Cerebrospinal fluid edema associated with shunt obstruction

Hiroaki Sakamoto, M.D., Ken Fujitani, M.D., Shouhei Kitano, M.D., Keiji Murata, M.D., and Akira Hakuba, M.D.

Departments of Neurosurgery, Osaka City University Medical School and Children's Medical Center of Osaka City, Osaka, and Department of Neurosurgery, Shimada Municipal Hospital, Shizuoka, Japan

The authors report four hydrocephalic children with cerebrospinal fluid (CSF) edema extending along the ventricular catheter of an obstructed CSF shunt. Three of the patients exhibited massive CSF edema along the ventricular catheter, yet they manifested neither ventricular enlargement nor apparent periventricular CSF edema despite increased intraventricular pressure. These findings suggested ventricular tautness. The remaining patient, who had dilated ventricles with periventricular CSF edema, displayed CSF edema in a limited area along the ventricular catheter. Replacement of the obstructed peritoneal catheter of the shunt resulted in rapid improvement of the edema in all patients. In the three patients with massive CSF edema, however, a small lesion remained in the subcortical white matter along the ventricular catheter as demonstrated by computerized tomography and/or magnetic resonance imaging 3 to 5 years after shunt revision. It is concluded that shunt obstruction may result in massive CSF edema along the ventricular catheter in hydrocephalic children who have ventricular tautness after installation of the shunt causing irreversible although usually asymptomatic damage to the affected area of the brain.

Key Words: hydrocephalus • shunt complication • cerebrospinal fluid edema • intracranial pressure • children

As hydrocephalus develops, excess cerebrospinal fluid (CSF) usually accumulates in the ventricles, dilating them by increased intraventricular pressure. Excess CSF in the ventricles can also migrate into the infraparenchyma, especially the periventricular white matter such as that area around the horns of the lateral ventricles, via disrupted ependyma. Obstruction of the shunt in hydrocephalic patients, which causes the hydrocephalus to recur, can present different manifestations. In the slit-ventricle syndrome, the ventricles fail to dilate and remain small or slit-like despite increased intracranial pressure. Shunt obstruction can also induce CSF edema along a catheter inserted into the ventricles rather than in the periventricular space.

We treated four hydrocephalic children who displayed CSF edema along the ventricular catheter in association with shunt obstruction. Three of them had massive CSF edema along the catheter without enlargement of the ventricles despite increased intraventricular pressure. In this report, we discuss the mechanism for the development of massive CSF edema at such an atypical site in relation to ventricular tautness after shunt installation.

Summary of Cases

Patient Population

Between 1980 and 1992, four children with hydrocephalus of various etiologies were found to have CSF edema along the catheter inserted into a lateral ventricle (Table 1). All patients had undergone placement of a ventriculoperitoneal shunt, with the ventricular catheter inserted from the coronal site. The ventricular catheters had inflow holes varying in length from 1.8 to 2.5 cm. A shunt system with a medium-pressure valve was installed in each patient. The patients presented with headache, vomiting, or a subcutaneous fluid collection along the shunt catheter. Two patients (Cases 1 and 2) developed massive CSF edema twice along the same ventricular catheter. At onset, the patients were aged 17 years or less, and five of the six presentations occurred in patients less than 3 years of age. The time between shunt installation and onset of symptoms varied between 1 and 16 months.

The intraventricular pressure, measured via a flushing device directly connected to the ventricular catheter while the patients were under mild sedation, was increased in all patients at admission. Analysis of the ven-
Trabecular fluid revealed no significant increase in cell count or protein content. Radiography of the shunt with contrast material introduced through the flushing device verified in all four patients that, although the ventricular catheter was patent, the peritoneal catheter was completely or partially obstructed.

Computerized tomography (CT) performed before the onset of CSF edema revealed small or slit-like ventricles in all patients. Admission CT scans showed that three patients (Cases 1, 2, and 3) had small or slit-like ventricles with a low-density area along the ventricular catheter of the shunt (Figs. 1, 2A and B, and 3A). This low-density area was observed predominantly in the white matter from the subependymal site of the lateral ventricle to the subcortical area. At the onset of symptoms in Cases 1, 2, and 3, CT showed a round lesion with lower density along the ventricular catheter surrounded by a large area of less low density (Figs. 1, 2B, and 3A). It was also revealed that the patent ventricular catheter on the right side was too short in Case 1 (Fig. 1) and that the lateral ventricles of Cases 2 and 3 were quite small (Figs. 2A and B, and 3A). These findings indicated that the inflow holes in the catheter tip were lying partially in the intraparenchymal outside the lateral ventricles. Computerized tomography, performed after injection of contrast material into a flushing device in Cases 1 and 2, revealed clear opacification of the round lesion (Fig. 2C). The CT findings indicated that the low-density area along the ventricular catheter corresponded to CSF edema and that the round lesion with lower density corresponded to a cystic lesion containing CSF. No abnormal enhancement was observed around the CSF edema area on contrast-enhanced CT scans. Magnetic resonance (MR) imaging on admission in Case 3 clearly demonstrated a cystic lesion at the center of the massive CSF edema (Fig. 3B). In Case 4, CT scans showed a small low-density area along the ventricular catheter and enlargement of the ventricles accompanied by a periventricular low-density region (Fig. 4A).

**Treatment and Outcome**

The obstructed peritoneal catheter was replaced with a new one without revising the existing patent ventricular catheter. Symptoms and signs disappeared within 1 day of shunt revision in all patients. In Cases 1, 2, and 3, CT scans performed 1 week after shunt revision showed a significant decrease in size of both the CSF edema and the cystic lesion along the ventricular catheter. In Case 4, CT showed that the CSF edema along the ventricular catheter had completely disappeared 2 days after shunt revision and that the ventricles had returned to a small size in conjunction with disappearance of the periventricular CSF edema (Fig. 4B).

In follow-up periods of 2 to 7 years (mean 5 years),

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**TABLE 1**

Summary of clinical data for four patients with CSF edema*  

<table>
<thead>
<tr>
<th>Factor</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>age (yrs), sex</td>
<td>7, M myelomeningocele</td>
<td>7, M achondroplasia</td>
<td>19, M neurofibromatosis (after removal of acoustic neuroma)</td>
<td>6, M occipital encephalocele</td>
</tr>
<tr>
<td>cause of hydrocephalus</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>age at shunting</td>
<td>(1) 10 days, (2) 3 mos</td>
<td>(1) 8 mos, (2) 17 mos</td>
<td>17 yrs</td>
<td>2 yrs</td>
</tr>
<tr>
<td>age at onset of symptoms</td>
<td>(1) 3 mos, (2) 15 mos</td>
<td>(1) 16 mos, (2) 3 yrs</td>
<td>17 yrs</td>
<td>3 yrs</td>
</tr>
<tr>
<td>interval between shunting &amp; onset</td>
<td>(1) 9 mos, (2) 12 mos</td>
<td>(1) vomiting, subcutaneous fluid collection, (2) vomiting</td>
<td>1 no</td>
<td>1 yr</td>
</tr>
<tr>
<td>presentation</td>
<td>(1 &amp; 2) subcutaneous</td>
<td>(1) vomiting, subcutaneous fluid collection, (2) vomiting</td>
<td>headache, unconvincing spell</td>
<td>drowsiness, vomiting</td>
</tr>
<tr>
<td>ventricular pressure</td>
<td>(1) not measured, (2) &gt; 40 cm H2O</td>
<td>(1) &gt; 20 cm H2O, (2) not measured</td>
<td>&gt; 25 cm H2O</td>
<td>&gt; 30 cm H2O</td>
</tr>
<tr>
<td>ventricular size at onset</td>
<td>(1) small, (2) small</td>
<td>(1) slit-like, (2) slit-like</td>
<td>slit-like</td>
<td>dilated</td>
</tr>
<tr>
<td>degree of CSF edema along catheter before shunt revision</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 wk after revision</td>
<td>(1) massive, (2) massive</td>
<td>(1) massive, (2) massive</td>
<td>masssive</td>
<td>mild</td>
</tr>
<tr>
<td>duration between onset &amp; shunt revision</td>
<td>(1) mild, (2) mild</td>
<td>(1) mild, (2) mild</td>
<td>mild</td>
<td>none</td>
</tr>
<tr>
<td>intraparenchymal lesion on follow-up CT or MRI</td>
<td>(1) yes, (2) yes</td>
<td>(1) yes, (2) yes</td>
<td>yes</td>
<td>no</td>
</tr>
<tr>
<td>follow-up period</td>
<td>(1) 3 mos, (2) 7 yrs</td>
<td>(1) 3 yrs, (2) 6 yrs</td>
<td>2 yrs</td>
<td>3 yrs</td>
</tr>
</tbody>
</table>

* CSF = cerebrospinal fluid; CT = computerized tomography; MRI = magnetic resonance imaging. (1) and (2) refer to first and second presentation.
Cerebrospinal fluid edema from shunt obstruction

CT scans of Cases 1, 2, and 3 showed a small low-density lesion along the ventricular catheter in the subcortical white matter (Fig. 2D). In Case 3, MR imaging performed 3 years after shunt revision demonstrated a small lesion along the ventricular catheter (Fig. 3C). However, the patients experienced no symptoms or signs originating from the lesions and their shunts were functioning well. In contrast, a CT scan of Case 4 showed no lesion along the ventricular catheter 3 years after revision.

Discussion
Massive CSF Edema Along the Ventricular Catheter

Our study showed that excessive CSF due to shunt obstruction accumulated in the white matter along the ventricular catheter, rather than in the more common sites of the intraparenchyma, such as the area around the anterior or posterior horn of the lateral ventricles. Cases 1, 2, and 3 demonstrated that, despite increased intraventricular pressure, the ventricles remained small or slit-like without periventricular CSF edema (Figs. 1, 2A and B, and 3A and B). These findings suggest that the ventricles became taut after shunt installation. Case 4, however, displayed ventricular dilatation associated with periventricular CSF edema under increased intraventricular pressure, and CSF edema along the ventricular catheter was much less significant than in the other three patients (Fig. 4A). In hydrocephalic patients with shunt placement, ventricular tautness will exaggerate intraparenchymal CSF migration along the ventricular catheter, thus inducing massive CSF edema along the catheter.

The CT scans obtained after contrast material was injected into the ventricular catheter indicated that the edema fluid along the catheter was ventricular CSF (Fig. 2C). There was no evidence of an enhanced lesion around the CSF edema; this suggested no significant increase in local vascular permeability, which would have induced vasogenic edema fluid.

Flow of intraventricular CSF into brain parenchyma might be attributed to the length of the patent ventricular catheter bearing the inflow holes. If the ventricles remain small or slit-like even with shunt obstruction, adhesion around the site where the ventricular catheter penetrates the ependyma may prevent the outflow of ventricular CSF into the intraparenchyma through the penetration hole. When the ventricular catheter is inserted from the coronal site, the catheter tip which contains inflow holes is so long that the holes may lie partially in the lateral ventricle and partially in the intraparenchyma. Increased intraventricular pressure may drive the ventricular CSF into the ventricular catheter through the inflow holes, and the CSF may then escape into the intraparenchymal region through the inflow holes. Therefore, if ventricular catheters are inserted from the coronal site, the catheter tip with inflow holes should be adjusted for small or slit-like ventricles after shunt installation, especially in pediatric patients.

Massive CSF edema along the ventricular catheter, with or without a cystic lesion demonstrated on CT scans, indicates the active stage of intraparenchymal CSF migration induced by shunt obstruction. Intraparenchymal CSF accumulation can be regarded as a clinical example of the direct infusion edema model. A high infusion rate of CSF into the brain parenchyma results in massive edema associated with a tissue cleft, which becomes a cystic lesion containing the infusion fluid. In the acute stage of shunt obstruction, increased intraventricular pressure (the cause of CSF migration) forces the intraventricular CSF into the intraparen-
H. Sakamoto, et al.

Fig. 3. Case 3. A: Computerized tomography scans obtained at admission showing slit-like ventricles. B: T1-weighted magnetic resonance (MR) images (upper row) clearly showing a cystic lesion with lower intensity at the center of the massive cerebrospinal fluid (CSF) edema. The T2-weighted MR images (lower row) show that the lesion ends at the subcortical white matter. No CSF edema is visible around the anterior or posterior horns of the lateral ventricles. C: T1-weighted (upper row) and T2-weighted (lower row) MR images obtained 3 years after shunt revision showing a small lesion along the ventricular catheter.

Chyma. Migration of CSF induces massive CSF edema along the catheter and also makes a tissue cleft, which becomes a cystic lesion containing CSF at the center of the CSF edema.

There have been several reports of hydrocephalic children with shunts in place who display a cystic lesion, with or without significant CSF edema, along the ventricular catheter.1,12,15,16,17 of seven patients presenting with massive CSF edema along the ventricular catheter in association with shunt obstruction, six were younger than 17 years. Pediatric patients develop ventricular tautness after shunt installation more frequently than adults.8,9,18 This may be one of the reasons why pediatric patients more often show massive CSF edema along the ventricular catheter.

Treatment and Outcome

The obstructed shunt should be replaced as soon as possible in patients with CSF edema lying along the ventricular catheter. Particularly if there is no ventricular dilatation, the patient may have less ability to compensate for intracranial accumulation of CSF resulting from shunt obstruction. For patients with ventricular tautness, replacement of the ventricular catheter may be dangerous.14,18 In addition, because shunt obstruction usually occurs in the distal catheter, we suggest

Fig. 4. Computerized tomography scans in Case 4. A: Scan obtained 1 day after the onset of shunt obstruction showing cerebrospinal fluid (CSF) edema (arrows) along the catheter in association with the periventricular CSF edema. B: Scan obtained 2 days after shunt revision. The CSF edema has resolved.

182 J. Neurosurg. / Volume 81 / August, 1994
Cerebrospinal fluid edema from shunt obstruction

prompt replacement of only the obstructed distal catheter to relieve increased intracranial pressure and CSF edema, so long as the ventricular catheter is verified to be patent by shunt radiography. Of the four patients presented here, two patients developed massive CSF edema twice due to shunt obstruction. We treated the patients by replacing the obstructed peritoneal catheter with a new catheter offering similar pressure resistance. If overdrainage of CSF induces ventricular tautness after shunt placement in hydrocephalic children, upgraded replacement of the shunt system or installation of an anti-siphon device can prophylactically reduce the recurrence of this shunt-related complication.15,16

Massive CSF edema along the ventricular catheter may result in an asymptomatic but irreversible lesion in the affected area. Shunt revision improved both symptoms and signs and also reduced CSF edema. In one patient (Case 4) in whom no ventricular tautness was displayed, the CSF edema along the catheter completely resolved shortly after shunt revision (Fig. 4B). However, in Cases 1, 2, and 3, where ventricular tautness was present, massive CSF edema resulted in an irreversible parenchymal lesion visible on CT scanning or MR imaging (Figs. 2D and 3C), even though the shunt functioned well after revision.

Conclusions

Massive CSF edema observed along the ventricular catheter of a shunt strongly implies obstruction of the distal catheter, especially in children with taut ventricles. The obstructed shunt should be revised as soon as possible. If it is not, the brain tissue affected by the massive edema may develop irreversible changes.

References


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