Papillomas of the Choroid Plexus

SAMI I. NASSAR, M.D., AND LESTER A. MOUNT, M.D.

Department of Neurological Surgery, Columbia University, College of Physicians and Surgeons,
Service of Neurological Surgery, Columbia-Presbyterian Hospital,
The Neurological Institute, New York, New York

Papillomas of the choroid plexus are solitary reddish grey tumors attached to the choroid plexus. The irregular surface is covered by a thin transparent capsule, and the firm tumor occasionally is partly calcified. Microscopically, papillomas resemble normal choroid plexus, being composed of columnar or cuboidal epithelium supported by highly vascularized connective tissue. They are usually benign but may undergo malignant transformation. They are occasionally multiple and may metastasize by way of the cerebrospinal fluid regardless of their ventricular location.

In order of frequency papillomas are located in the fourth ventricle, atrium of the lateral ventricle, and the third ventricle. In infants and the younger age groups they are most often found in the lateral ventricle; in adults, in the fourth ventricle. Although they occur in any age group, even in the newborn, they show a predilection for the younger age group with the greatest incidence in the first decade of life. They are slightly more common in males and have been found in a brother and sister. Papillomas of the choroid plexus account for 0.4 to 0.6% of all brain tumors.

The clinical picture of a papilloma of the fourth ventricle is one of increased intracranial pressure without localizing features except for ataxia, nystagmus, and cranial nerve palsies. In infants, the symptoms are those of progressive hydrocephalus with enlargement of the head, apathy, irritability, and a failure to thrive. In adults, the hydrocephalus is caused by obstruction to the circulation of cerebrospinal fluid, while in infants it frequently is due to the increased secretory surface created by the tumor which causes a concomitant increase in the production of cerebrospinal fluid.

Subarachnoid hemorrhage is rarely the presenting symptom in an adult.

The diagnosis of papilloma of the choroid plexus is usually established by an air study, usually ventriculography. A brain scan will reveal the tumor if it is larger than 2 cm. If the clinical picture is one of subarachnoid hemorrhage, the diagnosis rests principally on arteriography. A tumor stain with well-defined choroidal vessels feeding the neoplasm is usually demonstrated. If arteriography is inconclusive, then ventriculography is indicated.

The treatment is surgical removal of the tumor. Operative treatment, however, carries with it a high mortality and morbidity. Most series reported give a mortality of 27% to 45%. The risk is higher when the papilloma is in the fourth ventricle in close proximity to the brain stem. The benefit of x-ray therapy to these tumors remains inconclusive; some authors believe it reduces the vascularity of the tumor. Others believe its main value is in diminishing the overproduction of cerebrospinal fluid, which may also be the result of the decreased vascularity. In general, however, radiotherapy is not indicated in this benign form of tumor but should probably be used when there are invasive characteristics or malignant transformation.

Analysis of Cases

After reviewing 17 cases of papillomas of the choroid plexus treated at the Neurological Institute of New York, we divided them into two groups: tumors arising in the third and fourth ventricles, and those arising in the lateral ventricles.

Papillomas of the Third and Fourth Ventricles (14 Cases). Five papillomas originated in the third ventricle and nine in the fourth. The average age of the patients was 33 years, with a range of 11 to 52 years. The duration of symptoms was 1.9
years with extremes of 2 months to 5 years. All patients had headache, nausea, and vomiting, and unlocalized signs of increased intracranial pressure, namely, papilledema and a decrease in visual acuity. When the tumors arose in the fourth ventricle, nystagmus and gait disturbances were prominent signs. In all patients the diagnosis was achieved by air study, usually ventriculography, which demonstrated the presence of a mass lesion causing obstructive hydrocephalus. The cerebrospinal fluid pressure was high, ranging between 380 and 600 mm of water with an average of 460 mm. The mean ventricular cerebrospinal fluid protein was 26 mg%, with a low of 9 mg% and a high of 208. In the latter case the fluid was also bloody and xanthochromic. Although subarachnoid hemorrhage as an initial precipitating symptoms was not encountered in any of our patients, the cerebrospinal fluid was blood-tinged and xanthochromic in two.

**Surgical treatment: third ventricle papillomas (5 cases).** Treatment of the third ventricle papillomas was total excision of the tumor in four patients and partial removal in one, all approached through the lateral ventricle. One patient had no significant neurological deficit 1 year after operation, but was then lost to follow-up. Three patients followed for over 10 years are doing well. The patient with partial excision died in the immediate postoperative period from midbrain and thalamic hemorrhage (Table 1).

**Surgical treatment: fourth ventricle papillomas (9 cases).** Gross total excision of the mass through a suboccipital craniectomy was accomplished in three patients. One of them died in the postoperative period; the other two are alive and well 16 and 11 years post-operatively, with only a mild decrease in visual acuity as a residual deficit. Subtotal excision of the tumor was done in six patients. Two of them died 1 year later from recurrence; of the remaining four, one (followed for 5 years) is doing well except for mild ataxia, two have been lost to follow-up, and one has a significant neurological deficit (Table 2).

**Papillomas of the Lateral Ventricles (3 Cases).** Two patients had papillomas arising in the right lateral ventricle and one in the left. The average age of the patients was 8 months, and the average duration of symptoms, 1 month. All three had enlarged heads and signs of increased intracranial pressure consistent with the clinical picture of congenital hydrocephalus. The diagnosis was made by ventriculography in two cases and by pneumoencephalography in one case. In two cases there was uniform enlargement of the ventricular system without any evidence of obstruction to the cerebrospinal flow, and in one case obstruction of the foramen of Munro with unilateral enlargement of the involved ventricle. In two cases the cerebrospinal fluid was blood-tinged.

**TABLE 1**

<table>
<thead>
<tr>
<th>Tumor Location</th>
<th>No. of Cases</th>
<th>Average Age</th>
<th>Surgical Excision</th>
<th>Total Excision</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Third ventricle</td>
<td>5</td>
<td>38 yrs</td>
<td>4 Total, 1 Subtotal</td>
<td>4 improved; doing well after 1-yr (1 case) &amp; 10-yr (3) follow-up</td>
<td>1 died, hemorrhages in midbrain and brain stem.</td>
</tr>
<tr>
<td>Fourth ventricle</td>
<td>9</td>
<td>33 yrs</td>
<td>3 Total, 6 Subtotal</td>
<td>2 improved, doing well after 11 &amp; 16-yr follow-ups; 1 died, brain stem hemorrhages</td>
<td>2 lost to follow-up; 2 died from recurrent tumor; 1 poor, severe residual signs; 1 improved, mild cerebellar dysfunction</td>
</tr>
<tr>
<td>Lateral ventricle</td>
<td>3</td>
<td>8 mos</td>
<td>3 Total, 0 Subtotal</td>
<td>2 poor, mental retardation; 1 died</td>
<td></td>
</tr>
</tbody>
</table>
Papillomas of the Choroid Plexus

**TABLE 2**

*Choroid plexus papilloma of the fourth ventricle*

<table>
<thead>
<tr>
<th>Tumor Location</th>
<th>No. of Patients</th>
<th>Surgical Excision</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fourth ventricle (9 cases)</td>
<td>1</td>
<td>total</td>
<td>improved; doing well 16 yrs later</td>
</tr>
<tr>
<td></td>
<td>1</td>
<td>total</td>
<td>improved; doing well 11 yrs later</td>
</tr>
<tr>
<td></td>
<td>1</td>
<td>total</td>
<td>died; brain stem hemorrhages</td>
</tr>
<tr>
<td></td>
<td>1</td>
<td>subtotal</td>
<td>improved; mild cerebellar dysfunction</td>
</tr>
<tr>
<td></td>
<td>1</td>
<td>subtotal</td>
<td>poor; severe mental changes and cerebellar residual signs</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>subtotal</td>
<td>died; recurrent tumor growth</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>subtotal</td>
<td>lost to follow-up</td>
</tr>
</tbody>
</table>

The treatment of the three cases of lateral ventricle tumors consisted of transcortical gross total excision of the mass. One patient died during the immediate postoperative period from respiratory failure. The two remaining patients were left with significant mental retardation and neurological deficit (Table 3).

Seven patients received postoperative radiotherapy but had no significant alteration in the course of the disease.

We are reporting the case of one patient who had a new presenting symptom, a localized enlargement of one side of the head.

**Case Report.** The patient was a 6-month-old baby girl, the product of a full-term uncomplicated pregnancy and delivery, who had developed normally. The parents noted, however, that the child had had an asymmetrical head since birth, with a prominent right side; 4 weeks before admission the prominence apparently increased. There was no history of trauma, vomiting, fever, or convulsions.

**Examination.** The general physical examination was normal and the only neurological abnormality was gross asymmetry of the head. There was a 2 cm bulge about 10 cm in diameter, in the right posterior parietal region; the skull here was thin and easily depressed, with a "ping pong" ball consistency. The anterior fontanelle measured 5 × 5 cm and was soft and pulsating. The circumference of the head was 46 cm. X-rays of the skull showed thinning and scalloping of the right parietal bone but no radiological evidence of generalized increased intracranial pressure. A pneumoencephalogram revealed a large mass projecting into the atrium and body of the right lateral ventricle, displacing the right occipital horn laterally and downward and the aqueduct of Sylvius and the posterior part of the third ventricle to the left. The radiological interpretation was an extra- and intraventricular tumor.

**Operation.** On November 18, 1966, a right parietooccipital craniotomy was performed under general anesthesia. A spherical, semi-encapsulated, reddish-gray tumor, 5 cm in diameter, was found in the atrium of the right lateral ventricle extending into the temporal horn, the occipital horn, and the body of the lateral ventricle. It had moderate vascularity, was firm in some areas but soft in others, and was attached to the

**TABLE 3**

*Choroid plexus papilloma of the lateral ventricle in three patients*

<table>
<thead>
<tr>
<th>Treatment</th>
<th>No. of Patients</th>
<th>Result</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gross total excision</td>
<td>1</td>
<td>poor</td>
<td>mental retardation and akinetic seizures</td>
</tr>
<tr>
<td>(average age of patient, 8 mos)</td>
<td>1</td>
<td>poor</td>
<td>postoperative obstruction of atrium; shunt procedure necessary; severe cortical atrophy</td>
</tr>
<tr>
<td></td>
<td>1</td>
<td>died</td>
<td>postoperative respiratory failure</td>
</tr>
</tbody>
</table>
choroid plexus. The tumor was totally removed.

The pathological diagnosis was choroid plexus papilloma. The immediate postoperative course was febrile, which was thought to be secondary to blood in the ventricular system. The patient then made a good recovery and when last seen 9 months postoperatively was doing well, with a continued pattern of normal growth and development.

Discussion

Papillomas of the choroid plexus occur predominantly in the first decade of life. In contrast to other series, 77% of the 17 reported patients at the Neurological Institute in the past 30 years were in the older age group, with an average age of 34 years. The most common location in children was in the lateral ventricle and in adults in the fourth ventricle.

When these tumors occur in infants they are invariably accompanied by a progressively enlarging head and must be differentiated from patients having communicating or obstructive hydrocephalus of other etiology. Matson\textsuperscript{14} stresses the need for determining the etiology of the hydrocephalus by radiological diagnostic tests so that proper treatment can be given. Hydrocephalus secondary to papilloma of the choroid plexus recedes when the offending lesion is removed surgically. In our reported case there was some enlargement of the head but no hydrocephalus to suggest oversecretion of cerebrospinal fluid by the tumor and no obstruction to the flow of cerebrospinal fluid by the neoplasm. The localized unilateral expansion of the skull was apparently due to the mass effect of the tumor, which in its posterior location was partially prevented from growing inward by the falx and tentorium.

Radiologically, these tumors exhibit no characteristic appearance. When they grow in the fourth ventricle, they may simulate an ependymoma, medulloblastoma, astrocytoma, or other type of tumor in the posterior fossa. In the lateral ventricle they may grow to a large size and project through the atrial wall of the ventricle into the cerebral substance. In the differential diagnosis one must therefore consider an astrocytoma, oligodendroglioma, ependymoma, or intraventricular meningioma. A papilloma may also simulate a porencephalic cyst, which was the original interpretation of the plain x-ray films of the skull in our case. Papillomas of the third ventricle must be differentiated from midline congenital tumors, gliomas, and colloid cysts when they rise in the anterior part of the ventricle.

Papillomas of the choroid plexus are vascular and, if large, may show an abnormal collection on radioisotope scan studies or a tumor cloud in the arteriogram. These studies may differentiate such lesions from less vascular tumors in the same site, but the diagnosis can be established only by operation.

The great majority of these tumors are histologically benign; complete excision is indicated, although the tumor location and vascularity sometimes precludes total extirpation. The value of x-ray therapy as an adjunct treatment is questionable.

Morbidity, mortality, and neurological deficit increase with the size of the tumor. The result of operations on papillomas of the choroid plexus therefore depends to a considerable extent on early diagnosis, accurate localization, and direct surgical excision.

Summary

We have reviewed the problem of papilloma of the choroid plexus, particularly as seen in 17 cases at the Neurological Institute of New York. One of these cases had the unusual presenting symptom of asymmetrical enlargement of the head.

References

7. Drucker, G. A. Papillary tumor of the

20. Schlesinger, E. B. Personal communication.