Papillomas of the Choroidal Plexus

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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

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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 mid-brain 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

Results of surgical treatment of choroid plexus papillomas of the third, fourth, and lateral ventricles

<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</td>
<td>1</td>
<td>4 improved; doing well after 1-yr (1 case) &amp; 10-yr (3) follow-up</td>
</tr>
<tr>
<td>Fourth ventricle</td>
<td>9</td>
<td>33 yrs</td>
<td>3</td>
<td>6</td>
<td>2 improved, doing well after 11 &amp; 16-yr follow-ups; 1 died, brain stem hemorrhages</td>
</tr>
<tr>
<td>Lateral ventricle</td>
<td>3</td>
<td>8 mos</td>
<td>3</td>
<td>0</td>
<td>2 poor, mental retardation; 1 died</td>
</tr>
</tbody>
</table>

1 died, hemorrhages in midbrain and brain stem.
2 lost to follow-up; 2 died from recurrent tumor; 1 poor, severe residual signs; 1 improved, mild cerebellar dysfunction