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Cranial Arteriovenous Hemangioma in a Neonate

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Benign hemangiomas and vascular malformations are the most common tumor of infants and children [1]. Skull involvement is rare compared with soft-tissue involvement. A plethora of classifications applied to these lesions has led to an imperfect and suboptimal understanding of them. Burrows et al. [2] describe angiographic differences between hemangiomas and vascular malformations that corroborate clinical and laboratory differences previously reported. Enzinger and Weiss [1] state that arteriovenous hemangioma as a specific entity bridges the distinct categories of vascular malformation and hemangioma, often exhibiting features of both lesions. We report a rare neonatal cranial arteriovenous hemangioma with elements of both hemangioma (dense angiographic stain) and vascular malformation (osseous irregularity and calcified phleboliths).

Case Report

A 5-week-old girl was admitted to our hospital with an enlarging left suboccipital mass that was first noted at birth. The pregnancy had been uncomplicated, and the infant was born at term with forceps assistance to terminate a prolonged labor. Routine physical and neurologic examination were otherwise normal at the time of admission and at birth. The suboccipital mass was firm and rubbery with a palpable bony edge. No apparent pain was elicited on palpation of the mass, but the palpation pressure was observed to transmit to the anterior fontanel. Laboratory values were unremarkable except for an elevated urinary VMA of 112 units (normal <30) and a serum alpha fetoprotein level of 1240 units (normal <20).

Noncontrast CT showed a 5 × 5 cm high-density left occipital extraaxial mass with both intra- and extracranial components (Fig. 1A). The mass contained multiple foci of calcification that were proven histologically to represent phleboliths. After IV contrast administration, the mass enhanced densely and homogeneously (Fig. 1B). Bone windows demonstrated irregular spiculated bone architecture (Fig. 1C). Hydrocephalus was present as a result of either kinking of the aqueduct or obstruction of the fourth ventricular outlet. Left common carotid and left vertebral arteriography showed the mass to be densely staining with areas of contrast pooling and fed by branches of the left occipital and middle meningeal arteries (Fig. 1D). There was no evidence of rapid arteriovenous shunting or enlarged draining veins.

Neurosurgical operation showed the lesion to have two major components by gross inspection. The first and most external portion was a "hypertrophic bony component" related to the calvarium, which had a natural dissection plane separating it from the dural or intracranial portion. The second, inner component was a highly vascular soft-tissue tumor that revealed cavernous sinuses with apparent endothelial lining when grossly incised. No clear dissection plane was present between this component and hypertrophied dura.

Histopathologic study revealed a lesion of variable-sized thick- and thin-walled vascular channels (Fig. 1E) with occasional luminal obliteration (Fig. 1F). Prominent stromal fibrosis and metaplastic ossification and chondrification were observed (Fig. 1G). The endothelium lining the vessels was benign. Anastomosing vascular channels were seen partially separated by thickened fibrous bridges resulting in a complex sievelike pattern. An abundance of mast cells was identified by Giemsa stain. There was no evidence of malignancy. The final diagnosis was arteriovenous hemangioma.

Discussion

Benign vascular lesions are very common in children, with the vast majority of cases needing no treatment of any kind. Burrows et al. [2] reviewed the preoperative angiograms of 14 children who had cellular analysis of resected vascular lesions, and described angiographic and osseous differences between hemangiomas and vascular malformations. These radiographic differences correlated well with previously noted histologic, cellular, and clinical differences between these two vascular lesions [3].

Hemangiomas are benign endothelial-cell neoplasms. They account for 7% of all benign tumors, and are the most common tumor of infancy and childhood [1, 4]. They usually appear 1–2 months after birth, and may rapidly proliferate over 8–12 months. Without treatment, involution and regression occur in more than 90% of patients [2]. These tumors have a predilection for the head and neck. Histology reveals circumscribed masses of endothelial cells, with or without vascular lumena. A high mast-cell count is present [3]. Plain films show a localized soft-tissue mass without calcifications or osseous changes. On angiography, a lobular neoplasm is seen with a staining parenchymal component. Vascular feed-
Fig. 1.—A, Noncontrast-enhanced CT scan shows high-density left occipital extraxial mass with intra- and extracranial components and multiple foci of calcification. The fourth ventricle is displaced and temporal horns are dilated.
B, Contrast-enhanced CT scan shows marked homogeneous enhancement of the mass.
C, Bone windows show osseous irregularity and spiculation in left occipital bone.
D, Left common carotid arteriogram shows dense tumor stain. Supply originates from branches of left occipital and middle meningeal artery (not shown). There is no evidence of rapid arteriovenous shunting.
E, Representative section of mass shows numerous thick-walled vascular channels filled with red blood cells (arrow).
F, Stromal fibroblast proliferation compresses and obliterates vascular channels (arrows).
G, Low-power view shows foci of metaplastic ossification and chondrification (arrow).

ers are slightly dilated but taper normally. (Treatment is conservative, with steroids and radiation used occasionally.) Features typical of hemangioma in our case include a densely staining lobular mass angiographically and the presence of endothelial-cell proliferation with numerous mast cells. Features in this case not characteristic of hemangioma are the bone involvement characterized by markedly irregular osseous architecture and calcification and the presence of the lesion at birth. Although hemangiomas may be present at birth, in more than 50% of cases these lesions do not appear until 1 or 2 months after birth.

Vascular malformations are errors of vascular morphogenesis and are always present at birth [2]. They may be capillary, venous, arterial, lymphatic, or any combination thereof.
Growth of the malformation is commensurate with the patient, and may respond to hormonal modulation—e.g., puberty. Histology reveals collections of abnormal vessels with no evidence of endothelial-cell proliferation. The mast-cell count is normal [3]. Radiology shows a soft-tissue mass with osseous changes in a large majority of cases. Phleboliths are common in capillary venous malformations. Bone size may be increased with or without cortical thickening, erosions, and coarsened trabeculae. Angiography shows ectatic, abnormal vascular channels, with no parenchymal stain. Arteriovenous shunting is common. Pooling of contrast is noted in the capillary venous type. Selected cases are amenable to embolization or resection. The osseous irregularity and calcified phleboliths in our case are characteristic of vascular malformation, whereas the angiographic appearance of dense parenchymal stain coupled with the lack of arteriovenous shunting is atypical.

A diagnosis of neuroblastoma was entertained preoperatively in this infant because of the presence of a densely enhancing extraxial mass associated with osseous irregularity, calcification, and elevated urinary VMA. The possibility of a rare sarcoma of bone with chondroid foci of calcification was also raised but considered unlikely in a neonate. The markedly irregular spiculated bony appearance in this benign lesion perhaps is explained by synchronous development of the calvarium and the arteriovenous hemangioma, which may be a reactive phenomenon. This is in contradistinction to bone that has developed normally but is subsequently invaded by growth of malignant tissue.

The findings in this lesion of angiographically staining parenchyma and osseous changes with calcified phleboliths well support Enzinger and Weiss's [1] concept that the arteriovenous hemangioma, as an explicit lesion, bridges the distinct categories of (1) vascular malformations and (2) benign hemangiomas.

REFERENCES