Onset of Dandy-Walker Syndrome in Adult following Head Trauma: A

Title

Author(s)
Kaminogo, Makio; Ochi, Akira; Moroki, Jiro; Shibata, Shobu

Citation

Issue Date
1999-06-23

URL
http://hdl.handle.net/10069/16135

NAOSITE: Nagasaki University’s Academic Output SITE
http://naosite.lb.nagasaki-u.ac.jp
Onset of Dandy-Walker Syndrome in Adult following Head Trauma: A Case Report.

Makio KAMINOGO,1) Akira OCHI,1) Jiro MOROKI,2) Shobu SHIBATA 1)

1) Department of Neurosurgery, Nagasaki University School of Medicine
2) Department of Neurosurgery, Yamaguchi Central Hospital, Yamaguchi

Although head trauma is suggested to precipitate hydrocephalus in adult-onset Dandy Walker syndrome (DWS), the clear mechanism is not verified. A 56-year-old female recovered completely after evacuation of acute epidural hematoma until development of dementia 7 days after operation. At day 20, she underwent ventriculo-peritoneal shunting to treat progressive hydrocephalus and has been free from neurologic symptoms for 2 years. The serial CT examinations indicate that after head trauma without subarachnoid hemorrhage, hydrocephalus can develop in a case of previously silent DWS. Rapid changes of intracranial pressure in trauma may affect cerebrospinal fluid outflow through a DWS-related valve mechanism at the foramina of Luschka, which results in hydrocephalus.

Key words: Dandy Walker syndrome, hydrocephalus, head trauma, and adult

Introduction

The Dandy-Walker syndrome (DWS) is developmental anomaly consisting of a posterior fossa cyst (cystic dilation of the fourth ventricle) and hypoplasia of the cerebellar vermis. Clinical expression of DWS usually occurs early in life, its major manifestations are caused by hydrocephalus which is considered a consequence, not a part, of the DWS. Occasionally, patients first become symptomatic with increased intracranial pressure in adult life. Head trauma has been suggested to precipitate such hydrocephalus in adults, but the pathophysiology of DWS-associated hydrocephalus in relation to head trauma has not been clarified. We present an adult with DWS in whom symptomatic hydrocephalus developed soon after evacuation of an acute epidural hematoma.

Case report

A 56-year-old female was referred to us because of altered consciousness (Glasgow Coma Scale: E3, V4, M6) and mild right hemiparesis developing about 12 hrs after head trauma. She had a normal developmental history and had been in good health. She fell down second-floor stairs late one evening and consulted a local physician immediately afterward. No neurologic deficit was evident, and she returned home after treatment of a left parietal scalp laceration. She returned to the physician the next morning because of progressive headache and drowsiness, and was referred urgently to us. In addition to the left parietal scalp laceration, a soft tissue mass about 3 cm in diameter was found in the occipital area. Her head circumference measured 57.3 cm. (After full recovery, she indicated that she had been aware of the occipital mass prior to injury but had not asked a physician about it.) Computed tomography (CT) on admission demonstrated a left epidural hematoma, a small contusion in the right temporal lobe, a large posterior fossa cyst, hypoplasia of the vermis, an occipital bone defect, and a cystic scalp mass communicating with the posterior fossa cyst through the bone defect. No subarachnoid hemorrhage (SAH) or hydrocephalus was evident (Fig. 1). The epidural hematoma was evacuated immediately. During surgery the dura mater was opened to examine the brain surface, but no subdural hematoma or SAH was disclosed. Consciousness became clear and left hemiparesis disappeared on the day following surgery, when CT indicated that decompression had been obtained both supratentorially and...
Fig. 1 Computed tomograms on admission (12 hrs after head trauma). A left epidural hematoma, a small contusion in the right temporal lobe, a posterior fossa cyst communicating with the fourth ventricle, hypoplasia of the vermis, an occipital bone defect, and an occipital meningocele communicating with the posterior fossa cyst were revealed.

Fig. 2 Computed tomograms on the day after evacuation of the hematoma. Enlargement of the subdural or subarachnoid space was depicted infratentorially.

Fig. 3 Computed tomograms 13 days after head injury revealed hydrocephalus and enlargement of the posterior fossa cyst.

Fig. 4 Magnetic resonance imaging shortly before ventriculo-peritoneal shunt placement verified patency of the aqueduct and indicated anterior displacement of the brainstem due to enlargement of the posterior fossa cyst.

infratentorially according to enlargement of subdural and/or subarachnoid spaces (Fig. 2). Six days after the head injury, she again began to complain of headache and nausea, and disorientation and urinary incontinence were noted at about day 10. CT on day 13 revealed dilation of lateral and third ventricles (Fig. 3). The posterior fossa cyst also had enlarged. She underwent ventriculo-peritoneal (V-P) shunting on day 20 after patency of the aqueduct had been verified using magnetic resonance imaging (MRI, Fig. 4). Postoperatively
her neurologic symptoms resolved and her occipital scalp mass became smaller. MRI 10 days after shunt-
ing revealed patency of the aqueduct, hypoplasia of
the corpus callosum, and disappearance of previously
noted anterior displacement of the brainstem (Fig. 5).
She was discharged with no neurologic deficit 17 days
after V-P shunt placement. Follow-up CT demonstrated
reduction in size of the lateral ventricles (Fig. 6). The
posterior fossa cyst also was smaller. She remained
asymptomatic with a normal neurologic examination 2
years after surgery.

Discussion

In most cases of DWS, especially in children, symptoms
and signs are caused by hydrocephalus, which re-
results from but is not part of the malformation. Late
onset of symptomatic hydrocephalus in adult life occa-
sionally has been reported. These cases follow one
of two patterns, either new appearance of hydrocepha-
lus with normal head circumference or loss of com-
pen sation in early arrested hydrocephalus with a large
head. The etiology of the late-onset cases is not
clear, but in some reported cases head trauma had pre-
ceded onset of symptomatic hydrocephalus. Lipton et
al. reviewed seven cases of adult-onset DWS, pointing
out that hydrocephalus appeared to be precipitated by
minor head trauma in two of the seven patients. On
reviewing previous reports of DWS following head
trauma, we noted that several cases were discovered
incidentally by neuroradiologic examination after head
injury. Some of these patients exhibited no neurologic
symptoms, while others had neurologic signs or
symptoms not related to DWS.

Three well-documented DWS cases exhibited symp-
tomatic hydrocephalus after head trauma (Table). Two
manifested papilledema 2 to 3 weeks after minor head
injury. The third patient, with pre-existing mild
mental retardation, exhibited gradually progressing im-
pairment of recent memory and of vision including
diplopia. However, little documentation of actual
onset of hydrocephalus was available in these three
cases. Ours is the first report of adult-onset DWS with
onset of symptoms of hydrocephalus after head
trauma in which serial CT and MRI were performed in
the acute posttraumatic stage.

In the present case, head circumference was normal
and no developmental delay was evident. CT immedi-
ately after evacuation of the hematoma revealed nor-
mal size of the third ventricle and the anterior horns
of the lateral ventricles, though the bodies of the lat-
eral ventricle was slightly enlarged (probably because
of the hypoplastic corpus callosum). These findings
suggested that hydrocephalus did not develop until
after head trauma in our case. Occipital meningocele
such as in our patient is not a rare accompanying mal-
formation in DWS. She had been aware of the occipi-
tal mass but did not consult a physician, because it
did not progressively enlarge. As it shrank signifi-
cantly after V-P shunting and was covered with nor-
mal skin and thick subcutaneous tissue, we did not re-
pair it.

Posttraumatic hydrocephalus is not common compli-
cation of head injuries. Cardoso et al. retrospectively
reviewed 2374 severely head injured patients and
Table. Adult onset of symptomatic hydrocephalus following head trauma in Dandy-Walker syndrome

<table>
<thead>
<tr>
<th>Source</th>
<th>Age/Sex</th>
<th>Head trauma</th>
<th>Size of head</th>
<th>Developmental delay</th>
<th>ICP* elevation</th>
<th>Cerebellar or brainstem signs</th>
<th>Clinical evolution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sahr (8)</td>
<td>16y.o./M</td>
<td>neck-strength</td>
<td>?</td>
<td>(-)</td>
<td>(+)</td>
<td>(+)</td>
<td>acute</td>
</tr>
<tr>
<td></td>
<td></td>
<td>contest</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Coleman</td>
<td>17y.o./M</td>
<td>collision with relatively</td>
<td>(-)</td>
<td>(+)</td>
<td>(-)</td>
<td>acute</td>
<td></td>
</tr>
<tr>
<td>et al (5)</td>
<td>other boy</td>
<td>large</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stovall</td>
<td>30y.o./M</td>
<td>struck by car</td>
<td>large</td>
<td>(+)</td>
<td>(-)</td>
<td>(+)</td>
<td>chronic</td>
</tr>
<tr>
<td>et al (13)</td>
<td>while walking</td>
<td>(67.3 cm)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

ICP*, intracranial pressure

pointed out that hydrocephalus demonstrating large ventricles without enlargement of the sulci and new development of symptoms was found only in 17 patients (0.7%). They also pointed out that SAH and intracranial infection following head injury were the major causes of the traumatic hydrocephalus. However, intraoperative findings and CT imagings did not indicate the apparent SAH to induced subsequent hydrocephalus. Moreover, there was no evidence of inflammation causing hydrocephalus. Since arachnoid cysts can bleed after minor trauma, unsupported vessels around the occipital cyst are postulated to tear easily resulting in hemorrhage into the posterior fossa cyst or the subdural or subarachnoid space in the posterior fossa. Such bleeding may be responsible for disturbance of cerebrospinal fluid (CSF) outflow from the fourth ventricle to the perimedullary subarachnoid space or CSF flow within the subarachnoid space. Hirsch et al. point out that hydrocephalus develops frequently in the 2 months following birth and postulate that bleeding at the time of delivery also may represent an important cause of infantile-onset DWS. However, no neuroradiologic verification is available for this hypothesis. While the present case is the first in which CT was reported from the acute stage of head trauma in a patient with DWS who subsequently developed symptomatic hydrocephalus, CT did not demonstrate intracranial hemorrhage which would interfere with CSF flow. In one of the three reported cases, CSF was examined by lumbar puncture in the acute posttraumatic stage; again, no subarachnoid hemorrhage was verified. Massive cerebral contusion and cerebral edema can be also proposed as causes of blockage of CSF flow in subarachnoid space, however, serial CT examinations indicated only small cerebral contusion in our patient.

A second explanation is that after head trauma obstructive mechanisms other than bleeding develop in the CSF outlets from the fourth ventricle to the subarachnoid space. Foramina leading from the ventricular system are not typically patent in patients with DWS; the foramen of Magendie reportedly is obstructed in most cases. CSF is speculated to exit via an undetectable outlet or patent foramina of Luschka. Alternatively, the foramina of Luschka may be patent intermittently, with a ball-valve mechanism. In one adult-onset case related to head trauma, intraoperative inspection indicated that neither the foramen of Magendie nor the foramina of Luschka appeared patent. In that case, head trauma was postulated to directly affect CSF outflow which previously had been maintained through incompletely occluded foramina. In our case, CT the day after the evacuation of hematoma revealed widening of the subdural or subarachnoid space in the posterior fossa, an indication of rapid decompression in the posterior fossa following evacuation of the epidural hematoma. Compression and subsequent decompression of the foramina of Luschka may have occurred with
development of epidural hematoma, and its evacuation, affecting the valve mechanisms in the foramina of Luschka to result in hydrocephalus. Symptoms due to increased intracranial pressure appeared after a neck-strength contest in one reported case.\(^9\) Since increased venous and CSF pressure can occur during exertion, this transient change in ICP also may have interfered with a complicated valvular communication in the foramina of Luschka.

Yet, another explanation for delayed onset of hydrocephalus is that silent aqueductal stenosis may become symptomatic after head trauma, since aqueductal stenosis has accompanied DWS in some cases.\(^1\) However, in the present case, MRI disclosed patency of the aqueduct and the occipital meningocele communicating with the occipital cyst shrank after V-P shunt placement, indicating good communication between the lateral ventricles and the occipital cyst.

In summary, the present study cannot precisely demonstrate the cause of DWS-associated hydrocephalus developing after head trauma in adult life. However, CT indicated that hydrocephalus may develop after head trauma without SAH or intracystic hemorrhage in an adult case of previously asymptomatic DWS. Valvular communicating mechanisms in the foramina of Luschka may be vulnerable to transient changes in ICP accompanying head trauma.

### References

8. Sahs AL. Congenital anomaly of the cerebellar vermis. *Arch Pathol* 32:52-63, 1941
17. Ishimitsu H, Namba S, Nakasone S. An adult case of a Dandy-Walker syndrome. *Brain and Nerve* 31:583-589, 1979 (abstract in English)