Acute respiratory arrest following partial suboccipital cranioplasty for cerebellar ptosis from Chiari malformation decompression

Report of 2 cases

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Cerebellar ptosis is a rare complication following Chiari malformation decompression, and generally is the result of a very large suboccipital craniectomy. This can lead to the descent of the cerebellum through the craniectomy defect, which in turn may result in cerebellar herniation through the surgical defect as well as the reestablishment of contact between the cerebellar tonsils and the brainstem. In addition, dorsal adherence of the herniated cerebellum to the dura mater or dural patch and an associated obstruction of cerebrospinal fluid flow at the cervicomedullary junction may ensue. Such a result is not desirable, in that it reproduces or mimics the pathoanatomical relationships that existed prior to the surgical decompression. (DOI: 10.3171/FOC.2008.25.12.E12)

Key Words • cerebellar ptosis • Chiari malformation • respiratory arrest

Partial suboccipital cranioplasty is effective in treating cerebellar ptosis. We report respiratory arrest following partial suboccipital cranioplasty for cerebellar ptosis secondary to CM decompression in 2 patients. The first patient was a 50-year-old woman with a 1-year history of recurrent symptoms following decompression of CM performed 2 years previously. The second patient was a 66-year-old woman with a 4-year history of progressively worsening symptoms that were characteristic of her CM symptoms experienced 7 years prior (Table 1). Admission MR imaging revealed the presence of severe cerebellar ptosis with excessive cerebellar tonsil descent (Fig. 1 left) in both patients. These patients both underwent a standard partial suboccipital cranioplasty performed by different senior surgeons at our institution (Fig. 1 right). A large suboccipital craniectomy was identified in both intraoperatively, with the cerebellar hemisphere protruding out of the decompression window (Fig. 2). Fifteen and 72 hours postoperatively (in Cases 2 and 1, respectively), the patients experienced similar episodes of apnea and subsequent respiratory arrest. Postoperative MR images and emergency CT scans were negative for bleeding, infarct, and blockage of the CSF pathway (Fig. 3). Chest radiographs revealed no evidence of pulmonary aspiration in either patient (Fig. 4).

Case Reports

Case 1

History. This 50-year-old right-handed woman presented with a recurrent and intractable headache, and with a gait problem that had continued for 6 months. She had undergone CM decompression in another hospital ~2 years before.

Examination. On examination, the patient was alert and oriented but dehydrated, and she was in a wheelchair due to headache with nausea and vomiting, imbalance, and dizziness. The MR imaging showed no evidence of ventriculomegaly and syrinx, but it did show cerebellar hemispheres with tonsils descending down through the marked occipital craniotomy (Fig. 1 left).

Operation. Intraoperatively, a large (5 × 4–cm)
boundary of the former suboccipital craniectomy was found, and the descended cerebellar tonsils and the cervical spinal cord were clearly seen intact (Fig. 2). A good CSF flow from the foramen of Magendie was noticed after the dura mater was opened at the cervicomедullary junction in the midline. Alloderm duraplasty was used to close the dura with a 4–0 suture in a watertight fashion. Attention was then turned toward the creation of the new posterior fossa floor for treatment of ptosis and cerebellar support. This was fashioned from titanium mesh and MMA, and secured with 4 screws around the perimeter of the cranioplasty. The cranioplasty was formed in such a way as to reduce the large craniectomy, from ~4–5 cm down to a 2 × 2–cm craniectomy. Care was taken not to allow any sharp edges and or any compression of the cerebellum or cervicomедullary junction by the cranioplasty. The cranioplasty was seated well and secured over the duraplasty. The incision was then irrigated and closed in layered fashion with a running absorbable suture for skin closure.

Postoperative Course. Postoperatively, the patient was able to be out of bed and walking without assistance on postoperative Day 1 and ambulated on Day 2. On Day 3, the patient suddenly suffered respiratory distress and was intubated immediately. Chest radiographs confirmed there was no infiltrate, and a head CT scan was negative in the posterior fossa, with no evidence of acute hydrocephalus or infarct. The patient recovered, was extubated within 24 hours, and was discharged on Day 7. Two weeks after being discharged the patient walked into the clinic, with preoperative headache and imbalance having completely resolved. The patient remains well on annual follow-up examinations.

Case 2

History. This 66-year-old right-handed woman presented with a 4-year history of mild headache, dizziness, difficulty in swallowing, 2 years of gait imbalance, and 1 year of progressive numbness in the face and upper extremities. The patient reported that she had recently experienced occasional urinary incontinence. She had received a diagnosis of CM-I and had undergone her CM decompression 7 years earlier at a different institution.

Examination. The patient was alert and oriented, with stumble gait. She showed absent gag reflex on the right side. Admission MR imaging showed tonsil herniation without syrinx.

Operation. During the operation, the cerebellar tonsils were indeed found protruding in a conical fashion through the foramen magnum and pushing on the medulla at the level of the obex. There was no CSF seen initially emitting from the obex. With the aid of bipolar cautery, cerebellar tonsils were coagulated in both a caudal and a cephalad direction. Once the tonsils were cauterized, CSF began emitting from the obex of the fourth ventricle. Some subarachnoid adhesions were loosened using the microscissors. A small piece of ligamentum nuchae was removed, and this was used as a pericranial graft. The graft was sutured in place with 4–0 Nurolon sutures in a
continuous running fashion. Then, using the drill with the M8 bit, we made circumferential divots in the preexisting shelf of bone to accept the MMA and create a mortise and wedge type of interface to hold the glue. Once mixed and in the hardening stages, we conformed the MMA to create a shelf that contained the contents of not only the duraplasty but also the posterior fossa, and allowed a generous ring of foramen magnum (~ 2 × 2 cm). The MMA was undermined under the divots, thus conforming to the valleys of the divots.

Postoperative Course. The patient did well throughout the postanesthesia care, and was transferred to the regular neurosurgery nursing unit. Approximately 15 hours postoperatively, the patient experienced acute respiratory failure. Results of the emergency CT scan were negative (Fig. 3), and the chest radiograph revealed no evidence of pulmonary aspiration (Fig. 4). The patient was then reintubated and placed on ventilation in the intensive care unit. Extubation failed twice in this patient, and a prolonged ventilation weaning period was anticipated. Subsequently, on postoperative Day 8, the patient underwent a tracheostomy and percutaneous endoscopic gastrostomy. Since then, she has been fed by tube via percutaneous endoscopic gastrostomy. The patient was admitted to the subacute care facility for rehabilitation, and then was transferred to a nursing home, where she is living dependently.

Discussion

The treatment of CM-I remains challenging and controversial. This is especially so regarding surgical options. These include the size of the suboccipital craniectomy, a decompression with or without an opening of the dura mater, an opening of the dura with or without closing or patching with dural graft, tonsilllectomy, and so on.1,2,12,19,21,22,24 Overall, surgery has proven to be quite effective.5,18,24 The most frequently reported complications following CM surgery include pseudomeningocele,16,25 meningitis,4 CSF leakage,25 and cerebellar ptosis.3 The latter is a complication with potentially severe consequences, and its incidence has probably been underestimated. Cerebellar ptosis is generally the result of a prior suboccipital craniectomy that is > 4 × 4 cm from the estimated edge and center of the foramen magnum. Patients with cerebellar ptosis characteristically present with intractable headaches, neck pain, suboccipital burning sensation, dizziness, nausea, swallowing problems, and imbalanced gait. Partial suboccipital cranioplasty has been used to treat the cerebellar ptosis because symptomatic cerebellar ptosis almost always follows excessive decompression of CM.8

We attribute the symptoms of cerebellar ptosis in part to a damaged brainstem resulting from chronic compression. This results in a further worsening of the ptosis and associated symptoms; the latter include respiratory arrest. An awareness of the increased potential for such a complication should minimize the incidence of catastrophic sequelae.

The goal of surgery for CM-I is to decompress the cerebellar tonsils and the upper cervical spinal cord and to restore the flow of CSF in the subarachnoid space in the region of the cervicomedullary junction. Treatment of the adult form of this condition presents a much greater challenge to the neurosurgeon. Most authors agree that treatment should consist of a suboccipital craniectomy and upper cervical laminectomy to decompress the malformation at the foramen magnum.5,7,18,24 However, CM decompression may lead to cerebellar ptosis if the suboccipital craniectomy is excessive.3,8,26 In some patients the material used for dural grafting may also play a role, often due to dural tension related to scarring. Although it is recognized that some surgeons favor a wide posterior fossa craniectomy, others have opted for a small craniectomy in the treatment of CM-I.10 The most common management strategy selected for this complication is partial cranioplasty, with or without intradural exploration. Duraplasty was performed in both of our cases to attain adequate restoration of CSF flow from the fourth ventricle.
TABLE 1: Characteristics in 2 patients with respiratory arrest following suboccipital cranioplasty for cerebellar ptosis

<table>
<thead>
<tr>
<th>Age</th>
<th>Previous CM Op</th>
<th>Preop Symptoms</th>
<th>Onset of RA (hrs)</th>
<th>Preop Hydrocephalus</th>
<th>Change in Ventricle Size</th>
<th>Postop Complications</th>
<th>Aspiration/Infection</th>
</tr>
</thead>
<tbody>
<tr>
<td>50, F</td>
<td>2</td>
<td>intractable HA, neck pain, extremity weakness</td>
<td>72</td>
<td>shunt placed</td>
<td>no</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>66, F</td>
<td>4</td>
<td>constant HA, neck pain, vertigo, swallowing difficulty, incontinence, spastic gait, absent gag reflex</td>
<td>15</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>no</td>
</tr>
</tbody>
</table>

* HA = headache; RA = respiratory arrest

Alloderm was sutured as dural graft in one case, and a piece of ligamentum nuchae was used in the other. The previous craniectomy sizes were found > 4 × 4 cm from the edge of the foramen magnum in both cases. The crescent-shaped cranioplasty reduced the craniectomy to ~ 2 × 2 cm in both cases. The patients partially recovered and were transferred from the intensive care unit to a ward. A respiratory arrest occurred at 15 and 72 hours following the surgeries in Cases 2 and 1, respectively.

Respiratory failure following suboccipital craniectomy frequently involves an intraoperative injury to the brainstem, or secondary compression or herniation from the postoperative hematoma and via acute obstructive hydrocephalus. Postoperative imaging and emergency CT scans after respiratory arrest in the 2 patients presented here demonstrated no evidence of intraoperative trauma, postoperative bleeding, or hydrocephalus (Fig. 3). In 1983, Park et al.\textsuperscript{17} reported a series of 45 infants with myelomeningocele and hydrocephalus, in which 28 (62%) of the patients were alive and 17 (38%) had died at the last follow-up assessment after decompression of CM. The majority of deaths were attributed to respiratory failure. Paul et al.\textsuperscript{18} reviewed 71 cases of CM-I. All patients underwent suboccipital craniectomy and C1–3 laminectomy. Respiratory depression was the most frequent postoperative complication (14%). One patient died of sleep apnea 36 hours after the operation. Additionally, acute respiratory arrest has been reported previously at 2, 3, and 42 hours, and as late as 2 months after suboccipital craniectomy for CM-I.\textsuperscript{5,9,14,27} Sudden respiratory deterioration due to the initial procedure has been described, possibly as a result of the loss of respiratory center neurons in the morbid condition, with this loss possibly due to congenital deformity or lengthy compression.\textsuperscript{15,23} Although there was no obvious tether identified intraoperatively on the spinal cord or brainstem in either case, one wonders if stretching on the vulnerable cervicomедullary junction from the elevation of the cerebellum during the cerebellar shelving procedure is possibly responsible. Besides, it might be that some of the compromised vasculature (including arteries and veins) played a certain role in the catastrophic complication as well, which has been reported sporadically in patients with CM-I.\textsuperscript{11,13,20}

Both patients presented here, however, had neurological abnormalities for years following suboccipital craniectomy and C-1 laminectomy due to their CM. We have been unable to find any report of sudden, unprovoked respiratory arrest following suboccipital cranioplasty in adults with cerebellar ptosis. In both of our patients, the radiological and operative findings suggested compression of the brainstem and spinal cord due to the cerebellar ptosis. The patient in Case 1 has a shorter history regarding both the initial diagnosis of CM and the recurrent symptoms from cerebellar ptosis, whereas the one in Case 2 was observed to have a greater disturbance of lower cranial nerves. The latter may indicate that the secondary injury from the recurrent cerebellar ectopia might add to the neurological impairment, in turn making the involved brainstem more susceptible to further injury and subsequent respiratory arrest.

Conclusions

The risk of sudden respiratory arrest as well as gradual respiratory deterioration must be considered in planning the management protocol in patients with cerebellar ptosis. Indeed, they may have a postoperative course and ultimate outcome that is less successful than expected with a de novo CM decompression operation.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Acknowledgment

The authors thank Ms. Christine Moore for her kind assistance with the preparation of this manuscript.

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