Giant Cranial Hemangiopericytoma: MR and Angiographic Findings

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Summary: The authors detail the MR and angiographic findings of a very large hemangiopericytoma in the skull of a 63-year-old woman. Angiography showed a marked tumor blush with early draining veins; MR showed heterogeneously increased T2 signal intensity and enhancement with gadolinium secondary to the richly vascularized tumor.

Index terms: Brain neoplasms, magnetic resonance; Brain neoplasms, angiography; Hemangiopericytoma

Hemangiopericytoma is a mesenchymal tumor of varying malignant potential. It rarely occurs intracranially and can resemble a meningioma when small to moderate in size. This tumor can produce predominantly lytic calvarial lesions and intense enhancement on computed tomography (CT) scans. Angiography reveals dural supply, irregular feeding vessels, “fluffy” staining, lack of early veins, and prolonged tumor circulation time. Magnetic resonance (MR) in our case shows heterogeneously increased T2 signal intensity and enhancement with Gd/DTPA secondary to the increased vascular permeability, compact cellular pattern, and prominent neovascular channels.

Case Report

A 63-year-old woman had noted a slowly enlarging mass at the top of her head for approximately 4 years. She attributed the abnormality to a remote injury, but became concerned because of recent increase in the size of the lesion. The patient denied any symptoms related to the mass other than a mild local pressure sensation.

Physical examination demonstrated a nontender, broadly based, pedunculated mass beneath the scalp, measuring 13 × 13 × 9 cm. The superficial scalp arteries were enlarged. No neurologic deficit was identified.

Anteroposterior and left lateral skull radiographs (Fig. 1) showed a large calvarial osteolytic defect with an overlying soft-tissue mass. A few calcific flecks were present at the base of the mass. A radionuclide bone scan showed increased activity at the margin of the large irregular area of destruction involving the skull vertex. No additional skeletal lesions were detected.

MR (Figs. 2A–2D) showed a large multilobulated soft-tissue mass at the skull vertex, slightly larger on the left side. The underlying calvarium was destroyed with the mass extending intracranially. Sharp demarcation between the mass and the underlying compressed parietal lobes was noted. The superior sagittal sinus was displaced but not thrombosed. Mild deformity of the lateral ventricles was present. The lesion was predominantly isointense to gray matter on T1-weighted and proton-density images. On T2-weighted images, the mass was slightly hyperintense to gray matter. Several focal areas of prolonged T1 and T2 signal intensity were noted, consistent with necrosis. Multiple areas of signal void within the mass were felt to represent neovascular channels.

Subselective external carotid angiography (Fig. 3A) demonstrated hypertrophied and tortuous occipital and superficial temporal arteries supplying the lesion with the base of the tumor receiving parasitic supply from the prominent middle meningeal arteries. The typical arterial spoke-wheel pattern of a meningioma was absent. In addition, irregular tumor vessels with occasional “cork-screw” pattern and a lobular, well-margined tumor with densely heterogeneous stain (Figs. 3A and 3B) were noted. Large vascular channels and arteriovenous shunting were present within the tumor. While the smaller endocranial portion of the tumor had recruited some arterial supply from both middle meningeal arteries, the angiographic pattern was most consistent with an extra-axial malignant mass based on the primary blood supply from branches of the external carotid arteries. The internal carotid artery studies showed no major contribution to the arterial supply of the tumor.

Preoperative embolization was performed with small fragments of gelatin sponge. These were delivered into the tumor via a 5-F catheter. Successful occlusion of left external carotid contribution and partial occlusion of the right external carotid supply was achieved.

At surgery, all visible tumor was removed. The tumor was extradural in location, but adherent to the adjacent dura at several points. Despite the embolization procedure, a large amount of intraoperative hemorrhage occurred.

Pathologic evaluation of the specimen showed a hemangiopericytoma. Histologically, two components to the tu-
Fig. 1. Lateral view of the skull shows destructive change associated with soft-tissue mass.

mor were identified: loose and compact cellular patterns corresponding to the two areas of increased signal intensity on T2-weighted images. Multiple vascular channels with palisading pericytes were noted. Positive reticulin staining was present.

The postoperative course included evacuation of a large epidural hematoma and removal of redundant scalp tissue, simultaneously. The patient developed a recurrence in the region of the previous surgery, approximately 18 months later, which demonstrated an increase in malignant characteristics on microscopic evaluation.

Discussion

Stout and Murray first described hemangiopericytoma as a clinical entity in 1942 (1). The tumor has been diagnosed in multiple areas of the body. It accounts for less than 2% of soft-tissue sarcomas (2) and originates from cells termed pericytes, which are contractile cells surrounding capillaries.

The biologic behavior of the tumor appears to be independent of the primary site of involvement. It is a soft-tissue sarcoma of variable malignant potential. Neither the lack of mitoses nor absence of anaplasia ensures a benign course (3, 4). Goellner et al reviewed 26 cases of meningeal hemangiopericytomas and found no relationship between the histologic division and clinical course (5). Fatal results due to the tumor occurred in seven of nine patients with borderline histology and 12 of 17 with a malignant histology (5).

Approximately one quarter of hemangiopericytomas occur in the head and neck (6). Recent reports demonstrate a young age of onset with a slight predilection for males (7). Intracranial hemangiopericytomas are mostly supratentorial in location, attached to the falx, tentorial, or dural sinuses (7). The typical presentation is a slowly enlarging painless soft-tissue mass. In three quarters of the cases, the tumor is well circumscribed or encapsulated (8). The tumor is firm on examination and may grow as large as 10 centimeters in diameter (9). Few tumors have reached the size that was attained in our patient.

A poor response to radiation therapy or chemotherapy is typical. Therefore, the primary treatment is surgical excision. Local recurrence occurs in 50% of cases (6, 9) and is associated with a poor prognosis. When metastases are present, they most commonly involve the liver, lungs, and skeletal system.

The plain film findings are nonspecific. Calcification within the tumor is infrequent (4, 10). Osseous changes related to hemangiopericytomas may be primary or secondary. CT findings are nonspecific. The tumor is typically a broad-based lesion attached to the dural surface. The tumors demonstrate variable attenuation (11) and homogeneous contrast enhancement on CT. When small, they resemble meningiomas. However, larger tumors can have thick, regular enhancing walls and nodular margins. The associated brain edema is generally mild. Distinction from a metastatic lesion, anaplastic meningioma, or a lymphoma can be difficult based on CT alone.

Hemangiopericytoma is a hypervascular tumor that typically has a prolonged heterogeneous tumor blush with angiography (4). Findings on angiography typical of this tumor include tiny, irregular feeding vessels, “fluffy” vascular stain, absence of early draining veins, and a prolonged circulation time through the tumor (12).

The MR characteristics in this case had signal isointense with brain parenchyma on T1-weighted images and slightly brighter than gray matter on proton density-weighted images. On T2-weighted images, the tumor was heterogeneous, with areas isointense to gray matter, and other areas with varying degrees of greater signal intensity. This is a reflection of the compact cellular and loose cellular components on pathologic examination. Diffuse and heterogeneous tumor enhancement occurred, following Gd/DTPA administration, secondary to the richly vascularized tumor with penetrating capillaries, permitting accumulation of the contrast in the expanded
Fig. 2. A, Coronal proton density (2600/20/2), and B, coronal T2-weighted (2600/80/3), show a multilobulated mass with destruction of the calvarium, compression of brain parenchyma without invasion, and vascular channels (arrows). Variable signal characteristics are noted primarily on the T2-weighted image corresponding to the variable cellular components (small arrows delineating compact, curved arrows loose components).

C and D, Sagittal T1-weighted (600/25/2) before and sagittal T1-weighted (800/26/1) after Gd/DTPA administration reveals marked enhancement of the mass.

Fig. 3. A, Lateral view following selective left external carotid injection shows hypertrophy of the superficial temporal (small arrow) and occipital arteries (large arrow). There is mild enlargement of the middle meningeal artery branches (arrowhead).

B, Lateral view in capillary phase. Marked tumor blush is present, as well as venous shunting (arrow).
extracellular space (13). Multiple regions of flow-void that were demonstrated on the MR examination reflect the large vascular channels present in this tumor.

References

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