

Occult Cerebrospinal Fluid Fistula between Ventricle and Extra-Ventricular Position of the Ventriculoperitoneal Shunt Tip

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Abstract-

Purpose: Ventriculoperitoneal (VP) shunt or ventriculoatrial shunt is a common operation for treatment of hydrocephalus. Usually, shunt series (plain radiographs of the skull, neck, chest and abdomen) and brain computed tomography (CT) are used to monitor the status of hydrocephalus and VP shunt. However, does the result of a brain CT really reflect the status of the hydrocephalus and shunt function? In patients with VP shunt, brain CT image only may lead to misdiagnosis of the status of the hydrocephalus and result in inadequate treatment plan.

Case Report: The authors reported a 6-year-old patient with occult cerebrospinal fluid (CSF) fistula between ventricle and extra-ventricular position of the VP shunt tip on CT scan and resulted in inappropriate shunt removal.

Conclusion: The patient was diagnosed to have shunt-dependent hydrocephalus with inadequate shunt removal. In this kind of patient, further studies may reduce unnecessary morbidity or mortality.

Key Words: hydrocephalus, cerebrospinal fluid fistula, ventriculoperitoneal shunt malfunction

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INTRODUCTION

Cerebrospinal fluid (CSF) diversion as ventriculoperitoneal (VP) shunt is a common treatment for hydrocephalus. Malfunction of the VP shunt may result in severe mortality or morbidity at very short intervals. Thus, regular follow-up after shunt surgery is required for these kinds of patients. History taking, physical examination, and diagnostic neuroimaging, including shunt series (plain radiographs of the skull, neck, chest and abdomen) and brain computed tomography (CT)

play a significant role in diagnosing shunt malfunction. Shunt fracture, disconnection of the shunt, shunt kinking and shunt tip out of the ventricle are regarded as radiologic evidence of shunt malfunction⁽¹⁾. However, does the result of the neuroimaging really reflect the status of the shunt apparatus and the status of the hydrocephalus? Use only brain CT image to diagnose the status of hydrocephalus and VP shunt function may lead to inappropriate treatment and cause lethal complications. The authors present a 6-year-old boy with occult CSF fistula between ventricle and extra-ventricular position

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of the VP shunt tip on CT scan.

CASE REPORT

This 6-year-old male patient was born with gestational age (GA) 37+ weeks by a G1P1 mother who was

diagnosed rheumatic heart disease with regular medication. Fetal hydrocephalus was diagnosed since GA 24 weeks by ultrasonography. After delivery, brain echo revealed increased intracranial pressure (ICP) and brain CT revealed dilated lateral ventricles and third ventricle (Fig. 1). Thus, emergent VP shunt insertion was done the



Figure 1. Brain CT without contrast revealed dilated bilateral lateral ventricles and third ventricle. Hydrocephalus can be diagnosed via the image findings.



Figure 2. Plain radiography of chest including abdomen revealed intra-abdominal location of the peritoneal part of the VP shunt.

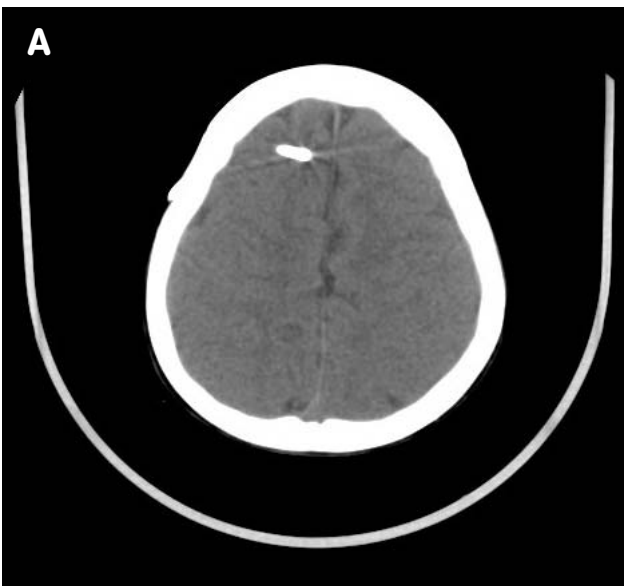


Figure 3. Brain CT without contrast revealed extra-ventricular position of the ventricular tip of the VP shunt. No connection between the VP shunt tip and ventricle can be seen.

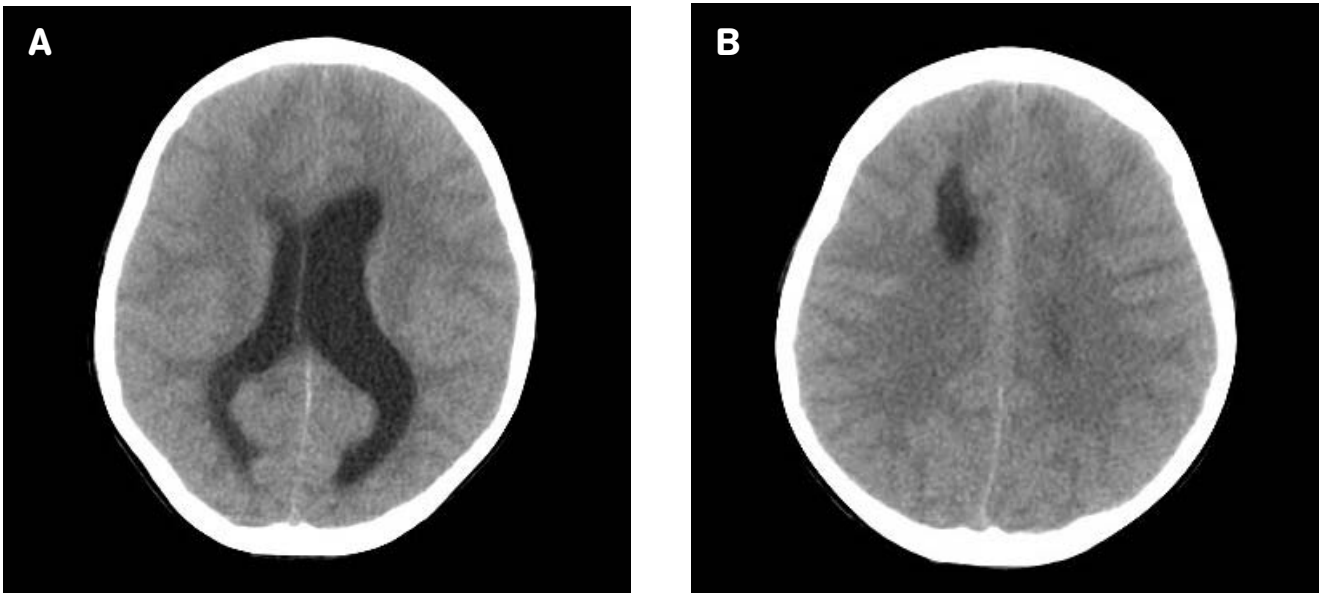


Figure 4. Brain CT after operation revealed dilated bilateral ventricles. A cyst cavity was seen at the location of previous shunt tip. A small fistula can be seen between the ventricles and extra-ventricular position of the previous VP shunt tip.

next day. After operation, he had regular follow-up at our outpatient clinic. He had normal development and good performance at school. Except for mild spastic gait under regular rehabilitation, no other neurologic deficit was found. Because of the performance status and no newly developed neurologic deficit after operation, his family wondered about the possibility of removing the VP shunt. On 2008/01/30, follow-up brain CT revealed VP shunt tip at extra-ventricular position (Fig. 2). The family then asked for shunt removal based on the cosmetic aspect and no symptoms of increased ICP. As a result, no function of the VP shunt was suspected and VP shunt removal was arranged. Physical examination at admission revealed clear consciousness, Glasgow coma scale (GCS): E4V5M6 with mild spastic gait, which has been noted since birth. Muscle power over four extremities was 5/5. Pupil size was isocoric as 3/3 with prompt light reflex. Laboratory data were all in normal range. Chest X-ray including abdomen revealed intra-peritoneal position of peritoneal part of the VP shunt (Fig. 3).

Removal of VP shunt was done smoothly in the morning on 2008/03/13. After operation, intermittent vomiting with deteriorated consciousness (GCS from E3V4M6 at 13:00 to E1V2M5 at 21:30) were noted.

Pupil size changed to 5/3 at 21:30. Emergent brain CT revealed dilated lateral ventricles and third ventricle. Also, cystic cavity and a CSF tract between the ventricle and the cavity were found at the location of previous VP shunt tip (Fig 4). Emergent VP shunt insertion was arranged right after the CT scan. After operation, the patient's GCS returned to E4V5M6 within 2 days. He was discharged one week after operation and recovery to the preoperative status.

DISCUSSION

VP shunt placement as a primary treatment for hydrocephalus child or infant is one of the most common operations in neurosurgery. After the operation, regular follow-up was recommended. During the follow-up period, different statuses of hydrocephalus can be determined. McLone et al. explained four categories of the hydrocephalus status after shunting procedure: (1) no hydrocephalus; (2) arrested hydrocephalus; (3) uncompensated hydrocephalus; and (4) compensated hydrocephalus⁽²⁾. As McLone et al. described, no hydrocephalus means that the patient has normal ventricles in the long-term absence of a functioning shunt. When the

patient presents no hydrocephalus, shunt may be removed. When the patient is in the status of arrested hydrocephalus or compensated hydrocephalus, further clinical follow-up is recommended. In this patient, follow-up brain CT at outpatient clinic revealed normal size ventricle with extra-ventricular position of the VP shunt tip. No connection between the tip of the VP shunt and ventricle could be seen via brain CT. Thus, this patient could be categorized to no hydrocephalus group according to the brain CT image. Vinchon et al. described another term as asymptomatic shunt malfunction for patient who had no increased ICP symptoms with suspected VP shunt malfunction. And they suggested that in patients with asymptomatic shunt malfunction, early shunt revision before symptoms occurred is a better treatment than further observation⁽³⁾. Sgours et al described long-term follow-up for shunted infantile hydrocephalus patients and found that patients with a catheter obviously positioned out of the ventricle system can be considered to be shunt independent if asymptomatic⁽⁴⁾. As the conclusion mentioned above, removal of the VP shunt was arranged in this patient. However, rapid deterioration and the appearance of hydrocephalus occurred within 9 hours after shunt removal. Emergent brain CT revealed dilated ventricle and CSF extended to the site of previous VP shunt tip location. Thus, the neuroimaging led to the misdiagnoses of not having hydrocephalus, shunt independent hydrocephalus and asymptomatic shunt malfunction in this patient.

When the VP shunt insertion is performed at infancy in a hydrocephalus patient, follow-up brain CT may reveal the tip of the VP shunt migrated gradually because of the patient's growing taller. During the process of migration, fistula formation may take place like this patient. Takei et al reported that subependymal glial proliferation was much more prominent in shunted cats than in normal cats⁽⁵⁾. In the same experiment it also revealed that the gliosis was more prominent in effective shunted animals than in those with ineffective shunts. Peri-ventricular gliosis may develop while the patient is growing up with migration of the shunt tip. As the tip migrates, a fistula may develop from the ventricle to the tip of the shunt. Because the fistula exists, CSF drainage

via VP shunt continued. In reviewing the English literatures, this is the first case reported about fistula formation between the extra-ventricular positioned VP shunt tip and ventricular system, and the fistula is not visible by brain CT scan. No fistula was visualized via the brain CT may contribute to several factors. First, the diameter of the fistula may be so small that it results in difficult visualization of the tract in CT scan. Second, the dynamic of the CSF drainage may alter the result of brain CT. When the ICP is elevated, the process of CSF drainage takes place while the process stops at low ICP condition. In this patient, the fistula collapsed at the time the CT was performed. Third, the patient's position and the activity before CT scan may also play a role to influence the result of the images. In the patients with asymptomatic shunt malfunction, different opinions were reported. Mazza et al suggested that surgery is not indicated for patients with asymptomatic shunt failure⁽⁶⁾. Langmoen et al suggested shunt revision for patients with asymptomatic shunt failure because symptoms of increased ICP may eventually develop⁽⁷⁾. Vinchon et al suggested frequent follow-ups for this kind of patients in short intervals instead of operations until shunt obstruction became overt or ventricular dilatation could be seen⁽³⁾. Like this patient, as no symptom occurred, further follow-up may be a better management than shunt removal.

Because of the possibility of the existence of the fistula in this kind of patients, further examinations before shunt removal are required. Chang reported that magnetic resonance imaging (MRI) can be used to detect CSF flow to 1.7ml/hour⁽⁸⁾. Thus, this may be a tool to detect the status of the shunt function before revision. Also, shuntogram using variable iodinated contrast material or radiotracers can be used for this purpose. However, false negative shuntogram was reported⁽⁹⁾. Further adjustment or standardization for the use of shuntogram or the protocol of the shuntogram is needed. Different diagnostic tools can be used to avoid misdiagnosis of the existence of the fistula between the VP shunt tip and ventricle. Tapping reservoirs to evaluate if there is still CSF flow from the ventricle site may be a simplest way to evaluate the function of the VP shunt.

Patients with shunted congenital hydrocephalus are not uncommon. Shunt tip may migrate gradually from the ventricle toward cortex when a patient grows up. As a result, this phenomenon may be much more common than expected. This may contribute to neurosurgeons' preference of shunt revision rather than shunt removal in this situation which leads to the masking of the phenomenon of fistula existence. To know the exact incidence of this phenomenon, further studies, such as MRI or shuntogram for this kind of patients are required.

In conclusion, this is the first reported congenital hydrocephalus patient with an invisible fistula based on brain CT between the extra-ventricular positioned VP shunt tip and ventricular system. Evaluating a child with a suspected VP shunt malfunction is important. In an asymptomatic shunted patient, the diagnosis of malfunction VP shunt via only one series of brain CT maybe not reliable. Further follow-up or image studies instead of immediate shunt removal or shunt revision may be a better and safer way for both the patient and the neurosurgeon. A definite diagnosis of the shunt without function is absolute necessary before removal of the shunt.

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