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Radiologic-Pathologic Correlation

Capillary Hemangioma of the Meninges

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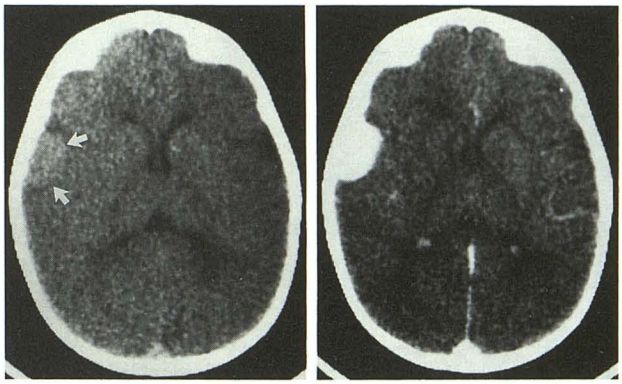
Clinical History

A Hispanic male infant, aged 17 months, presented with a new onset of focal motor seizures. Generalized seizures began at 12 months of age but had become focal with left facial twitching and tonic extension of the left arm. Neurologic examination was normal and developmental history revealed mild generalized delay. There were no dermatologic abnormalities. Family history was positive for a generalized seizure disorder in the mother that was well controlled with diphenylhydantoin and phenobarbital. Computed tomography (CT) of the brain, with and without postcontrast study, was performed at an outside hospital (Fig. 1). Magnetic resonance (MR) imaging of the head with contrast was obtained at our institution (Fig. 2), followed by selective arteriography (Fig. 3). The preoperative diagnosis was meningioma.

Right temporal craniotomy exposed a vascular-appearing mass attached to the dura by a fibrous stalk and supplied by a large middle meningeal artery. The mass protruded into the subdural space and measured 3 cm × 3 cm (Fig. 4). It was easily devascularized by ligation of the feeding vessels and was then excised en bloc. Pathologic examination established the diagnosis of capillary hemangioma.



A B

Fig. 1. CT without (*A*) and with (*B*) intravenous contrast infusion.
A, There is a dural-based lentiform mass with a smoothly convex inner border (*arrows*). It is of higher attenuation than adjacent gray matter.
B, It enhances intensely and homogeneously after contrast administration.

The postoperative course was uneventful except for the occurrence of early postoperative seizures that were easily controlled with phenobarbital.

General

Hemangiomas are the most common tumor of the head and neck in children, presenting in 10% to 12% of white infants. The majority of hemangiomas are of two types, capillary and cavernous (1). Capillary hemangiomas are the most common and are characterized by the appearance of a red or blue cutaneous papule in the first 6 weeks of life, a period of rapid growth, and spontaneous involution, usually by 6 years of age (2). They are most common

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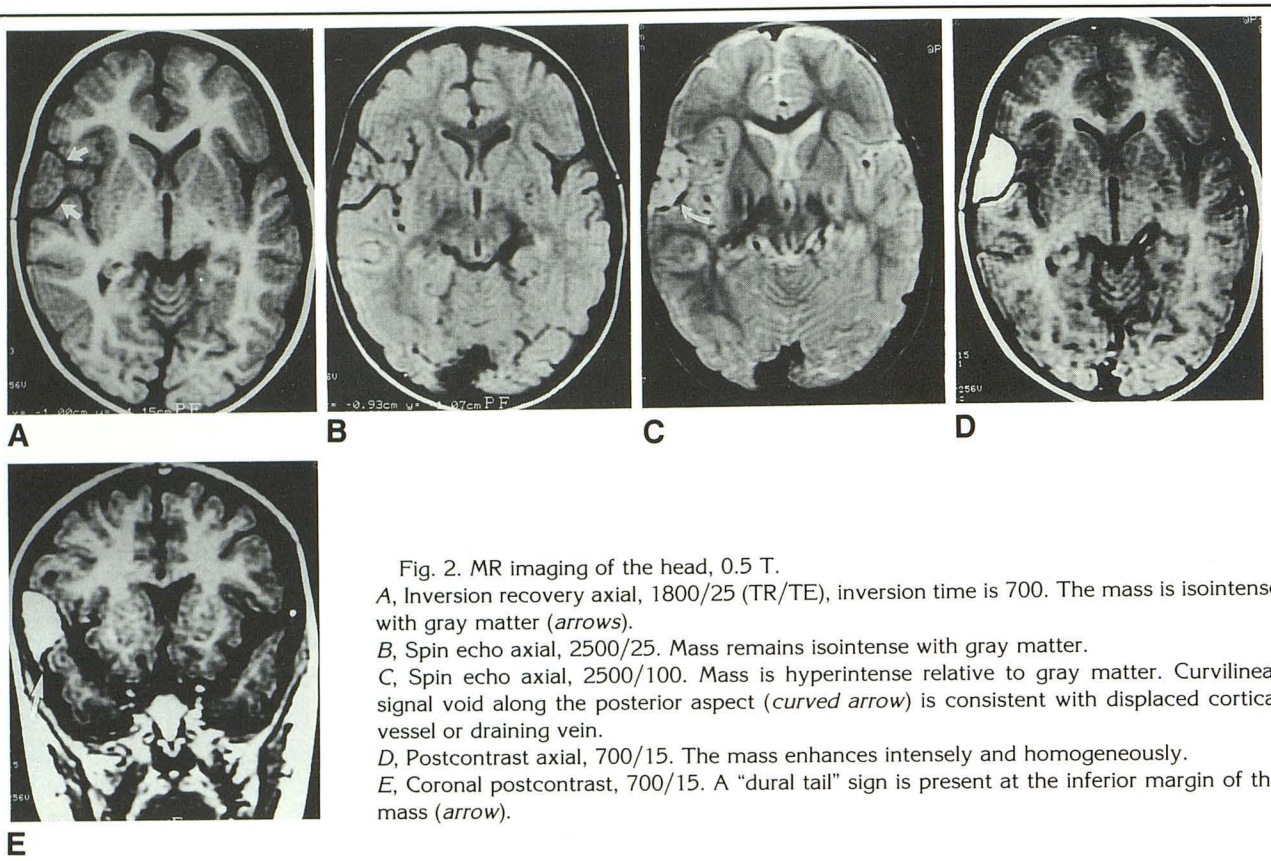


Fig. 2. MR imaging of the head, 0.5 T.
A, Inversion recovery axial, 1800/25 (TR/TE), inversion time is 700. The mass is isointense with gray matter (*arrows*).
B, Spin echo axial, 2500/25. Mass remains isointense with gray matter.
C, Spin echo axial, 2500/100. Mass is hyperintense relative to gray matter. Curvilinear signal void along the posterior aspect (*curved arrow*) is consistent with displaced cortical vessel or draining vein.
D, Postcontrast axial, 700/15. The mass enhances intensely and homogeneously.
E, Coronal postcontrast, 700/15. A "dural tail" sign is present at the inferior margin of the mass (*arrow*).

in females, and in adults may show size changes related to ovarian hormonal cycles and pregnancy (3). Cavernous hemangiomas frequently occur in children but do not involute spontaneously (3–6).

Location

The vast majority of capillary hemangiomas arise in the skin, scalp, or oral mucosa, typically appearing within a few months of birth (2). The cutaneous lesions can be quite disfiguring during the period of growth, and capillary hemangiomas of the oral cavity may bleed intermittently. The orbits are also a frequent site, particularly the palpebrae, often with extension to the rectus muscles and the intraconal space (4). Mucosal capillary hemangiomas of the upper airway, particularly the nasal cavity, occur in adults during the 4th and 5th decades.

Cavernous hemangiomas occur in subcutaneous, intramuscular, deep fascial, and visceral locations and may infiltrate deep soft tissues extensively. They can be associated with the Kasabach-Merritt syndrome, a microvascular coagulation within the tumor resulting in consumptive coagulopathy and thrombocytopenia (4–6).

Intracranial cavernous hemangiomas are common and are usually discovered in adulthood, either incidentally or during evaluation of seizures. They are usually parenchymal, but occasionally occur in extraaxial locations. In the bony walls of the central nervous system, hemangiomas of the cranium are nearly always cavernous, while vertebral hemangiomas are more often capillary in type (5). Intraosseous capillary hemangiomas have been reported in the frontal and sphenoid bones (5–7). Involvement of the central nervous system was reported in three cases of intraspinal capillary hemangiomas (8–10). One report describes an intracranial Masson hemangioma, an uncommon variety of hemangioma (11).

Pathology

Gross

The tumor was a spherical, smooth-surfaced, purple/red, firm, 2.2 cm × 2.5 cm ×

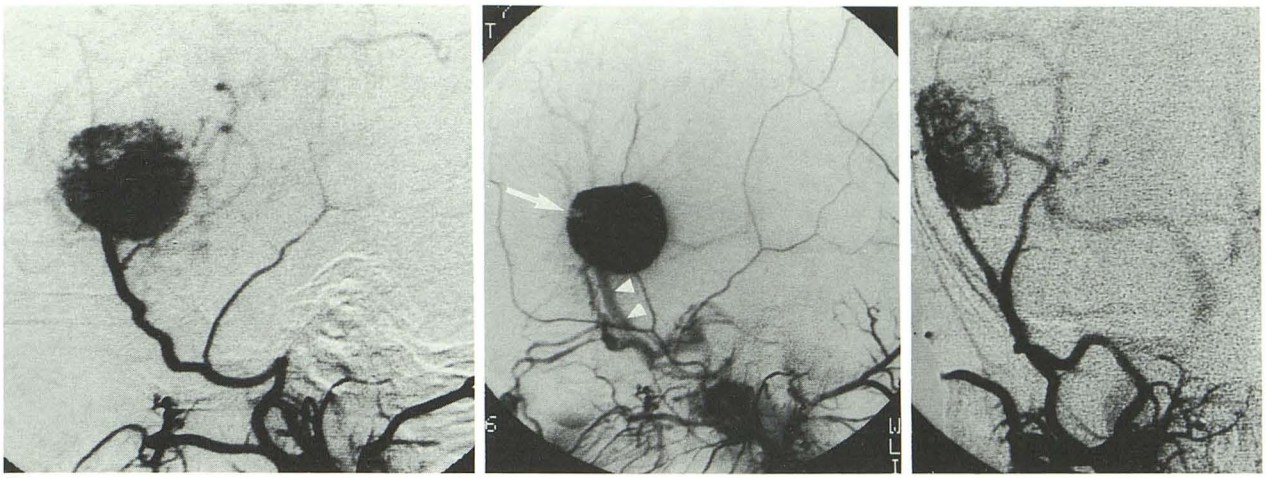


Fig. 3. Selective external carotid arteriogram, lateral view, early (*A*) and late (*B*) arterial phases.
A, A markedly enlarged middle meningeal artery supplies the tumor, which stains intensely.
B, Early draining veins are present (*arrowheads*). A small defect in the stain (*arrow*) may represent an early area of degeneration.
C, Anteroposterior view, early arterial phase. The penetrating vessels branch out in a radial pattern supplying individual lobules within the mass.

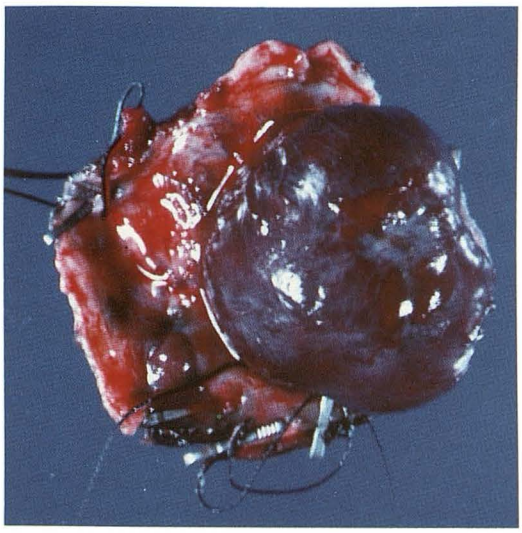
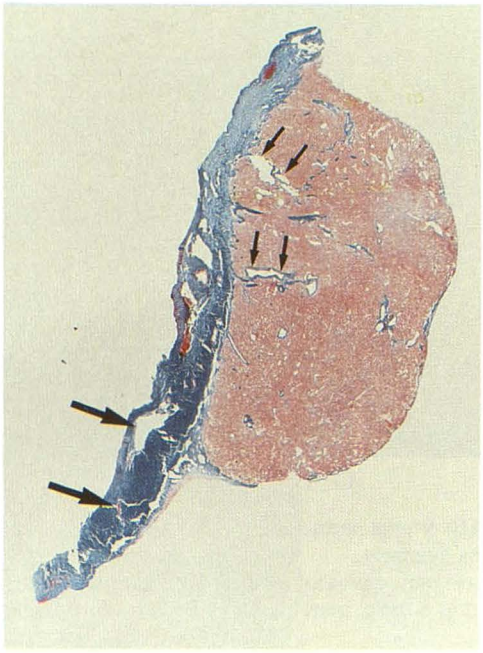


Fig. 4. Gross surgical specimen with attached dura. Tumor shape and color are characteristic of hemangioma, but highly vascularized meningioma, or another well-demarcated vascular tumor, may present a similar appearance.

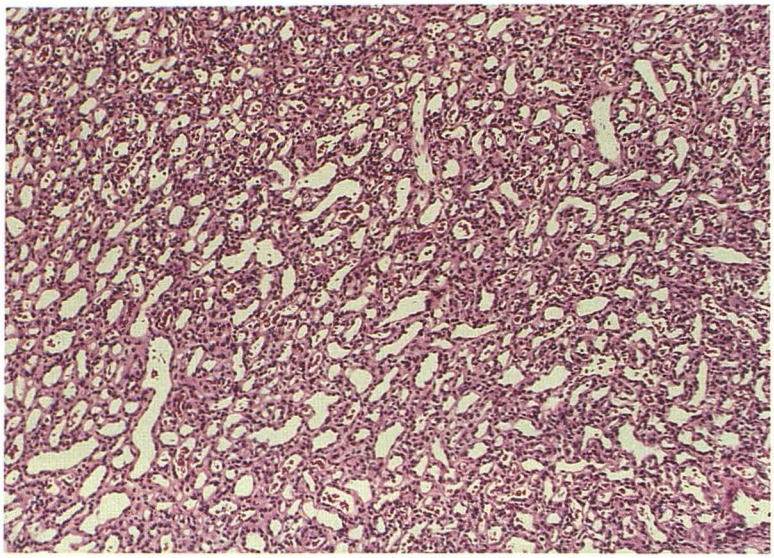
1.3 cm mass with a broad-based dural attachment (Fig. 4). Sectioning showed a uniformly spongy, dark red, oozing cut surface. Its color and character were typical of a vascular tumor.

Microscopic

The histopathology was diagnostic of capillary hemangioma. The tumor consisted of closely packed congeries of mature capillary-like vessels lined by flattened endothelium and separated by a scant amount of connective tissue stroma (Fig. 5). Encapsulation by dense connective tissue continuous with the inner aspect of the dura accounted for its grossly smooth surface (Fig. 5A). Thin-walled tiny feeding arteriolar and draining venous vessels were present along the inner dura; they communicated with centrally placed delicate branching vessels within the mass (Fig. 5A). Masson trichrome stain confirmed the presence of delicate smooth muscle within the walls of the feeding and draining vessels; some vessels showed mild mural and perivascular collagenization. Elastin stains showed elastic laminae within the dura-associated arteriolar vessels. Ectatic thin-walled vessels were also scattered in the subcapsular periphery of the mass. These more prominent vessels corresponded to the marginal, radial arteries and draining vein seen on angiography (Figs. 3B and 3C). Prominent vessels in the dural margins corresponded to the "dural tail" on MR imaging (Fig. 5A). Vascular dilatation and perivascular and mural collagenization are changes secondary to relatively high blood flow through and/or pressure within vascular lesions. Such high flow was demonstrated angiographically by the enlarged middle meningeal artery, intense staining, and early venous drainage. Reticulin stain showed delicate encirclement of each closely apposed capillary (Figs. 5E and 5F), characteristic of hemangioma. These findings



A



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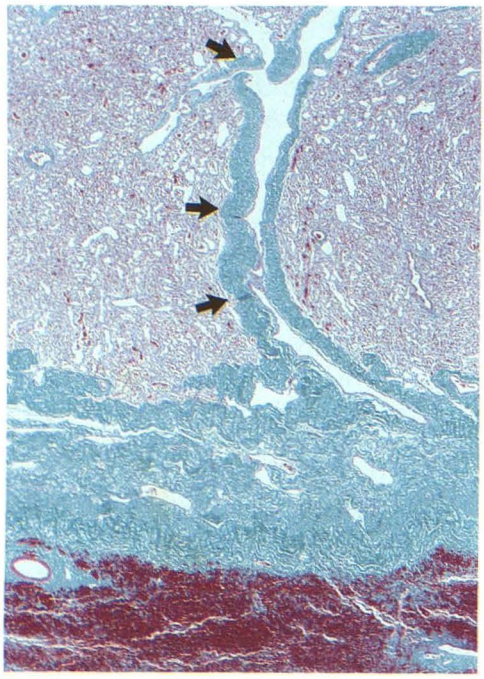


C

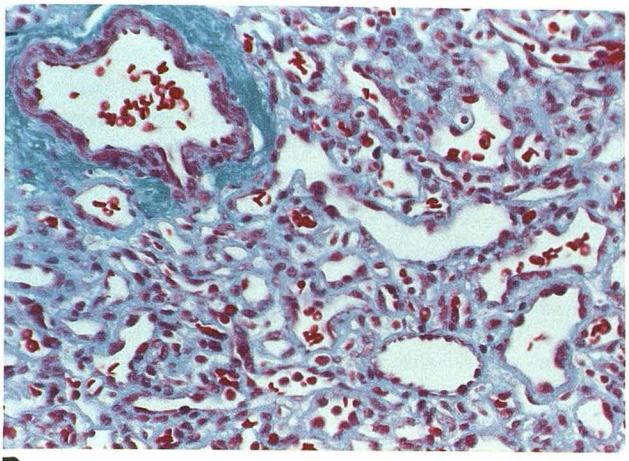
Fig. 5. *A*, Low-power photomicrograph of the capillary hemangioma, Masson trichrome stain, demonstrating dural origin and encapsulation (*blue* staining connective tissue), tiny feeder and prominent draining vessels (*small arrows*) and fine capillary meshwork of the tumor parenchyma. Increased dural vascularity (*large arrows*) corresponds to dural tail on MR.
B, Low-power photomicrograph, hematoxylin-eosin stain, revealing closely packed capillary-like, vascular spaces lined by flattened endothelium and surrounded by delicate connective tissue.
C, Low-power photomicrograph of dura-associated central draining vein of the tumor, Masson trichrome stain. Note collagenization of the vein wall (*arrows*) and connective tissue around the tributaries.

excluded other vascular tumors known to present in this location (12–14).

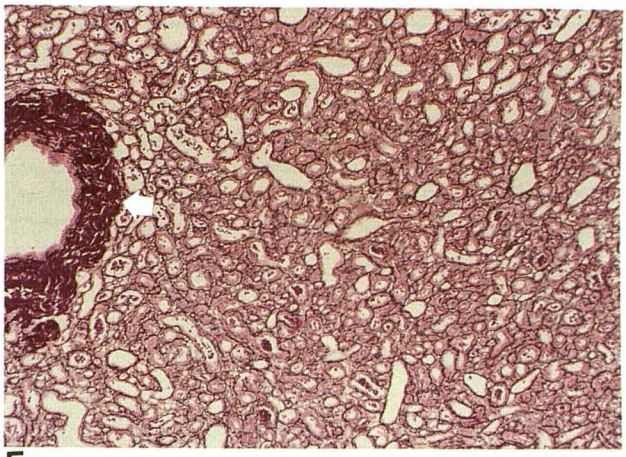
Pathologic Differential Diagnosis

On gross examination, meningioma generally presents a tough, pink-gray, whorled, or trabeculated cut surface. Foci of gritty calcification, recent or old hemorrhage, mucoid degeneration, or metaplastic cartilage or bone may be present. Highly vascularized meningiomas may, however, have a spongy red appearance (13, 15, 16).

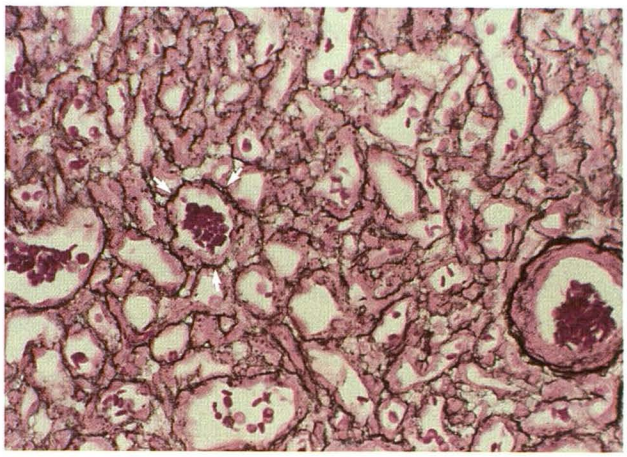
Histologically, meningioma, including the highly vascularized variant, was excluded by the absence of a spindled or meningothelial



D



E



F

Fig. 5. *D*, High-power photomicrograph demonstrating mature capillary-like spaces containing erythrocytes, Masson trichrome stain. Scant intervening connective tissue is present. The left upper corner shows branch of a small feeder arteriole with smooth muscle wall (*pink*) bordered by connective tissue (*blue*).

E, Low-power photomicrograph, and *F*, high magnification (reticulin stain) showing reticulin deposition outlining each capillary (*thin arrow*) and its scant deposition within the stroma. A portion of a draining vein is included at left border in (*thick arrow*) *E*.

TABLE 1: Mass lesions of the dura (excluding skull base)

Common
Meningioma
Hematoma
Uncommon
Abscess
Metastasis
Vascular malformation
Postoperative fibrosis
Rare
Sarcoma
Pachymeningitis
Hemangiopericytoma
Fibroma
Sarcoidosis
Capillary hemangioma

cell component. No whorled or storiform pattern, psammoma body formation, intranuclear vesicular inclusions or foci of foam cells, myxoid degeneration, or metaplastic cartilage or bone formation were seen (13, 15, 17). Furthermore, reticulin deposition in meningioma, though it outlines tumor blood vessels, effectively segregates clusters of intervening meningothelial cells (13); this pattern is unlike that seen in capillary hemangioma (Figs. 5E and 5F).

The differential diagnosis also included vascular lesions of the dura, namely vascular malformations and hemangiopericytoma (Table 1), which were excluded by histopathologic examination. Russell and Rubenstein consider vascular malformations as hamartomatous lesions and categorize them as arteriovenous malformation, venous malformation, capillary telangiectasia, and cavernous hemangioma (13). Enzinger and Weiss consider cavernous hemangioma a benign vascular tumor, but they acknowledge that the distinction between benign neoplasm and malformation is not always clear (3). However, all of these lesions exhibit much greater vessel caliber and structural variability than is seen in capillary hemangioma. Arteriovenous malformation shows dilated, abnormal, thick-walled vessels with altered elastin

TABLE 2: Diagnostic features of dural masses (excluding skull base)

	CT Findings	MR Findings	Angiographic Findings	Comments
Meningioma	Round, convex or flat dural mass. Intense contrast enhancement. Bone changes common: hyperostosis, erosion.	Iso- or hypointense to gray on T1, highly variable SI on T2WI. Intense enhancement. "Dural tail" sign usually present. Cerebral edema often present.	Supplied by meningeal vessels with occasional pial recruitment. Radial pattern of arterial feeders, dense staining persists late into venous phase.	The commonest meningeal neoplasm. Usually occurs in middle aged females.
Metastasis	Enhancing dural mass. Adjacent bone destruction common.	Slightly brighter than CSF on T1WI. Intense enhancement.	Usually hypovascular.	Miliary seedling more common than isolated mass.
Hematoma	Mass of increased or diminished attenuation. Convex or concave inner border. Enhancing wall or membranes in chronic stages.	Signal intensity a function of age. Subacute and chronic hematoma bright on T1—almost specific.	Avascular mass.	History of trauma or acute onset of severe headache.
Abscess/empyema	Low attenuation extraaxial mass with variable rim enhancement. Sinusitis usually present.	Slightly hyperintense relative to CSF on T1 and bright on T2WI. Marked rim enhancement.	Avascular mass. Possible early draining veins.	Fevers, headaches, meningismus, history of sinusitis or surgery.
Vascular malformation	Enhancing round or tubular structures. Adjacent hematoma or subarachnoid hemorrhage. Calcifications often present.	Signal voids, areas of hemorrhage.	Arteriovenous shunting. Enlarged feeding vessels and draining veins.	Acute onset of severe headache. May present with seizures.
Postoperative fibrosis	Craniotomy defects. Dural abnormalities usually not seen.	Thickened, enhancing dura subjacent to a craniotomy site.	No angiographic findings.	History of previous surgery.
Pachymeningitis	Thickened, enhancing tentorium or other dural regions.	Hypointense dural thickening with marked enhancement.	Narrowing of dural sinuses or carotid siphon.	Headaches, cranial nerve deficits, elevated sed rate.
Hemangiopericytoma	Lobulated mass, intense homogeneous enhancement, bone erosion. May have narrow dural attachment.	Intratumoral signal voids.	Multiple irregular feeding vessels.	Includes most tumors previously identified as angioblastic meningioma.
Capillary hemangioma	Slightly hyperdense mass with intense enhancement.	Hyperintense on T2WI, intense enhancement, draining veins.	Enlarged meningeal feeders, intense staining, lobular architecture.	Infants or very young children.

Note: SI = signal intensity; T1WI = T1-weighted images; T2WI = T2-weighted images.

and smooth muscle fibers in the arteries and collagenization of veins; no intervening capillaries are present. Venous malformation consists of a proliferation of varicose veins. Capillary telangiectasia displays interposed brain tissue between the variably sized, ectatic capillaries. Cavernous hemangioma shows centrally crowded, widely sized, thin-walled vascular spaces and peripheral hemosiderosis and infiltration by reactive gliotic brain tissue (13). Calcification and phlebolith formation are common (3). Hemangiopericytoma is a neoplastic proliferation of pericytes, the supportive cells surrounding capillaries. The tumor is highly vascular with a sinusoidal appearance and gaping, irregularly shaped (so-called staghorn) vascular spaces separated by the neoplastic spindle cells. Reticulin stains show reticulin deposition surrounding every pericyte in the intercapillary space and encircling tumor vessels (18).

Imaging

CT

Capillary hemangiomas are well circumscribed masses of high attenuation without

calcifications, and exhibit intense contrast enhancement. Our case followed this pattern. The intense enhancement may be attributed to their highly vascular structure with enhancement of the blood pool, and absence of a blood-brain barrier. Cavernous hemangiomas may contain phleboliths and have more unpredictable enhancement characteristics, sometimes enhancing only faintly or after a prolonged delay (4, 11).

MR

The MR features of 6 capillary hemangiomas of the nasal cavity were reported by Dillon et al (19). The tumors were of intermediate signal intensity on T1-weighted images and of increased intensity on T2-weighted images. The long T2 may reflect the long T2 of unclotted blood, which comprises a substantial portion of the mass. In this respect, capillary hemangiomas were not quite as intense as cavernous hemangiomas, which possess much larger vascular spaces in proportion to the size of the tumor (20). In two of Dillon et al's cases, areas of T2 shortening were seen correlating with clotted blood at pathologic examination. All tumors demonstrated intense homogeneous enhancement following intravenous gadolinium-DTPA infusion, which is expected because of their highly vascular architecture in conjunction with slow flow. Signal voids were not present as commonly seen with other hypervascular tumors or vascular malformations.

Our case manifested the same signal characteristics as extracranial capillary hemangiomas. The lesion was isointense with gray matter on T1-weighted images and hyperintense on T2-weighted images (isointense with cerebrospinal fluid). Intense homogeneous contrast enhancement occurred, with a "dural tail" sign. The dural tail appears to be representative of prominent meningeal vasculature. A signal void on the inner margin of the lesion was consistent with a prominent draining vein or displaced cortical vessel, but not a hemosiderin rim. On pathologic examination, we found an arterialized draining vein at this site on the margin of the tumor. There were no MR findings indicative of prior hemorrhage.

Angiography

The angiographic features of pediatric hemangiomas and vascular malformations were reported by Burrows et al (21). Their series included six hemangiomas. While differentiation between cavernous and capillary types was not made, the clinical characteristics of the lesions described were typical for capillary hemangiomas. All hemangiomas were characterized by sharp margins and intense persistent staining, usually in a lobular pattern. The tumors were supplied by slightly enlarged branches of normal systemic arteries. In three of the cases, branches of the feeding arteries encompassed the lesion, forming a so-called equatorial network with smaller feeding vessels branching at right angles. In two cases, the feeding arteries divided immediately into individual branches feeding each lobule. Direct arteriovenous shunting was not observed in this series, but has been reported. In the venous phase, small venous branches seemed to drain each lobule, joining into large veins at the base of the mass.

Our case exhibited the typical angiographic features of capillary hemangioma, eliminating from consideration cavernous hemangioma or vascular malformation. However, we were unable to eliminate meningioma based on any imaging findings, and that remained our preoperative diagnosis. A similar difficulty has been alluded to by other authors (22, 23).

Radiologic Differential Diagnosis

The differential diagnosis for dural masses is summarized in Tables 1 and 2. The most common meningeal tumor by far is meningioma. However, meningiomas represent less than 2% of intracranial tumors in childhood and commonly occur within the ventricles. Although rare in this age group, the imaging findings compelled us to consider meningioma as the likeliest diagnosis. Hematoma could be excluded on the basis of MR signal characteristics and the intense, solid contrast enhancement. The angiographic appearance was inconsistent with metastasis, carcinoma, vascular malformation, fibroma, or sarcoma. A hemangiopericytoma might still be considered, but none of

the characteristic features described by Buetow et al (24) were present, including multilobulated contour, a narrow dural base or "mushroom" shape, intratumoral signal voids on MR imaging, multiple irregular feeding vessels on angiograms, and bone erosion.

Summary

Hemangiomas are the most common tumor of the head and neck in children, including intracranial neoplasms. Capillary hemangioma in turn is the commonest type of hemangioma. Our case establishes that its anatomic distribution may include the intracranial compartment. We were unable to distinguish capillary hemangioma from meningioma based on imaging findings alone.

References

1. Burns AJ, Kaplan LC, Mulliken JB. Is there an association between hemangioma and syndromes with dysmorphic features? *Pediatrics* 1991;88:1257–1267
2. Batsakis JG, Rice DH. The pathology of head and neck tumors: vasoformative tumors. Part 9A. *Head Neck Surg* 1981;3:231–239
3. Enzinger FM, Weiss SW. Benign tumors and tumorlike lesions of blood vessels. In: *Soft tissue tumors*, 2nd ed. St. Louis: CV Mosby, 1988:491–497
4. Torres C, Kelley JK, Watts FB. Capillary hemangioma of the orbit: the role of computed tomography. *J Am Osteopath Assoc* 1987;87:82–86
5. Suss RA, Kumar AJ, Dorfman HD, Miller NR, Rosenbaum AE. Capillary hemangioma of the sphenoid bone. *Skeletal Radiol* 1984;11:102–107
6. Rothstein J, Maisel RH, Miller R, Tubman D. Mixed cavernous and capillary hemangioma of the frontal bone. *Ear Nose Throat J* 1985;64:43–49
7. Hook SR, Font RL, McCrary JA, Harper RL. Intraosseous capillary hemangioma of the frontal bone. *Am J Ophthalmol* 1987;103:824–827
8. Hanakita J, Suwa H, Nagayasu S, Suzuki H. Capillary hemangioma in the cauda equina: neuroradiological findings. *Neuroradiology* 1991;33:458–461
9. Mawk JR, Leibrock LG, McComb RD, Trembath EJ. Metameric capillary hemangioma producing complete myelographic block in an infant. *J Neurosurg* 1987;67:456–459
10. Tatsumi H, Kambara K, Fujii S, et al. A case report of dumbbell shaped cervical hemangioma. *Orthopedics* 1987;38:1871–1875
11. Chen TJ, Kuo TT. Giant intracranial Masson's hemangioma. *Arch Pathol Lab Med* 1984;108:555–556
12. Dehner LP. Vascular tumors. In: *Pediatric surgical pathology*, 2nd ed. Baltimore: Williams & Wilkins, 1987:874–883
13. Russell DS, Rubenstein LJ. *Pathology of tumors of the nervous system*, 5th ed. London: Edward Arnold, 1989:27–746
14. Chow L, Chow W, Fong DT. Epithelioid hemangioendothelioma of the brain. *Am J Surg Pathol* 1992;116:619–625
15. Kepes JJ. Topography and gross characteristics of meningiomas. In: *Meningiomas. Biology, pathology, and differential diagnosis.* New York: Masson, 1982:4–149
16. Davis RL, Robertson DM. *Malformations of intracranial vessels*, 2nd ed. Baltimore: Williams & Wilkins, 1991:627–631
17. Sheporaitis LA, Osborn AG, Smirniotopoulos JG, Clunie DA, Howieson J, D'Agostino AN. Intracranial menigioma: radiologic-pathologic correlation. *AJNR: Am J Neuroradiol* 1992;13:29–37
18. Enzinger FM, Weiss SW. Hemangiopericytoma. In: *Soft tissue tumors*, 2nd ed. St. Louis: CV Mosby, 1988:596–613
19. Dillon WP, Som PM, Rosenau W. Hemangioma of the nasal vault: MR and CT features. *Radiology* 1991;180:761–765
20. Itoh K, Nishimura K, Togashi K, et al. MR imaging of cavernous hemangioma of the face and neck. *J Comput Assist Tomogr* 1986;10:831–835
21. Burrows PE, Mulliken JB, Fellows KE, Strand RD. Childhood hemangiomas and vascular malformations: angiographic differentiation. *AJR: Am J Roentgenol* 1983;141:483–488
22. Atallah NK, Nassar SI. Calvarial hemangioma with blood supply from branches of the internal carotid artery. *J Neurosurg* 1971;34:823–826
23. Dickens JR. Cavernous hemangioma of the sphenoid wing. *Arch Otolaryngol* 1978;104:58
24. Buetow MP, Buetow PC, Smirniotopuolos JG. Typical, atypical, and misleading features in meningioma. *Radiographics* 1991;11:1087–1106