Cerebral Hemangioblastoma

Case Report

E. Rivera, M.D., AND J. L. Chason, M.D.

Department of Pathology, Detroit General Hospital and Wayne State University
School of Medicine, Detroit, Michigan

Hemangioblastomas of the central nervous system are said to constitute from 1.1 to 2.4 per cent of all intracranial neoplasms. While almost all have occurred in the cerebellum, they have occasionally been found in the pons, medulla and spinal cord. Cushing and Bailey inferred that hemangioblastomas do not occur in the cerebrum, while Russell and Rubinstein consider these tumors to be extremely rare in the cerebral hemispheres. We are reporting a case of supratentorial hemangioblastoma.

Case Report

An 11-year-old Hungarian boy was first hospitalized on March 17, 1958, because of decreased visual acuity noted 1 month earlier by his teacher. He had had severe bitemporal headaches for 2 years, occurring daily for 1 year. Ataxia and a tendency to drop objects had been present during the last 4 months.

Physical examination. Blood pressure was 115/75; pulse was 80 and regular. Visual acuity was O.D. 3/200, O.S. 10/200. There was bilateral papilledema, more marked on the right, and perivascular exudates. Nystagmus was present on right lateral gaze. The other cranial nerves were intact. There were multiple "lapses of posture" and tic-like movements of the face. Muscle tone was generally decreased. There was weakness, slight spasticity, incoordination and fast pointing on the left. The Romberg sign was negative. The tendon reflexes were normal on the right and 3+ on the left. There was an extensor plantar response on the left.

Lumbar puncture on March 20 revealed an opening pressure of 450 mm. of water and closing pressure of 350 mm. The cerebrospinal fluid was normal. Ventriculography on March 27 showed displacement of lateral ventricles to the left and downward in the right parietal area. Right carotid angiograms on April 1 disclosed bowing of the anterior cerebral artery to the left. An abnormal vascular pattern suggestive of tumor stain was seen in the superior medial portion of the right fronto-parietal region.

Operation. At exploratory craniotomy on April 7, a mass deep in the right parietal area could only be partly removed.

Pathological Findings. The surgical specimen consisted of 15 gm. of grey, moderately firm but friable tissue. The tumor was made of normal delicate vascular channels primarily of capillary type with blood in the lumina of the larger vessels. The endothelial cells lining the vessels were uniformly plump and slightly spindle shaped, with clear cytoplasm and rounded or oval nuclei. Hyperchromatism and mitotic figures were rare. Reticulum stains showed a fine circle of argyrophilic fibers about the endothelial cells and enclosing groups of the cells in the vascular walls (Fig. 1). The diagnosis was hemangioblastoma.

Postoperative Course. The immediate postoperative course was marked by a left hemiplegia which responded slowly to physiotherapy. From May 12 to June 20 the patient received 2,700 r to the right parieto-occipital region. During the following 6 years, after temporary initial improvement, loss of vision became complete. There were, however, no other new neurological symptoms until July 14, 1964, when he was admitted because of the recurrence of headaches accompanied by nausea and vomiting. A soft bulge was present at the site of previous craniotomy. On neurological examination optic atrophy was marked; there was a right 6th nerve palsy, left central facial paresis, and generalized hyperreflexia. The Babinski sign was present on the left. Further surgery was refused. He died following the aspiration of food on August 1, 1964, 8 years after the initial onset of symptoms.

Postmortem Examination. The significant findings were confined to the brain, lungs, spleen and adrenal glands.

Received for publication March 2, 1966.

FIG. 1. Reticulum stain displaying a fine network of fibers around endothelial cells.
The brain weighed 1,450 grams. Occupying the parasagittal portion of the right frontal and parietal lobes was a firm, well-demarcated, grey-white mass, 7.5 cm. in maximum diameter (Fig. 2). It contained many 1 to 3 cm. elongated, oval, red to red-brown areas. The medial and superior portions of the mass were attached to the dura. The corpus callosum was markedly thinned. The right lateral ventricle was compressed and displaced downward and medially, and the 3rd ventricle was slightly tilted to the left. Microscopically, the tumor exhibited the same features described in the biopsy specimen. In addition, characteristic "pseudo-xanthomatous

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author</th>
<th>Year</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Gross Appearance</th>
<th>Duration of Illness</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Bennett</td>
<td>1946</td>
<td>*</td>
<td>*</td>
<td>Rt. Frontal</td>
<td>*</td>
<td>*</td>
</tr>
<tr>
<td>2</td>
<td>Bennett</td>
<td>1946</td>
<td>*</td>
<td>*</td>
<td>Lt. Frontal</td>
<td>*</td>
<td>*</td>
</tr>
<tr>
<td>3</td>
<td>Bennett</td>
<td>1946</td>
<td>*</td>
<td>*</td>
<td>Rt. Temporal</td>
<td>*</td>
<td>*</td>
</tr>
<tr>
<td>4</td>
<td>Kautzky &amp; Vierdt</td>
<td>1953</td>
<td>*</td>
<td>*</td>
<td>Lt. Frontal</td>
<td>*</td>
<td>*</td>
</tr>
<tr>
<td>5</td>
<td>Stein et al.</td>
<td>1960</td>
<td>12</td>
<td>F</td>
<td>Rt. Frontal</td>
<td>Cystic</td>
<td>4.5 mos.</td>
</tr>
<tr>
<td>6</td>
<td>Stein et al.</td>
<td>1960</td>
<td>49</td>
<td>M</td>
<td>Lt. Temporal</td>
<td>Solid</td>
<td>3 wks</td>
</tr>
<tr>
<td>7</td>
<td>Bras include</td>
<td>1961</td>
<td>63</td>
<td>M</td>
<td>Lt. Parietal</td>
<td>Solid</td>
<td>*</td>
</tr>
<tr>
<td>8</td>
<td>Rivera &amp; Chason</td>
<td>1966</td>
<td>11</td>
<td>M</td>
<td>Rt. Fronto-parietal</td>
<td>Solid</td>
<td>8.3 yrs.</td>
</tr>
</tbody>
</table>

* Not recorded.
cells" were present in isolated clusters, often associated with areas of necrosis.

The 190-gram spleen was firm and red. Sections showed the typical appearances of amyloidosis. The adrenal glands were similarly affected. The lungs exhibited gross and microscopic evidences of an aspiration pneumonia.

Discussion

Hemangioblastomas are histologically benign tumors originating from remnants of the mesoderm that grow into the central nervous system during the 3rd fetal month. When hemangioblastomas occur in the central nervous system, they are usually found in the cerebellum, much less commonly in the pons or medulla and rarely are noted above the tentorium. Two cases of supratentorial hemangioblastomas are included in the series of Stein et al. and Bennett reported 3, but no details are given. Brasseur has recently reported one more case, and accepts the one of Kautzky and Vierdt reported in 1953. Table 1 summarizes the cases reported to date. The case reported by Rochat has been excluded because of insufficient photomicrographic documentation, and the case of Barnard and Walshe does not belong in this category.

The classical description of this tumor is that of Cushing and Bailey, which distinguished between two main forms, namely, cystic and solid; the latter is further subdivided into capillary, cellular and cavernous, depending upon the predominating histological pattern. These tumors have a characteristic reticulum network considered to be peculiar to them. Also characteristic of these lesions are the "pseudo-xanthomatous cells," originally described by Lindau, and grown in tissue cultures by Brasseur. These cells seem to represent degenerative cellular changes.

Our case did not show other vascular malformations as seen in Lindau-von Hippel disease or the polycythemia often associated with cerebellar hemangio-blastomas (the hemoglobin was 9.5 and 10.1 on 2 separate occasions). There was no satisfactory explanation for the amyloidosis of the spleen and adrenal glands.

Summary

Hemangioblastomas are benign tumors (vascular malformations rather than true neoplasms) occasionally found in the cerebellum and very rarely in the cerebrum. We have reported a case of hemangioblastoma and have reviewed the literature. An unusual feature of our case was its association with amyloidosis of the spleen and adrenal glands.

References