Hydrocephalus Due to Diffuse Villous Hyperplasia of the Choroid Plexus

—Case Report—

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Abstract

An 8-month-old female presented with hydrocephalus caused by cerebrospinal fluid (CSF) overproduction due to bilateral choroid plexus enlargement, which was clinically diagnosed as diffuse villous hyperplasia of the choroid plexus, but differentiation from bilateral choroid plexus papilloma was difficult. She initially underwent ventriculoperitoneal shunt surgery, but developed marked retention of ascites. Therefore, the peritoneal end of the shunt was removed for external drainage, but excessive CSF (1,500 ml/day) was collected. Computed tomography and magnetic resonance imaging revealed marked symmetric enhancement of the choroid plexuses in the bilateral lateral ventricles. Thallium-201 chloride single-photon emission computed tomography showed pronounced uptake on both early and delayed images, and good washout. CSF examination revealed no abnormalities such as atypical cells, and a ventriculoatrial shunt was inserted, achieving good control of the hydrocephalus.

Key words: choroid plexus, hyperplasia, hydrocephalus, cerebrospinal fluid overproduction, ventriculoatrial shunt

Introduction

Diffuse villous hyperplasia of the choroid plexus (DVHCP) is a rare condition involving enlargement of the entire choroid plexus, which remains histologically normal, resulting in severe hydrocephalus due to cerebrospinal fluid (CSF) overproduction. DVHCP must be differentiated from the rare occurrence of bilateral choroid plexus papilloma (CPP), but discrimination between the two diseases still presents problems. We treated a patient under a diagnosis of DVHCP based on clinical findings, but differentiation from bilateral CPP was difficult due to the lack of pathological specimens.
An 8-month-old female was brought to our hospital as an emergency case from a local hospital due to an acute increase in her head circumference, tightness of the lower limbs, and poor sucking ability on September 30, 2009. She was born in distress by Caesarian section on the 4th day of the 34th week of pregnancy in December 2008. Computed tomography (CT) showed intraventricular bleeding and hydrocephalus, but she was discharged after 1 month, and she was observed on an outpatient basis. After about 2 months, she developed tightness of the lower limbs, gradually lost energy, and demonstrated poor sucking reflex, so she was taken to another hospital. Since emergency magnetic resonance (MR) imaging revealed marked hydrocephalus, she was transported to our hospital.

On admission, her head circumference was 49.5 cm (+2 standard deviation), compared to 43.1 cm 1 month earlier. The anterior fontanelle was tense, and sunset phenomenon, tightness of the lower limbs, and scalp vein distention were observed. Heart rate, respiratory rate, blood pressure, and body temperature were normal. Blood examination showed no abnormalities in any parameter including electrolytes, renal function, and liver function. CT revealed marked ventricular dilation and periventricular lucency (Fig. 1).

Emergency ventriculoperitoneal (VP) shunt surgery was performed. The surgical procedure was completed without problems. On the day following the operation, marked abdominal distention occurred. Abdominal CT revealed massive accumulation of free fluid in the abdominal cavity (Fig. 2). We considered that her peritoneum could not adequately absorb the CSF, and removed the peritoneal end of the shunt to allow external drainage. The initial pressure was established at 100 mm above the patient’s external acoustic meatus. Since 200 ml CSF/hour drained from the external acoustic meatus, intermittent clamping was performed to prevent excessive outflow. However, 200 ml CSF/hour continued to drain. CSF examination showed no abnormal values or atypical cells.

CT and MR imaging, both with contrast medium, showed prominent enhancement of the symmetrically enlarged bilateral choroid plexuses (Fig. 3). Thallium-201 chloride single-photon emission computed tomography (TI SPECT) revealed marked uptake on both early and delayed images and good washout with a negative retention index (Fig. 4). After 2 weeks, her general condition worsened, with hyponatremia, methicillin-resistant Staphylococcus aureus pneumonia, and meningitis, prompting performance of a tracheotomy. However, improvement was observed after antibiotic administration intravenously and into the CSF. The diagnostic significance of the TI SPECT was unclear at this point, but...
DVHCP was the most likely diagnosis based on the CT, MR imaging, and CSF findings. After resolution of the meningitis, a ventriculotriangular (VA) shunt was inserted. She was transferred to the previous referring hospital, and has been followed up on an outpatient basis. Her intracranial pressure has been well controlled. No morphological changes were observed in the bilateral plexuses on follow-up CT 6 months after the operation.

**Discussion**

DVHCP was first reported by Davis⁹ in 1924 before the introduction of CT, and only 18 patients with hydrocephalus caused by CSF overproduction due to bilateral choroid plexus enlargement have been reported, including findings employing imaging techniques (Table 1).¹,³,⁵,⁷,¹⁰,¹³,¹⁶,¹⁸,¹⁹,²¹,²² In most patients, VP shunt surgery was performed but failed, resulting in marked ascites. This suggests difficulty in establishing the diagnosis of this disease in the initial stage.

Bilateral CPP is characterized by asymmetric enlarged choroid plexuses⁵,¹² which tend to be separated into lobules,⁹,¹³ and cyst formation in a few cases.¹⁹ However, the enlarged choroid plexuses were symmetric and resembled DVHCP on imaging in some cases,¹² enabling diagnosis only after pathological examination. Most patients diagnosed with DVHCP¹,³,⁵,⁷,¹⁰,¹³,¹⁴,¹⁶,¹⁸,¹⁹,²¹,²² showed symmetric choroid plexus enlargement, but some exhibited irregular enlargement²¹ or cyst formation.⁶ In addition, no pathological findings were available in 3 patients including our patient, so the diagnosis of DVHCP was based on imaging findings.³,¹⁴ Differential diagnosis is possible based on imaging findings,¹⁴ and VA shunt surgery is the simplest and most appropriate choice,²² considering the risks of complications of resection such as hemianopsia, nystagmus, and bleeding during the operation due to the rich blood flow in the choroid.¹⁵,¹⁶ Indeed, 3-year follow-up examination showed good control of hydrocephalus and absence of morphological changes in the choroid plexus in previous patients.²,⁴ Similar results were obtained in our patient, although the follow-up period was only 6 months. We also evaluated other options (biopsy or resection), but selected VA shunt without biopsy and resection, considering the risks of shunt failure due to bleeding-related increase in the CSF protein level when a shunt becomes necessary, infection due to long-term external drainage, and electrolyte abnormalities due to the removal of a large volume of CSF from the body.³ However, pathological findings may be needed to establish the diagnosis.⁵,¹³ Initial VA shunt was subsequently followed by tissue resection and pathological examination due to the development of complications such as thrombosis and infection, resulting in the diagnosis of bilateral CPP.¹¹,¹³ In most previous patients, the diagnosis of DVHCP was established based on the histological findings.¹,³,⁵,⁷,¹⁰,¹³,¹⁴,¹⁶,¹⁸,¹⁹,²¹,²² However, pathological examination suggested DVHCP on the tissue surface but CPP in the lobular lesion, confounding the pathological diagnosis of DVHCP in one case.¹¹

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Age at initial procedure</th>
<th>Radiographical features</th>
<th>Initial (and 2nd) procedures</th>
<th>Last procedure</th>
<th>Clinical diagnosis</th>
<th>Histological findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gudeman et al. (1979)</td>
<td>3 yrs</td>
<td>SE</td>
<td>VPS, VAS</td>
<td>resection</td>
<td>papilloma</td>
<td>papilloma</td>
</tr>
<tr>
<td>Welch et al. (1983)</td>
<td>7 yrs</td>
<td>SE</td>
<td>VPS</td>
<td>resection</td>
<td>DVHCAP</td>
<td>NCP</td>
</tr>
<tr>
<td>Buchholz and Pittman</td>
<td>29 mos</td>
<td>SE</td>
<td>VPS</td>
<td>EC</td>
<td>DVHCAP</td>
<td>NCP</td>
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<tr>
<td>Hirano et al. (1994)</td>
<td>7 yrs</td>
<td>SE</td>
<td>VPS</td>
<td>resection</td>
<td>DVHCAP</td>
<td>NCP</td>
</tr>
<tr>
<td>Britz et al. (1996)</td>
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<td>SE</td>
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<td>VAS</td>
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<td>NCP</td>
</tr>
<tr>
<td>Phillips et al. (1998)</td>
<td>14 mos</td>
<td>SE, CF</td>
<td>VPS, VAS</td>
<td>EC, VPS</td>
<td>DVHCAP</td>
<td>NCP</td>
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<td>10 mos</td>
<td>SE, lobular</td>
<td>VPS, VAS</td>
<td>resection</td>
<td>papilloma</td>
<td>papilloma</td>
</tr>
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<td>Erman et al. (2003)</td>
<td>3 yrs</td>
<td>ASE, lobular</td>
<td>resection</td>
<td>(resection)</td>
<td>papilloma</td>
<td>papilloma</td>
</tr>
<tr>
<td>D’Ambrosio et al. (2003)</td>
<td>3 mos</td>
<td>SE</td>
<td>VPS</td>
<td>resection</td>
<td>DVHCAP</td>
<td>NCP</td>
</tr>
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<td>Fujimoto et al. (2004)</td>
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<td>VPS</td>
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<td>NCP</td>
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<tr>
<td>Aziz et al. (2005)</td>
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<td>SE</td>
<td>VPS (at 2 mos)</td>
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<td>NCP</td>
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<td>24 mos</td>
<td>SE</td>
<td>VPS</td>
<td>EC, resection, VPS</td>
<td>DVHCAP</td>
<td>NCP</td>
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<tr>
<td>Iplikcioglu et al. (2000)</td>
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<td>VAS</td>
<td>DVHCAP</td>
<td>NCP</td>
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<td>Smith et al. (2007)</td>
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<td>SE</td>
<td>VPS</td>
<td>resection, VPS</td>
<td>DVHCAP</td>
<td>NCP</td>
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<td>VPS</td>
<td>resection, VPS</td>
<td>DVHCAP</td>
<td>NCP</td>
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<td>VPS, VAS</td>
<td>resection</td>
<td>papilloma</td>
<td>papilloma</td>
</tr>
<tr>
<td>Catalleo et al. (2010)</td>
<td>9 days</td>
<td>SE</td>
<td>VPS, EC</td>
<td>resection, VPS, VAS</td>
<td>DVHCAP</td>
<td>NCP</td>
</tr>
<tr>
<td>Present case</td>
<td>8 mos</td>
<td>SE</td>
<td>VPS</td>
<td>VAS</td>
<td>DVHCAP</td>
<td>no</td>
</tr>
</tbody>
</table>

ings suggested DVHCP in our patient, CPP remains a possibility. Therefore, long-term follow-up observation is necessary despite the absence of morphological changes after 6 months.

DVHCP may progress to CPP and finally to choroid plexus carcinoma (CPC), suggesting the importance of measuring the MIB-1 index.7 The MIB-1 index is nearly 0% in the normal choroid plexus, 0.2–17.42% in CPP, and 4.14–29.74% in CPC.4,5,20 The MIB-1 index was 4% in a previous case,7 and 3% (left) and 0.5% (right) in another case, suggesting the transition period from DVHCP to CPP in both patients. The pathological condition may have reflected this transition from DVHCP to CPP in patients with histological findings differing between specimen sites,11 atypical imaging findings for pathological conditions,10,21 or uptake on TI SPECT despite DVHCP suggested by CT and MR imaging, as in our patient.

In the majority of patients2,3,5,7,10–16,18,19,21,22 including ours, VP shunt surgery was performed first, but failed due to marked ascites. Subsequently, resection or endoscopic coagulation was performed in most patients to inhibit CSF production, followed by radical or shunt surgery.1,3,5,7,10–13,15,16,18,19,21,22 The advantage of these procedures is that histological diagnosis is possible, so CPP is not overlooked. However, the histological findings may differ between sites, which causes confusion.11 In addition, there are risks of neurological complications,22 and adverse effects of invasiveness, such as bleeding.21 On the other hand, a method in which excessive CSF is returned to the systemic circulation using a VA shunt has been proposed,14 which is noninvasive and straightforward, but whether the direct return of a large volume of CSF to the systemic circulation is appropriate remains questionable. The most important disadvantage is that definite histological diagnosis cannot be established. In our patient, excessive CSF was also directly returned to the systemic circulation using a VA shunt, but long-term follow-up observation is necessary.

The present and previous cases of hydrocephalus caused by CSF overproduction due to bilateral choroid plexus enlargement considered to be DVHCP suggest that a full disease description, diagnostic criteria, and treatment methods have not yet been established, requiring further studies.

References


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