Hemangioblastoma presenting with intraparenchymatous hemorrhage

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The authors report six cases of hemangioblastoma presenting with apoplectic symptoms but with no history related to the tumor. In each case, computerized tomography disclosed an intraparenchymatous hemorrhage, which was located supratentorially in four and in the cerebellum in the remaining two. Angiography revealed an abnormal vascular blush in two cases, but no abnormal vessels or tumor blush in the other four. In all cases, a solid tumor with abnormal vessels, such as red veins and feeding arteries, was found within or adjacent to the hemorrhage at surgery. The possibility of hemangioblastoma should be kept in mind as a cause of intraparenchymatous hemorrhage, particularly subcortical. Evacuation of the hematoma should be carefully carried out, and the whole hematoma wall should be thoroughly investigated for abnormal vessels or a solid mass.

KEY WORDS • hemangioblastoma • supratentorial hemorrhage • cerebellum • apoplexy

Apoplectic presentation of a brain tumor due to hemorrhage is a well known phenomenon, however, it is uncommon in cases of hemangioblastoma, although asymptomatic minor bleeding is not unknown. In several major hemangioblastoma series, few patients developed clinically significant hemorrhage from the tumor, and case reports regarding this phenomenon are rare.

During the past 5 years, we have treated six patients who presented with apoplectic symptoms caused by hemorrhage from hemangioblastoma. Computerized tomography (CT) disclosed an intraparenchymatous hemorrhage in each case. These six cases are presented in this report, and the clinical significance of hemorrhage from this type of tumor is discussed.

Summary of Cases

Fourteen patients with lobar hematoma and two with cerebellar hematoma were operated on in our institute during the past 5 years. None of these patients showed abnormal vessels or tumor blush on angiography. Hemangioblastoma was found in four of these patients and angioma in nine. In the remaining three, no angioma or tumor was verified pathologically. The four patients with hemangioblastoma and no angiographic vascular blush, and an additional two patients with vascular blush on angiography, are the basis of the present study.

At admission, the patient's ages ranged from 2 to 74 years, with a mean age of 44 years. There were four males and two females (Table 1). Initial clinical manifestations were disturbance of consciousness, generalized convulsive seizures, nausea, vomiting, and focal neurological deficits. These symptoms developed suddenly in all patients. There were no medical histories related to the tumor. There was no familial history of erythrocytosis. No retinal angioma could be demonstrated at ophthalmoscopic examination in any case. Body CT scanning was carried out in all patients and disclosed multiple renal cysts in one (Case 5).

The hematoma was located in the left parieto-occipital region in four cases and in the cerebellum in the remaining two (Fig. 1). Computerized tomography with contrast infusion failed to reveal an enhanced mass within or near the hematoma in any case. An irregular area of enhancement was noted on the superior vermis in Case 1. An abnormal vascular blush with feeding arteries and draining veins was demonstrated on angiography in two cases (Cases 1 and 4, Fig. 2 left). In the other four, angiograms revealed no abnormal vessels or tumor blush, but mass signs were evident (Fig. 2 right).

At surgery, solid tumor associated with abnormal
Hemangioblastoma with intraparenchymatous hemorrhage

TABLE 1

Summary of six cases of hemangioblastoma presenting with hemorrhage

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Symptoms &amp; Signs</th>
<th>Site of Hematoma</th>
<th>Angiogram</th>
<th>Size of Nodule (cm)</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2, M</td>
<td>disturbance of consciousness, stridor, seizures</td>
<td>superior vermis</td>
<td>tumor blush</td>
<td>1 x 1 x 1</td>
<td>fair</td>
</tr>
<tr>
<td>2</td>
<td>49, M</td>
<td>dizziness, nausea, vomiting, astasia, hypermetria</td>
<td>lt cerebellar hemisphere</td>
<td>negative*</td>
<td>1 x 0.5 x 1</td>
<td>good</td>
</tr>
<tr>
<td>3</td>
<td>32, F</td>
<td>disturbance of consciousness, hemiparesis, seizures</td>
<td>lt parieto-occipital</td>
<td>negative*</td>
<td>not totally excised</td>
<td>good</td>
</tr>
<tr>
<td>4</td>
<td>35, M</td>
<td>headache, nausea, homonymous hemianopsia</td>
<td>lt parieto-occipital</td>
<td>tumor blush</td>
<td>1 x 1 x 1</td>
<td>good</td>
</tr>
<tr>
<td>5</td>
<td>74, M</td>
<td>Gerstman’s syndrome, hemiparesis</td>
<td>lt parieto-occipital</td>
<td>negative*</td>
<td>5 x 3 x 0.5</td>
<td>good</td>
</tr>
<tr>
<td>6</td>
<td>72, F</td>
<td>disturbance of consciousness, hemiparesis, hemihypalgesia, homonymous hemianopsia, dyscalculia</td>
<td>lt parieto-occipital</td>
<td>negative*</td>
<td>3 x 1 x 0.5</td>
<td>good</td>
</tr>
</tbody>
</table>

* No abnormal vessels or tumor blush were seen, but mass effect was visualized on angiography.

vessels (such as red veins and small or dilated feeding arteries) was identified within or adjacent to the hematoma in all cases. A number of tangled abnormal arteries and veins mimicking an arteriovenous malformation were noted over the cerebellar surface in Case 1. The solid tumor was removed totally in all patients but one (Case 3). The outcome of the operation was good in all patients but one (Case 1), who developed disturbance of consciousness, hemiparesis on the left side, and truncal ataxia after surgery. However, this young child recovered consciousness 1 month following the operation and is now able to walk without aid.

Surgical specimens were examined histologically using hematoxylin and eosin, elastica van Gieson, and reticulin stains. All six tumors were diagnosed as typical hemangioblastoma. According to Silver and Hennigar’s classification, five tumors were classified as transitional and the sixth as juvenile type (Fig. 3).

Illustrative Case

Case 2

This 48-year-old man suffered a sudden attack of dizziness, nausea and vomiting, and astasia on June 11, 1983. He was admitted to the neurosurgical ward 3 days later.

Examination. Neurological examination at admission revealed dysmetria of the extremities on the left side. The patient was alert but unable to stand. Nystagmus was noted on left lateral gaze. No choked disc was found. Blood pressure was 142/110 mm Hg, and heart rate was 72 beats/min. Otherwise, the neurological and physical examination was normal. The patient had been taking antihypertensive medication for a year until 1 month before admission. A CT scan demonstrated a moderate-sized hematoma in the left cerebellar hemisphere. There was no enhancement on infusion of contrast material (Fig. 1B). Vertebral angiography disclosed only mass effect, with no abnormal vessels or tumor blush (Fig. 2 right).

Operation. A suboccipital exploration was undertaken on June 27, 1983. The hematoma was evacuated
with the aid of an operative microscope. A small vascular nodule, 1 × 0.5 × 1 cm in size, was found on the inferomedial side of the hematoma wall. After removal of this mass, the fourth ventricle was opened. Other parts of the hematoma wall were smooth and yellowish in color.

**Postoperative Course.** The postoperative course was uneventful. Ten days after surgery, the patient's cerebellar ataxia had resolved, and he was able to walk.

**Discussion**

Spontaneous lobar or cerebellar hematoma may be caused by several underlying diseases such as hypertension, angioma, aneurysm, tumor, or blood dyscrasias. However, hemangioblastoma has rarely been documented as a cause of these types of hematoma. In the present series, hemangioblastoma was not expected preoperatively to be the cause of hemorrhage, and the CT scans failed to reveal any enhanced mass associated with the hematoma. Nevertheless, careful search during surgery revealed a vascular tumor with abnormal vessels within the hematoma wall in all six cases. The possibility of hemangioblastoma should be kept in mind as a potential cause of spontaneous lobar or cerebellar hematoma, even in cases with normal angiography and no enhancing mass visible on CT scanning.

Among our six cases, no abnormal vessels or tumor blush was seen on the angiograms in four. This comprises 25% of the total series of 16 operated cases with spontaneous lobar or cerebellar hematoma with normal angiograms. In only three of the 16 cases was the cause of hemorrhage not verified. Thus, the discovery rate of the underlying pathology was about 81%, which is greater than that of other series. This high rate might be related to our microsurgical technique, or to a bias of the patient population in our institute.

The reason why the feeding and draining vessels and the tumor blush were not visualized on the angiograms, although they were found at surgery, in four of our cases may be that the tumor was partially destroyed by the sudden hemorrhage, and the hematoma compressed the contributing vessels. Similar mechanisms have been proposed for spontaneous regression of cerebral arteriovenous malformations and angiographically occult angiomas. However, angiography several months after hemorrhage may disclose vascular pathology not seen on the initial study.

Pathologically, five tumors were diagnosed as transitional and the remaining lesion as a juvenile type, characterized by the presence of thin-walled, tightly packed capillaries and a varying number of dilated vessels. In the five transitional tumors, the same pattern was found in approximately half of each specimen (Fig. 3). These vascular structures seem very likely to be related to intratumoral hemorrhage.

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**References**

Hemangioblastoma with intraparenchymatous hemorrhage

FIG. 3. Photomicrographs of the operative specimens. Reticulin stain, × 85. A: Case 1. Thin-walled capillaries and dilated vessels are tightly packed (juvenile type). B to F: Cases 2 to 6, respectively. Thin-walled capillaries and dilated vessels are intermingled with stromal cells (transitional type).
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