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

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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. Phenol-sulfonphthalein 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).