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This information is current as of April 19, 2024.

AJNR Am J Neuroradiol 1991, 12 (4) 756-760
<http://www.ajnr.org/content/12/4/756.citation>

MR Findings in Extracerebral Cavernous Angiomas of the Middle Cranial Fossa: Report of Two Cases and Review of the Literature

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Extracerebral cavernous angioma is a rare entity; only 45 cases have been reported [1–32]. Thirty-seven of the angiomas, including the two cases reported here, were found in the parasellar region of the middle cranial fossa [1–26]. Differentiation from parasellar meningioma is known to be difficult, since the CT findings are similar. We report the MR findings in two cases of extracerebral cavernous angioma of the middle cranial fossa and review the literature.

Case Reports

Case 1

A 34-year-old woman had diplopia that worsened over a 1-year period. At a local hospital, the diagnosis of a left parasellar meningioma was made with CT and angiography. The tumor was partially removed and the histologic diagnosis was cavernous angioma. One year after surgery, the patient was referred to our hospital for a second operation. Neurologic examination on admission revealed complete palsy of the left abducens nerve and facial hypoesthesia in the region of the ophthalmic and infraorbital nerves.

CT of the brain demonstrated a well-defined, minimally hyperdense mass, 3 cm in diameter, in the left parasellar region (Fig. 1A). After injection of contrast material, marked enhancement with minimal central hypodensity was seen (Fig. 1B). Angiography was normal. MR imaging was performed on a 1.5-T superconducting magnet. The left parasellar tumor was isointense relative to gray matter on T1-weighted images, 450/15/4 (TR/TE/excitations) (Fig. 1C). The tumor filled the left cavernous sinus, laterally displacing the anterior temporal lobe. On T2-weighted images, 2500/90/1, the tumor was homogeneously bright, similar to CSF (Fig. 1D). After administration of gadopentetate dimeglumine (0.1 mmol/kg), the tumor showed marked homogeneous enhancement (Fig. 1E). At surgery the tumor was found to be located within the cavernous sinus and appeared reddish and angiomatous. It was successfully removed, although there was profuse bleeding during the procedure. The pathologic specimen demonstrated a structure composed of entwined vascular spaces of various sizes, lined with a single layer of endothelial cells separated by loose connective tissues. The findings were consistent with cavernous angioma.

Case 2

A 65-year-old man had slowly progressive diminution in left visual acuity over a 15-year period. No other neurologic signs referable to cranial nerve involvement were present.

CT demonstrated a high-attenuation mass in the left parasellar region. Prominent homogeneous enhancement was seen after contrast administration. Erosion of the left half of the dorsum sellae was noted. Cerebral angiography demonstrated medial displacement and stretching of the cavernous portion of the left internal carotid artery, but the tumor appeared avascular (Fig. 2A). The tumor was isointense relative to gray matter on T1-weighted MR images (Fig. 2B) and hyperintense relative to brain on T2-weighted images (Fig. 2C). Marked homogeneous enhancement was seen after administration of contrast material (Figs. 2D and 2E).

The preoperative diagnosis was parasellar meningioma. At surgery the cavernous sinus was filled by a hyperemic tumor, which was successfully removed. Histologic examination revealed blood-filled sinusoidal vascular spaces lined with a single layer of endothelium. The diagnosis was cavernous angioma.

Discussion

Intracerebral cavernous angioma has been considered a common disease ever since CT has been available to detect it; it represents approximately one fourth of cerebral vascular malformations [33]. However, intracranial extracerebral cavernous angioma is a rare entity most often located in the middle cranial fossa [1–26]. Its origin is unknown, but invariably it is closely associated with the dura mater of the cavernous sinus. In many cases, the tumor capsule is identical to the dura mater of the base of the middle cranial fossa and is speculated to arise from the dural tissue [21]. Kamijo et al. [7] reported a cavernous angioma with its own capsule that was located between the meningeal and endosteal layers of the dura mater of the cavernous sinus. To our knowledge, 35 extracerebral cavernous angiomas of the middle cranial fossa

Received December 10, 1990; revision requested January 8, 1991; revision received February 21, 1991; accepted February 25, 1991.

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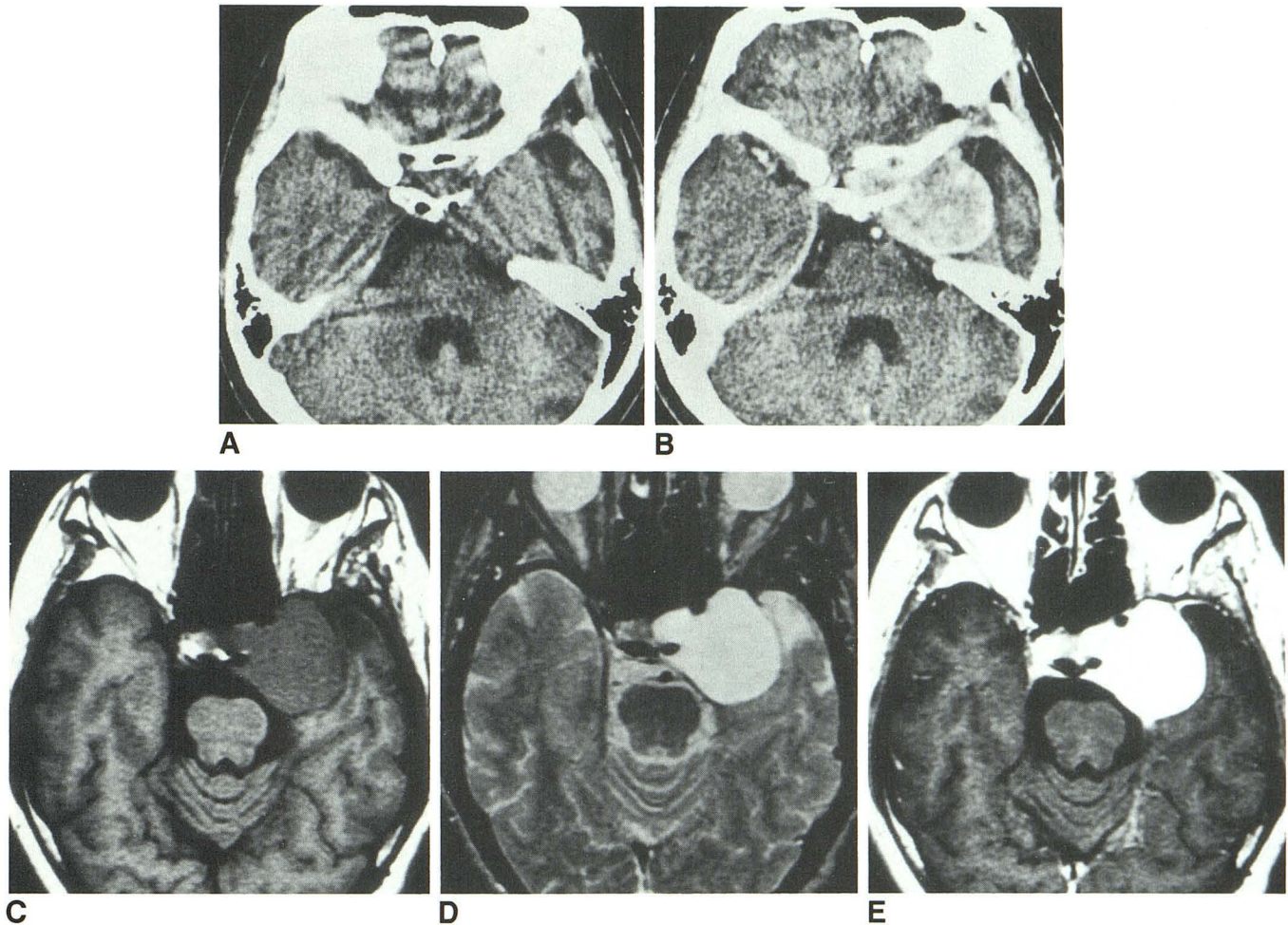


Fig. 1.—Case 1: 34-year-old woman.

A, Unenhanced CT scan. Minimally hyperdense mass, 3 cm in diameter, is in left parasellar region, although it is not well defined owing to linear artifacts. There is evidence of previous craniotomy at left anterior temporal bone.

B, Enhanced CT scan. Tumor is densely enhanced with minimal relative central hypointensity.

C, Unenhanced T1-weighted MR image (450/15/4). Left parasellar tumor is well demarcated and isointense relative to gray matter, displacing anterior part of temporal lobe laterally. Medial part of tumor partially extends into sella.

D, T2-weighted image (2500/90/1). Tumor is homogeneously hyperintense relative to brain parenchyma. Bright signal at tip of left temporal lobe represents old contusion caused by previous operation.

E, Axial enhanced T1-weighted image (450/15) shows marked homogeneous tumor enhancement.

have been reported in addition to the two cases reported here. Other rare locations for this tumor include the tentorium cerebelli [23, 27–31] and the dura mater of the cerebral convexity [32].

In contrast to its intracerebral counterpart, extracerebral cavernous angioma occurs predominantly in middle-aged women. Among 37 cavernous angiomas reported to involve the middle cranial fossa, 86% (32/37) were in women, 72% (23/32) of whom were 30–59 years old. Another interesting aspect of this tumor is that 68% (25/37) of reported cases were in Japanese people. The most common initial symptoms of extracerebral cavernous angioma of the middle cranial fossa are ocular, including diplopia, impaired visual acuity, and visual field defects [33]. Facial hypoesthesia, facial weakness, and exophthalmos have been reported also [21, 33].

Although the neurologic symptoms usually are insidious, acute visual disturbance [20] and acute onset of severe headache suggestive of subarachnoid hemorrhage have been reported also [10, 21]. Exacerbation of symptoms during pregnancy, followed by disappearance after delivery, is known [6, 10, 23], and has also been documented in parasellar meningioma [34].

CT findings of extracerebral cavernous angiomas of the middle cranial fossa have been described in 21 cases [11–26]. CT demonstrates a well-circumscribed hyperdense mass with homogeneous dense contrast enhancement. On angiography, cavernous angiomas of the middle cranial fossa are avascular or show faint to moderate blush, but dilated feeding arteries, the meningo-hypophyseal trunk, and the middle meningeal artery are visualized rarely [1–26]. Numaguchi et al.

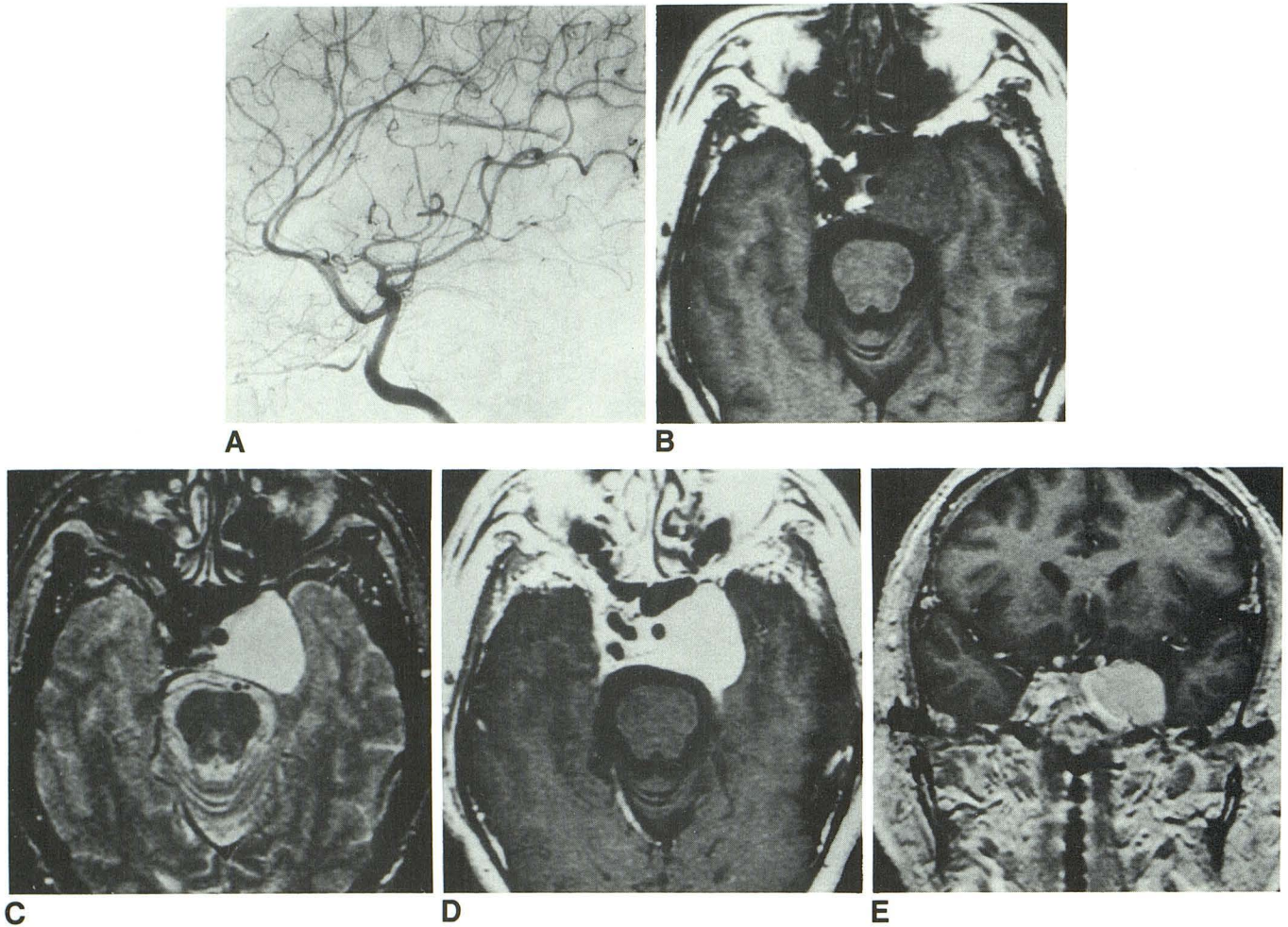


Fig. 2.—Case 2: 65-year-old man.

A, Left internal carotid angiogram. Distal part of internal carotid artery is stretched. Tumor appears avascular.

B, Unenhanced T1-weighted MR image (450/15). Tumor is well demarcated and isointense relative to gray matter.

C, T2-weighted image (2500/90). Tumor appears homogeneously hyperintense relative to brain.

D and E, Enhanced T1-weighted axial (450/15) and coronal SPGR (45/5/4, 30° flip angle) images. Tumor is homogeneously enhanced. Left internal carotid artery is displaced medially.

[13] reported that prolonged injection angiography could facilitate the diagnosis of cavernous angiomas, but it is not pathognomonic.

MR findings in cavernous angiomas of the middle cranial fossa have been reported in three cases [24–26], of which the earliest two reports did not contain detailed radiologic descriptions. Pozzati et al. [24] reported a case of cavernous angioma of the middle cranial fossa eroding the tegmen tympani and described MR findings of hyperintensity on T2-weighted images. Voci et al. [25] reported three cases of extracerebral cavernous angiomas, and MR findings were available in one. In their report, a parasellar cavernous angioma appeared to be hyperintense on T2-weighted images; no information was provided about T1-weighted images. Most recently, Kaard et al. [26] reported detailed MR findings of a parasellar cavernous angioma. The tumor was of homogeneous hypointensity on T1-weighted images and of a hyperintensity similar to that of CSF with some hypointense foci on

T2-weighted images. There was no description of an enhanced study.

In our two cases, both angiomas were isointense relative to gray matter on T1-weighted images and showed marked homogeneous contrast enhancement. On T2-weighted images, the tumors appeared homogeneously hyperintense. These findings are consistent with the previous reports on MR findings of extracerebral cavernous angiomas.

It is difficult to differentiate cavernous angioma of the middle cranial fossa from parasellar meningioma, since their CT and angiographic presentations as well as clinical pictures are similar. Among 15 cases for which preoperative diagnoses were available [2, 5, 7, 10, 11, 13, 14, 16–19, 21], only five were diagnosed correctly [2, 11, 14, 17, 19], and the remaining 10 cases were misdiagnosed as meningiomas. The preoperative diagnoses in our two cases were also parasellar meningioma. Meanwhile, correct preoperative differentiation between cavernous angioma and meningioma could be of

great importance for surgical planning; extracerebral cavernous angiomas have a strong tendency to bleed profusely during surgery, and some deaths due to unmanageable bleeding have been reported [1, 4, 7, 9, 16, 21, 26].

It is well known that meningiomas appear to be less intense generally than gliomas and other brain neoplasms, and they are often isointense or hypointense relative to brain on both T1- and T2-weighted images [35, 36]. Elster et al. [37] analyzed the signal intensity of 40 histologically proved meningiomas and showed that fibroblastic and transitional subtypes were isointense or hypointense relative to brain, while meningiomas composed predominantly of syncytial or angioblastic elements could be markedly hyperintense on T2-weighted images [37]. Therefore, it might still be difficult, from CT and MR findings alone, to differentiate cavernous angioma of the middle cranial fossa from parasellar meningioma of a syncytial or angioblastic subtype. When the distinction is unclear, angiographic findings would be helpful; angioblastic meningioma tends to show irregular hypervascularity and often is associated with arteriovenous shunting [38], while cavernous angioma usually shows no or only mild abnormalities and feeding arteries are rarely visualized. Other morphologic features such as shape or location of the lesion appear to be of no help in differentiating cavernous angioma from parasellar meningioma. As to plain radiographic findings in the skull base, the presence of hyperostosis or blistering suggests meningioma, but erosive, destructive change is common in both tumors.

Other parasellar tumors to be differentiated from cavernous angiomas include trigeminal schwannomas and epidermoid tumors. Approximately 50% of trigeminal schwannomas arise as extradural middle fossa masses, and their MR findings are similar to those of cavernous angiomas of the middle cranial fossa in terms of signal intensity; they exhibit homogeneous bright signal on T2-weighted images and show marked contrast enhancement [39, 40]. However, schwannomas often have areas of decreased attenuation on enhanced CT owing to cystic degeneration, and similar findings are expected on MR. Their location and configuration could also be of help for differentiation; schwannomas are centered more posteriorly than parasellar hemangiomas are and could extend into the posterior fossa or down to the foramen ovale. Demonstration of an intact Meckel cave would be helpful to argue against trigeminal schwannoma, but it was not identified in either of our two cases because of compression by the hemangiomas. Initial symptoms also provide an important clue for differential diagnosis; trigeminal schwannomas most often manifest with facial hypoesthesia or pain and are accompanied only rarely by ocular symptoms [40]. As for epidermoid tumor, it also has low signal intensity on T1-weighted images and high intensity on T2-weighted images, but it usually appears less homogeneous on both T1- and T2-weighted images and often has lobulated margins conforming to the cisternal space with minimal mass effect. It is easily differentiated from other tumors because of its lack of significant contrast enhancement on CT and MR [39, 41].

As for extracerebral intracranial cavernous angiomas in a location other than the middle cranial fossa, two reports on

MR findings are available. Matsumoto et al. [30] described a tentorial cavernous angioma with a dumbbell configuration that was isointense relative to brain parenchyma and had a hypointense rim on T1-weighted images and that was hyperintense on T2-weighted images. Quattrocchi et al. [31] documented MR findings of cavernous angioma in the posterior fossa arising from the tentorium cerebelli. The tumor was hypointense on T1-weighted images and hyperintense on T2-weighted images, and showed marked homogeneous enhancement after contrast administration. These MR findings are nearly identical to those of cavernous angiomas of the middle cranial fossa in terms of signal intensity. Furthermore, similar findings have also been reported in cavernous angiomas of the liver. It is well established that hepatic cavernous angiomas show marked hyperintensity relative to liver parenchyma on T2-weighted images and that this finding is of use for differentiating cavernous angioma from hepatocellular carcinoma [42, 43]. Soft-tissue angiomas also have been reported to show similar bright signal on T2-weighted images [44]. Salient prolongation of the T2 relaxation time seems to be a common denominator in most cavernous angiomas, probably reflecting the signal from pooling blood with very slow flow in the vascular channels of the lesion. Intracerebral cavernous angiomas, however, have MR appearances dramatically different from those of extracerebral cavernous angiomas: inhomogeneous mixed signal intensity on both T1- and T2-weighted images, usually with peripheral hypointensity on T2-weighted images.

The difference in MR appearances between extra- and intracerebral cavernous angiomas is likely to be reflected in pathologic findings. Both extra- and intracerebral cavernous angiomas comprise serpiginous vascular lumens lined with a single layer of endothelium. In extracerebral cavernous angiomas, including our two cases, stromal connective tissues surrounding the vascular spaces are scant and loose, vascular lumens are spacious, and there is little hemosiderin and few calcium deposits [6, 10, 14, 21, 27]. In intracerebral cavernous angiomas, the luminal walls are circumscribed by prominently thickened fibrous tissue and gliosis, the vascular lumens are narrow and often occupied by thrombi in different stages of organization, and hemosiderin and calcium deposits are frequently found in the stroma [45]. These findings, likely resulting from vascular accidents in angioma, probably cause inhomogeneity on MR appearances. The reason why intracerebral cavernous angiomas are complicated more often than their extracerebral counterparts is uncertain.

In summary, bright signal on T2-weighted images in an extraaxial middle cranial fossa mass, in conjunction with characteristic CT and angiographic findings, should suggest the diagnosis of cavernous angioma.

ACKNOWLEDGMENT

We thank M. Mukai, Department of Pathology, Keio University Hospital, for thorough review of the pathologic specimens and useful advice.

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