Cerebrospinal fluid overproduction and hydrocephalus associated with choroid plexus papilloma

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With the use of a ventricular perfusion technique, a cerebrospinal fluid formation rate of 1.4 ml/min was determined in a 5-month-old child with a choroid plexus papilloma and hydrocephalus. This rate was slightly greater than four times the rate observed in normal children. The clinical course, as well as a measured absorption rate, indicated that the patient's capacity for absorption was normal.

Key Words • choroid plexus papilloma • hydrocephalus • ventricular perfusion • cerebrospinal fluid overproduction

It is generally assumed that choroid plexus papillomas are associated with overproduction of cerebrospinal fluid (CSF). Documentation of this overproduction has depended on continuous drainage to measure CSF formation. Rates determined by this method, however, may be in error since during the collection period the amount of fluid absorbed is unknown and the CSF volume might not remain constant. In addition, whether the diffuse ventricular dilatation that is frequently associated with these tumors can occur in the face of a normal capacity for absorption has not been definitely established.

Using the ventricular perfusion technique, we studied a patient with a choroid plexus papilloma of a lateral ventricle.

Case Report

A 5-month-old boy entered the hospital because of an enlarging head. One week before admission he had developed vomiting, irritability, and a squint. The prenatal and natal history were normal. At 12 weeks of age the head circumference measured 43 cm (97 percentile). Growth and development were otherwise appropriate for his age.

When the child was admitted to the hospital, the head circumference was 46.5 cm. Other abnormal physical findings were: frontal bossing, a hyperresonant skull percussion note, a full anterior fontanel, and a limitation of abduction of both eyes. Bilateral subdural punctures were negative. Ventriculography demonstrated ventricular dilatation with communication to the spinal subarachnoid space and basal cisterns. The left ventricle was larger than the right, without shift of midline structures. No mass was seen within the ventricles, but the trigonal...
regions were not outlined. A small amount of gas had been injected into the subarachnoid space at the convexity before ventricular filling. This space was excessive (Fig. 1). The ventricular fluid was clear and colorless with a total protein content less than 10 mg%, a sugar content of 50 mg%, and 27 red blood cells and 2 lymphocytes. Following this study, the patient developed a massive subgaleal effusion with chemosis, repeated vomiting, and progressive lethargy.

A ventriculoperitoneal shunt with a low pressure Holter valve was inserted. The effusion cleared, but two days after operation a CSF leak developed at the abdominal incision. After additional sutures were placed to stop this leak, the anterior fontanel became more tense. Ventricular pressure measured through the shunt reservoir was 180 mm Hg. Inability to control the hydrocephalus led to a shunt revision 1 week after the initial procedure.

At operation the shunt was found to be mechanically functional, but large amounts of fluid could be expressed from the peritoneal tubing by compressing the abdomen. The distal end of the shunt was converted to an atrial position. A choroid plexus papilloma was suspected, and a selective cerebral arteriogram was done. A vascularized mass was demonstrated in the region of the left trigone supplied by the posterior and anterior choroidal arteries (Fig. 2). A \textsuperscript{99m}Tc brain scan without potassium perchlorate pretreatment showed a well-defined area of increased activity in the left parietal region. A ventriculolumbar perfusion was done. The methods and results of this study are described below.

After the perfusion study the tumor was excised through a high left parietal cortical incision. It was contiguous with normal choroid plexus and extended from the trigone to the tip of the frontal horn of the left lateral ventricle. Histological examination of the specimen revealed a choroid plexus papilloma (Fig. 3). After the operation there was a right hemiparesis which began to clear by the second week. The shunt was ligated and divided 12 days after operation, and at the time of discharge 1 week later there were no signs of increased intracranial pressure. Six months after removal of the tumor the hemiparesis had almost completely resolved. The child could hold objects with either hand, stand, and walk with support. There were no signs of
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increased intracranial pressure; the head circumference was 47.8 cm (50th percentile).

Perfusion Study

Ventriculolumbar perfusions have been used to determine the rate of CSF formation and absorption in patients with unobstructed CSF pathways\(^2,13\) and in patients with hydrocephalus.\(^8\) The study carried out in this patient employed the same methods with some slight modifications.

The patient was premedicated with Demerol Compound CHMC* supplemented during the course of the study to maintain continuous mild sedation; he slept during most of the study. Under local anesthesia, the shunt was ligated distal to the valve through a small scalp incision. With the patient lying on his side, artificial CSF (Elliot's B solution)‡ containing 10 \(\mu\)Ci of \(^{125}\)I human serum albumin‡ per liter was infused by a constant infusion pump into the right lateral ventricle through a No. 21 gauge needle placed percutaneously into a Rickham shunt reservoir. Fluid was collected from the lumbar subarachnoid space through a No. 21 gauge spinal needle. The perfusion pressure relative to the patient’s midline was determined by the height of the orifice of the outflow tubing. Inflow and outflow rates were determined by weighing aliquots of the respective fluids. Ventricular and lumbar pressures were monitored continuously with pressure transducers. The concentration of \(^{125}\)I albumin was determined in a well-type scintillation counter at 35% efficiency.

An initial ventricular pressure measurement, made approximately 90 minutes after shunt occlusion, was 410 mm H\(_2\)O. Before starting the perfusion, the ventricular pressure was lowered to 130 mm by draining CHF from the lumbar subarachnoid space. The rates of formation and absorption and the volume of distribution of \(^{125}\)I albumin were calculated from equations derived by Heisey, et al.\(^4\)

The concentration of \(^{125}\)I albumin in the lumbar effluent as a function of time is shown in Fig. 4. It was apparent that as the endogenous CSF was replaced by the artificial fluid, the concentration of \(^{125}\)I albumin gradually increased. At steady state, the concentration of \(^{125}\)I albumin in the effluent was about 50% that of the inflow. The rate of formation was calculated to be 1.43 ml/min while the rate of absorption was 0.59 ml/min. The volume of distribution was 66.2 ml, suggesting that a reduction in ventricular volume had occurred.

*Demerol Compound CHMC manufactured by Children's Hospital Medical Center, Boston, Massachusetts. (Not commercially available; it contains 6.25 mg phenergen, 6.25 mg thorazine, 25 mg demerol per cc).

‡Artificial CSF (Elliot's B solution) made by Abbott Laboratories, North Chicago, Illinois.

†\(^{125}\)I human serum albumin made by Mallinckrodt Chemical Works, St. Louis, Missouri.
FIG. 4. $I^{125}$ albumin activity in the lumbar effluent ($C_e$) and the inflow ($C_i$) per unit volume and time plotted as a function of time; CPM = counts per minute.

Discussion

Reports of CSF overproduction in patients with verified choroid plexus papillomas have to date relied on either nonquantitative clinical evidence or rates determined by the collection of ventricular fluid by drainage. Ray and Peck\(^1\) reported a case of an infant with initially unrecognized choroid plexus papillomas of both lateral ventricles who became severely dehydrated after insertion of a lumboureteral shunt and developed massive ascites following revision of a peritoneal shunt. Vigouroux\(^1\) estimated a nasal outflow of 800 ml/day in a patient with a fourth ventricular plexus papilloma and CSF rhinorrhea. Johnson\(^2\) reported a rate of 45 ml/hour of ventricular drainage, at a pressure of 450 mm H$_{2}$O in a patient whose tumor was also in the fourth ventricle. Using the same method, Fairburn\(^3\) collected 500, 400, and 960 ml/day on three consecutive days at a pressure of 50 mm H$_{2}$O in a dehydrated infant with a lateral ventricular papilloma. Although these reports infer overproduction of CSF associated with choroid plexus papillomas, the CSF production does not appear much greater than that measured in a group of patients with unobstructed CSF pathways.

The measured rate of absorption in our case, 0.59 ml/min at 130 mm H$_{2}$O, was nearly equal to the mean absorption rate, 0.61 ml/min at the same pressure, in children with unobstructed CSF pathways\(^2\) and was considerably greater than that reported for children with communicating hydrocephalus.\(^8\) The clinical course in this patient also implied a normal capacity for absorption.

References

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This study was supported in part by National Institutes of Health Grants NS 05172; NS-HD 09704, and the Children's Hospital Medical Center Mental Retardation and Human Developmental Research Program Grant HD 03773.

This paper was presented at the annual meeting of the American Association of Neurological Surgeons, Los Angeles, California, April, 1973.

Dr. Eisenberg is the recipient of Special Fellowship 1 F10 NS 2340-01 NSRB, and Dr. Lorenzo the recipient of Career Development Award HD 18519.

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