Isolated Posterior Fossa Hypertension and Brainstem Compression Caused by Entrapped Dandy-Walker Cyst: A Case Report

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Abstract

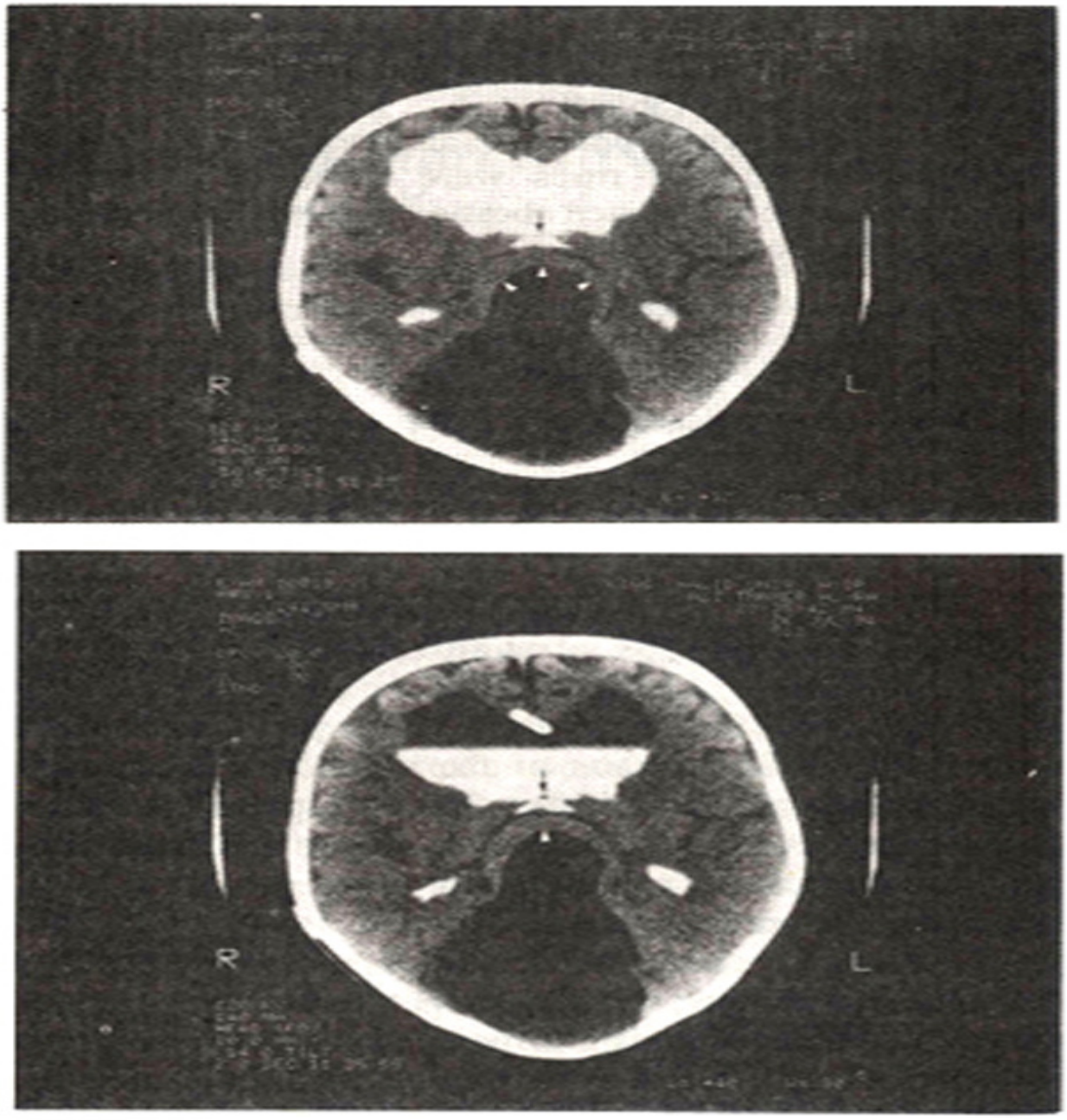
Lateral ventricular shuntmg alone is often considered to be adequate treatment for hydrocephalus caused by Dandy-Walker syndnme. A patient is presented mwhom progressive spastic tetraparesis and signs of severe brainstem compression developed due to an entrapped posteriorfossa cysts, in spite of an adequately 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 hypeitension 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-Walkersyndrom1. Ventriculoperitoneal (VP) shunt, therefore, is an adequate treatment for majority of these cases with hydrocephalus. However, lack of this communication ma minority of patients, may lead to failures of VP shunting, sometimes with serious sequelae.

Case Report

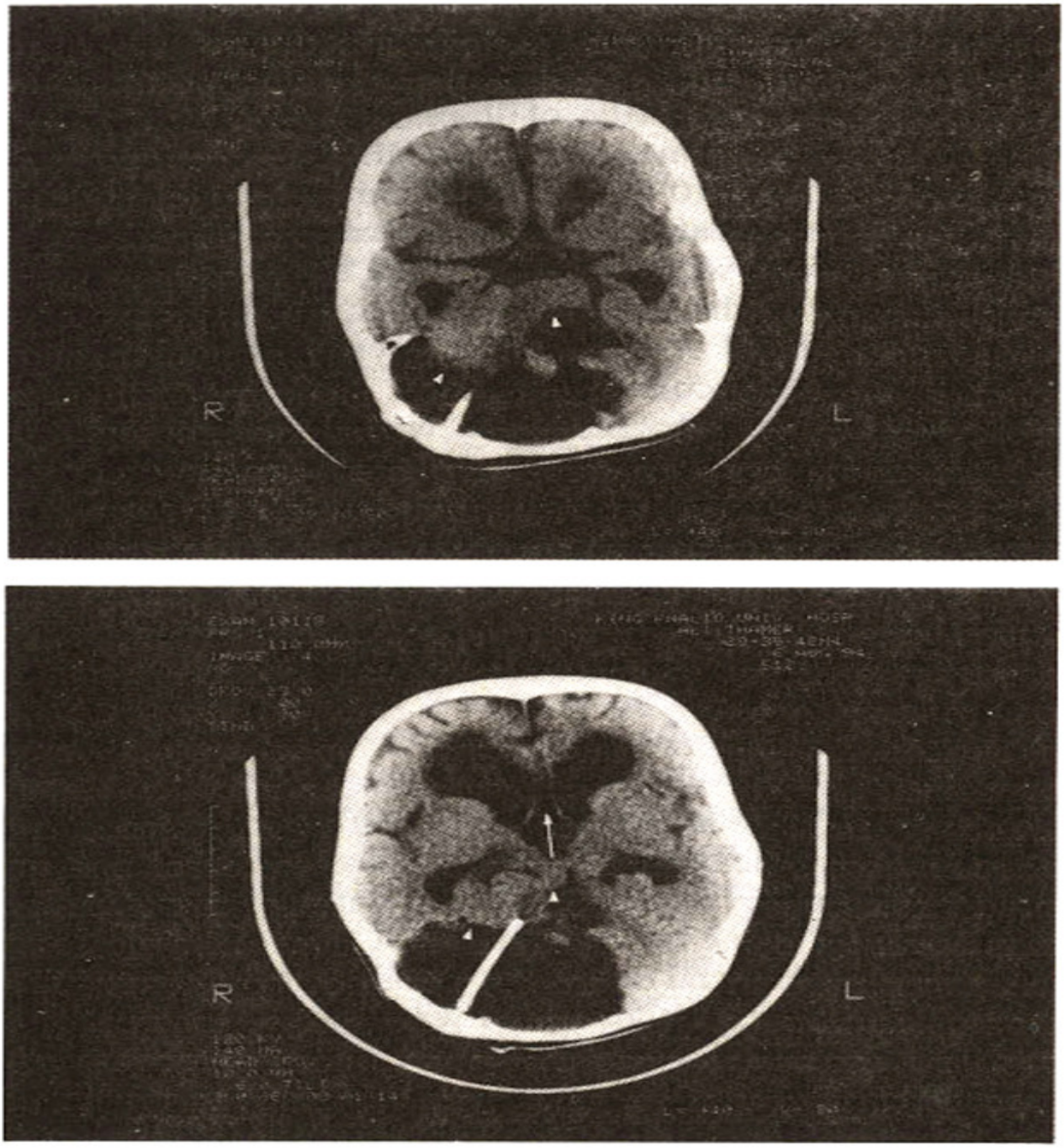
This 4 years old child was admitted with a history of fncreasing occipital headaches, inability to stand or walk and slurred speech for many months. During the week prior to admission, the mother noticed increasing stiffness of limbs, squint and tendency to sleep all the time. Past history revealed that the child hadaVP 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 tendonjerks were noticed in the lower limbs. Local examination of head revealed a prominent occipital region and a functioning shunt (confinned later by investigative procedures). Computed tomographic (CT) scan alongwith metrizamide ventriculography (Figures 1a and b)



Figures 1 (a) and 1 (b). CT - ventriculography early (a) and delayed (b) films showing opacification of the lateral and third ventricle but not the dilated 4th ventricle, indicating obstruction at the level of the aqueduct. Note: (1) asymmetrical hypoplasia of both cerebellar hemispheres; (2) marked compression and flattening of the brainstem in the anteroposterior diameter (arrowhead) by the entrapped posterior fossa cysts and (3) the 3rd ventricle (arrow) that is compressed and displaced from behind and reduced to a slit in coronal (side-to-side) plane.

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 spasticily 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)



Figures 2 (a) and 2 (b). Postoperative CT scans after cystoperitoneal shunt showing an increase in the size of cerebellum and brainstem (arrowheads), reduction in the size of the ventricular cyst and restoration of the normal antero-posterior disposition of the 3rd ventricle (arrow).

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 nonnal cerebrospinal fluid movements[2]. In hydrocephalus associated with the Dandy-Walker syndrome, the aqueduct was patent in 87 percent of the reported cases as deterniinedby postmortem examination, intra-operative visualization or investigative procedures[1]. In a much smaller number, the aqueduct may be anatomically closed orfunctionally occluded by compression from the herniated superior vermis or fmm extension of the cyst into the incisura[3]. Ventricular shunting is the preferred primary treatment forthis condition[3,4]. The question of the patency of the aqueduct must be answered pnor placement of a shunt. A reliable method for demonstrating the functional patency of the aqueduct is to inject a small amount bf 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 fmm the lateral ventricle and the cyst should be used to avoid complications from an entrapped posterior fossa cyst.

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