the resection in our case was total, the patient was not subjected to radiotherapy.

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. The best donor source of autologous bone is split-thickness cranium, but it is nearly impossible to perform the procedure in children younger than 3 years old because of the thinness and the underdeveloped diploe of the skull. The second choice for autologous bone grafts is split rib grafts. There is no minimum patient age for harvesting of rib, although this procedure requires a second operative site and a longer operating time. In addition, regardless of the source, autologous bone grafts can undergo partial absorption.[21,23] Allograft materials used for cranioplasty are metallic mesh plates, methylmethacrylate, hydroxyapatite, and high-density porous polyethylene.[18,23] Methylmethacrylate is easy to use and does not affect CT or MR imaging results, in contrast to metallic mesh plates. It may, however, cause severe tissue inflammation, and its tremendous exothermic reaction during setting may be harmful to underlying neural tissue.[3,23] Hydroxyapatite does not achieve its final strength for several hours or days after the initial setting, thus the implant can be cracked by the continuous pulsation of CSF transmitted through the dura mater during this time.[16,23] We preferred high-density porous polyethylene for the cranioplasty material in our case; its porous character allows for rapid fibrovascular and soft tissue ingrowth and eventual incorporation of bone, which strengthens the implant as well as decreasing the risk of infection.[12,15]

Conclusions

Although it is extremely rare in the newborn, cavernous hemangioma of the calvaria should be included in the differential diagnosis of calvarial lesions, and intraoperatively, utmost attention should be paid to avoid blood loss, which could be fatal in infants.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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