Isolated fourth ventricle as a complication of ventricular shunting

Report of three cases

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Signs of cerebellar dysfunction combined with signs suggestive of shunt malfunction developed in three children with obstructive hydrocephalus. Shunt function was normal. In all cases, the cerebellar signs persisted and computerized tomography scans revealed enlargement of the fourth ventricle. Shunting of the fourth ventricle returned the patients to normal function.

KEY WORDS □ isolated fourth ventricle □ hydrocephalus □ ventriculoperitoneal shunt □ computerized tomography

Scans obtained by computerized tomography (CT) not only facilitate the diagnosis of lesions previously demonstrable by conventional contrast studies but also may show lesions that conventional studies would fail to reveal.

Three patients with obstructive hydrocephalus treated by cerebrospinal fluid (CSF) bypass systems presented for evaluation of symptoms suggestive of shunt malfunction. On examination, each patient had cerebellar signs but no signs of raised intracranial pressure (ICP). Plain films were taken to ascertain proper anatomical location of the proximal and distal ends of the shunt and its continuity. Shunt scans, when performed, showed good CSF flow. Computerized tomography scans were performed to exclude a posterior fossa lesion or subdural hematoma and to confirm the ventricular size. Much to our surprise, they showed an isolated and dilated fourth ventricle.

Case Reports

Case 1

This premature baby girl was transferred to our hospital shortly after birth on August 6, 1972, because of respiratory distress. Septicemia (E. coli) was present, and the child showed evidence of intraventricular hemorrhage. Her head enlarged rapidly, and subsequent ventriculography revealed complete obstruction of the outlets of the fourth ventricle and enlargement of the entire ventricular system. A left ventriculoperitoneal (VP) shunt was placed on August 24, 1972. Seven revisions were required during the next 4 years. The patient's condition was complicated by a development of a seizure disorder.

The patient was readmitted in December, 1976, because of irritability, intermittent headache and vomiting, severe ataxia, and increasing frequency of the seizures. A CT scan
Fourth ventricle enlargement after shunting

revealed tremendous enlargement of the fourth ventricle (Fig. 1 left). At operation on January 5, 1977, a ventricular catheter was inserted into the fourth ventricle and connected with a Y connector to the left VP shunt. A CT scan taken 3 weeks later showed diminution in size of the fourth ventricle (Fig. 1 right). Since her fourth ventricle has been shunted, the child has had no symptoms and her seizures have been well controlled with medication; she still needs assistance to walk.

Case 2

This baby boy was treated for sepsis and hyperbilirubinemia; he was then transferred to our hospital in October, 1971, at the age of 6 weeks, because his head was enlarging rapidly. A ventriculogram revealed dilation of the entire ventricular system, with obstruction of the foramina of Magendie and Luschka. A right ventriculoatrial (VA) shunt was performed. Three revisions were carried out during the next 3 years.

In March, 1977, the boy was readmitted here because of intermittent headaches, nausea, and lack of coordination. Radio-nuclide shunt scan showed that the shunt was functioning well, but CT scan revealed dilation of the fourth ventricle, with small lateral ventricles. On July 27, 1977, a catheter was inserted into the encysted fourth ventricle and connected to the original bypass system. The child is now making good progress; he has no headaches and his coordination is normal.

Case 3

This infant was born prematurely in July, 1974; he required treatment for a moderately severe respiratory distress syndrome, associated with cerebral anoxia and hyperbilirubinemia. Intraventricular hemorrhage was documented. His head circumference increased precipitously during the sixth week of life, and a ventriculogram (Fig. 2 left) demonstrated dilation of the entire ventricular system with obstruction at the outlets of the fourth ventricle. A left VP shunt was inserted in August, 1974.

Malfunction of the child's shunt first became apparent when he was aged 2½ years. He was readmitted in May, 1977, 2 months after his second revision, with recurrence of vomiting and lethargy, and ataxia so severe that he could no longer sit up unaided. A CT scan (Fig. 2 right) revealed considerable dilation of the fourth ventricle and small lateral ventricles. On May 13, 1977, a ventricular catheter was inserted into the fourth ventricle and was joined with a Y connector to the ex-
Fig. 2. Case 3. Ventriculogram performed in August, 1974, lateral view, showing the patent aqueduct with dilation of the occipital horns of the lateral ventricles and the dilated fourth ventricle. Right: Computerized tomography scan, May, 1977, revealing marked enlargement of the fourth ventricle with small lateral ventricles.

isting shunt. Postoperatively, the ataxia resolved promptly and the child could walk by the eighth postoperative day. Six months later there were no indications of cerebellar dysfunction.

Discussion

These cases are presented to bring attention to yet another complication of shunting of the lateral ventricles. Ataxia suggested a lesion in the posterior fossa, and CT scan readily identified the isolated fourth ventricle. The prompt clinical improvement in these cases has alerted us to the desirability of a CT scan whenever these clinical signs and symptoms develop in patients who have a shunt for relief of hydrocephalus.

Blockage of a shunt is usually associated with signs and symptoms of increased intracranial pressure. The positive physical findings in these patients were limited to cerebellar dysfunction, but initially the full significance of these cerebellar signs was not appreciated. The clinical picture was suggestive of a progressive problem. In Case 3, for example, the patient deteriorated from mild ataxia until he was unable to sit. The decrease in the size of the fourth ventricle as symptoms abated further supports the proposition that encysting of the fourth ventricle was the etiology of our patients' problems.

Recognizing that the isolated fourth ventricle can cause symptoms and signs of cerebellar malfunction, we now perform a CT scan when confronted with this possible diagnosis. In our first two cases, CT scans were obtained to evaluate ventricular size and to rule out subdural collections of fluid or posterior fossa lesions. Results showing small or slit-like lateral ventricles and an enlarged fourth ventricle led to the diagnosis of an isolated fourth ventricle. In the third case, with our prior experience, the diagnosis of the encysted fourth ventricle was made before confirmation by CT scan.

Initially, all three children had obstruction of the fourth ventricle outlets secondary to intraventricular hemorrhage. Foltz and Shurtleff reported spontaneous occlusion of the aqueduct of Sylvius after insertion of a ventricular shunt. The initial ventriculograms, coupled with Foltz's work, led us to surmise that the fourth ventricle was isolated due to an inflammatory aqueduct stenosis consequent on the presence of a foreign body (the shunt tube) in the lateral ventricle. The supposition is further supported by the description of Raimondi, et al., of two patients with Dandy-Walker cysts in whom
cerebellar signs and symptoms developed after shunting of the lateral ventricle only. In these latter cases, the children's condition improved after the posterior fossa cyst had been shunted, leading the authors to advocate dual shunting as the initial treatment for this condition. Carmel, et al., reported similar complications, but they advocated dual shunting only when "a lack of adequate communication between the dilated fourth ventricle and lateral ventricle system was suspected." We certainly would not advocate fourth ventricle shunting nor dual shunting as the initial treatment of these patients since they have a much smaller fourth ventricle initially than do patients with the Dandy-Walker malformation and all had a patent aqueduct before shunt insertion.

 Epstein et al., have demonstrated the efficacy of the CT scan in the follow-up study of patients with treated hydrocephalus. The entity we have described would be difficult, if not impossible, to demonstrate by conventional neuroradiological techniques. The CT scans in these patients easily confirmed the diagnosis of an isolated fourth ventricle and consequently enabled prompt and proper treatment.

References

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