Primary intracranial meningeal and spinal hemangiopericytoma: radiologic manifestations.

D R Osborne, P Dubois, B Drayer, M Sage, P Burger and E R Heinz

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Primary Intracranial Meningeal and Spinal Hemangiopericytoma: Radiologic Manifestations

Meningeal hemangiopericytomas are rare but aggressive tumors. All eight patients in this series developed recurrences, three with distant metastases. In six patients a lytic destructive lesion of bone was evident. CT scans in three patients demonstrated diffusely enhancing lesions. No tumor calcification was present. In five of six patients angiography was characterized by multiple irregular tumor vessels, an intense prolonged blush, and the absence of early draining veins. Differentiation of these tumors from meningiomas is discussed.

Hemangiopericytoma is a rare vascular tumor deriving from capillaries and pericytes [1–4]. Although it is a well established entity clinically and histologically, its occurrence in the intracranial meninges and spine has been controversial and, until recently, disputed [5–7]. Earlier investigators classified hemangiopericytomas as angioblastic variants of meningioma [5–7]. They are now considered separate both clinically and histologically [6, 8–12]. Hemangiopericytomas of the intracranial meninges and spine have a high rate of recurrence after primary therapy, occasionally metastasize, and tend to occur in a younger group of patients than do meningiomas [6–8, 11, 13, 14]. This report evaluates the radiologic findings in cases of primary hemangiopericytomas of the intracranial meninges and spine and investigates those features which may allow radiologic differentiation from meningioma.

Materials and Methods

Clinical and radiologic findings of eight cases of histologically proven hemangiopericytomas of the intracranial meninges and spine referred to Duke University Medical Center over a 20 year period were reviewed. Histologic diagnosis was by the Section of Neuropathology, Duke University Medical Center.

Results

All patients had an extraaxial mass. There were six patients with intracranial meningeal hemangiopericytomas, four supratentorial and two in the posterior fossa. The two lumbosacral spinal tumors presented with extradural masses (table 1).

On plain film examination, six of the patients had a lytic cranial or spinal lesion due to bone destruction (fig. 1A). None of the tumors generated a sclerotic bone response, and no tumor calcification was demonstrated.

A diffusely enhancing tumor was present in all patients who had had CT (figs. 2A, 3A, 3B and 4A). In one of these patients, bolus-contrast-enhanced rapid-sequence CT demonstrated areas of vascular pooling and variable matrix enhancement (fig. 2B) not seen on the conventional-enhanced examination. Tumor calcification was not seen.
<table>
<thead>
<tr>
<th>Case No. (Age, Gender)</th>
<th>Primary Site</th>
<th>Plain Film</th>
<th>CT, Scintigraphy, or Myelography</th>
<th>Angiography</th>
<th>Clinical Course</th>
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<tr>
<td>1 (54, M)</td>
<td>R occipital meninges</td>
<td>Destructive lesion R occipital bone</td>
<td>CT: diffusely enhancing right occipital tumor</td>
<td>Densely persistent tumor blush; no e.d.v.; Supply: middle meningeal and occipital arteries</td>
<td>Recurrence inferior surface of tentorium at 3 years</td>
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<td></td>
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<td></td>
<td>Irregular peripheral tumor vessels; dense persistent tumor blush; no e.d.v.; Supply: superior cerebellar, posterior vertebral meningeal arteries</td>
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<td>2 (32, M)</td>
<td>R occipital meninges</td>
<td>Destructive lesion R occipital bone</td>
<td>CT: diffusely enhancing posterior fossa tumor</td>
<td>Irregular peripheral tumor vessels. Dense persistent tumor blush; no e.d.v.: venous pooling; Supply: meningeal and posterior inferior cerebellar arteries</td>
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<td>Pulmonary metastases at 9 years; posterior fossa occipitocervical recurrence at 18 years</td>
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<td>3 (43, F)</td>
<td>Foramen magnum</td>
<td>Eroded foramen magnum</td>
<td>CT: diffusely enhancing posterior fossa tumor</td>
<td>Dense persistent tumor blush; no e.d.v.; Supply: meningeal and posterior inferior cerebellar arteries</td>
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<td>Lytic metastasis L clavicle at 1 year; recurrence tentorium at 2 years</td>
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<td>4 (35, F)</td>
<td>L middle cranial fossa floor</td>
<td>Destructive lesion, floor middle cranial fossa and petrous apex</td>
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<td>Dense persistent tumor blush; Supply: meningeal artery</td>
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<td>Recurrence middle cranial fossa at 2 years</td>
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<td>5 (55, F)</td>
<td>L parietal meninges</td>
<td>Lytic lesion L parietal bone</td>
<td>Radionuclide scan: tumor uptake</td>
<td>Dense persistent tumor blush; no e.d.v.; Supply: meningeal, superficial temporal and occipital arteries</td>
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<td>Recurrence L parietal region at 7 years</td>
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<td>6 (47, F)</td>
<td>R sphenoidal ridge</td>
<td>Normal</td>
<td>Radionuclide scan: tumor uptake</td>
<td>Dense persistent tumor blush; Supply: middle meningeal and middle cerebral arteries</td>
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<td>Local recurrence and subarachnoid seeding at 1 year</td>
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<td>7 (38, F)</td>
<td>Sacrum</td>
<td>Lytic lesion sacrum</td>
<td>Myelogram: L5-S1-S2 extradural tumor</td>
<td>Pulmonary metastases</td>
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<td>Local recurrence at 6 years</td>
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<td>8 (43, F)</td>
<td>Lumbosacral junction</td>
<td>Normal</td>
<td>Myelogram: Lumbosacral extradural tumor</td>
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Note. — e.d.v. = early draining veins.

Fig. 1.—Case 7. Spinal hemangiopericytoma. A, Lytic lesion of left side of sacrum (arrows). B, Myelogram. Extradural mass effect on sacral subarachnoid sac (arrows).

Angiography demonstrated a highly vascular extraaxial mass with multiple irregular, predominantly peripheral early filling tumor vessels but no early draining veins (figs. 2C and 4B). In five of the six patients with angiography there was persistent and dense tumor blush (figs. 4C and 4D). Vascular pooling of contrast material was present in one patient. The blood supply to the tumors was from branches of the meningeal arteries, but in four instances an additional supply
Fig. 3.—Case 1. Right occipital supratentorial hemangiopericytoma. A, Unenhanced CT scan. Tumor demonstrated by rounded area of slightly increased attenuation. B, Tumor enhancement with intravenous contrast. C, Vertebral angiogram. Capsular branches from calcarine branch of posterior cerebral artery cap lesion (arrowheads). No tumor stain demonstrated on vertebral angiograms, but was present on external carotid study.

Fig. 4.—Case 1. Tentorial recurrence posterior fossa. A, Enhanced CT scan. B, Left vertebral angiogram, early arterial phase. Enlargement of posterior meningeal artery (arrowheads) which supplies periphery of tumor posterosuperiorly, while branches of superior cerebellar artery (arrows) supply tumor "capsule" anteriorly. C and D, Left vertebral angiogram, late arterial phase. Tumor blush. No early draining veins.

from either an intracranial internal carotid or vertebral artery was demonstrated (fig. 4B).

The two patients with spinal tumors had their extradural masses demonstrated at myelography (fig. 1B).

Surgery was the initial treatment in the eight patients, curative resection being attempted in five. Tumor recurrence developed in all eight, from as early as 1 year to as late as 18 years after surgery. Recurrence was local in four, adjacent to but not at original site in four, and in three patients distant metastases developed in the lungs and skeleton (fig. 2D).
Discussion

Stout and Murray [1, 2] coined the term hemangiopericytoma to describe a soft tissue neoplasm in which there are pericytic tumor cells tightly packed in and around capillaries and lymphatics. Reports of this tumor occurring intracranially and in the spine are very infrequent [6-8, 13]. There is one case report of a primary tumor involving the choroid plexus of a lateral ventricle [15]; in all others, the location has been primarily in the intracranial and spinal meninges. Kernohan and Uhlen [8] reported on 26 hemangiopericytomas to emphasize their distinct histologic features and high recurrence rate after primary treatment. Kruse [7] noted recurrence in six of eight cases he reported, while Goellner et al. [13], reviewing the Mayo Clinic experience (26 cases), found a recurrence rate of 80% and metastases in 23%. The aggressive nature of the tumor is a feature that further differentiates the hemangiopericytoma from meningiomas, even hemangioblastic meningiomas [11, 14] and is confirmed again in our study. In the spine, reports of primary hemangiopericytomas are even less frequent, but here also recurrences are common [7, 16-18]. Recurrence may be delayed and indolent. One of our patients (case 2) developed pulmonary metastases 9 years after primary resection of his occipital tumor. These metastases have been present untreated a further 9 years, at the end of which time two local recurrences of the occipital tumor have developed.

Radiologic analyses of the meningeal hemangiopericytomas is scant. Plain film examination has not been specifically addressed in previous reviews. In six of our patients there was a lytic bony lesion. No patient had a tumor that generated a sclerotic bone response. Lytic cranial or vertebral lesions have been noted incidentally and mentioned in case report summaries in six other cases [7, 17, 19].

Marc et al. [20] reported the angiographic findings in five intracranial hemangiopericytomas. They described multiple irregular feeding vessels, an intense “fluffy” stain, lack of early draining veins, and a prolonged tumor circulation. There was a dual arterial supply from meningeal arteries and branches of the internal carotid. They believed that the angiographic architecture was sufficiently different to allow a highly probable diagnosis of hemangiopericytoma. Our cases support their view; further, the angiographic architecture in their series and ours is similar to that described for hemangiopericytoma occurring elsewhere in the body [3, 19]. In six of our patients who had angiography, the tumor was supplied primarily from meningeal vessels as befits a tumor of the meninges, but a dual arterial supply was present in four of these. In three of these (cases 1-3) the intracranial internal carotid or vertebral supply was most marked on the angiographic examination of the tumor recurrence. Tumor spread in these three patients was adjacent to but not at the site of the original tumor. In one patient (case 6) subarachnoid seedings were observed at operation. This is the probable mode of spread of tumor in cases 1-3 and is the most likely explanation for the dual arterial supply.

The appearance of an intracranial meningeal hemangiopericytoma on CT examination has not been reported previ-ously. The unenhanced study demonstrated the tumor as an area of slightly increased attenuation in one patient, but in the other two there was no more than evidence of mass effect. Our three cases displayed a homogenously enhancing tumor after contrast material; in one of these, bolus-contrast-enhanced rapid-sequence CT demonstrated venous pooling and a variable matrix enhancement.

No tumor calcification was seen in our series on the plain film examination or on CT in the three patients who had had this study. It has occasionally been reported in hemangiopericytomas elsewhere in the body, although never with an intracranial or spinal lesion.

Differentiation of hemangiopericytoma from meningioma is important because of the aggressive nature of hemangiopericytoma. Although the intracranial and spinal origin of both tumors is similar, a lytic cranial or spinal lesion, absent tumor calcification, and consistent angiographic features should raise the suspicion of hemangiopericytoma in a young patient. Careful histologic analysis is necessary, however, before any firm differentiation can be made.

REFERENCES

16. Pittlyk PJ, Dockery MB, Miller RH. Hemangiopericytoma of the


