Multiple choroid plexus papillomas of the lateral ventricle distinct from villous hypertrophy

Case report

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Except for villous hypertrophy of the choroid plexus that may not be true tumors, multiple choroid plexus papillomas are extremely rare. The authors report a case involving multiple choroid plexus papillomas that were distinct from villous hypertrophy. These lesions were localized, one in the atrium of the right lateral ventricle and the other in the inferior horn of the left lateral ventricle. A review of the literature revealed that this case represented the first reported case of true multiple choroid plexus papillomas documented by findings on magnetic resonance imaging.

KEY WORDS • choroid plexus • papilloma • brain neoplasm • hydrocephalus • metastasis • magnetic resonance imaging

MUltiple choroid plexus papillomas are not considered to occur infrequently. In 1970, Rovit, et al., 29 reported the incidence to be 3.7% in 245 cases, 11 of which were their own cases and the rest collected from the literature. However, multiple choroid plexus papillomas are actually very rare lesions that were not encountered in several large series. 3,4,14,15,22,24,25,32,33 Furthermore, the majority of examples described in the literature 5,8,13,17,19–21,23,34,35 appeared to represent villous hypertrophy of the choroid plexus, originally described by Davis, 6 and may not have been true tumors. We report the case of a patient with multiple choroid plexus papillomas that were distinct from villous hypertrophy. The review of the literature revealed that no such case has been documented previously by using computerized tomography (CT) scanning or magnetic resonance (MR) imaging.

Case Report

History. This 16-year-old girl was referred to our service in 1993 because of the suspicion of gigantism and because neuroimaging showed enlargement of the sella turcica.

Examination. The patient measured 170 cm in height and weighed 70 kg; her head circumference was 58.5 cm. A physical examination revealed no abnormalities and no apparent neurological deficit. Computerized tomography scans demonstrated dilation of the lateral, third, and fourth ventricles, and two separate tumors: one in the atrium of the right lateral ventricle and the other in the inferior horn of the left lateral ventricle. A review of the literature revealed that this case represented the first reported case of true multiple choroid plexus papillomas documented by findings on magnetic resonance imaging.

Operation. On September 29, 1993, the patient underwent a right parietal craniotomy. A reddish, cauliflower-like mass located within the atrium of the right lateral ventricle was exposed through a small cortical incision of the postmarginal gyrus. The mass was freely mobile within the ventricle except for the portion attached to the normal choroid plexus. There was no continuity of the mass to the ventricular wall or any extension of the mass or seeding along the ventricular wall to the foramen of Monro. The mass was totally removed after dissection from the choroidal plexus.

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Postoperative Course and Second Operation. Four weeks after her initial surgery, the patient underwent a left temporal craniotomy. The mass located within the trigone of the left lateral ventricle was exposed through the small cortical incision of the inferior temporal gyrus. The mass was freely mobile within the ventricle, much like the previously removed mass, except for the small section attached to the normal choroid plexus. Again, there was no extension of the mass or seeding along the ventricular wall. The tumor was totally removed after dissection from the choroid plexus. The postoperative course was uneventful.

Histopathological Examination. On histopathological examination both tumors were found to consist of a monotonous papillary structure composed of epithelioid cells forming a single layer over a fibrovascular stalk. These findings were consistent with the diagnosis of benign choroid plexus papilloma (Fig. 3). Follow-up CT scans obtained 1 month after the second surgery revealed normalization of ventricular size. When the patient was last seen 3 years after the second surgery, she demonstrated no neurological deficits. Magnetic resonance imaging revealed no local recurrence or distant metastasis suggestive of seeding.

Discussion

Rovit, et al., found nine cases of multiple choroid plexus papillomas in the literature. Since then, only 11 additional cases of multiple choroid plexus papillomas have been described. Most of these cases involved bilateral tumors found in the lateral ventricle; six of the cases reviewed by Rovit, et al., and nine of the cases reported subsequently had separate papillomas in both of the lateral ventricles. Davis reported such a case in 1925 and named these lesions villous hypertrophy of the choroid plexus, suggesting that these lesions are not true tumors. Most of the authors in more recent reports described their cases as villous hypertrophy.

A review of the cases of bilateral choroidal plexus papillomas indicated that, although an exact distinction between hypertrophy and papilloma was not possible, these lesions had many features in common. First, without exception, the lesions arose symmetrically within both lateral ventricles. The lesions were often diffusely enlarged choroid plexuses themselves and did not form an impressive mass. Even when the lesions appeared to form localized masses, enhanced coronal and sagittal MR images revealed the diffusely enlarged choroid plexus of both lateral ventricles. Second, production of a large volume of CSF, reaching four or five times that of normal production, was frequently mentioned, irrespective of the lesion size. This feature clearly indicates that these lesions maintain normal physiological function of the choroid plexus. Third, these lesions always caused severe progressive hydrocephalus and elevated intracranial pressure. Information on most of the cases reported earlier was derived from postmortem examinations in which the suspected cause of death was severe progressive hydrocephalus. It is clear that this is the consequence...
of the production of a huge volume of CSF, because resolution of hydrocephalus can be achieved by excision of lesions. Gudeman, et al., described in detail a dramatic reduction in CSF production after the removal of bilateral choroid plexus papillomas. Finally, most of these lesions occurred in neonates or infants younger than 15 months of age; the only exceptions were two cases in which the children were 3.5 and 7 years of age, respectively. These lesions should, therefore, be kept in mind as an important cause of neonatal or infantile hydrocephalus; however, their relatively small size may prevent their precise diagnosis, and these common features imply that previously reported cases of bilateral choroid plexus papillomas may represent villous hyper trophy or lesions triggered by a similar pathogenetic mechanism.

The present case does not resemble any of these previously reported cases. The lesions in our patient formed localized masses in asymmetrical sites, one in the atrium and the other in the inferior horn, without diffuse enlargement of the choroid plexus. Although the patient developed significant ventricular dilation, it was evident that the production of CSF had not been sufficient to cause severe progressive hydrocephalus, because no signs of elevated intracranial pressure were noted. In addition, the patient was far older than most of the patients previously reported. These differences strongly suggest that the lesions examined in the present case are unique, unlike those of the previously reported bilateral choroid plexus papillomas.

There is no evidence of a causal relationship between villous hypertrophy and bilateral choroid plexus papillomas. Although the obvious genetic factors have not been implicated in the development of choroid plexus tumors including villous hypertrophy, the fact that many of these tumors are found in neonates and infants indicates that these are congenital. There has been a report of choroid plexus tumor associated with von Hippel-Lindau disease. In this case, examination of the choroid plexus tumor tissue revealed loss of an allele on chromosome 3. There are also some reports of choroid plexus tumors associated with Aicardi’s syndrome and Li-Fraumeni syndrome. Bergasagel, et al., have suggested causal roles for polyoma virus and simian virus 40 in the development of choroid plexus tumors. Further neurogenetic and molecular biological studies are required to elucidate these developmental mechanisms.

Among the 245 cases of choroid plexus papillomas reviewed by Rovit, et al., two had separate tumors in the third or fourth ventricle in addition to the lateral ventricle and one had separate tumors in the third and fourth ventricles. Pascual-Castroviejo, et al., and Tomita, et al., reported third and lateral ventricular choroid plexus papillomas. Wilkins and Rutledge also described another case in which the tumors were located in the right lateral ventricle and fourth ventricle (Table 1). Although no more details are available, it is clear that the lesions in these cases are located in asymmetrical sites, suggesting that they are distinct from villous hypertrophy. The present case appears to belong to this rare group of multiple choroid plexus papillomas.

At times, choroid plexus papillomas have been noted to extend into the contiguous ventricular chambers or cisterns, to seed along the CSF pathways, and to...
expand within either the cerebral or cerebellar hemispheres. Such seeding occasionally occurs despite a finding of benign papilloma without mitosis. However, in the present case, no seeding within the CSF system or extension through the foramen of Monro was observed on preoperative neuroimaging studies or intraoperatively. No recurrence suggestive of seeding was noted during the postoperative follow-up period of 3 years. Considering the normal direction of flow of CSF, isolated metastasis from one lateral ventricle to the contralateral lateral ventricle is improbable in cases in which there is no extension to the foramen of Monro. Furthermore, the fact that each of the masses was attached to the normal choroid plexus in the present case also supports the inference that these two masses arose independently.

In conclusion, it seems evident that, contrary to widely held belief, multiple choroid plexus papillomas other than villous hypertrophy are extremely rare. Nevertheless, the present case confirms that multiple choroid plexus papillomas distinct from villous hypertrophy do exist. Metastasis via the usual mechanisms is improbable as the pathogenesis of such lesions. It may still be possible, however, that isolated metastasis can occur through mechanisms unique to this type of tumor. Any conclusive findings regarding this possibility await further accumulation and investigation of cases.

References


TABLE 1

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Cases</th>
<th>Age, Sex</th>
<th>Site of tumor</th>
<th>Hydro VH</th>
</tr>
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<tbody>
<tr>
<td>Matson &amp; Crofton, 1960</td>
<td>6</td>
<td>15 mos, M†</td>
<td>both lat vent</td>
<td>+ +</td>
</tr>
<tr>
<td>Wilkins &amp; Rutledge, 1961</td>
<td>1</td>
<td>3 mos, F‡</td>
<td>both lat vent</td>
<td>+ –</td>
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<tr>
<td>Rovit, et al., 1970</td>
<td>6§</td>
<td>—</td>
<td>both lat vent</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>1</td>
<td>—</td>
<td>rt lat vent + IV vent</td>
<td>+</td>
</tr>
<tr>
<td></td>
<td>1</td>
<td>—</td>
<td>lat vent + III vent</td>
<td>+</td>
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<tr>
<td></td>
<td>1</td>
<td>—</td>
<td>III vent + IV vent</td>
<td>+</td>
</tr>
<tr>
<td>Gudeman, et al., 1979</td>
<td>1</td>
<td>3.5 yrs, F</td>
<td>both lat vent</td>
<td>+ +</td>
</tr>
<tr>
<td>Laurence, 1979</td>
<td>1</td>
<td>6 mos, F</td>
<td>both lat vent</td>
<td>+ +</td>
</tr>
<tr>
<td>Kitami, et al., 1982</td>
<td>2</td>
<td>2.1 yrs, M</td>
<td>both lat vent</td>
<td>+ +</td>
</tr>
<tr>
<td></td>
<td>7 mos, F</td>
<td>both lat vent</td>
<td>+ +</td>
<td></td>
</tr>
<tr>
<td>Pascual-Castroviejo, et al., 1983</td>
<td>1</td>
<td>4 mos, M</td>
<td>lt vent + III vent</td>
<td>+ –</td>
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<tr>
<td>Welch, et al., 1983</td>
<td>1</td>
<td>neonatal, F</td>
<td>both lat vent</td>
<td>+ +</td>
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<tr>
<td>Tomita, et al., 1988</td>
<td>2</td>
<td>8 mos, M</td>
<td>both lat vent</td>
<td>+ +</td>
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<td></td>
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<td></td>
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<td>1</td>
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<td>+ +</td>
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<tr>
<td>Hirano, et al., 1994</td>
<td>1</td>
<td>7 yrs, F</td>
<td>both lat vent</td>
<td>+ +</td>
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</tbody>
</table>

* Hydro = hydrocephalus; vent = ventricle; VH = villous hypertrophy; + = present; – = absent.
† Reported by Davis.
‡ Reported by Ray and Peck.
§ Possibly six cases reported by Matson and Crofton.
Multiple choroid plexus papillomas


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