Acute external hydrocephalus complicating craniocervical decompression for syringomyelia–Chiari I complex

Case report and review of the literature

Paolo Perrini · Alexander Rawlinson · Richard Alfred Cowie · Andrew Thomas King

Abstract The occurrence of subdural cerebrospinal fluid collections and ventricular dilatation (external hydrocephalus) after foramen magnum decompression is extremely rare. The authors report on a 37-year-old man who developed symptomatic subdural fluid collections (SFC) after uncomplicated foramen magnum decompression for Chiari I–syringomyelia complex. Postoperative magnetic resonance imaging revealed a supratentorial SFC with prominent midline shift. In addition, bilateral infratentorial SFCs extending supratentorially through the tentorial notch, pseudomeningocele and initial shrinkage of the syrinx were observed. Few days after evacuation of supratentorial collection, the patient experienced increasing headache and a computerized tomography scan demonstrated a contralateral subdural collection as well as ventricular dilatation. A programmable ventriculoperitoneal shunt was placed with resolution of supratentorial collection and progressive improvement of infratentorial collections and pseudomeningocele. Although previous reports described the occurrence of extra-axial fluid collections and hydrocephalus after foramen magnum decompression for Chiari malformation, to our knowledge, this is the only report of acute external hydrocephalus after foramen magnum decompression requiring urgent evacuation of SFC in order to reduce the mass effect.

Keywords Chiari I malformation · External hydrocephalus · Foramen magnum decompression · Syringomyelia

Introduction

Foramen magnum decompression (FMD) of the cerebellar tonsils at the craniocervical junction is generally considered the most effective treatment for Chiari I malformation with or without associated syringomyelia. The most frequently reported complications of this procedure include CSF leak [8, 10, 12], meningitis [8, 12] and pseudomeningocele [9, 10]. Hydrocephalus is a rare complication of FMD and its association with subdural fluid collections (SFC) is exceptional. Although the term external hydrocephalus has been initially used to describe extracerebral fluid collections involving the subarachnoid or subdural spaces in presence of increased intracranial pressure in infants [1, 2], recent reports suggest that this condition can occur in adult patients after trauma, haemorrhage and aneurysm surgery [3, 5, 14]. Recently, the occurrence of external hydrocephalus after FMD for Chiari Malformation has been described [4, 7, 11]. We report on an unusual case of acute external hydrocephalus associated with pseudomeningocele complicating FMD for syringomyelia–Chiari I complex in an adult patient. This is a rare complication that must be considered should clinical deterioration occur in the presence of subdural fluid collection after posterior fossa decompression.

Case report

History and examination A 37-year-old man with Klippel–Feil Syndrome presented with a 1-year history of strain-related headache, burning-type dysesthesias in both arms...
and intermittent swallowing difficulty. A neurological examination revealed hypoesthesia for light touch, pain and temperature in both arms. Deep tendon reflexes were reduced in the arms and mildly hyper-reflexic in the lower extremities. The remainder of the examination was normal.

Magnetic resonance imaging (MRI) demonstrated a Chiari I malformation, syringobulbia limited to the medulla and syringomyelia extending from C2–C3 to T1. In addition, congenital fusion of the C7 and T1 vertebrae was appreciated (Fig. 1a). No hydrocephalus was observed (Fig. 1b).

**Operation** The patient underwent FMD in which a suboccipital craniectomy and laminectomy of C1 and C2 with intradural exploration were performed. A thickened arachnoid veil was detected at the outlet of the fourth ventricle. This was dissected and lysed to establish spontaneous CSF flow from the fourth ventricle. No duroplasty was performed and the dura was left open and stitched laterally to the muscles.

**Postoperative course** After the operation the patient experienced immediate improvement of strained-related headache and was discharged home after 6 postoperative days. Two weeks after the initial surgery, the patient developed progressive swelling on the wound site associated with dizziness and gradually increasing weakness of the right arm.

**Second admission and operations** The patient was afebrile and had normal vital signs. Physical examination revealed swelling on the occipitocervical wound. No CSF leak was detected. The result of neurological examination was significant for 3/5 weakness of the right arm. MRI of the brain showed a SFC in the left fronto-parietal-occipital region with prominent midline shift. The subdural collection was following the intensity of CSF in all sequences (Fig. 2a,b). In addition, bilateral infratentorial SFC with the same signal intensity was detected (Fig. 2a). MRI of the cervical spine revealed a significant pseudomeningocele, along with initial shrinkage of the syrinx (Fig. 2c). Following emergency burr-hole evacuation of subdural CSF collection, the patient’s strength of the arm rapidly normalized. A computerized tomography (CT) scan performed the day after the burr-hole trephination, demonstrated resolution of midline shift and air–fluid level at the subdural space (Fig. 3a). On postoperative Day 7, the patient developed increasing headache and mild difficulty with concentration and memory. A repeat CT scan disclosed a small contralateral low-density fluid collection in the frontal region along with dilatation of all four ventricles (Fig. 3b). No modification of infratentorial SFC was observed. The patient was taken to surgery where a
A Codman Hakim programmable valve with opening pressure adjusted to 70 mm H2O was placed. Postoperatively, the symptoms rapidly improved, and a CT scan revealed resolution of the supratentorial SFC and initial decrease in size of the ventricles (Fig. 3c). The patient was discharged on postoperative Day 10.

At the 4-month follow-up, the patient was headache-free and reported improvement of dysesthesic symptoms as well as resolution of swallowing difficulty; however, the hypoesthesia in both arms was unchanged. Follow-up images obtained at this time revealed resolution of both the hydrocephalus and the supratentorial SFC. In addition, further shrinkage of the syrinx cavity and reduction of infratentorial SFCs and of pseudomeningocele were observed (Fig. 4).

**Discussion**

The occurrence of SFCs after FMD for Chiari I malformation with or without associated syringomyelia is an extremely rare entity. In fact, only six cases were found in the literature and all cases were symptomatic and presented within 2 weeks following surgery (Table 1). A tear in the arachnoid membrane made at the time of surgery and accumulation of CSF resulting from an obstruction of free CSF flow have been reported to play a pivotal role in the development of this condition. The original hypothesis proposed by Ranjan and Cast [11] was that a tiny hole in the arachnoid acts as a slit valve mechanism allowing egress of CSF in the subdural space. In this scenario, the CSF dissects the dura-arachnoid interface layer accumulating over the superior surface of the cerebellum and eventually extending supratentorially through the tentorial notch. The surgical techniques proposed to prevent the CSF from dissecting into the subdural space are clipping the arachnoid to the dura or widely fenestrating the arachnoid in order to avoid the ball–valve mechanism [4, 11]. Our report seems to neatly question this latter hypothesis because we widely open the arachnoid to perform an intradural dissection.

The option of leaving open the dural layer after FMD for Chiari malformation was originally proposed by Williams [13]. Recently, some authors reported favourable results after FMD without duroplasty [6, 10]. In fact, the role of duroplasty in avoiding the occurrence of SFCs remains unclear. According to our review, all previously reported cases of SFCs after FMD underwent duroplasty.

In external hydrocephalus, the ventricular system dilates despite the presence of SFCs. Elton et al. [4] proposed that the SFC accumulating over the tentorial surface of the cerebellum compresses the cerebral aqueduct causing obstructive hydrocephalus. Our case differs from the other reported cases in that ventricular dilatation developed after evacuation of the supratentorial SFC despite the infratentorial fluid collection was unchanged in size. At that time, a
contralateral supratentorial fluid collection was evident suggesting an underlying disturbance in CSF absorption. We suggest that when an arachnoidal rent is produced the blood-contaminated CSF might interfere with the absorption of CSF leading to its accumulation. This occurrence can be transient or can progress to excessive CSF accumulation in different intracranial compartments. In addition, the CSF accumulated in the subdural space can shift among these compartments resulting in contralateral subdural collections and/or ventricular dilatation (internal hydrocephalus). It can be hypothesized that the delayed occurrence of internal hydrocephalus was caused by significant CSF drainage at the time of FMD. In this case, the progressive ventricular enlargement occurred after a period of low intraventricular pressure, which also preceded the occurrence of SFCs. Of note, our patient presented with a significant pseudomeningocele after FMD. Pare and Batzdorf [9] have reported an association between the postoperative persistence of syringomyelia and pseudomeningocele. This occurrence suggests that the pseudomeningocele may prevent the normalization of CSF flow dynamics at the craniocervical junction because of dissipation of the systolic pulse pressure into the distensible pseudomeningocele cavity [9]. Although in our case, initial shrinkage of the syrinx was observed after FMD, we hypothesize that the dysfunction of CSF flow caused by the pseudomeningocele cavity played a role in the persistence of external hydrocephalus.

To our knowledge, the current case is the only report of acute external hydrocephalus after FMD requiring urgent evacuation of SFC in order to reduce the mass effect. Because of the rarity of external hydrocephalus after FMD, the most appropriate and effective treatment is not known. However, of five reported cases in which a SFC was associated with ventricular dilatation, three required temporary ventricular drain, one required permanent shunting and one resolved spontaneously. It is interesting to note that the only other case requiring permanent shunting presented with both infra and supratentorial SFCs, as the present patient suggested a severe CSF flow disturbance.

**Conclusions**

The clinical course of SFCs following Chiari decompression is variable, ranging from spontaneous resolution to ventricular dilatation and to progressive clinical deterioration due to hydrocephalus or increasing mass effect by

<table>
<thead>
<tr>
<th>Author and year</th>
<th>Age (yr)/sex</th>
<th>Presentation</th>
<th>Surgical technique</th>
<th>Location of SFC</th>
<th>Internal hydrocephalus</th>
<th>Pseudomeningocele</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Elton et al. 2002 [4]</td>
<td>10/M</td>
<td>Chiari I</td>
<td>FMD + DO + AO + DU</td>
<td>Infratentorial</td>
<td>Present</td>
<td>Present</td>
<td>EVD (2 times)</td>
<td>6 months, resolution of SFC and hydrocephalus</td>
</tr>
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<td>14/F</td>
<td>Chiari I–Syringomyelia</td>
<td>FMD + DO + AO + DU</td>
<td>Infratentorial</td>
<td>Present</td>
<td>Absent</td>
<td>EVD</td>
<td>6 months, resolution of SFC and hydrocephalus</td>
<td></td>
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<td>14/F</td>
<td>Chiari I–Syringomyelia</td>
<td>FMD + DO + AO + DU</td>
<td>Infratentorial</td>
<td>Present</td>
<td>Present</td>
<td>EVD</td>
<td>1 month, Pseudotumor cerebri and LP shunt; 2 months, resolution of SFC and hydrocephalus</td>
<td></td>
</tr>
<tr>
<td>Marshman et al. 2005 [7]</td>
<td>17/F</td>
<td>Chiari I–Syringomyelia</td>
<td>FMD + DO + DU</td>
<td>Infra and supratentorial</td>
<td>Present</td>
<td>Absent</td>
<td>EVD, VP shunt</td>
<td>3 months, resolution of SFCs and hydrocephalus</td>
</tr>
<tr>
<td>59/M</td>
<td>Chiari I</td>
<td>FMD + DO + DU</td>
<td>Infratentorial</td>
<td>Present</td>
<td>Absent</td>
<td>Conservative</td>
<td>3 months, resolution of SFCs and hydrocephalus</td>
<td></td>
</tr>
<tr>
<td>Present case</td>
<td>37/M</td>
<td>Chiari I–Syringomyelia</td>
<td>FMD + DO + AO</td>
<td>Infra and supratentorial</td>
<td>Present</td>
<td>Present</td>
<td>Drainage of SFC, VP shunt</td>
<td>4 months, resolution of SFCs and hydrocephalus</td>
</tr>
</tbody>
</table>

AO Arachnoid opening, DO dural opening, DU duroplasty, SFC subdural fluid collection, EVD external ventricular drain, F female, FMD foramen magnum decompression, LP lumbar peritoneal shunt, M male, mo month, VP ventriculoperitoneal
enlarging subdural collection. The accumulation of further case reports and longer follow-up data is needed to better define this pathological entity regarding its pathophysiology, treatment options and long-term clinical outcome.

References


Comments

Jörg Klekamp, Quakenbrück, Germany

The authors describe an unusual complication after foramen magnum decompression for Chiari type I malformation. It is well established that Chiari I malformation causes CSF flow obstruction at the foramen magnum which may lead to syringomyelia. However, little is known about the intracranial consequences of this flow obstruction. Surely, there must be alterations of intracranial CSF flow. However, whereas obstructive hydrocephalus is a regular feature of Chiari type II malformation, it appears in only about 10% of patients with Chiari I. In other words, the intracranial CSF flow abnormalities are well compensated in most cases.

With decompression at the foramen magnum, a CSF passage is provided that leads to a decrease of the syrinx in the overwhelming majority of patients. However, flow patterns inside the cranial cavity are changed as well. Most patients suffer from low pressure symptoms for a few days due to the intraoperative loss of CSF. Postoperative scans taken at this stage almost always demonstrate a dilatation of the subarachnoid space in the posterior fossa. But this phenomenon resolves quickly and has never caused any long-term problems in my experience. On the other hand, about 3% develop a postoperative hydrocephalus without resolution of the syrinx even though the CSF passage at the foramen magnum is patent. This may happen immediately after surgery or weeks later. With placement of a ventricular shunt this problem resolves and the syrinx decreases.

I think that the patient presented here belongs into this category. The only unusual feature is that the raised CSF pressure led to subdural and large soft tissue CSF collections first, before a ventricular dilatation developed. I don’t think that performing a duroplasty would have prevented this complication. At the moment, we can only speculate why this complication may develop. This case report emphasizes that all patients operated for a Chiari type I malformation should be monitored postoperatively for signs of increased intracranial pressure.
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