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Abstract

Lateral ventricular shunting alone is often considered to be adequate treatment for hydrocephalus caused by Dandy-Walker syndrome. A patient is presented whom progressive spastic tetraparesis and signs of severe brainstem compression developed due to an entrapped posterior fossa cysts, in spite of an adequately functioning lateral ventricular shunt. Addition of a cystoperitoneal shunt resulted in rapid resolution of symptoms and deficits. This case illustrates that potentially fatal brainstem compression and dangerous posterior fossa hypertension may develop if the posterior fossa cyst does not communicate with the lateral ventricles, where the shunt is placed.

Introduction

Dilated 4th ventricle communicates with the lateral and 3rd ventricle in most of the cases of Dandy-Walker syndrome. Ventriculoperitoneal (VP) shunt, therefore, is an adequate treatment for majority of these cases with hydrocephalus. However, lack of this communication in a minority of patients, may lead to failures of VP shunting, sometimes with serious sequelae.

Case Report

This 4 years old child was admitted with a history of increasing occipital headaches, inability to stand or walk and slurred speech for many months. During the week prior to admission, the mother noticed increasing stiffness of limbs, squint and tendency to sleep all the time. Past history revealed that the child had a VP shunt for hydrocephalus during infancy. Examination revealed a drowsy child with bilateral 6th nerve paresis and nystagmus on looking laterally. There was no papilloedema. Spastic tetraparesis with more marked upper motor neuron signs in the way bilaterally upgoing plantars, clonus and exaggerated tendon jerks were noticed in the lower limbs. Local examination of head revealed a prominent occipital region and a functioning shunt (confirmed later by investigative procedures). Computed tomographic (CT) scan along with metrizamide ventriculography (Figures 1a and b)
showed no communication of the ventricles with the posterior fossa cyst. A second shunt (cystoperitoneal) was added to the previously present and functioning VP shunt. Operative findings included: tense posterior fossa dura and markedly raised pressure of fluid in the cyst. Composition of the cyst fluid (including the protein content) was found to be similar to that of normal ventricular cerebrospinal fluid. Postoperatively, there was steady improvement in the child’s neurological status.
Child became more alert with resolution of limb spasticity and upper motor neuron signs during the first week and could walk with help one month after the insertion of the second shunt. Postoperative CT scan (Figures 2a & b) showed reduction in the size of the cyst and an increase in the size of cerebellum and brainstem suggesting decompression of these structures.
**Discussion**

Intelligent and successful treatment of hydrocephalus depends upon an accurate knowledge of the location and type of obstruction to normal cerebrospinal fluid movements\(^2\). In hydrocephalus associated with the Dandy-Walker syndrome, the aqueduct was patent in 87 percent of the reported cases as determined by postmortem examination, intra-operative visualization or investigative procedures\(^1\). In a much smaller number, the aqueduct may be anatomically closed or functionally occluded by compression from the herniated superior vermis or from extension of the cyst into the incisura\(^3\).

Ventricular shunting is the preferred primary treatment for this condition\(^3,4\). The question of the patency of the aqueduct must be answered prior to placement of a shunt. A reliable method for demonstrating the functional patency of the aqueduct is to inject a small amount of a dilute water-soluble contrast agent into the lateral ventricle and follow with CT scanning\(^5\) (Figure la & b). If the lateral, the 3rd and the dilated 4th ventricles communicate, then a single VP shunt is placed. If there is an anatomical or functional block of the aqueduct, then lateral ventricular shunt alone may be inadequate and hazardous\(^1\). Potentially fatal brainstem compression or symptoms suggestive of dangerously high pressure in the posterior fossa may be caused by the entrapped Dandy-Walker cyst; as happened in our patient. Not all authors have experienced this complication, nor do they agree that ventricular shunting alone can lead to disaster, even in the presence of aqueductal occlusion\(^3,6\). If ventriculography shows an occluded aqueduct (Figure la & b), a double shunt from the lateral ventricle and the cyst should be used to avoid complications from an entrapped posterior fossa cyst.

**References**