Hemangioma calcificans

Case report

ROBERT E. HARBAUGH, M.D., DAVID W. ROBERTS, M.D., AND JONATHAN D. FRATKIN, M.D.

Section of Neurosurgery and Department of Pathology, Dartmouth-Hitchcock Medical Center, Hanover, New Hampshire

A 44-year-old woman presented with intraventricular hemorrhage and a calcified avascular mass lesion in the anterior third ventricle. This lesion proved to be a densely calcified variant of cavernous hemangioma, namely, hemangioma calcificans. Twelve previous cases of this lesion have been reported in the neurosurgical literature. Association with spontaneous intraventricular hemorrhage has not been previously reported. The clinical presentation, radiological appearance, surgical findings, and histopathology of this lesion are discussed and a brief review of the literature is presented.

KEY WORDS • cavernous hemangioma • hemangioma calcificans • intracranial calcification • intracranial hemorrhage

Hemangioma calcificans is the rarest variant of cavernous hemangioma (cavernoma).² Twelve cases have been reported previously in the neurosurgical literature,²⁻⁴,⁶ with only Penfield and Ward³ presenting more than a single case. Previous cases have been manifested clinically by focal neurological deficits and/or seizure disorders. We describe a case of hemangioma calcificans of the anterior third ventricle presenting as intraventricular hemorrhage in a 44-year-old woman. A review of the literature concerning the clinical presentation, radiological appearance, pathology, and surgical treatment of such lesions is presented.

Case Report

This 44-year-old right-handed woman noted the explosive onset of bitemporal headache and nausea on December 2, 1982. She subsequently developed neck stiffness, low-back and radicular leg pain, anorexia, and malaise. Symptoms persisted and on December 9, 1982, she was evaluated by her family physician. A lumbar puncture was performed revealing bloody xanthochromic cerebrospinal fluid under elevated pressure. She was transferred to our institution for further evaluation and therapy. Her medical history was unremarkable for neurological disease.

Examination. She was a thin middle-aged woman who complained of headache and neck stiffness. Her general physical examination was unremarkable except for marked meningismus. Funduscopy revealed no papilledema, hemorrhages, or venous pulsations. She was lethargic but fully oriented, and conversation was appropriate. Cranial nerve examinations were completely normal as were her sensory and motor examinations. Deep-tendon reflexes were normoactive and symmetrical, and no pathological reflexes could be elicited. There was no evidence of cerebellar dysfunction.

Computerized tomography (CT) with and without intravenous contrast enhancement revealed a densely calcified, lobulated, unenhancing mass lesion in the region of the anterior third ventricle and foramina of Monro. Mild ventriculomegaly and intraventricular hemorrhage was noted (Fig. 1). Pancerebral angiography demonstrated an avascular calcified mass lesion (Fig. 2).

Operation. Diphenylhydantoin and dexamethasone were started prophylactically, and on December 13, 1982, a bifrontal craniotomy with a transcaldosai approach to the left lateral ventricle was performed. Clot was evacuated from the left frontal horn and a well demarcated, multilobulated, purple-red mass was seen to protrude through the foramen of Monro. Hemosiderin staining was present around the mass, and a gliotic pseudocapsule made separation of tumor from surrounding brain relatively easy. No unusually large feeding or draining vessels were encountered. Portions of
FIG. 1. Transaxial (left) and coronal (right) computerized tomography scans showing a calcified anterior third ventricle mass. There is mild lateral ventricular enlargement and intraventricular blood.

R. E. Harbaugh, D. W. Roberts, and J. D. Fratkin

the mass were bone-hard and other areas had a rubbery consistency. The soft component appeared multicystic with blood and thrombus within the cysts. Gross total excision of tumor was achieved.

Postoperative Course. The patient awoke rapidly after surgery without discernible neurological deficit. The following morning she was transferred to the neurosurgical ward. About 4 hours later a rapid deterioration in level of consciousness occurred and repeat CT scan showed marked obstructive hydrocephalus. A subcutaneously tunneled external ventricular drain was placed, with immediate improvement in neurological status. External ventricular drainage was continued for 12 days. The drain was clamped and a CT scan 24 hours later showed no hydrocephalus. The drain was removed. Following her episode of acute hydrocephalus, the patient was noted to be confused and partially

TABLE 1

Summary of data in 13 cases of hemangioma calcificans*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Cases</th>
<th>Age (yrs), Sex</th>
<th>Location of Tumor</th>
<th>Presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Penfield &amp; Ward, 1948</td>
<td>5</td>
<td>21, M</td>
<td>rt temporal</td>
<td>seizures</td>
</tr>
<tr>
<td></td>
<td></td>
<td>63, F</td>
<td>rt temporal</td>
<td>seizures</td>
</tr>
<tr>
<td></td>
<td></td>
<td>21, M</td>
<td>rt temporal</td>
<td>seizures</td>
</tr>
<tr>
<td></td>
<td></td>
<td>31, M</td>
<td>rt temporal</td>
<td>seizures</td>
</tr>
<tr>
<td></td>
<td></td>
<td>34, M</td>
<td>lt temporal</td>
<td>seizures</td>
</tr>
<tr>
<td>Shafey, et al., 1966</td>
<td>1</td>
<td>16, F</td>
<td>lt occipital</td>
<td>seizures</td>
</tr>
<tr>
<td>Runnels, et al., 1969</td>
<td>1</td>
<td>30, M</td>
<td>rt frontal, temporal, parietal</td>
<td>seizures, lt hemiparesis</td>
</tr>
<tr>
<td>Segall, et al., 1974</td>
<td>1</td>
<td>29, M</td>
<td>rt frontal</td>
<td>seizures</td>
</tr>
<tr>
<td>DiTullio &amp; Stern, 1979</td>
<td>1</td>
<td>59, F</td>
<td>multiple: rt frontal, rt temporal, lt occipital, hypothalamus</td>
<td>seizures</td>
</tr>
<tr>
<td>Ramina, et al., 1980</td>
<td>1</td>
<td>45, F</td>
<td>rt temporal, parietal, occipital</td>
<td>seizures, lt hemiparesis, dementia</td>
</tr>
<tr>
<td>Sanchis Fargueta, et al., 1981</td>
<td>1</td>
<td>20, F</td>
<td>lt temporal</td>
<td>seizures</td>
</tr>
<tr>
<td>Occhiogrosso, et al., 1983</td>
<td>1</td>
<td>35, M</td>
<td>quadrigeminal plate</td>
<td>seizures, hydrocephalus</td>
</tr>
<tr>
<td>Harbaugh, et al., 1984</td>
<td>1</td>
<td>44, F</td>
<td>anterior third ventricle</td>
<td>intraventricular hemorrhage</td>
</tr>
</tbody>
</table>

FIG. 2. Arterial (left) and venous (right) phases of a lateral left carotid angiogram showing an avascular calcified mass in the anterior third ventricle (arrows).
Hemangioma calcificans

amnestic for recent events. These problems gradually resolved and she was discharged home on January 5, 1983, where she continued to do well.

On pathological examination, the surgical specimen consisted of a purple-red mass of hyalinized non-arterial vascular channels with areas of thrombosis and conspicuous calcification. The histopathological diagnosis was hemangioma calcificans (Fig. 3).

Discussion

Cavernous hemangiomas (cavernomas) have been extensively reviewed.6-10 They are commonly diagnosed in patients between the third and sixth decade, usually in males. The cerebrum is most often involved, in which case the lesions are usually subcortical and often perirolandic. Other favored sites include the pons and walls of the third ventricle. Multiple lesions are present in about 15% to 25% of cases.6,10

The rarest variant of cavernous hemangioma is characterized by conspicuous calcification and bone formation, and has been designated "hemangioma calcificans."6-3,7 The 13 reported cases of this lesion (including ours) are summarized in Table 1. As with the more common types of cavernoma, hemangioma calcificans is usually found in the cerebrum or walls of the third ventricle. The age at diagnosis is also similar to that associated with the more common types of cavernous hemangioma. No clear sexual preference is shown in the small number of reported cases. Unlike noncalcified cavernomas, hemangioma calcificans has not previously been reported with intracranial hemorrhage.

The proper management of this lesion has been debated in the literature. Voigt and Yaşargil10 suggested that readily accessible cavernous hemangiomas be resected because of the risk of intracranial hemorrhage. Penfield and Ward advocated surgical excision of hemangioma calcificans because of their epileptogenic properties and potential for growth. Four of the five patients reported by these authors underwent surgery for hemangioma calcificans with excellent results in seizure control. Their only unoperated case was that of a 63-year-old woman with a 30-year history of seizures, who died in status epilepticus. Our patient, like the other reported cases, has done well after total or subtotal excision of this lesion.1-5,7-9 DiTullio and Stern,4 however, proposed that hemangioma calcificans represented a more benign form of cavernoma without documented risk of hemorrhage. They recommended that surgical intervention be reserved for patients with medically intractable seizure disorders or documented growth of the lesion, or to resolve a diagnostic dilemma.

The case reported here demonstrates that hemangioma calcificans can present as spontaneous intracranial hemorrhage. Previous reports document the epileptogenic potential of such lesions and the favorable outcome with surgical excision. In addition, confident diagnosis of intracranial calcification as hemangioma calcificans is difficult without pathological confirmation. Based on these observations, we would advocate surgical exploration and excision in patients suspected of having this lesion.

References


Fig. 3. Photomicrograph of the tumor. It is composed of non-arterial vascular channels with areas of dense calcification and bone formation. H & E, x 125.