Patients with Chiari malformation Type I presenting with acute neurological deficits: case series

Clinical article

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Object. A subset of patients with Chiari malformation Type I (CM-I) presented with acute onset of a neurological deficit. In this study the authors summarize their experience with these patients’ clinical presentation, imaging results, timing of surgery, and outcome following decompression.

Methods. The authors reviewed clinical records, imaging studies, and operative notes from all patients undergoing posterior fossa decompression for CM-I at St. Louis Children’s Hospital from 1990 to 2008. Of the 189 patients who underwent surgery, 6 were identified with the acute onset of a neurological deficit at presentation.

Results. All 6 children (age range 3–14 years, 3 boys and 3 girls) had either syringomyelia (5 patients) or T2 signal changes in the spinal cord (1 patient) and CM-I on initial MR imaging. Three patients presented after minor trauma (1 with paraparesis, 2 with sensory deficits). Three patients presented without a clear history of trauma (1 with abrupt onset of spontaneous dysphagia and ataxia, 2 with sensory deficits). Decompression was performed at a mean 7.7 ± 4.9 days after symptom onset (7.0 ± 1.6 days after neurosurgical evaluation). In 1 patient, symptoms had resolved by the time of surgery; in the remainder of the patients, clear improvements were noted within 2 weeks of surgery, with complete resolution of symptoms by 12 months postoperatively. Follow-up MR images were obtained in 4 patients, demonstrating improvement in the extent of the syrinx in each patient.

Conclusions. Children with CM-I and syringomyelia can develop acute spinal cord or bulbar deficits with relatively minor head or neck injuries. The prognosis for symptomatic improvement in the observed deficit is good, with each patient in our series showing resolution of deficits over time. However, based on this relatively limited experience, the authors suggest that patients who present with an acute neurological deficit and are found to have CM-I be managed with early posterior fossa decompression. Patients with CM-I and syringomyelia may be at higher risk of acute neurological deficit than those without a syrinx. (DOI: 10.3171/2010.11.PEDS1097)

Key Words • posterior fossa decompression • syringomyelia • Chiari malformation Type I • neurological deficit

Chiari malformation Type I is defined as cerebellar tonsillar herniation greater than 5 mm inferior to the foramen magnum.1 Anatomically, this herniation is also associated with decreased posterior fossa volume, decreased CSF in the posterior fossa, and variable skull base dysplasia.24,30 Epidemiological studies of CM-I suggest that approximately 0.56%–0.97% of the population shows > 5 mm tonsillar herniation on MR imaging.2,10,27 Either explicit or vague neurological symptoms attributable to hindbrain herniation are present in 63%–69% of patients with radiologically proven CM-I.2,10 Presenting symptomatology is highly variable and includes headache syndromes and brainstem and/or cerebellar dysfunction, including cranial nerve signs, neck pain, and spinal cord dysfunction.24 Classically, these symptoms are nonspecific, but can progress, resulting in the need for surgical management weeks to months after development of the first symptom.22,25,26 Clinical presentation also varies with age. Young children tend to present with oropharyngeal symptoms and older patients tend to present with headaches and progressive sensory or motor findings.1,2,13,15,26

Although the classic symptom pattern is evident for most cases, case reports describe 8 adult and 9 pediatric patients with various focal neurological findings includ-
Chiari malformation Type I with acute neurological deficit

ing esotropia, unlar neuropathy, carpal tunnel syndrome, focal motor deficits, vertigo, motor weakness and respiratory

decline, hearing loss, and trigeminal neuralgia presenting progressively over a subacute to chronic time course.⁴,⁵, ⁸, ¹⁰–¹⁸, ₂⁸, ²⁹, ³¹, ³⁴, ³⁵, ³⁷

A smaller number of case reports describe CM-I presenting with neurological deficits including quadriaparesis, balance difficulty, and hemiparesis over a 24- to 72-hour period.⁴⁹, ⁵⁶

Methods

We reviewed clinical records, imaging studies, and operative notes from all patients undergoing posterior fossa decompression for CM-I at St. Louis Children's Hospital from 1990 to 2008. Approval by the Human Research Protection Office at Washington University in St. Louis was obtained prior to the initiation of this study. Of the 189 patients who underwent surgery, 6 (3.2%) were identified with acute onset of a neurological deficit at presentation. Whereas a total of 189 patients underwent posterior fossa decompression from 1990 to 2008, records for those patients treated without surgery were only reliably present from 1994 to 2008, during which time 498 patients with CM-I were examined at St. Louis Children's Hospital. The lack of accurate records throughout the entire time period precludes precise determination of the proportion of all patients with CM-I presenting with acute onset of symptoms.

Results

All 6 children (age range 3–14 years, 3 boys, 3 girls; Table 1, Fig. 1) demonstrated spinal cord changes and CM-I on initial MR imaging. Five patients had syringomyelia, and 1 had T2 hyperintensity at the cervicomedullary junction. One patient showed T2 changes in the brainstem (Fig. 1G). Three patients presented after a clear history of minor trauma (1 with paraparesis, 2 with sensory deficits). One patient reported a mild blow to the head during a flag football game, another reported a fall from standing immediately preceding onset of symptoms, and the third patient performed a flip on a trampoline, landing on her feet prior to the onset of symptoms.

Three patients presented without a clear history of trauma (1 with abrupt onset of spontaneous dysphagia and ataxia, 2 with sensory deficits). One patient in this group reported a vague history of falls, but no clear blow to the head or neck. Decompression was performed at a mean 7.7 ± 4.9 days after symptom onset, and at a mean 7.0 ± 1.6 days after neurosurgical evaluation. In addition to suboccipital craniectomy, duraplasty was performed in each case. In 1 patient, symptoms had resolved by the time of surgery; in the remainder of the patients, clear improvements were noted within 2 weeks of surgery, with complete resolution of symptoms by 12 months postoperatively. Follow-up MR images were obtained in 4 patients, demonstrating improvement in the syrinx in each patient (Fig. 1).

Discussion

In this paper we describe our experience with acute onset of neurological deficits in 6 patients subsequently found to have CM-I. Five of these patients had an associated syrinx in the cervical spinal cord. The sixth patient had T2 hyperintensity in the cervical cord, a state that has been suggested by several authors to be a presyrinx state.¹¹, ¹⁹, ²⁰ In the extant literature, a total of 4 pediatric patients are described with acute onset of focal neurological symptoms secondary to CM-I with or without precipitating trauma.⁹, ³⁶ While the retrospective nature of this report prohibits an estimation of the true risk of acute neurological deficit in the setting of CM-I and/or syringomyelia, we believe that the proportion of patients with CM-I presenting after head and/or neck trauma with relatively acute neurological deficits may be underreported.

The technical details of surgical management of CM-I are well known, and will not be discussed here. Indications for surgery include symptoms from brainstem dysfunction, spinal cord dysfunction, and/or headaches localized to the occiput and worsened with a Valsalva maneuver. Given these indications, operative management provides excellent results.¹⁵, ³², ³⁶ In this paper, we present 6 pediat-

### TABLE 1: Patients presenting to St. Louis Children's Hospital with acute neurological symptoms from CM-I*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Days Btwn Sx Onset &amp; Op</th>
<th>Evidence of Syrinx</th>
<th>Trauma</th>
<th>Neurological Deficit</th>
<th>FU (mos)†</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>12, F</td>
<td>2</td>
<td>yes</td>
<td>history of falls</td>
<td>quadriparesis</td>
<td>26</td>
</tr>
<tr>
<td>2</td>
<td>13, F</td>
<td>5</td>
<td>yes</td>
<td>no</td>
<td>lt UE &amp; LE paresthesias</td>
<td>12</td>
</tr>
<tr>
<td>3</td>
<td>3, M</td>
<td>3</td>
<td>no, but showed increased T2 signal intensity in the spinal cord</td>
<td>no</td>
<td>vocal cord paresis</td>
<td>14</td>
</tr>
<tr>
<td>4</td>
<td>14, M</td>
<td>14</td>
<td>yes</td>
<td>blow to head during football game</td>
<td>rt UE paresthesia</td>
<td>11</td>
</tr>
<tr>
<td>5</td>
<td>10, M</td>
<td>14</td>
<td>yes</td>
<td>fall from standing</td>
<td>paraparesis</td>
<td>24</td>
</tr>
<tr>
<td>6</td>
<td>13, F</td>
<td>8</td>
<td>yes</td>
<td>flip on trampoline, landing on feet</td>
<td>rt hemianesthesia</td>
<td>5</td>
</tr>
</tbody>
</table>

* FU = Follow-up; LE = lower extremity; UE = upper extremity.
† In all patients, a complete resolution of symptoms was found by 12 months postoperatively.
ric patients who experienced the acute onset of focal neurological symptoms. The majority of our patients (5 of 6) presented with a syrinx (Table 1). This correlation, while by no means definitive, leads us to suggest that this group may be a higher risk group for development of acute neurological deficits with relatively minor head or neck trauma than patients with CM-I as a whole. With appropriate early surgical intervention, the prognosis for resolution of acute mild to moderate deficits is excellent. All 6 patients in our series experienced resolution of symptoms at follow-up.

Chiari malformation Type I is known to cause significant compression of the brainstem and cervical spinal cord. Additionally, some authors have posited that symptoms in patients with CM-I may be aggravated by increased intracranial pressure, and that CM-I itself may alter the normal capacity of CSF for moderating increasing intracranial pressure. Callaway et al. argued in 1996 that sports should be avoided in patients with CM-I and syringomyelia, indentation of the medulla, or obliteration of the subarachnoid space. Vaccaro et al. similarly argued that sports are contraindicated in patients with CM-I and basilar invagination. Miele et al. as well as Bailes and Cantu argued that CM-I found on concussive evaluation and symptomatic CM-I should also be a contraindication to contact sports. Despite these recommendations, none of these authors have defined which sports are of particular concern. While the current study is small, our population does have several patients that were injured and found to have a CM. At the very least, we believe that this relationship requires further study.

Return-to-play guidelines for neurological diseases are controversial because there is a dearth of evidence-based recommendations and a lack of guidelines from any national governing body. The senior author in this study counsels patients with CM-I and a syrinx to discontinue contact sports until treatment. Patients with CM-I and a syrinx need to be counseled about a possible increase in risk of neurological injury with minor trauma. After surgical decompression, return-to-play counseling should be carefully considered, as there is little guidance from the literature. Imaging confirming resolution of syringes in the spinal cord and brainstem may provide adequate reassurance that contact sports may be resumed. Alternatively, clinical improvement after surgery and an appropriate postoperative period of restricted activity may also prove sufficient for advancement to unrestricted activity.

Conclusions

Chiari malformation Type I can rarely present with acute neurological findings with or without precipitating trauma. Once identified, early surgical management leads to excellent outcomes. Return to contact sports for patients with CM-I and associated syringomyelia should be carefully considered by the treating physician, and may involve imaging postoperatively.
Chiari malformation Type I with acute neurological deficit

Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: Powers, Smyth. Acquisition of data: Yarbrough, Powers. Analysis and interpretation of data: Yarbrough, Powers, Limbrick. Drafting the article: Yarbrough. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors. Statistical analysis: Yarbrough. Administrative/technical/material support: Yarbrough. Study supervision: Park, Leonard, Smyth.

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