Extraaxial Parasellar Cavernous Hemangioma

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Intracranial cavernous hemangiomas (cavernomas) are uncommon vascular hamartomas that develop infrequently in an extracerebral location, with the middle cranial fossa being the most frequently reported site [1, 2]. Accurate presurgical diagnosis is not often achieved, and because of their location and propensity to bleed profusely during surgery, these tumors are usually impossible to resect completely.

We present a case of intracranial extracerebral cavernous hemangioma that illustrates the difficulty of differentiating this tumor diagnostically from other parasellar masses, in particular from meningioma. The MR, CT, and angiographic features of this lesion are described.

Case Report

A 56-year-old woman was referred with a 7-month history of progressively worsening right temporooccipital headaches (present for many years) and visual deterioration.

Neurologic assessment showed a bitemporal visual field defect, bilateral temporal optic disk pallor, and visual acuity of 6/30 and 6/12 in the right and left eyes, respectively. On general examination the patient was noted to be obese. Her serum prolactin level was 1895 mU/l (normal = 650 mU/l) and the serum follicle-stimulating hormone and luteinizing hormone levels were slightly reduced. The patient was euthyroid, and the adrenocortical trophic hormone stimulation test was normal.

Craniotomy demonstrated a dumbbell-shaped mass involving the sella turcica with extension into the suprasellar region and the left middle cranial fossa, with marked uniform contrast enhancement (Fig. 1). Minor erosion of the left petrous apex was noted.

On selective cerebral arteriography (left internal, left external carotid, left vertebral, and right common carotid) the mass was hypovascular with arterial supply from cavernous and hypophyseal branches of the left internal carotid artery and the left middle meningeal artery. Several small contrast pools within the mass were identified, and a faint, diffuse blush in the late cavernous phase of the left external carotid artery injection was shown (Fig. 2).

Sagittal and coronal T1-weighted, 600/20/2 (TR/TE/excitations), and axial T2-weighted, 2310/100/1. MR studies of the cranium were obtained on a 1.5-T unit. Slice thickness of 5 mm with a field of view of 200 mm was used. Matrix size was 256 × 256. The T2-weighted images were cardiac-gated and flow-compensated. On the T1-weighted sequence the tumor, of low uniform signal intensity, occupied the sella with a large lobulated parasellar extension indenting the lamina terminalis and the basal ganglia. The tumor further extended into the left middle cranial fossa (Fig. 3). On the T2-weighted sequence the tumor demonstrated increased signal intensity on both the first and second echoes, with the intensity being somewhat similar to CSF. Several small hypointense foci were present within the central portion of the mass (Fig. 4). No separate pituitary gland was identifiable.

Craniotomy revealed a bluish, vascular, encapsulated extradural tumor that bled profusely on biopsy and proved impossible to excise. Frozen-section biopsy confirmed a vascular tumor. Hemostasis was achieved and the skull closed. A left ptosis and complete oculomotor nerve palsy developed postoperatively. Transfusion of two units of packed cells was required. Seven days postoperatively the patient collapsed and died from a massive pulmonary embolus, confirmed at autopsy.

Postmortem examination revealed a trilobar, well-circumscribed, encapsulated, dark red-purple tumor measuring approximately 6 × 4 cm (Fig. 5). The tumor appeared to be arising from the left cavernous sinus and extended above and into the pituitary fossa, compressing the gland, and into the middle cranial fossa.

The optic nerves stretched over the top of the tumor and the adjacent vascular structures were displaced. Histologic sections demonstrated the tumor to be composed of numerous cavernous blood-filled and endothelial-lined spaces. These were separated by an edematous fibrous connective tissue stroma, containing occasional hemosiderin-laden macrophages. The lining endothelial cells showed no atypia. The final diagnosis was of cavernous hemangioma.

Discussion

Cavernomas are vascular hamartomas that uncommonly affect the CNS; they occur more commonly in other organ systems, such as the gastrointestinal tract, bones, and skin [3].

CNS cavernomas typically occur in the cerebral hemispheres but have been reported in the basal ganglia, ventricles, and spinal cord. Rarely are they extracerebral in location [1, 2]. Cavernomas represent one of the major categories of cerebrovascular malformations, consisting of thin-walled ectatic blood vessels of varying dimensions with an intervening stroma of fibrous connective tissue [1, 3]. Their natural history

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is one of slow growth (over many years); hemorrhage may occur into intracerebral cavernomas [3].

Before 1989, only 41 cases of intracranial extracerebral cavernomas involving the middle cranial fossa were reported in the world literature [2, 4], with the majority of these reported in the Japanese literature.

Intracerebral cavernomas are most often located supratentorially; in 16–33% of patients they are subcortical and multiple [5]. They are often discovered to be a cause of epilepsy and are found during investigation of focal seizures.

Extracerebral cavernomas of the middle fossa have a different clinical presentation, radiologic appearance, and surgical outlook than do intracerebral cavernomas [1]. As in this case, their clinical history is most often one of insidious onset of symptoms referable to compression of adjacent structures. Visual symptoms such as visual field defects, loss of motor activity, diplopia, exophthalmos, and optic atrophy are common. Other symptoms include headache, facial sensory defects, obesity, and amenorrhea [2, 6]. Presentation is usually after the third decade, and there is a female preponderance [6]. Neuroradiologic findings include erosion of the sella turcica and base of the middle fossa on skull examination.

Calcification is usually not detected, in contrast to intracerebral cavernomas [1, 2, 6]. Angiographic findings are typically of a hypovascular mass that may demonstrate pooling of contrast material and a tumor blush on the later venous phase. Identification of feeding vessels from both internal and extracranial (middle meningeal artery) carotid branches has been reported [1, 2]. In our case, small arteries supplying the lesion from the left internal carotid artery and the left middle meningeal artery were identified. Pools of contrast medium were seen in the mass and a faint diffuse blush on the late venous phase. CT appearance is of an isodense or hypodense lesion with homogeneous, marked enhancement following contrast administration. The CT enhancement pattern and angiographic findings are often indistinguishable from those of meningiomas [2]; however, in our opinion a hypodense lesion on the nonenhanced CT scans may be a useful differentiating feature from meningioma.

Despite a hypovascular appearance an angiography (caused by the small caliber of internal vessels, slow circulation, and thrombosis) [3], cavernomas bleed profusely during surgery. Total resection is usually not possible, although partial resection and rare cases of successful resection have
been reported [3]. Presurgical radiotherapy may be of benefit [1]. To our knowledge only one other study describing the MR appearance of an extracerebral cavernoma has been reported [4], and the features described there are in accord with our own findings. Intracerebral cavernomas are described as having a surrounding hypointense rim, in particular on T2-weighted sequences, caused by the presence of hemosiderin. Internal inhomogeneity, when present, is due to hemorrhage resulting from hematomas in evolution. Foci of low signal intensity caused by calcification or hemosiderin deposits have been reported in intracerebral lesions [4, 5]. In our case of an extracerebral cavernoma the appearance was of an almost uniformly high intensity on the T2-weighted sequence. We believe that the lack of a surrounding rim of low signal intensity, one of the hallmark features of an intraaxial cavernous hemangioma, can be explained by the absence of hemosiderin-laden macrophages around the lesion, which characteristically are found surrounding the intraaxial variety. Scattered hypointense foci distributed centrally in the larger portion of the mass are most likely due to deposits of hemosiderin. Less likely is the possibility that this represents a signal void due to blood flow within the tumor [7] or foci of calcification (not detected on histologic sections). These appearances are not typical of a meningioma or a pituitary tumor [8].

Cavernomas of the middle cranial fossa should be considered in the differential diagnosis of parasellar lesions, in particular when considering the possibility of meningioma. Surgical resection is often impossible owing to the vascular nature of these tumors and their tendency to bleed profusely.

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REFERENCES