Intraventricular cavernous hemangioma

Case report

HIDEAKI IWASA, M.D., ITSUO INDEI, M.D., AND FUMIAKI SATO, M.D., PH.D.

Department of Surgical Neurology, Jichi Medical School, Minamikawachi-Machi, Tochigi-ken, Japan

A case of intraventricular cavernous hemangioma in a neonate is reported. In over 200 previously reported cases of intracranial cavernous hemangioma, 11 histologically verified cases have been collected and analyzed. The present case is the second occurring in a neonate.

KEY WORDS • cavernous hemangioma • intraventricular tumor • ventricle • neonate • hydrocephalus

To our knowledge, there has been only one previous histologically confirmed report of an intraventricular cavernous hemangioma in a newborn baby. Voigt and Yasarigil reported a review of 164 cases of cavernous hemangiomas, four (2.5%) of which were intraventricular in origin.

We have treated the case of a newborn girl who was found to be hydrocephalic at birth and in whom computerized tomography (CT) scans revealed a mass in the lateral ventricle. Cavernous hemangioma was the histological diagnosis. On the 8th day after her birth, we removed the entire tumor without difficulty. At present (2 years 9 months later), she can walk and even run without assistance, and is able to talk in monosyllables and phrases. In this report, we stress the importance of CT scanning for hydrocephalic newborn babies and children.

Case Report

This baby girl was born on July 26, 1978, by Cesarean section because of cephalopelvic disproportion. Her mother had no history of illness, medication, or irradiation during the course of her pregnancy. At birth, the baby had asphyxia, and it was noticed that she had a large head. Her Apgar score at birth was 6.

Examination. The head circumference was 42 cm, and the fontanels were tense and bulging. Her limbs were less active than normal, although the left side was more active than the right. Pupils were equal in size and reacted promptly to light on both sides, she responded to light and sounds normally. Plantar reflexes were extensor on both sides and fundi were normal. No congenital nevus was seen on her body.

Skull x-ray films showed separation of sutures but no abnormal calcification. Plain CT on admission showed high-grade hydrocephalus and a round mass of high density (mean value 13 Hounsfield units), approximately 3 cm in diameter, at the trigone of the left lateral ventricle (Fig. 1). The posterior portions of both lateral ventricles were greatly enlarged, more so on the left than the right. A brain technetium-99m revealed abnormal accumulation around the left trigonum. Electroencephalography showed irregular theta and delta waves in both hemispheres, more pronounced on the left side than the right. Angiograms were not available.

Blood analysis showed the following results: white blood cells 18,400 × 10^6/liter; red blood cells 436 × 10^6/liter, hemoglobin 14.6 gm/dl; reticulocytes 7%; platelets 27.7 × 10^9; total protein 5.6 gm/dl; blood sugar 180 mg/dl; hemoglobin S negative; C-reactive protein negative; syphilis testing negative; Na 143 mM/liter; K 4.9 mM/liter; C1 110 mM/liter.

Operation. A left temporoparietal craniotomy was performed with the patient under general endotracheal anesthesia and in the supine position. On opening the dura, we found a paper-thin temporal cortex which was bulging and hemispherical in shape, expanded with cerebrospinal fluid (CSF). The cortex was incised, and a copious amount of CSF was discharged. In the floor of the ventricle at the level of the trigone there was a solid, spongy, well encapsulated mass, dark red and blue in color, a portion of which seemed to be attached
firmly to the wall of the inferior horn of the left lateral ventricle; however, there was no feeding artery (Fig. 2). A draining vein was seen leading out of the tumor to the inner surface of the body of the lateral ventricle. The tumor was easily dissected from the ventricular wall, and there was a slight defect (approximately 1 cm in diameter) in the ependyma. Oozing was minimal. Total removal of the tumor was performed without difficulty. A shunt was placed between both lateral ventricles and the peritoneal cavity.

**Pathological Examination.** Histologically, the tumor was composed of clustered vascular spaces of various sizes (Fig. 3). A few microscopic foci of calcification were seen in the vascular lumina and interstitium along with thrombosis and old hemorrhage. This is compatible with typical cavernous hemangioma.

Cerebrospinal fluid, collected during surgery from both lateral ventricles separately, was examined. The results of the examination are shown in Table I. Protein levels in the left ventricular CSF were remarkably high.

**Postoperative Course.** The postoperative course was uneventful. Head circumference just after operation was less than 40 cm. Limb movement was much more active than preoperatively. The postoperative CT (Fig. 1 right) showed a decrease in size of both lateral ventricles and an area of low density in the left temporal lobe. No tumor shadow was seen. At present, the patient is 2 years and 9 months old. She can walk and even run without assistance, and is able to talk in monosyllables and phrases.

**Discussion**

The classification of cavernous hemangiomas proposed by Russell and Rubinstein has been generally accepted. According to their classification, cavernous hemangiomas are not true neoplasms and show no potential for metastasis. They vary in size from a few millimeters to several centimeters. Occasionally, cysts

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**Fig. 1.** *Left:* Preoperative plain computerized tomography scan. A pingpong ball-sized mass is shown in the left trigone. Very large lateral ventricles can be seen, the left side larger than the right. *Right:* Postoperative enhanced scan. No tumor mass was seen before or after injection of contrast material. Ventricles have decreased in size.

**Fig. 2.** Operative photograph showing the tumor at the trigone of the left lateral ventricle. The wall of the inferior horn and the body of the lateral ventricle are seen. The draining vein is not visible in this photograph.

**Fig. 3.** Photomicrograph of the tumor section showing cavernous hemangioma. H & E, × 100.
Intraventricular cavernous hemangioma

form within the tumor. Microscopically, they are composed of packed thin-walled vascular channels and are not separated by parenchyma. Calcification of cysts or vessels is not uncommon, but has appeared on conventional skull films in only four reported cases. Skull films of our patient did not show any abnormal calcification, but plain CT showed high-density areas with a mean value of 13 Hounsfield units.

Voigt and Yaşargil33 collected 164 cases of cavernous hemangioma up to 1976. In 1979, Namba, et al., added 13 cases, including one case of their own. We have collected another 38 reported cases, there were 11 cases (4.7%) of histologically verified ventricular cavernous hemangiomas among more than 200 cases (Table 2). Of these 11 patients were under 2 months old. Voigt and Yaşargil33 reported four intraventricular cases (2.5%) among 164 cases of cavernous hemangioma. Namba, et al., described eight cases arising from the cerebral ventricles. Five of these originated from lateral ventricles, another from the third ventricle, and the remaining two were so-called “choroid plexus angiomas.” Towfighi, et al., reported 28 cases of choroid plexus angiomas, including their own patients. McGuire, et al., described one case of bilateral angioma of the choroid plexus. Doe, et al., reported four cases of angiomas of the choroid plexus. McConnell and Leonard reported a patient with a very small cavernous angioma of the choroid plexus, but we included it in the group of choroid plexus angiomas. We omitted cases of choroid plexus an-

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Authors, Year</th>
<th>Age, Sex</th>
<th>Symptoms</th>
<th>Operation</th>
<th>Outcome</th>
<th>Localization</th>
<th>Size of Lesion</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Dandy, 1928</td>
<td>31 yrs, M</td>
<td>weakness lower extremities, blurred vision</td>
<td>suboccipital craniectomy</td>
<td>doing well 7 months after op</td>
<td>fourth ventricle</td>
<td>not mentioned, probably size of hazel nut</td>
</tr>
<tr>
<td>2</td>
<td>Merritt, 1940</td>
<td>16 yrs, F</td>
<td>headache, nausea, vomiting, diplopia after head injury</td>
<td>not mentioned</td>
<td>not mentioned</td>
<td>later ventricle</td>
<td>hen’s egg size</td>
</tr>
<tr>
<td>3</td>
<td>Arnstein, et al., 1951</td>
<td>3 days, M</td>
<td>regurgitation of fluids, tracheoesophageal fistula</td>
<td>—</td>
<td>died 2 days postop; at autopsy: large cavernous hemangioma in lateral ventricle with recent hemorrhage, fistula, &amp; multiple other anomalies</td>
<td>lateral ventricle</td>
<td>not mentioned</td>
</tr>
<tr>
<td>4</td>
<td>Lattermann, 1952</td>
<td>68 yrs, F</td>
<td>obesity, virilism, Morgagni's syndrome</td>
<td>—</td>
<td>pancreatitis &amp; cholecystitis, died; at autopsy: cavernous hemangioma in third ventricle</td>
<td>anterior portion of third ventricle</td>
<td>about 1 cm in diameter</td>
</tr>
<tr>
<td>5</td>
<td>Schneider &amp; Liss, 1958</td>
<td>33 yrs, F</td>
<td>headache, convulsion</td>
<td>rt temporoparietal craniotomy, total removal</td>
<td>not mentioned</td>
<td>lateral ventricle</td>
<td>10 cm in diameter</td>
</tr>
<tr>
<td>6</td>
<td>Jain, 1966</td>
<td>15 yrs, M</td>
<td>throbbing headache, papilledema</td>
<td>rt frontal craniotomy, total removal of cystic mass</td>
<td>not mentioned</td>
<td>anterior horn</td>
<td>5 cm in diameter</td>
</tr>
<tr>
<td>7</td>
<td>Numaguchi, et al., 1977</td>
<td>43 yrs, M</td>
<td>choked disc, tinnitus, monoparesis, hearing loss</td>
<td>craniotomy, total removal</td>
<td>alive 5 yrs after op</td>
<td>trigone</td>
<td>large</td>
</tr>
<tr>
<td>8</td>
<td>Coin, et al., 1977</td>
<td>36 yrs, F</td>
<td>convulsion</td>
<td>not clear</td>
<td>asymptomatic 2 yrs except for homonymous hemianopia</td>
<td>rt trigone</td>
<td>not mentioned</td>
</tr>
<tr>
<td>9</td>
<td>Giombini &amp; Morello, 1978</td>
<td>21 yrs, M</td>
<td>absence attacks</td>
<td>suboccipital craniectomy biopsy</td>
<td>died suddenly 2 mos postop</td>
<td>floor of fourth ventricle</td>
<td>1 cm in diameter</td>
</tr>
<tr>
<td>10</td>
<td>Pau &amp; Orunesu, 1979</td>
<td>56 yrs, ?</td>
<td>subarachnoid hemorrhage</td>
<td>not mentioned</td>
<td>died 3 hrs after admission</td>
<td>temporal horn</td>
<td>not mentioned</td>
</tr>
<tr>
<td>11</td>
<td>Iwasa, et al., 1983</td>
<td>3 days, F</td>
<td>enlarged head</td>
<td>lt temporal craniotomy, total removal</td>
<td>talks in monosyllables, able to run</td>
<td>lt trigone</td>
<td>3 cm in diameter</td>
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giomas because the lesions associated with the choroid plexus were very small, and because numerous microscopically abnormal dilated vessels within the choroid plexus were mixed with small arteries, veins, and capillaries. We also omitted the cases of Barsky\(^1\) and Guerard\(^6\) because they were not the cavernous angioma type discussed here.

Voigt and Yaşargil\(^1\) reported that 22 (13.6\%) of their 164 cases of cavernous hemangioma were multiple, and Giombini and Morello\(^9\) also showed a high incidence of the solitary type. Russell and Rubinstein\(^7\) noted that solitary cases were about three times more common than multiple cases.

Hydrocephalus was the first symptom in our patient. Dandy\(^6\) reported 45 cases of venous abnormalities and angiomas of the brain; five of these lesions were cavernous angiomas, and four of the patients showed initial symptoms of hydrocephalus. One patient had a fourth ventricular cavernous hemangioma. McGuire, \textit{et al.},\(^{17}\) reported one neonatal case in which the only symptom was hydrocephalus. We were not able to find any other patients whose first symptom was hydrocephalus.

Among the 11 cases of intraventricular cavernous hemangiomas that we have collected (Table 2), the first symptom was headache in three cases, epilepsy in three, papilledema in three, and motor weakness of the limbs in one case. Voigt and Yaşargil\(^1\) stated that the initial symptoms are commonly epileptic seizures, acute headache, and subarachnoid or intracerebral hemorrhage.

We found 11 reports in the literature dealing with the CT findings in the 17 cases of intracranial cavernous hemangiomas.\(^4,5,7,11,19-21,23,25,30,32\) All 18 cases, including our own, showed high-density areas. The CT density was reported in only six cases,\(^4,11,21,25,32\) and varied from 20 to 700 Hounsfield units. In our case, the density was 13 Hounsfield units.

We did not find any feeding arteries during surgery in our patient, but we did find a draining vein leading out of the tumor to the inner surface of the body of the left lateral ventricle. Protein content in the CSF of the left lateral ventricle was remarkably high (3040 mg/dl) in this case. This fact may indicate that the left inferior horn was blocked at the trigone by the tumor. We do not believe that the tumor secreted any protein. Histological sectioning did not show any secretory structure.

Acknowledgment

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Address reprint requests to: Hideaki Iwasa, M.D., Department of Surgical Neurology, Jichi Medical School, Minamikawachi-Machi, Kawachi-gun, Tochigi-ken, Japan 329-04.