Extra-axial cavernous hemangiomas involving the dural sinuses

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Extra-axial cavernous hemangiomas are rare lesions previously associated with unacceptable mortality and morbidity rates that precluded surgical resection. The authors analyze the clinical presentation, surgical results, and histology of eight intrasinus cavernous hemangiomas: six located in the cavernous sinus, one in the petrosal sinus, and one in the torcula. Magnetic resonance imaging is the best radiographic test for surgical planning. Successful tumor removal was achieved in six cases with no mortality and low morbidity. In the remaining two patients, only subtotal resection was achieved because of massive hemorrhage in one and the misdiagnosis of a pituitary adenoma leading to a transsphenoidal approach in the other. For hemangiomas arising within the cavernous sinus, extradural removal of the sphenoid bone facilitated preservation of the neurovascular structures. Since the clinical and histological characteristics of these lesions are distinct from intra-axial cavernous hemangiomas, a more appropriate term may be “sinus cavernoma” to indicate that these lesions are primarily intrasinus in origin.

KEY WORDS • cavernous hemangioma • dural sinus • cavernoma • vascular tumor

Extra-axial cavernous hemangiomas are rare lesions that usually present with mass effect, including headache and cranial nerve deficits. They are typically misdiagnosed preoperatively as meningiomas and, during surgery, massive hemorrhage can occur. In fact, successful tumor removal has rarely been reported. In contrast, intra-axial cavernous hemangiomas often present with multiple episodes of small parenchymal hemorrhages and seizures. Surgical resection of these lesions is typically performed with low morbidity and mortality rates. Therefore, despite similar pathological diagnosis, intra- and extra-axial cavernous hemangiomas are in fact distinct clinical entities.

There are approximately 22 published surgical reports of attempted resection of extra-axial cavernous hemangiomas, with a reported perioperative mortality rate of 38%. The small number of reported cases has prompted this analysis of our experience with eight extra-axial cavernous hemangiomas. The clinical characteristics of these lesions are defined, and an operative approach to expose the intracavernous neurovascular structures facilitating resection is advocated. Furthermore, it is hypothesized that extra-axial cavernous hemangioma may represent a primary vascular sinus lesion and that “sinus cavernoma” may be a more appropriate term to distinguish it from intra-axial cavernous hemangiomas.

Summary of Cases

Clinical Material

Between the years 1975 and 1989, eight patients underwent resection of extra-axial cavernous hemangiomas at the Mayo Clinic. Their clinical presentation is summarized in Table 1. The majority of clinical symptoms and signs were referable to the cranial nerves, which were either encased or compressed by the mass (Table 2). The preoperative diagnosis included menin-
TABLE 1
Clinical data in eight cases of cavernous hemangioma

<table>
<thead>
<tr>
<th>Factor</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>sex (M:F)</td>
<td>4:4</td>
</tr>
<tr>
<td>age range (yrs)</td>
<td>25-64</td>
</tr>
<tr>
<td>presenting symptoms</td>
<td></td>
</tr>
<tr>
<td>headache</td>
<td>5</td>
</tr>
<tr>
<td>diplopia</td>
<td>4</td>
</tr>
<tr>
<td>facial numbness</td>
<td>3</td>
</tr>
<tr>
<td>ptosis</td>
<td>2</td>
</tr>
<tr>
<td>symptoms associated with pregnancy</td>
<td>2</td>
</tr>
<tr>
<td>extremity weakness</td>
<td>1</td>
</tr>
<tr>
<td>tinnitus</td>
<td>1</td>
</tr>
</tbody>
</table>

TABLE 2
Examination results in eight cases of cavernous hemangioma

<table>
<thead>
<tr>
<th>Deficit</th>
<th>Preop Findings</th>
<th>Follow-Up Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>cranial nerve deficits</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2nd nerve</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>3rd nerve</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>4th nerve</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>5th nerve</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>6th nerve</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>7th nerve</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>8th nerve</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>monoparesis</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>galactorrhea</td>
<td>1</td>
<td>0</td>
</tr>
</tbody>
</table>

The eight surgical specimens were formalin-fixed and paraffin-embedded. Five-micron sections were stained by hematoxylin and eosin as well as for collagen (Masson’s trichrome), elastic tissue (Verhoeff’s elastic van Gieson), axonal processes (Bielschowsky), and iron (Prussian blue).

Surgical Results

Complete removal was achieved in six of the eight cases. In the two patients in whom only subtotal removal of tumor from the cavernous sinus was accomplished, the limiting factors were the misdiagnosis of a pituitary adenoma leading to performance of a transsphenoidal approach in one and a massive intraoperative hemorrhage in the second. The one posterior fossa cavernous hemangioma originating from the petrosal sinus was removed without difficulty. The torcular lesion was completely removed but repair of the sagittal sinus with a dural graft was required. Because of the vascular nature of these malformations, intraoperative blood loss was always a significant factor, requiring two to eight units of blood for hemodynamic stability in each patient.

In six cases there were no new neurological deficits and at last examination significant recovery of preoperative deficits had occurred (Table 2). One patient experienced a new postoperative ophthalmoplegia which has slowly resolved and another patient had increased numbness in the fifth nerve distribution. In one of the two patients with subtotal tumor resection, postoperative radiation therapy was administered (4000 rad) which caused a significant reduction in lesion size by the 1-year follow-up examination.

Analysis of the radiographic workup suggested that magnetic resonance (MR) imaging was most useful in demonstrating the intracavernous aspect of the tumor, thereby assisting in surgical planning. However, MR images could not differentiate between intracavernous tumors extending outward and extracavernous tumors, such as meningiomas, extending into the sinus. Angiography demonstrated encasement of the carotid artery with late capillary staining along with meningohypophyseal feeding arteries.

Histological Findings

Five specimens were generous in size to permit as-
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Assessment of gross anatomical relationships (Fig. 1). All of the lesions were cavernous hemangiomas. Microscopically, dural attachment was clearly visible in four specimens; the membrane appeared to represent a portion of the “capsule.” Each lesion consisted of endothelial-lined vascular spaces varying from capillary to cavernous in dimension. There was also well-formed vasculature: presumably, feeding arteries and draining veins (Fig. 2). In contrast to these vessels, the substance of the angioma consisted of cavernous vasculature devoid of musculature and possessing only a small quantity of poorly organized elastic tissue. Collagen made up the bulk of the vessel walls and was particularly prominent in five instances. Thrombosis with features of organization was apparent in five specimens; hemosiderin deposition was absent in all but two cases, in which it was scant. Neither cholesterol deposits nor calcification were noted. In the larger specimens, some large myelinated nerves were found lying adjacent to the dura. No neural network was noted deep within the substance of the hemangioma to suggest innervation of its vasculature. No neural parenchyma was seen in any case.

Surgical Considerations

In each patient, the hemangioma appeared to arise within the vascular sinus. The large size of the majority of these lesions caused expansion of the sinus with the dura forming the outer tumor “capsule.” The most difficult cavernous hemangiomas to remove were those within the cavernous sinus, primarily due to their extreme vascularity and the encasement of neurovascular structures. Specifically, they consisted of vascular channels of varying caliber which had insinuated themselves around the carotid artery and cranial nerves. After a pterional craniotomy, the lesser sphenoid wing (including the anterior clinoid) was removed extradurally with a high-speed air drill. This exposed the dural reflections of the optic nerve and superior orbital fissure. The medial aspect of the greater sphenoid wing was also removed to expose both the carotid artery and the second division of the trigeminal nerve. The dura was then incised in a cruciate fashion and the sylvian fissure opened widely with coagulation of the temporal tip draining veins. Withdrawal of cerebrospinal fluid (CSF) through a lumbar needle also aided retraction. The dissection first proceeded by incision of the dura over-
lying the superior orbital fissure, with identification of the cranial nerves. This incision was then extended posteriorly over the roof of the cavernous sinus, including incision of the dural ring surrounding the carotid artery.

Piecemeal tumor removal was then used to identify the extent of involvement of the carotid artery and cranial nerves, using their relationships in both the superior orbital fissure and subarachnoid space to provide consistent orientation. As evidenced by the need for intraoperative blood transfusions, the most difficult aspect of tumor removal was hemostasis. The source of hemorrhage was the venous cavernous sinus and tumor arterial feeders. In well-oxygenated patients, distinguishing between these two sources was often difficult; however, early coagulation of the meningohypophyseal trunk facilitated hemostasis. Avitene pledgets were most useful in controlling venous sinus bleeding.

Illustrative Case

This 35-year-old right-handed woman presented with a 2-year history of headaches. Her neurological examination had been found normal on several occasions. Due to her persistent pain, contrast-enhanced computerized tomography was performed which demonstrated a large middle fossa tumor suggestive of a medial sphenoid wing meningioma (Fig. 3). Follow-up MR imaging revealed that the tumor was extending from the cavernous sinus outward into the middle fossa (Fig. 4). The patient was then referred to our institution. To better define the anatomical pathology, a transfemoral cerebral angiogram was performed which demonstrated that the main blood supply was from both the meningohypophyseal trunk and the middle meningeal artery. In addition, the cavernous and suprachiasmatic portions of the carotid artery were encased by the tumor.

As described above, the sphenoid wing was removed extradurally through a pterional craniotomy, including exposure of the first division of the fifth cranial nerve. Dural relaxation was then achieved with removal of CSF from a lumbar spinal drain. The distal middle cerebral artery was identified in the sylvian fissure and followed proximal to the carotid bifurcation which was encased by tumor medially. Tumor was dissected off the anterior choroideal and posterior communicating arteries; the dural sheath was then dissected over the superior orbital fissure, and the incision was extended posteriorly along the roof of the cavernous sinus. The carotid artery was then followed until the meningohypophyseal trunk was identified and coagulated. Next, the third, fourth, and sixth cranial nerves were localized, first in the superior orbital fissure and then in relation to the carotid artery. During the piecemeal tumor removal with bipolar cautery, Avitene (microfibrillar collagen) pledgets were used to control bleeding from the venous channels within the tumor. Eventually, the neurovascular structures were skeletonized, facilitating dissection of the tumor off the tentorial edge. After resection, intraoperative Doppler sonography confirmed patency of the carotid artery.

The patient's postoperative course was unremarkable. Despite structural continuity of the cranial nerves, the patient had a complete ophthalmoplegia along with numbness in the first division of the trigeminal nerve. At her 6-month follow-up examination, the ophthalmoplegia was resolving.

Discussion

Surgical Considerations

Extra-axial cavernous hemangiomas are extremely rare lesions. Approximately 22 cases have been reported, with a 38% mortality rate and with only three complete resections. Because of this high mortality rate and rare complete removal, radiation therapy has been proposed as the primary treatment. In contrast, the present report suggests that extra-axial cavernous hemangiomas can be safely approached. Extradural removal of the sphenoid wing with early exposure of the neurovascular structures within the cavernous sinus makes tumor removal possible with low morbidity. Accordingly, radiation therapy should be reserved for adjunctive treatment after subtotal resection or as a primary treatment in the elderly or debilitated patient.

A preoperative determination that the tumor is indeed intracavernous is necessary for successful tumor removal; this finding would alter the surgical approach decision in favor of medial sphenoid wing removal. For the more common medial sphenoid wing meningioma,
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Figure 4. Magnetic resonance images demonstrating that the lesion extends from the cavernous sinus into the middle fossa. The carotid artery is encased in the cavernous sinus and in the supraclinoid region.

removal of this bone is unnecessary. Therefore, the best diagnostic test for surgical planning is coronal MR imaging. In addition, preoperative angiography with trial balloon occlusion of the internal carotid artery is helpful in determining the patient’s ability to tolerate either temporary or permanent carotid artery occlusion. In most cases, the ipsilateral part of the neck and lower extremity are included within the draping in case an interposition saphenous vein graft becomes necessary.

Removal of the sphenoid wing and anterior clinoid is best approached extradurally with a high-speed air drill and using microscopic magnification. Use of the diamond burr facilitates hemostasis and helps prevent inadvertent tearing of the underlying dura and neurovascular structures. After the sylvian fissure is opened, the dura overlying the cavernous sinus is incised over the superior orbital fissure, which allows early identification of the cranial nerves. This incision is then extended posteriorly along the roof of the cavernous sinus to include the dural ring which tethers the carotid artery. With tumors extending along the free edge of the tentorium, the temporal lobe must be retracted in an anterior-posterior direction. The temporal lobe will tolerate this direction of retraction better than medial-to-lateral retraction. After this maneuver, the third nerve can be identified and protected. The tumor is then removed piecemeal with the use of a bipolar cautery and Avitene pledgets to control sinus bleeding. Early identification and coagulation of the meningohypophyseal trunk are important for hemostasis. Good communication between the neuroanesthesiologist and neurosurgeon and strict attention to blood loss are essential.

Terminology

The natural history of extra-axial cavernous hemangiomas is unknown. Although it is termed “cavernous hemangioma,” this lesion is clearly distinct from the more typical intra-axial cavernous hemangioma. The latter is classified as a low-flow vascular malformation and is usually small, presenting with multiple parenchymal hemorrhages and seizures. There has been some confusion regarding these lesions since terminology including “cavernous hemangioma” and “cavernous angioma” has been used interchangeably. On pathological examination, they consist of small often thrombosed endothelium-lined channels. The cavernous hemangiomas described here are large, highly vascular intrasinus lesions. Although they occur more commonly in the cavernous sinus, they can originate from other dural sinuses, as evidenced by this report.

The designation “cavernous hemangioma” for this extra-axial lesion may be a misnomer. Although these masses are part of a spectrum of vascular malformations, they have characteristics suggestive of a neoplasm, including mass effect, encasement of neurovascular structures, growth often in pregnancy, and radiographic features suggestive of a tumor. The term “vascular malformation” includes arteriovenous malformations, venous and cavernous hemangiomas, and capillary telangiectases. Although venous and cavernous hemangiomas are characterized by their solid na-
ture (that is, their lack of intervening parenchyma), the cavernous lesion lacks both musculature and an organized elastic layer. Unlike extra-axial cavernous hemangiomas, parenchymal hemangiomas have been well characterized. They comprise about 10% of vascular lesions of the central nervous system and, presumably due to the paucity of an arterial component and a tendency to thrombose, are often avascular on angiography. Approximately 25% of symptomatic cases present with hemorrhage. Microscopic calcification is common, but massive dystrophic calcification as in the "hemangioma calcificans" of Penfield and Ward is rare.

In contrast to parenchymal cavernous hemangiomas, this study demonstrates that extra-axial hemangiomas lack evidence of previous hemorrhage, an important observation in that hemorrhage and resultant reactive changes have been suggested as a basis for the growth of angiomas. It is unlikely that this mechanism plays a significant role in extra-axial lesions, since there was no evidence of prior hemorrhage. Alternative mechanisms of growth include: 1) capillary outgrowth at the periphery of the lesion; 2) gradual ectasia of vascular spaces; or 3) thrombosis with organization and progressive sclerosis. This is based on the observation that these cases demonstrated the presence of a capillary component, transitional vasculature smaller in caliber than the fully developed cavernous spaces, and thrombosis with organization.

Conclusions

Cavernous hemangiomas arising from within a dural sinus are highly vascular tumors that are exceedingly difficult to remove. In particular, those which arise within the cavernous sinus require careful surgical planning to preserve the encased neurovascular structures. In this location, early identification of the proximal carotid artery within the cavernous sinus by removal of the lesser sphenoid wing facilitates removal. Since the clinical and histological characteristics of these lesions are unique, a more appropriate term may be "sinus cavernoma" to distinguish these lesions from the more common intra-axial cavernous hemangiomas and to indicate that they are primary intrasinus lesions.

References


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