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Intracranial Hemangiopericytomas: MR and CT Features

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PURPOSE: To describe the MR and CT imaging features of hemangiopericytoma and to identify the characteristics that might distinguish them from meningioma. **METHODS:** We retrospectively reviewed the CT and MR findings in 34 pathologically proved cases of hemangiopericytoma. We evaluated the size, shape, and location of the tumor; the presence of hydrocephalus, edema, and mass effect; the type of dural attachment (broad-based or narrow-based) and bone changes (erosion, hyperostosis); and the tumor's density, signal, and contrast-enhancement characteristics. **RESULTS:** Thirty of 34 tumors were 4 cm or more in greatest dimension, 32 were lobular, and only seven were in the posterior fossa. Hydrocephalus was present in 18, edema in 30, and mass effect in 33. Twenty-three had broad-based dural attachment and 11 had narrow-based attachment. All 26 unenhanced CT scans showed hyperdense tumors; 19 were heterogeneous and seven homogeneous. All 27 contrast-enhanced CT scans showed enhancement; 17 were heterogeneous and 10 homogeneous. Bone erosion was present in 17 of 29 hemangiopericytomas imaged with CT. None had hyperostosis or tumor calcifications. On T1-weighted MR images, 13 of 17 tumors were isointense with cortical gray matter; on T2-weighted images, 10 of 17 were isointense. All 14 tumors imaged with contrast-enhanced T1-weighted MR imaging showed enhancement, and 13 of these were heterogeneous; eight of the 14 had a "dural tail" sign. **CONCLUSION:** Intracranial hemangiopericytomas are multilobulated, extraaxial tumors, sometimes associated with narrow-based dural attachment and bone erosion. Unlike with meningiomas, hyperostosis and intratumoral calcification are not present.

Index terms: Brain, neoplasms; Hemangiopericytoma

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Hemangiopericytomas were described in 1942 by Stout and Murray (1), in a series of nine cases, as tumors arising from the pericytes of Zimmerman, which are modified smooth muscle contractile cells surrounding capillaries. *Angioblastic meningioma* was the term used by Bailey et al (2) in 1928 to describe a meningeal

tumor observed in three cases. In 1954, Begg and Garret (3), in reviewing their single case of a hemangiopericytoma of the meninges and six cases of angioblastic meningioma described by Cushing and Eisenhardt (4), proposed that all of these tumors should be designated as hemangiopericytomas.

The current (1993) classification of the World Health Organization (WHO) has eliminated the term *angioblastic meningioma* in favor of *hemangiopericytoma*. Preoperative identification of these tumors is important because of their aggressive nature, high rate of local recurrence, and propensity for late, distant metastases (5-7). Guthrie et al (7) have recommended total surgical excision of intracranial hemangiopericytomas and postoperative irradiation before the first recurrence.

To determine the imaging characteristics of intracranial hemangiopericytoma, we reviewed the computed tomographic (CT) and magnetic

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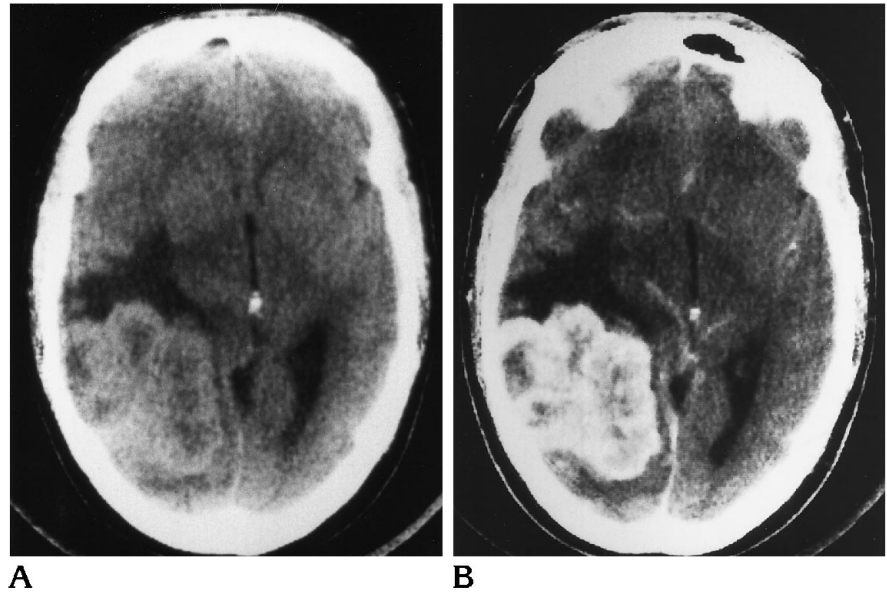
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Fig 1. A, Axial unenhanced CT scan at the level of the third ventricle shows a heterogeneous hyperdense extraaxial mass in the right occipital region, which is large and multilobulated. This mass straddled the right tentorium and exhibited edema and mass effect.

B, Axial contrast-enhanced CT scan shows heterogeneous dense enhancement. Pathologic diagnosis was anaplastic hemangiopericytoma with necrosis.



resonance (MR) findings in 34 patients with histologically proved tumors.

Materials and Methods

Thirty-four cases of intracranial hemangiopericytoma (26 primary, seven recurrent, and one metastatic from the abdominal wall) were collected from multiple institutions between 1979 and 1994. Case histories were reviewed and clinical data compiled by the authors. A previously reported case of hemangiopericytoma is included in our series (8). Hemangiopericytoma was diagnosed in 19 female and 15 male subjects, whose most frequent presenting symptom was headache. Patients were 14 to 73 years old; the mean age at initial presentation in female subjects was 46 years, the mean age in male subjects was 39 years.

All 34 hemangiopericytomas were surgically resected; of these, 28 surgical reports and one autopsy summary were available for review. All 34 tumors were confirmed as hemangiopericytomas on initial pathologic review. Thirty-two of 34 hemangiopericytomas were histologically re-evaluated for the purposes of this retrospective study by an experienced neuropathologist. These tumors were classified in accordance with the current criteria of the 1993 WHO system. Twenty-one were characterized as differentiated hemangiopericytomas and 11 were anaplastic hemangiopericytomas. The microscopic slides for the remaining two cases were not available for a second review.

All patients were examined with CT and/or MR imaging. CT protocols and MR sequence parameters, gradient field strength of the MR scanners, and dose of specific contrast agents varied among the referral institutions.

Obtained images included 26 unenhanced CT scans and 27 contrast-enhanced CT scans from a total of 29 patients who had unenhanced and/or contrast-enhanced CT examinations. There were 17 T1-weighted images and 17 T2-weighted images from a total of 18 patients who had

MR examinations (16 had both T1-weighted and T2-weighted studies, 14 had contrast-enhanced T1-weighted studies). These images were reviewed by a neuroradiologist and a research fellow in neuroradiology.

The CT and MR imaging features we assessed included tumor size, shape (round or lobular), and location (parasagittal, falcine, tentorial, anterior fossa, middle fossa, posterior fossa, vertex, sphenoid ridge); presence of hydrocephalus, edema, or mass effect; type of dural attachment (narrow based, with tumor diameter greater than dural attachment, or broad based, with tumor diameter equal to or smaller than dural attachment); bone erosion, hyperostosis, tumor calcification (present, absent, cannot be determined); and tumor contrast-enhancement characteristics, including presence or absence of a "dural tail" sign (9). Tumor density was noted on unenhanced CT scans. Signal intensity relative to cortical gray matter and serpentine signal voids (present or absent) were assessed on both T1-weighted and T2-weighted MR images.

Results

Twenty-eight surgical reports and one autopsy summary reported tumor invasion in 23 of 29 cases of hemangiopericytoma (10 venous, 10 bone, three nerve, and two falcine, with multiple sites of invasion noted in some cases), and no invasion noted in six of 29 cases. No clinical follow-up was available in most cases; however, subsequent to presentation, metastases occurred in at least three patients (pancreas, bone, and lung/liver, respectively).

Most of the hemangiopericytomas (30 of 34) were 4 cm or more in greatest dimension; the smallest was 2 cm and the largest was 9 cm. Thirty-two were lobular and two were round (Fig

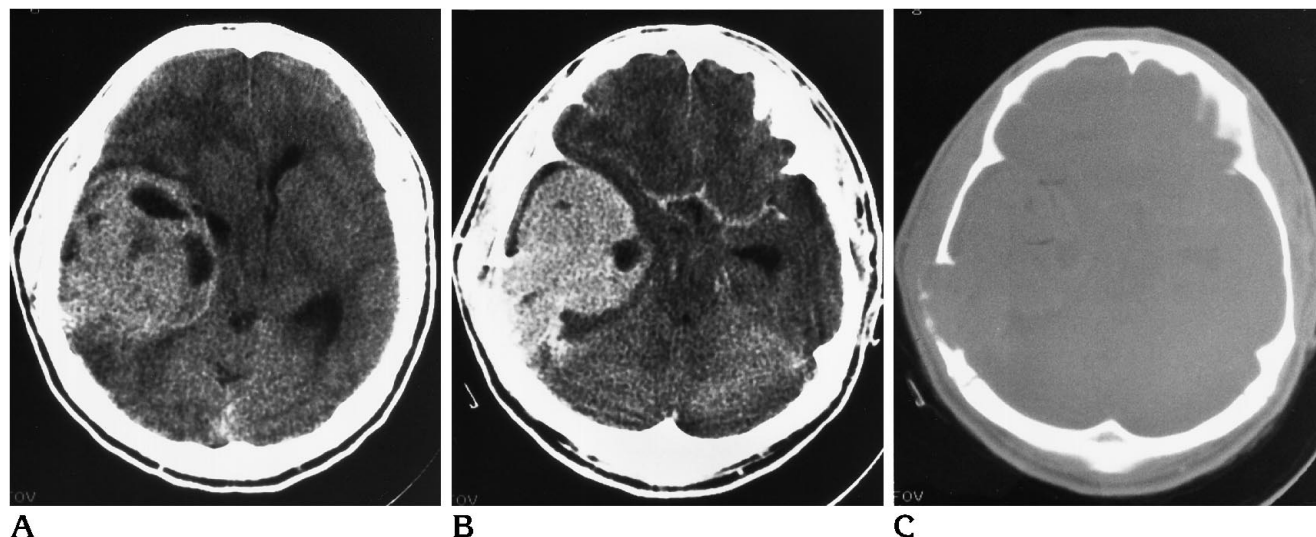


Fig 2. A, Axial unenhanced CT scan shows a very heterogeneous hyperdense extraaxial mass occupying the right middle fossa, causing moderate edema and severe mass effect.

B, Axial contrast-enhanced CT scan shows the narrow base of dural attachment of the mass (suggesting erosion of the temporal bone) and heterogeneous enhancement. Pathologic diagnosis was a differentiated hemangiopericytoma.

C, Axial bone window CT scan shows severe bone erosion of the mass and extension into the adjacent soft tissue.

1). Ten tumors were located in the middle fossa, seven were in the posterior fossa, six were fal-cine, four were in the anterior fossa, two were parasagittal, two were tentorial, two were at the sphenoid ridge, and one was at the vertex. Those located in the three fossae were in the basilar aspect of the fossae. Of the 34 cases of hemangiopericytoma, hydrocephalus was present in 18, edema in 30, and mass effect in 33. Twenty-three of the 34 hemangiopericytomas showed broad-based attachment to the dura and 11 had narrow-based attachment on CT and MR studies.

A total of 32 hemangiopericytomas were re-evaluated by our neuropathologist and designated as differentiated or anaplastic. Of 11 hemangiopericytomas that showed a narrow base of dural attachment on CT scans and/or MR images, seven were differentiated and four were anaplastic. Of 21 hemangiopericytomas that showed a broad base of dural attachment, 14 were differentiated and seven were anaplastic (Fig 2). None of the 32 hemangiopericytomas showed calcification histologically.

Nineteen of the hemangiopericytomas showed bone erosion on CT and/or MR studies. Fourteen of these were broad-based, of which eight were differentiated, five were anaplastic, and one did not receive a second review. Five of the 19 hemangiopericytomas with bone erosion were narrow-based; of which three were differ-

entiated and two were anaplastic. Of the remaining 15 of 34 hemangiopericytomas, seven did not show bone erosion; three of these were broad-based and four were narrow-based; six were differentiated and one was anaplastic. In the other eight, the presence of bone erosion could not be determined; seven of eight were broad-based and one was narrow-based; five were differentiated, two were anaplastic, and one did not receive a second pathologic review. Of 14 broad-based hemangiopericytomas showing bone erosion, eight were differentiated, five were anaplastic, and one did not receive a second pathologic review. None of the 29 cases imaged with CT had hyperostosis or tumor calcification.

Of a total of 26 unenhanced CT scans, 19 hemangiopericytomas appeared as heterogeneously hyperdense tumors with focal areas of hypodensity; the other seven tumors were homogeneously hyperdense. Of a total of 27 contrast-enhanced CT scans, 17 hemangiopericytomas had heterogeneous enhancement (four mild, five moderate, and eight marked); the other 10 had homogeneous enhancement.

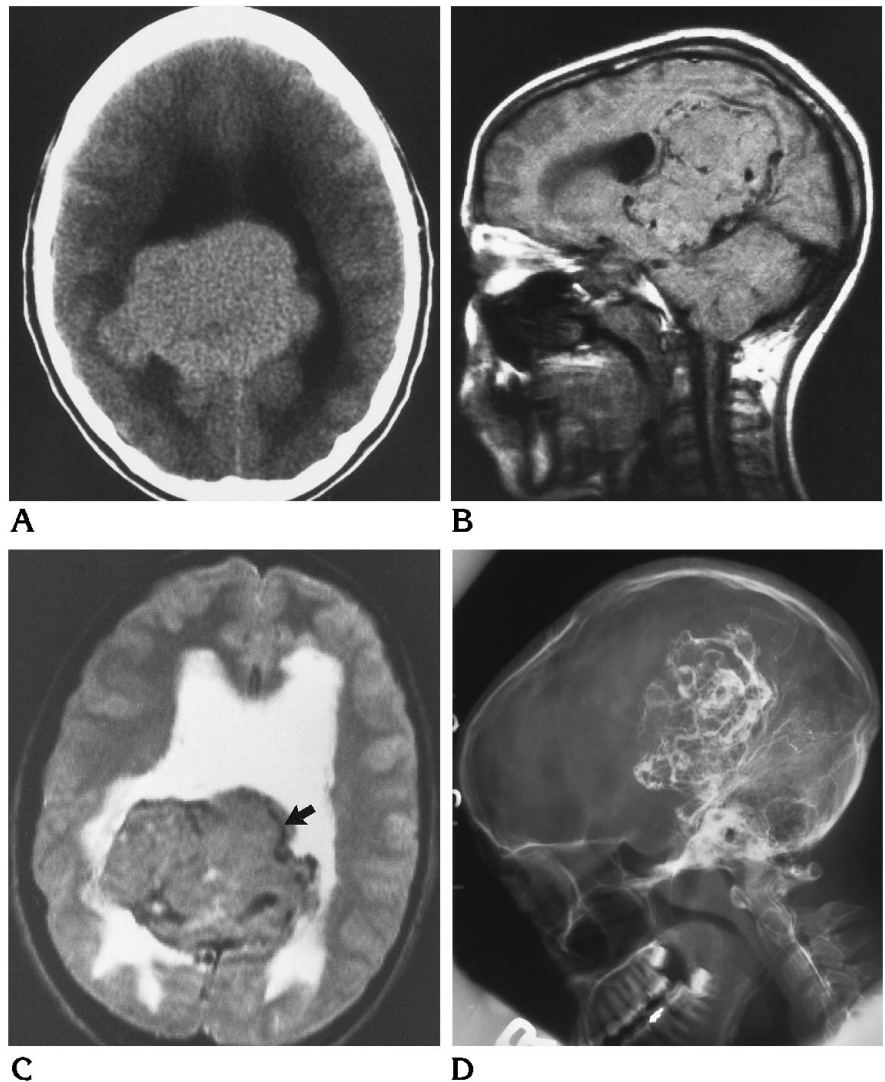
MR imaging was performed in 18 patients, and in 16 of these both T1-weighted and T2-weighted images were obtained. All 18 hemangiopericytomas had prominent internal serpentine signal voids, suggesting vessels (Fig 3). Calcifications were not present in any of these

Fig 3. A, Axial unenhanced CT scan shows a large, lobulated midline hyperdense mass.

B, Sagittal T1-weighted MR image shows that the bulk of the mass is isointense with cortical gray matter but has prominent internal signal voids that were not due to calcification (no calcification was present in the pathologic specimen).

C, Axial T2-weighted MR image shows a lobulated, bilateral falcine mass, which is heterogeneous, predominantly hypointense, with internal foci of hyperintensity, and has a broad base of dural attachment. Again, note multiple serpentine flow voids (arrow). Pathologic diagnosis was differentiated hemangiopericytoma.

D, Lateral view of right vertebral angiogram shows multiple large and irregular vascular channels within the mass.



tumors, either on unenhanced CT scans ($n = 26$) or on pathologic specimens ($n = 32$). For cases in which no CT studies were available ($n = 8$), we did not attempt to determine the presence of either calcification or hyperostosis. However, since these tumors do not calcify pathologically, hypointensities cannot be due to calcification.

Of a total of 17 T1-weighted MR images, 13 hemangiopericytomas were isointense with cortical gray matter (seven were homogeneously isointense and six were isointense with focal areas of hypointensity), two were hypointense, and two were hyperintense. Of a total of 17 T2-weighted MR images, 10 hemangiopericytomas were isointense with cortical gray matter (three had focal areas of hypointensity, four had foci of hyperintensity, and three were homoge-

neously isointense), four were hypointense with focal areas of hyperintensity, and three were homogeneously hyperintense.

Fourteen hemangiopericytomas were imaged with contrast-enhanced T1-weighted imaging, and all tumors enhanced. Thirteen showed heterogeneous enhancement and one showed homogeneous enhancement. Eight tumors showed a dural tail sign (Fig 4) and six did not (6).

Discussion

Intracranial hemangiopericytomas are neoplasms of pericytes that originate in the meninges and occur in a histologically pure form, without a meningioma component (10) (Fig 5). Hemangiopericytomas represent less than 1%

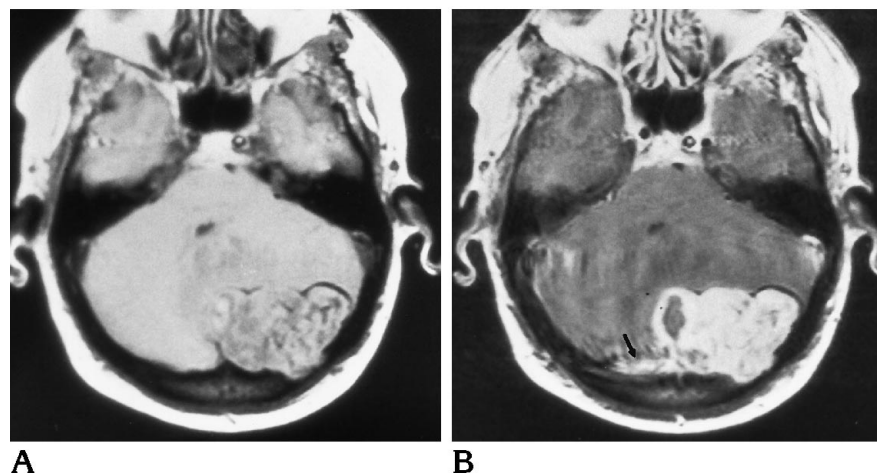


Fig 4. A, Axial T1-weighted MR image shows a heterogeneous extraaxial mass in the posterior fossa, with foci of hyperintensity and hypointensity.

B, Axial contrast-enhanced T1-weighted MR image shows a heterogeneously enhancing lobulated mass. A dural tail sign is seen posteriorly and extending to the right of the mass (*arrow*). Mass effect and edema are displacing the fourth ventricle anteriorly and to the right.

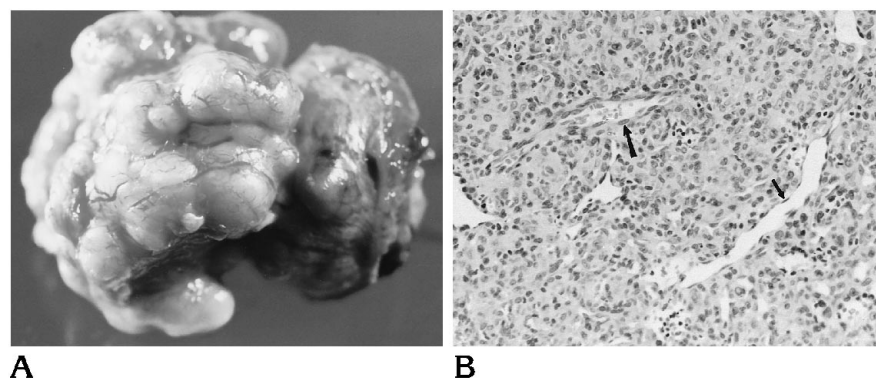


Fig 5. A, Gross image of a meningeal hemangiopericytoma shows the characteristic multilobularity.

B, Photomicrograph shows the characteristic appearance of a hemangiopericytoma: a single layer of endothelial cells lining the "staghorn" capillaries (*small arrow*). Oval or spindle-shaped cells (arising from pericytes) surround the vascular sheaths (*large arrow*).

of all central nervous system tumors (7). Surgically, hemangiopericytomas are described as well-demarcated masses attached to the dura, and are associated with profuse bleeding on resection (6, 11). These are aggressive lesions that tend to occur at an earlier age than other meningeal tumors, recur with high frequency, and metastasize extracranially, predominantly to bone, lung, liver, kidney, pancreas, and adrenals (5, 12). Postoperative radiation therapy and/or chemotherapy has been associated with increased survival time, regardless of the histologic subtype of hemangiopericytoma (5).

Intracranial hemangiopericytomas are detected at a mean age ranging from 37 to 44 years (6, 7, 11–14). In a pathologic series of 94 central nervous system hemangiopericytomas, Mena et al (5) reported a mean age at presentation of 47 years among female subjects and of 41 years among male subjects. The younger age at presentation in males is similar to that seen in our series. However, we found a slightly higher prevalence of intracranial hemangiopericytomas in females, which differs from earlier reports (5–7, 11–14). Goellner et al (6) reported

headache as the most frequently seen symptom in their series of 26 meningeal hemangiopericytomas, which agrees with our clinical findings.

In our series, intracranial hemangiopericytomas were multilobulated tumors, most of which were large (more than 4 cm in greatest dimension) at presentation. The presence and degree of hydrocephalus, edema, and mass effect were not, however, necessarily related to the size and/or location of the hemangiopericytomas. Although Elster et al (15) in their series of 40 meningiomas reported that the histologic subtypes of syncytial and angioblastic meningioma had the most severe edema, only four of 14 meningiomas from those two subtypes were angioblastic meningiomas.

The location of intracranial hemangiopericytomas is similar to that of meningiomas. Like meningiomas and other extraaxial masses, hemangiopericytomas are dural-based and show white matter "buckling" (16). New et al (17) described the three most common sites of 164 meningiomas as sphenoid/parasellar, lateral convexity, and superior parasagittal; no pathologic subtyping of meningioma was reported in

this series. Zimmerman et al (16) reported the three most common sites in 32 meningiomas as middle fossa, anterior fossa, and posterior fossa, with a basal predominance. Guthrie et al (7) reported that 27 of the 44 intracranial hemangiopericytomas in their series were supratentorial; the four most common sites of occurrence were parasagittal/falx, convexity, posterior fossa, and tentorial; none occurred as purely intraparenchymal masses. Pitkethly et al (12) described the most common sites of occurrence in the hemangiopericytic variant of angioblastic meningiomas as the frontal fossa, middle fossa, and posterior fossa. These findings and those of our series confirm that hemangiopericytomas occur in locations similar to meningiomas.

Approximately one third of our hemangiopericytomas showed a narrow base of dural attachment, with the remaining two thirds showing broad-based attachment. In a review of 21 intracranial hemangiopericytomas by Jaaskelainen et al (11), all eight tumors imaged by CT showed broad-based dural attachment. Buetow et al (18) reported that up to 85% of meningiomas in their series showed a broad base of dural attachment. Our series indicated that hemangiopericytomas may show a narrow base of dural attachment, a feature not typically seen in meningiomas. Furthermore, we found no relationship between histologic subtype of hemangiopericytoma and mode of dural attachment.

Bone erosion was a feature seen in more than half our cases of hemangiopericytoma. Osborne et al (14) described a series in which six of six intracranial hemangiopericytomas were associated with lytic destruction of bone on plain films. Cosentino et al (8) described a very large lytic vertex hemangiopericytoma seen on plain films and MR images, which we included in our series. Bydder et al (19) reported two of 12 cases of meningioma with MR evidence of bone erosion, although pathologic findings in these cases were not discussed. Meningiomas are typically described as dural-based tumors that are frequently associated with hyperostosis and calcifications (16–18, 20). None of our hemangiopericytomas showed hyperostosis on CT scans. On the other hand, bone erosion was seen in most of our anaplastic hemangiopericytomas but was present in only about half of the differentiated hemangiopericytomas. No relationship was noted between mode of dural attachment and bone erosion.

Reports of the CT appearance of intracranial hemangiopericytoma are scarce. Hemangiopericytomas, like meningiomas, are extraaxial lesions, but unlike meningiomas, they do not have tumor calcifications (7, 11, 14). In their review of 131 atypical-appearing meningiomas, Russell et al (21) described two cases imaged with CT: the first was a cystic, isodense, falcine mass with dense nodular enhancement of a focal nodule (angioblastic type); the second was a hyperdense convex mass, lacking calcifications, with heterogeneous enhancement (syncytial meningioma with angioblastic elements). Servo et al (22) summarized their CT findings in eight cases of intracranial hemangiopericytomas as follows: unenhanced CT scans showed isodense or slightly hyperdense, well-defined, sometimes nodular masses, without calcifications, and these lesions were associated with slight edema. These hemangiopericytomas were typically connected to the convexity or falx with a broad base, were often bilateral, and often showed dense, ring-shaped enhancement (22). Guthrie et al (7) reported that all four of their cases of hemangiopericytomas examined with CT had imaging features similar to meningiomas, without describing specifics. Osborne et al (14) described three of six intracranial hemangiopericytomas as “diffusely enhancing” on contrast-enhanced CT scans. Overall, these reports agree with our findings that intracranial hemangiopericytomas are heterogeneous, hyperdense, dural-based lesions that, unlike meningiomas, are not associated with calcifications or hyperostosis, and typically show heterogeneous enhancement on enhanced CT scans.

Descriptions of the MR imaging features of intracranial hemangiopericytoma are similarly limited. Guthrie et al (7) described the T1-weighted and T2-weighted MR imaging features in three of 44 intracranial hemangiopericytomas as isointense on both sequences. Cosentino et al (8) described a single giant intracranial hemangiopericytoma (included in our series). The mass was heterogeneous, isointense with gray matter on T1-weighted images, slightly hyperintense on T2-weighted images, with signal vessel voids. Our MR imaging findings concur with these reports: intracranial hemangiopericytomas were heterogeneous, predominantly isointense on T1-weighted and T2-weighted MR images, showed prominent internal vessel voids, and enhanced heterogeneously on contrast-enhanced MR images. In

our series, slightly more than half of the hemangiopericytomas examined with contrast-enhanced MR imaging showed a dural tail sign. Tien et al (9) concluded that the dural tail sign in a patient with no history of surgery or radiation therapy was highly suggestive, but not diagnostic, of meningioma. Aoki et al (20) described the "flare" sign as linear enhancement along the dura emanating from the margins of a meningioma. They believed that this sign was caused by hypervascularity, did not always indicate dural invasion by tumor, and might be shown in other meningeal tumors and nonneoplastic disorders that produce meningeal irritation (20).

In conclusion, intracranial hemangiopericytomas are dural-based hypervascular masses similar to meningiomas; however, histologically they are not meningiomas, and they often have different CT and MR imaging features. Intracranial hemangiopericytomas are rare, extraaxial, multilobulated masses that typically occur in patients in their fourth and fifth decades. Although meningiomas are frequently associated with dural invasion and with development of abnormal vessels (20), hemangiopericytomas are more aggressive, tend to recur even after gross total resection, and occasionally have extracranial metastases. Unlike meningiomas, which frequently show hyperostosis of adjacent bone and may have intratumoral calcifications on unenhanced CT scans, hemangiopericytomas show bone erosion and lack calcifications and hyperostosis. Meningiomas typically show a broad base of dural attachment on CT and MR studies; therefore, if a dural-based mass has a narrow attachment, one should consider the possibility of hemangiopericytoma. Unenhanced CT scans of hemangiopericytomas typically show hyperdense heterogeneous tumors, and T1-weighted and T2-weighted MR images typically show heterogeneous isointense tumors. Hemangiopericytomas show heterogeneous enhancement on both contrast-enhanced CT scans and MR images.

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