Intraorbital optic nerve hemangioblastoma with von Hippel-Lindau disease

Case report

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An intraorbital hemangioblastoma of the optic nerve is reported in a 23-year-old woman with von Hippel-Lindau disease. The absence of dural attachment and the microscopic findings were characteristic of hemangioblastoma. Four years later the patient developed a cystic hemangioblastoma in the left cerebellum which was successfully treated.

Key Words • hemangioblastoma • optic nerve • von Hippel-Lindau disease

Hemangioblastomas of the brain are most commonly found in the cerebellum; they appear less frequently in the spinal cord and medulla oblongata. A small number of supratentorial lesions have been reported. Lesions of the optic nerve are extremely rare: to our knowledge, only four such cases have been reported previously, in which three tumors were found by chance at autopsy and the fourth was surgically removed. We are now presenting the case of a patient with multiple hemangioblastomas located in the intraorbital portion of the optic nerve and in the cerebellum.

Case Report

This 23-year-old woman was first admitted to Kurume University Hospital on November 15, 1974, with complaints of visual loss and exophthalmos of the left eye. About 7 years earlier she had first noticed blurring in the left eye, and a diagnosis of von Hippel’s disease (angiomatosis retinae) was made. Since then, her vision had been getting worse and was finally lost when secondary glaucoma and cataract developed during the 4 years before admission. The family history, including that of living parents and three siblings, revealed no evidence of brain tumor or abnormality of the central nervous system.

Examination. Physical and neurological examinations were not remarkable except for pulsating exophthalmos, blindness, and absent direct light reflex in the left eye. There were no superficial skin lesions. Routine laboratory investigations disclosed no abnormal data. X-ray films showed that the left optic foramen was twice as large as the right (Fig. 1). A left carotid angiogram revealed a hypervascular mass in the orbit, supplied by a few branches from the enlarged, elongated ophthalmic artery (Fig. 2). The initial diagnosis, based on these findings, was probable intraorbital optic nerve hemangioma or meningioma.

First Operation. A left frontal osteoplastic craniotomy was performed under general anesthesia, and the left optic canal was exposed. The tumor was found to arise in the left optic nerve itself, and was mainly situated in the orbit. The tumor was not attached to the dura and had no capsule. It was completely removed, together with the intraorbital portion of the optic nerve, which was resected between the posterior wall of the eyeball and the point of entry into the optic canal. The tumor measured about 29 x 17 x 15 mm, and had a reddish-brown surface. Microscopic examination revealed numerous thin-walled vascular channels, separated by nests of large polygonal cells with swollen foamy cytoplasm (Fig. 4A). Reticulin stain showed a capillary structure and stroma cells surrounded by reticulin (Fig. 4B). Sudan III stain showed heavy but irregularly distributed deposits of lipid (Fig. 4C). The stump of the optic nerve was found adjacent to the tumor tissue (Fig. 4D). The findings were those of a capillary hemangioblastoma.
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Her postoperative course was uneventful except for blindness in the left eye. She was readmitted on February 20, 1978, with a 3-month history of occipital headache and vertigo. These symptoms were exacerbated when she lay on the left side. Computerized tomography revealed a large cystic lesion with a small area in the left cerebellar hemisphere that partially enhanced on injection of contrast medium (Fig. 5). Left vertebral angiography disclosed a cystic mass with a small vascular nodule.

Second Operation. At suboccipital craniectomy, a cystic hemangioblastoma with a mural nodule the size of a little fingertip was found in the left cerebellum. About 22 ml of cyst fluid was drained, and the vascular nodule was completely removed.

The patient had an uncomplicated postoperative course and was discharged 2 weeks after the procedure. Three years after the second operation she was living normally as a housewife, without neurological deficit except for blindness in the left eye.
Hemangioblastomas are commonly seen in the cerebellum, but their occurrence supratentorially is uncommon. An intraorbital hemangioblastoma of the optic nerve is an extremely rare tumor. Only four such cases have been described previously (Table 1). Of the five cases, four, including ours, were attributed to von Hippel-Lindau disease. Three tumors were found and two were surgically removed. In all cases, the tumors tended to be solid.

In the present case, x-ray films showed the left optic foramen to be twice as large as the right, and left carotid angiography revealed a hypervascular mass in the orbit. These findings were considered preoperatively to be consistent with hemangioma or angiovascular meningioma arising in the intraorbital optic nerve sheath. However, the operative findings showed that the tumor was not attached to the optic nerve sheath, and the main growth of this mass was within the intraorbital portion of the optic nerve. The histopathological findings fulfilled the criteria for diagnosis of a hemangioblastoma rather than angiovascular meningioma. Four years later, a cystic hemangioblastoma was found in the left cerebellum. The occurrence of cerebellar hemangioblastoma conforms with a diagnosis of von Hippel-Lindau disease. We consider that this optic nerve lesion was identical to hemangioblastomas found in other locations, such as the cerebellum or the brain stem.

In cases such as this, a careful long-term follow-up review should be undertaken to watch for the occurrence of another hemangioblastoma, since these tumors may be multiple.

### TABLE 1

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Patient’s Age (yrs) &amp; Sex</th>
<th>Gross Tumor Appearance</th>
<th>Infratentorial Occurrence</th>
<th>von Hippel-Lindau Disease</th>
<th>Site</th>
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<tr>
<td>Verga, 1930</td>
<td>57, ?</td>
<td>solid</td>
<td>yes</td>
<td>yes</td>
<td>rt intracranial</td>
</tr>
<tr>
<td>Schneider, 1942</td>
<td>?, ?</td>
<td>solid</td>
<td>yes</td>
<td>yes</td>
<td>intraorbital</td>
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<tr>
<td>Nobile, 1951</td>
<td>53, M</td>
<td>solid</td>
<td>yes</td>
<td>no</td>
<td>lt optic nerve</td>
</tr>
<tr>
<td>Stefani &amp; Rothenmund, 1974</td>
<td>43, M</td>
<td>solid</td>
<td>no</td>
<td>no</td>
<td>rt intracranial</td>
</tr>
<tr>
<td>In, et al., 1982</td>
<td>23, F</td>
<td>solid</td>
<td>yes</td>
<td>yes</td>
<td>lt intraorbital</td>
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</table>
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Fig. 5. Computerized tomography with contrast enhancement showing a cystic mass with an enhanced mural nodule in the left cerebellar hemisphere.

References


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