Slow Reduction in Ventricular Size after Removal of Choroid Plexus Papilloma

—Case Report—

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Abstract

A choroid plexus papilloma (CPP) was discovered in the right lateral ventricle of a 16-month-old girl who presented with signs of increased intracranial pressure following a minor head trauma. The tumor was accompanied by communicating hydrocephalus, which, at surgery, was found not to obstruct the flow of cerebrospinal fluid (CSF). Electron microscopy revealed the fine structure of the tumor to be essentially that of normal choroid plexus. Although the tumor appeared to have the potential to be highly secretory, there was no evidence of hypersecretion of CSF in our ultrastructural study. Removal of the tumor alone was sufficient to reduce the hydrocephalus after some intervals.

Because the volume of the CPP was two to three times that of normal choroid plexus and the dilated ventricle did not decrease in size immediately after surgery, it appears that communicating hydrocephalus with CPP does not result solely from hypersecretion of CSF by tumor cells. Additional causes may be an increase in the total CSF volume as a function of the tumor bulk and malabsorption of CSF due to a high CSF protein content.

Key words: choroid plexus papilloma, hydrocephalus, electron microscopy, CSF secretion, computed tomography

Introduction

Papillomas of the choroid plexus are relatively uncommon, accounting for 3–5% of brain tumors in children,¹³⁾ and are frequently associated with communicating hydrocephalus.¹¹⁾

They often arise in the lateral ventricles in children and in the fourth ventricle in adults.¹⁾ It is generally assumed that the communicating type of hydrocephalus associated with choroid plexus papillomas (CPP) is caused by overproduction of cerebrospinal fluid (CSF)^{5,8,12,14,18)} or by leptomeningeal fibrosis secondary to hemorrhage from the tumor.^{15,19)} On the other hand, Gainer *et al.*⁶⁾ and Sahar *et al.*²⁰⁾ reported hydrocephalus caused by ventricular obstruction by tumor. In the patient we report here, a young child with a CPP of the right lateral ven-

tricle, removal of the tumor alone resulted in a slow reduction in the size of the ventricles. We describe the fine structure of this CPP and discuss its relationship with the communicating hydrocephalus.

Case Report

A 16-month-old girl was admitted to the Soujinkai Hospital on October 11, 1984, with a 2-day history of vomiting. She had had a cold, with typical symptoms, in the preceding week. Five days before admission she had fallen on a staircase a distance of approximately 1 meter. She had not lost consciousness and did not develop a subcutaneous hematoma. Two days later she developed irritability and vomiting.

The pregnancy and delivery had been normal, and the child's development had also been normal until shortly after the trauma, when the family noted the irritability and vomiting.

On admission she was irritable and vomited frequently. Physical examination disclosed no paresis. The head circumference was 48.6 cm. The anterior fontanel was bulging and tense. A skull x-ray showed an enlarged vault with separation of the coronal sutures. Emergency CT was performed. The precontrast CT scan showed symmetrically enlarged lateral, third, and fourth ventricles, poorly defined sulci over the convexity, and a predominantly isodense, partially hyperdense tumor located in the cella-media portion of the right lateral ventricle. There was no indication of intracranial hemorrhage. On postcontrast CT the large tumor was strongly enhanced and its surface appeared irregular (Fig. 1). Right axial arteriography performed 1 day after admission disclosed a large staining mass fed by a posterior choroidal artery. By this time the patient had become lethargic, and within 2 days of admission was bedridden and unable to eat by herself. Because of the communicating hydrocephalus and the deterioration of consciousness, external CSF drainage was instituted *via* the lumbar route. The CSF was cytologically normal. The protein content was 50 mg/100 ml,

the sugar content 60 mg/100 ml, and the chloride content 118 mEq/1000 ml. After CSF drainage her consciousness cleared, the fontanel returned to normal, and she was able to eat by herself. However, at 5 days after admission her consciousness again deteriorated and her fontanel was tense. Emergency surgery was performed on October 15, 1984.

A right frontoparietal craniotomy was performed and, because the dura mater was bulging and very tense, it was opened after drainage of 40 cc of CSF through a ventriculostomy. The cortical surface appeared normal, and a cortical incision was made from the posterior parietal gyrus to the lateral ventricle. Separation of the cortical lips exposed the ventricular wall at a depth of approximately 2.5 cm. The tumor, which was visible in the trigone of the lateral ventricle, was attached to the glomus choroideum. It was soft, pink and yellow-gray, and shaped like a cauliflower. Except for its pedicle, which was attached to the choroid plexus, the tumor was freely mobile and did not directly obstruct the foramen of Monro. It was successfully and entirely removed by cutting the pedicle after coagulation. The wound was closed

Fig. 1 Preoperative precontrast (*upper*) and postcontrast (*lower*) CT scans. Note the marked enlargement of the third and lateral ventricles and the presence of a mixed iso- and high-density mass with an irregular surface after contrast enhancement. Also note the free foramen of Monro.

in the usual manner.

For electron microscopic examination, the histological specimens were immediately fixed by overnight immersion in chilled 2% glutaraldehyde and postfixes with 1% OsO₄ for 60 minutes. The tissues were then dehydrated in a graded series of ethanol and embedded in Epon. Ultrathin sections were prepared with a LKB ultramicrotome and stained with both uranylacetate and lead citrate. The specimens were observed with a Hitachi 100CX-1 electron microscope.

Light microscopy disclosed a single layer of well-differentiated cuboidal and cylindrical cells arranged in papillary formation on a loose stroma containing a thin blood vessel wall. No evidence of anaplasia, mitosis, or necrosis was found.

Under electron microscopy the lining cells were seen to be arranged in an orderly fashion (Fig. 2 *left*). The cytoplasm contained many mitochondria, granular endoplasmic reticulum, Golgi apparatus, free ribosomes, occasional fine filaments and coated vesicles, large aggregates of dense bodies with granular and lamellar internal structures, and multivesicular bodies (Fig. 2 *left, center*). The apical portions of these cells were covered by numerous microvilli, cilia, and basal bodies (Fig. 2 *right*). Frequently, the granular endoplasmic reticulum was concentrated and arranged in a lamellar pattern at the apical surface of the tumor cell (Fig. 2 *center*). The

appositive lateral surface of the cells exhibited tight junctions characterized by the obliteration and sealing off of the interstitial space and a zonula adherens characterized by widened interstitial clefts in the apical regions (Fig. 2 *right*). Many interdigitations and intercellular clefts were observed in the lateral and basal surfaces of the cells. The intercellular space was prominently dilated in the lateral surface of the cells (Fig. 2 *left*), and their basal surfaces were covered by a layer of continuous basal lamina (Fig. 3 *left*). In cross sections of the basal portions, many coated vesicles were observed near the infolding of the plasma membrane in the cell cytoplasm (Fig. 3 *right*).

The capillaries had well developed fenestrations and many pinocytotic vesicles. The perivascular extracellular spaces were dilated and contained loose, collagen-rich connective tissue (Fig. 4).

In summary, both light and electron microscopy disclosed features of choroid plexus papilloma.

The patient improved markedly after surgery and was discharged in a cheerful, healthy state on October 31, 1984. A CT scan obtained 14 days postoperatively revealed a reduction in the size of the ventricles, especially that of the right lateral ventricle. No residual tumor was detected on pre- or post-contrast CT scans. Small subdural collections of CSF were seen in the frontal region of the operated site. Some sulci had appeared over the convexities (Fig.

Fig. 2 Electron micrograph of the choroid plexus papilloma demonstrating cuboidal cells containing lobulated and infolded nuclei. *left*: Numerous microvilli and mitochondria (Mt) as well as occasional cilia (Ci), are visible at the apical ends. The granular endoplasmic reticulum (GER) is in a lamellated and concentrated arrangement, and there are numerous infoldings (*arrow*) and dilations (*arrowheads*) between the lateral surfaces. Bar = 3 μ m. *center*: Characteristic lamellar organization of the granular endoplasmic reticulum in the apical ends. A desmosome (Ds) and an aggregation of glycogen (Gy) containing membrane-bound dense bodies with a granular and lamellar internal structure are also seen. Bar = 0.5 μ m. *right*: Cytoplasm of an epithelial cell with tight junctions (Tj) and zonula adherens (Za), multivesicular bodies (Mv), cilia with basal bodies (Bb), numerous crossed cilia (Ci), numerous free ribosomes, and coated vesicles (Cv) in the apical portions. Bar = 0.5 μ m.

Fig. 3 Numerous infoldings of the plasma membrane are observed in the basal ends. *left*: The basal surface is covered by a layer of continuous basement membrane (B) facing the perivascular extracellular space. Bar = 0.5 μ m. *right*: A cross section of the cytoplasm, which contains numerous infoldings in the basal ends. Note the relationship between the infoldings (arrow) and the coated vesicles (Cv). Bar = 0.5 μ m.

5 *left*). A lumbar puncture was performed 8 days after surgery because of slight fever and signs of meningeal irritation. The fluid was cytologically normal, with a protein content of 20 mg/100 ml, a sugar content of 45 mg/100 ml, and a chloride content of 121 mEq/1000 ml.

Follow-up CT scans performed at 6 and 13 months after operation demonstrated further reduction in the size of the ventricles and, at 13 months, of the lateral ventricle in particular, no subdural collection of CSF, and no evidence of recurrent tumor (Fig. 5 *center, right*).

Discussion

The cause of communicating hydrocephalus associated with CPP is controversial because there are no obvious ultrastructural and ultrachemical differences between CPP and normal human choroid plexus.^{4,7,16,17,21,23)}

From a pathophysiologic standpoint, it is possible that in our case the CPP, which contained many endothelial cells, produced a large volume of CSF by plasma filtration, which was discharged from the cytoplasm of the tumor cells into the ventricular lumen in a process resembling that of normal choroid plexus.^{2,3,12,17,18)} Although CPP can produce CSF in this manner, there was no evidence of hypersecretion of CSF in any tumor cells that we examined ultrastructurally. Therefore, we speculate that in our patient the communicating hydrocephalus

Fig. 4 Relationship between the basal end of the epithelial cell and the vascular endothelium. *upper*: Fenestration (F) of the endothelium of the capillary and the continuous basement membrane (B) of the subendothelial and subepithelial regions, and collagen and loose tissue in the perivascular space. Bar = 0.5 μ m. *lower*: Numerous pinocytotic vesicles (Pv) are seen in the endothelium of the capillary and numerous coated vesicles (Cv) in the epithelial cytoplasm of the basal ends. Bar = 0.5 μ m.

may have been caused by an increase in the CSF volume as a function of the large size of the tumor, which was two to three times as large as the normal choroid plexus.

On the other hand, ventricular obstruction by the tumor^{6,20)} and leptomeningeal fibrosis secondary to hemorrhage from the tumor¹⁵⁾ are other possible causes of the hydrocephalus. In our case, however,

Fig. 5 Precontrast CT scans 5 days (*left*), 6 months (*center*), and 13 months (*right*) after surgery. Note the slow reduction in the size of the lateral ventricles.

the tumor was completely free within the ventricle and leptomeningeal fibrosis was not observed at the corticotomy site; there was also no evidence of intracranial hemorrhage. Therefore, it is unlikely that the communicating hydrocephalus in our case was caused by leptomeningeal fibrosis secondary to hemorrhage.

The slow regression of the hydrocephalus, which Turcotte *et al.*²²⁾ also reported in a similar case, may be attributable to a persistent CSF absorption abnormality, which may have resolved as the dilated ventricle gradually decreased in size. The electron microscopic findings for a CSF secretory function, a feature that Masuzawa *et al.*¹²⁾ noted in a case of CPP, were also observed in our case. However, the fact that follow-up CT scans demonstrated a slow reduction in the size of the ventricles after surgery suggests to us that the hydrocephalus could not have been caused only by hypersecretion of CSF by the tumor cells. Kühne *et al.*¹⁰⁾ reported three patients with acoustic neurinoma associated with communicating hydrocephalus who had a high CSF protein content preoperatively, so it is possible that a high CSF protein content causes malabsorption of CSF.

With CT scanning, the diagnosis of CPP is not difficult. A strongly enhanced ventricular mass having an irregular surface^{9,20,22)} and accompanied by communicating hydrocephalus is diagnostic of CPP. Our observations and those of others suggest that communicating hydrocephalus with CPP results from both an increase in the total volume of CSF through its production by the tumor and, in cases of a high CSF protein content, a reversible dysfunction in the absorption of CSF. Therefore, the hydrocephalus resolves only gradually after removal of the tumor.

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