PAPILLOMA OF THE CHOROID PLEXUS OF THE LATERAL VENTRICLES CAUSING HYDROCEPHALUS IN AN INFANT

Bronson S. Ray, M.D., and Fremont C. Peck, Jr., M.D.

Department of Surgery, New York Hospital-Cornell Medical College, New York City

(Received for publication February 27, 1950)

Papillomas of the choroid plexus may cause hydrocephalus by excessive production of cerebrospinal fluid though impressive clinical evidence in support of this concept has been meager. A unique case is presented here in which bilateral papillomas of the choroid plexus of the lateral ventricles were found in association with communicating hydrocephalus caused by overproduction of cerebrospinal fluid. The sequence of events in the management of the condition leading up to its recognition took a devious course which provides evidence of a kind not previously reported.

CASE REPORT

A female child, aged 2½ months, was admitted to the New York Hospital on Feb. 15, 1954.

History. The baby was born spontaneously after a full term normal pregnancy at another hospital on Nov. 29, 1953, and weighed 7 lbs. 10 oz. She was thought to have a somewhat enlarged head at the time of birth. Several cyanotic episodes occurred during the first few weeks of life, but otherwise she fed and cried normally. At 1 month of age the head measured 41 cm. in circumference and the chest 39.5 cm.

Examination. She was a well-nourished infant with a vigorous cry. There was obvious hydrocephalus. She could not hold up her head, which measured 45.5 cm. in circumference and showed prominent parietal bossing. The anterior fontanelle measured 6 × 7 cm. and was tense and bulging. There were cracked-pot resonance to percussion, convergent strabismus and horizontal nystagmus.

Subdural taps were negative. On ventricular tap clear, colorless fluid was obtained. Phenolsulfonphthalein injected in a lateral ventricle was promptly recovered from the opposite lateral ventricle and the lumbar subarachnoid space. Approximately 60 cc. of fluid were removed and replaced with air. Roentgenograms disclosed dilated ventricles and a cortical thickness of 3 cm. The protein content of the ventricular fluid was 13 mg. per cent.

The condition was regarded as a communicating hydrocephalus. Pyelography demonstrated the presence of two normally functioning kidneys.

1st Operation, Feb. 20, 1954. A right nephrectomy was performed, followed by a lumbar subarachnoid-ureteral shunt with a polyethylene tube.

Course. The baby tolerated the procedure well, but on the 3rd postoperative day she rapidly became dehydrated and went into shock. An immediate forced infusion of saline corrected the state, but over the next few days it became apparent that she was losing an excessive amount of electrolytes and water via the ureteral shunt. The addition of salt to the diet, plus the administration of large quantities of intravenous fluids, was necessary to prevent dehydration. Daily infusions of 500 to 1000 cc. of half normal saline were administered together with oral feedings. Each time an attempt was made to discontinue intravenous clyses or when they were given too slowly serious dehydration often bordering on shock occurred. In all, about ten such episodes took place in a period of 2 months. Chemical determinations during these episodes generally showed moderately low serum CO₂, normal chloride, slightly low to normal sodium and high potassium. All attempts to maintain adequate hydration by means other than daily infusions containing large quantities of salt met with failure. After 8 weeks of this therapy it was evident that no adjustment to the continued excessive fluid loss had occurred.
2nd Operation, April 20, 1954. The subarachnoid-ureteral shunt was transplanted to the peritoneal cavity in the hope that the fluid would be re-absorbed.

Course. The day after this procedure enormous abdominal distension occurred accompanied by vomiting and again the startling picture of dehydration. Over the next week, however, the clinical condition improved. The abdominal distension decreased somewhat, the appetite increased to a point far beyond what it had been and the anterior fontanelle remained soft. The baby was soon able to get along on oral feedings alone. There was no accurate estimation of fluid output during this period except that in the nurses' records it is noted that daily diaper changes averaged 10 per day while the ureteral shunt was functioning, and only 6 per day after it was placed in the peritoneal cavity.

The child was discharged 10 weeks after admission.

2nd Admission. Six weeks later the patient again entered the hospital because the polyethylene tube had become plugged with resulting sudden increase in intracranial pressure and size of head.

3rd Operation, June 11, 1954. A new tube was promptly inserted and a satisfactory recovery from the procedure took place.

3rd Admission. On Nov. 18, 1954, at the age of 1 year, the baby was readmitted because of slowly progressive abdominal enlargement. This condition had persisted since the last discharge and had reached the point where ascites had become a more critical problem than the hydrocephalus (Fig. 1). The head now measured 48 cm. and the abdomen 67 cm. in circumference.

Course. An abdominal paracentesis was performed to relieve the child's respiratory distress and improve her appetite; 3250 cc. of clear colorless fluid with a chloride content of 108 mg. per cent were obtained. On the day after the paracentesis another episode of shock from dehydration appeared and was corrected over a 24-hour period with 2100 cc. of parenteral fluids besides regular oral feedings. The child gained 1600 gm. in this period and the abdomen quickly refilled despite a tight abdominal binder.

When her condition had once again become stabilized after the paracentesis it was thought that the only therapeutic measure left was cauterization of the choroid plexus.

4th Operation, Oct. 29, 1954. A small right parieto-occipital craniotomy was performed and the right lateral ventricle was opened. In the region of the glomus of the choroid plexus was a multicystic globular mass. Implanted in numerous places in the walls of the cysts were pink buds of choroidal tissue. The cystic fluid was thin, clear and colorless. The portions of the choroid plexus that extended anteriorly in the temporal horn and body of the ventricle were large meaty looking processes having only occasional small cystic elements. That part of the choroid plexus at the margin of the foramen of Monro appeared normal. The entire tumor was removed, and measured 7 cm. in length by 3 cm. in its widest diameter (Fig. 2).
Course. Except for a persistent collection of fluid under the scalp the baby made a satisfactory recovery from the operation. There was no appreciable change in the degree of ascites, but some degree of overhydration occurred when parenteral fluids were given in amounts similar to those used before the operation.

5th Operation. On the 12th day after removal of the right ventricular tumor a similar surgical procedure was performed on the left side. A multicystic tumor of the choroid plexus similar to that in the opposite ventricle was excised. Just before closure of the wound, however, the child went into shock and expired on the operating table despite resuscitative measures.

Pathological Examination. Both papillomas (Figs. 3 and 4) revealed multicystic and papillary proliferation with no evidence of malignancy. There were masses of villous and frond-like structures. These usually possessed a stalk composed of a few strands of connective tissue and contained an engorged blood vessel lined by endothelium. The papillary processes were covered by acidophilic cuboidal cells containing rounded dark- and light-staining nuclei. There were no mitotic figures or pleomorphism. The cysts were lined by a single layer of cuboidal cells similar to but smaller than those in the villi.

Autopsy. There was a massive subarachnoid hemorrhagic clot extending over the basilar and temporal areas of the brain. This was the apparent cause of death. There was no blood in the ventricles and the source of the bleeding could not be determined. The choroid plexuses were absent in the lateral ventricles; those that remained were normal in appearance. The for-
amina of Monro, the aqueduct of Sylvius and the foramina of Luschka and Magendie were patent. The ependyma and basilar meninges were normal grossly and microscopically except for a slight degree of ependymitis at the juncture of the aqueduct and 4th ventricle.

The abdomen was greatly distended and the peritoneal cavity contained about 3,000 cc. of clear faintly yellow fluid. The polyethylene tube leading from the spinal subarachnoid space lay free in the peritoneal cavity, and was patent. The peritoneal surfaces were smooth and glistening, and the intestines were normal in appearance and position. The liver, spleen, biliary system, pancreas, kidney, adrenals, pelvic viscera and blood vessels appeared normal. The liver weighed 280 gm, and the spleen 26 gm. which are average for this age; they were of normal consistency and appearance on cut sections. Microscopic sections of the various abdominal organs showed essentially normal tissues. In brief there was no intra-abdominal disease to account for the large collection of fluid in the peritoneal cavity.

**DISCUSSION**

In the neighborhood of 100 papillomas (and angiomas) of the choroid plexuses have been reported in the literature. At least 10 per cent occurred in infants in the first year of life\(^2\) and the majority were accompanied by hydrocephalus. It is commonly accepted that cerebrospinal fluid is formed by the choroid plexuses and a priori reasoning readily leads to the conclusion that a papilloma of the structure, which in most cases is nothing more than a benign overgrowth, produces hydrocephalus by oversecretion. However, papillomas in the 4th ventricle may produce a de-
gree of ventricular obstruction which has made conclusions tenuous regarding the origin of hydrocephalus in these cases. In the cases of papillomas in the lateral ventricles Dorothy Russell\(^1\) appropriately raised a question of the possibility of accompanying ependymitis or basal meningitis as a cause of the hydrocephalus. When it is realized that intraventricular hemorrhage, xanthochromia or elevation of the protein content of ventricular fluid have been frequent findings in the reported cases it is to be expected that at least microscopic evidence of reaction in the ependyma and leptomeninges should be common.

More conclusive proof that papillomas of the choroid plexus may cause hydrocephalus has been dependent on actually determining an excessive production of cerebrospinal fluid or by demonstrating a cure of hydrocephalus by surgical excision of the abnormal plexuses.

In Fehr's\(^1\) 23-year-old patient with a papilloma of the choroid plexus in the 4th ventricle the removal of 200 to 300 cc. of spinal fluid daily failed to produce more than transient diminution in the increased cerebrospinal fluid pressure. Kahn and Luros,\(^3\) and Matson\(^4\) each reported subsidence of increased cerebrospinal fluid pressures and apparent cure after excision of papillomas in the lateral ventricles (Kahn and Luros’ patient was 20 years old and Matson’s was 4 weeks old). McGuire, Greenwood and Newton\(^4\) reported an apparent cure of hydrocephalus in a 3-month-old infant by excision of a bilateral angioma of the choroid plexus which in its microscopic description is not unlike what others have called papilloma.

In the case reported here the unfortunate operative death prevented concluding that the hydrocephalus was cured by excision of the tumors though it seems likely that such would have been the case had the child survived. However, the overproduction of cerebrospinal fluid is demonstrated beyond doubt, first, by the excessive loss of fluid through the ureteral shunt and second, by the development of massive ascites when the spinal subarachnoid shunt was transferred to the peritoneal cavity.

The failure to recognize the tumors in the beginning resulted from the use of an inadequate amount of air in performing the ventriculography. Also, the normal ventricular fluid gave no clue to the presence of the lesions. The failure to recognize earlier the significance of the excessive loss of spinal fluid through the ureter and into the peritoneal cavity is explained by the lack of previous experience of this kind.

**SUMMARY AND CONCLUSIONS**

This report presents the evidence for concluding that hydrocephalus resulted from the presence of benign papillomas of the choroid plexus in the lateral ventricles of an infant.

In the belief that the condition was a communicating hydrocephalus a spinal subarachnoid-ureteral shunt and later a peritoneal shunt was performed, each of which resulted in excessive loss of cerebrospinal fluid.

The lesson to be drawn is that it must be kept in mind that occasionally a papilloma (or an angioma) of the choroid plexus may be the cause of hydrocephalus. Suspicion of presence of the lesion should be raised by the finding of xanthochromia or excessive protein in the ventricular fluid, though these changes are not constant. Excessive loss of fluid through a ureteral shunt or the development of ascites with a peritoneal shunt should strongly suggest the presence of a tumor of the choroid plexus.
REFERENCES