Spinal cord injury without radiographic abnormality and Chiari malformation

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Spinal cord injury without radiographic abnormality and asymptomatic Chiari I malformation have an unusual coincidence. A young boy who had recently fallen from his porch was transferred to the neurosurgery service with a high cervical central cord syndrome. Careful study demonstrated no radiographic abnormality and, although the patient was previously quite well, magnetic resonance imaging revealed Chiari I malformation. Although expectedly uncommon, reports of three other similar cases support a less than independent relationship between these two processes. All four children, each aged 2 years, were premorbidly asymptomatic and were playing when they fell from low elevations; two were on a couch. All were evaluated by primary authorities 12 to 48 hours before definitive admission, and all had normal plain film examinations. Three of the four children suffered injuries in flexion, the fourth in extension. Three realized a 5-minute to 3-hour delay before the onset of symptoms, and three suffered gradual progression of deficit. Magnetic resonance imaging was the most commonly applied and productive diagnostic medium, demonstrating cerebellar ectopia in three of three cases. Two children were surgically treated, and all achieved at least a functional outcome. Similarities among these cases support a common mechanism of injury, and indicate careful counseling in children with asymptomatic Chiari I malformation and consideration of operative decompression in those children with progressive neurological injury and deficit.

KEY WORDS • spinal cord injury • pediatric spine • Chiari malformation • magnetic resonance imaging • children

Spinal cord injury is a common cause of morbidity and mortality in the United States. Large medical centers can experience well over 100 cases annually, however, spinal cord injury in the pediatric population, specifically between birth and 12 to 16 years of age, is relatively uncommon. Whether decreased exposure, increased resilience, or a combination of the two should be credited is as yet subject to speculation.

The pediatric population suffers its share of automobile accident-related injuries (especially considering automobile-pedestrian events), and has more falls from heights than the adult population. The traumatic forces involved can be quite severe, but even otherwise seemingly trivial injuries can lead to disastrous results.

Pediatric spinal cord injury tends toward greater consequence than its adult counterpart: injury is more often at higher vertebral levels, resulting in more severe disability; deficit more often shows delayed onset, making diagnosis more difficult; and spinal column disruption is more often at the cartilaginous endplates, causing deformity to be more threatening. In addition, pediatric spinal cord injury more often demonstrates radiographic abnormality. This further complicates diagnosis, treatment, and especially outcome since spinal cord injury without radiographic abnormality (SCIWORA) is more often complete.

Congenital cerebellar ectopia occurs rarely and is frequently grouped according to the extent of medullocerebellar displacement and associated anomalies. The eponymous Chiari I malformation describes the more benign case and includes those patients with displacement of non-neoplastic cerebellar tonsillar tissue below the foramen magnum but without fourth ventricular deformation or encephalocele. Symptoms and signs typically suggest dysfunction of cerebellar cortex and tracts, cervicomedullary long tracts, medullary cranial nerves and their nuclei, medullary vegetative centers, regions indirectly involved through the occasionally associated syrinx or hydrocephalus, and the activation of dural pain fibers. Although more often insidious, Chiari I malformation without syrinx or hydrocephalus can produce sudden effects during even physiological motion of the cervical spine. For any manifestations to appear before the second decade of life is, however, quite uncommon.

The incidence of SCIWORA in pediatric patients with Chiari I malformation would be quite small if each...
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Case Report

This 27-month-old Caucasian boy, with an otherwise uneventful gestational, delivery, and developmental history, was playing outside his parents’ house when he chased a dog off an elevated porch. The child fell 11 inches to the sidewalk below, landing on his chin and chest; he rolled over, sat up, and cried, apparently demonstrating good strength at the time. The boy’s father lifted him to the porch, walked him inside without difficulty, and lifted him onto a counter to be consoled. After approximately 5 minutes, the child began to move his left arm less than his right; after approximately 10 minutes, both arms lay limply at his side. He described no sensory complaints other than a tender chin. When lifted to the floor again, approximately 15 minutes after the fall, the child’s legs were unable to support his weight, prompting immediate transportation to the nearby emergency facility. After evaluation including a cervical spine radiography series, the patient’s cervical spine was considered “clear,” and he was admitted for overnight observation with the diagnosis of bilateral brachial plexus palsies. When the boy was slow to awaken the next morning and found to have paradoxical respiratory effort, he was referred to the University of Missouri Hospitals and Clinics for further care.

Examination. The child arrived in his parents’ arms with tachypneic (28 breaths/min) paradoxical respirations; he was distressed but demonstrated little other movement. His oral temperature was 38.6°C; no previous temperature elevation was reported. General examination revealed no traumatic lesions other than a small skin abrasion on his chin, no inflammation of the oropharyngeal mucosa or middle ear, and no tenderness of the cervical spine to spinous process palpation. His lungs showed rapid but diminished breath sounds punctuated by occasional upper airway rhonchi of collected secretions. On further examination, his cry was appropriate but weak and breathless in quality. The patient followed commands with his eyes and face; he had no cranial nerve deficits. He did not move his extremities either spontaneously or to command. The child flexed his lower extremities to pain; his upper extremities showed no response. He grimaced with pinprick in the lower extremities, requiring deeper pain for similar response in the upper extremities. Deep tendon reflexes were diminished in the lower extremities and absent in the upper extremities. Plantar responses were extensor, and there was no clonus at the ankles. His bladder was enlarged with urine and required catheterization. He was placed in a stiff cervical collar and slated for further laboratory and radiographic evaluation. The patient was scheduled for magnetic resonance (MR) imaging; however, he required endotracheal intubation for mechanical ventilatory support and airway care in the meantime.

Chemistries and urinalysis, including drug screen and latex agglutinations, were normal or negative. The blood leukocyte count was elevated to 14,600 cells/cu mm, with an excess of polymorphonuclear cells. The sputum harbored Haemophilus influenzae. Lumbar puncture was avoided until after radiographic and MR imaging studies.

Chest radiography revealed right lower lobe atelectasis which cleared with mechanical ventilation. Plain radiographs of the cervical spine revealed no fracture or subluxation. Flexion and extension views revealed no motion beyond physiological limits. A computerized tomography (CT) scan of the head was normal. Magnetic resonance imaging of the brain and spine revealed normal intensity configurations save for the craniocervical junction (Fig. 1). Cerebellar tonsillar tissue extended through the foramen magnum to the C-1 level. The lower medulla and cervical spinal cord were diffusely enlarged, tapering to normal at the cervicothoracic junction. Otherwise, the neural elements remained isointense on T1-weighted, T2-weighted, and gadopentetate dimeglumine-enhanced T1-weighted MR images.

Treatment and Outcome. The child was maintained in a customized stiff cervical collar and was treated aggressively for his pneumonitis. He was extubated on Day 11 after admission, allowing more intensive rehabilitation efforts. The patient was encouraged to wear the collar at all times for 12 weeks and his compliance seemed very good. At 9-month follow-up examination, the child was playful and mischievous. His neck was nontender, and its range of motion was normal. He had regained full and symmetrical strength in his lower extremities, continuing to chase the dog and use his feet.
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to unwrap candy. His upper extremities, however, re-
mained weak symmetrically, with less than antigravity
strength in his hands and biceps, triceps, and rhomboid
muscles; his deltoid muscles had only flicker strength.
Pain sensation was normal, and cerebellar function
was normal where strength allowed. Deep tendon re-
flexes were brisk in the lower extremities and attenuated
in the upper extremities; plantar responses were exten-
so. Although he had been weaned from diapers before
his fall, he continued to fail attempts to be weaned
again. At 30-month follow-up evaluation, the child's
motor function had continued to improve, especially
in the deltoid muscles which had returned to antigravity
strength. Deep tendon reflexes had become brisk in the
upper extremities. Although the patient had regained
fetal continence, he continued to have “occasional acci-
dents” holding his urine.

Discussion

Pediatric Spinal Cord Injury

Pediatric SCIWORA in a patient with otherwise
asymptomatic cerebellar ectopia is an unusual occur-
rence. Approximately one-half of nonpenetrating pe-
diatric spinal column injuries (musculoskeletal and/or
central nervous system (CNS)) manifest neurological
deficit; however, these constitute less than 10% of cord
injuries across all ages.\(^{10,12,23,24}\) Estimates of less
than 1% to greater than 9% have been reported, re-
vealing variability based mainly in pediatric age defi-
nition inconsistencies.\(^{12,16,24}\) Borderline early-teenaged
groups tend to have a relatively greater number of
traumatic cord injuries, swinging counts according to
their inclusion.\(^{10,23}\) Of nonpenetrating pediatric spinal
cord (CNS) injuries, those without radiographic abnor-
mality are reported to make up from 1% to 67%,\(^{10,12,16,23}\)
recently pared to 10% to 20%.\(^{6}\) Although inclusion age
again surely plays a large role in such variability, criteria
for and sophistication of analysis for determination of
radiographic normalcy is inconsistent as well.\(^{14,16}\)

Terminology in reference to spine (musculoskeletal
and/or CNS) injury can be quite challenging. The terms
“spine,” “spinal column,” “cord,” and “spinal cord”
can be easily misinterpreted. Especially in SCIWORA
where injury to the CNS may present in the absence of
apparent musculoskeletal injury, care should be exer-
cised in specifying neural injury, musculoskeletal in-
jury, or both.

Cerebellar Ectopia

Chiari I malformation without hydrocephalus or syr-
inx is seldom encountered during the first 10 years of
life and, for the most part, only then when symppto-
matic.\(^{4,13,15,19}\) With its rarity and a natural history as yet
mysterious, the incidence of asymptomatic pediatric
cerebellar ectopia is difficult to determine and is, at
best, the result of random survey and incidental finding
(RJ Lemire, personal communication, 1992).

Similar Cases

Independent distribution of pediatric SCIWORA and
otherwise asymptomatic Chiari I malformation would
be expected to yield a low coincidence rate. Indeed,
reported cases are few. The absolute number of cases
described, however, and the similarities among these
cases favor a less than independent relationship between
these two uncommon entities. Reported cases include
two children similar to our case and one child with
cerebellar ectopia and a nondisplaced atlas fracture on
CT.\(^{8,22,25}\) The four cases are summarized in Table 1.

The Importance of Age

The children studied show surprising conformity.
The gender ratio was 1:1, and all subjects were premor-
bidity well. All four children, moreover, were aged 2
years when their injuries occurred. Mechanisms postu-
lated for pediatric SCIWORA focus on the maturing
cervical spine, including its bone elements, ligaments,
muscles, and vessels, and their relationship to the rel-
atively large head they support and especially to the
young cord they enclose.\(^{3,6,10,16,23}\) Whereas the laxity of
the pediatric cervical spine may add some protection
against fracture and/or dislocation, the relatively unfor-
giving cervical cord suffers therefrom.\(^{6}\) Age underlies
much of the debate. Although children 8 years of age
and younger show incomplete ossification, those younger than 3 years of age tend toward an even
more immature pattern of injury.\(^{23}\) Both the quality
and quantity of traumatic forces to which children are
exposed differ from those of adults, and these differ-
ences play an important part in the relative infrequency
of pediatric spinal cord trauma.\(^{10,23}\) As carefully de-
scribed for one of the reported cases,\(^{22}\) the forces blamed
for all four injuries studied herein were relatively mild.
Interestingly, SCIWORA in the general population
tends to follow relatively more severe trauma.\(^{10}\) All four
children studied suffered falls, and the falls tended to
be from heights the child could reach unaided. Indeed,
two of the four children fell while playing on a couch.
The maturing child marks his/her developmental mile-
stones by leaps in mobility. The temporal junction
between this fresh but incoordinated play and the mat-
turation of the cervical spine lagging far behind puts the
young child at a decided natural disadvantage. Indeed,
when the cervical spinal cord is adjoined by ectopic ce-
rebellar tissue, this disadvantage seems to peak during
the 3rd year of life.

The Mechanism of Injury

Clues discovered in the description of each child's
injury are helpful in assigning a biomechanical displace-
ment pattern for each case. Whereas our patient landed
prone with trauma to his chin, extension injury is
suspect. Two children landed supine, suggesting flexion
injury, and the fourth child was found trapped in flex-
ion. Ligament folding may compromise the diameter
of the spinal canal in extension. In the cases studied,
however, flexion injury is more commonly described.
With such displacement, cord traction forces may be
to blame, although here again the canal compromise
from subluxation in lax spines may be involved.\(^{14}\)
Moreover, immature vasculature may be compromised
regardless of displacement.\(^{3}\) In either extension or
flexion, a mass of ectopic cerebellar tissue can worsen

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the compression forces, traction forces, and vascular compromise the child suffers.

**Presentation of SCIWORA**

Immediate paresthesias and subjective weakness, usually transient, can herald SCIWORA in some cases. Definite objective deficits often materialize only after a delay period that can be otherwise asymptomatic.\(^6,20,22\) Only one of the four children studied described immediate discomfort in his feet and toes;\(^22\) the remaining three apparently did not experience such herald sensations. These three children had 5-minute to 3-hour delays before the onset of their objective deficits. Indeed, the child who developed lower extremity paresthesias following his injury demonstrated only an "unusual gait" in the 2 hours before more objective weakness emerged, indirectly supporting the delayed pattern. Once the onset of symptoms had occurred, all of the children suffered a protracted course of deterioration lasting from 2 to 24 hours. Three children realized a gradual decline; although the fourth patient may also have experienced a gradual decline, the course may have been recognized as, and therefore described as, a stepped pattern.\(^3\) Although delayed onset is more simply explained as recurrent injury with each subsequent event predisposed to more severe injury by the previous event;\(^1,6,16,20,22\) the gradual course followed by most of the children fails to support this notion as the sole answer. Once a slowly expanding mass has been excluded, humoral and ischemic forces are suspect as guiding a gradual course.\(^3,6,17,22\) Indeed, an initial injury may set in motion a gradual deterioration punctuated by, sometimes accelerated by, and perhaps even facilitating subsequent injury.

The infrequency of pediatric spine trauma and more importantly the occasional delayed symptom onset can make early diagnosis difficult. All four children were referred for definitive evaluation between 12 and 48 hours after initial injury. Two were given working diagnoses other than cord injury, and two were discharged to home prior to referral. Even the most subtle symptoms should be carefully investigated.\(^1,6,20,22\)

The children demonstrated two patterns of presentation: greater weakness in the legs than the arms in one case and greater arm than leg weakness in three. One child (Case 3) showed signs suggestive of lower extremity spastic paraparesis. Interestingly, upper extremity weakness in excess of lower extremities.\(^1\) Arm weakness in excess of leg weakness requires careful anatomical consideration. Bilateral brachial plexus injury can produce profound weakness in the upper extremities, and concurrent root avulsion injury to the cord can lead to less pronounced, transient lower extremity findings. Dissociation of motor and sensory function in the arms would, however, be unlikely.\(^1\) Cervical central cord syndrome can, as well, lead to upper extremity weakness in excess of lower extremity weakness, although once again involvement of spinothalamic crossing fibers would make at least pinprick sensation dissociation less likely.\(^2\) Case 2 was described as having a cervical central cord syndrome and, indeed, our case (Case 1) demonstrated a similarly suggestive pattern; however, our patient real-

### TABLE 1

Comparison of cases studied*

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Case 1</th>
<th>Case 2(^1)</th>
<th>Case 3(^2)</th>
<th>Case 4(^3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>age (yrs), sex</td>
<td>2.25, M</td>
<td>2, F</td>
<td>2.5, M</td>
<td>2.75, F</td>
</tr>
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<td>premorbid status</td>
<td>well</td>
<td>well</td>
<td>well</td>
<td>well</td>
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<tr>
<td>event</td>
<td>fall from porch (11 in.)</td>
<td>fall from couch</td>
<td>handstand from couch</td>
<td>fall from bed (18 in.)</td>
</tr>
<tr>
<td>first complaint</td>
<td>extension</td>
<td>flexion</td>
<td>flexion</td>
<td>flexion</td>
</tr>
<tr>
<td>symptom delay</td>
<td>5 min</td>
<td>2 hrs</td>
<td>0</td>
<td>3 hrs</td>
</tr>
<tr>
<td>type of course</td>
<td>gradual</td>
<td>gradual</td>
<td>gradual</td>
<td>stepped</td>
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<tr>
<td>time course (hrs)</td>
<td>24</td>
<td>12</td>
<td>2</td>
<td>7</td>
</tr>
<tr>
<td>referred</td>
<td>plexus</td>
<td>GBS</td>
<td>discharge</td>
<td>observe</td>
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<tr>
<td>time to referral (hrs)</td>
<td>12-24</td>
<td>12-24</td>
<td>48</td>
<td>32</td>
</tr>
<tr>
<td>weakness pattern</td>
<td>arm &gt; leg</td>
<td>arm &gt; leg</td>
<td>arm &lt; leg</td>
<td>arm &gt; leg</td>
</tr>
<tr>
<td>pinprick test</td>
<td>accurate</td>
<td>—</td>
<td>accurate</td>
<td>accurate</td>
</tr>
<tr>
<td>respiratory dysfunction</td>
<td>yes</td>
<td>—</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>micrurition dysfunction</td>
<td>yes</td>
<td>—</td>
<td>yes</td>
<td>no</td>
</tr>
<tr>
<td>plain x-ray film</td>
<td>normal</td>
<td>normal</td>
<td>normal</td>
<td>normal</td>
</tr>
<tr>
<td>myelography</td>
<td>—</td>
<td>—</td>
<td>Chiari malformation</td>
<td>—</td>
</tr>
<tr>
<td>computerized tomography</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>fracture</td>
</tr>
<tr>
<td>MRI of foramen magnum</td>
<td>Chiari malformation</td>
<td>Chiari malformation</td>
<td>—</td>
<td>Chiari malformation</td>
</tr>
<tr>
<td>MRI of cord</td>
<td>abnormal</td>
<td>normal</td>
<td>surgery</td>
<td>—</td>
</tr>
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<td>surgery</td>
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<td>meningiitis</td>
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<tr>
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<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>outcome†</td>
<td>useful motor function</td>
<td>useful motor function</td>
<td>recovery</td>
<td>useful motor function</td>
</tr>
</tbody>
</table>

* MRI = magnetic resonance imaging; GBS = Guillain-Barré syndrome; — = none/not described.
† Outcome defined according to Frankel, et al.\(^9\)
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ized at least partial motor and sensory dissociation with preservation of upper extremity pain sensation. Unfortunately, the sensation level in Case 2 was not described. Selective anterior cervicomedullary injury can, in theory, produce varied weakness patterns whether uncrossed corticospinal fibers or differentially crossing corticospinal fibers are to blame in humans. Moreover, such weakness can spare sensory function. An anterior cervicomedullary paralysis, as a single unifying theme, can be applied to each child's presentation; one case bears probable MR imaging confirmation; the case described as cervical central cord syndrome may indeed have only medullary involvement if sensory sparing was realized; and the child with spastic paraparesis may have suffered more caudal medullary injury.5,8,22 Cerbellar ectopia is common to all four children, including our patient. Although the early attenuation of sensation and reflexes in the upper extremities and the diffuse cervical cord enlargement on MR imaging tend to support some component of cervical central cord injury, and although the early excess deltoid muscle weakness tends to support some component of lower motor neuron injury, the return of sensation in excess of strength in our patient also supports anterior cervicomedullary injury.

Radiography and Magnetic Resonance Imaging

Plain cervical radiographs were considered normal for each child. Early study of SCIWORA would consider such normality sufficient for inclusion as a SCIWORA; the roles played by myelography and CT in the search for pathology remain uncertain.17 Chiari 1 malformation was discovered by myelography in one case and nondisplaced fracture was demonstrated on CT in another. Strictly defined, both are radiographic abnormalities although they need not necessarily be considered part of the initial evaluation. Magnetic resonance imaging of the cervical spine was, aside from plain radiography, the most commonly applied diagnostic medium. Three children were studied by MR imaging and in all three cerebellar ectopia was demonstrated on the images. Moreover, imaging of the spinal cord proper was quite revealing. The MR image for one child (Case 4) demonstrated spinal cord intensity changes thought to add anatomical confirmation to the theory of Bell's cruciate paralysis.8 The images for another child (Case 2), described as having cervical central cord syndrome but postulated above to suffer anterior cervicomedullary injury, failed to demonstrate cervical spinal cord changes, thus lending support to the latter.22 Intensity changes in both the medulla and the spinal cord were demonstrated on MR imaging for our patient, who presented with clinical symptoms of both cervical central cord syndrome and anterior cervicomedullary paralysis. Not only was MR imaging the most commonly used and accurate diagnostic medium at the foramen magnum, it also aided the understanding of the neuroanatomical pathology of the lesions involved. With increasing use of MR imaging in the evaluation of CNS trauma, more cases of spinal cord injury with only magnetic abnormality may be discovered.

Surgical Intervention

Surgical and nonsurgical treatment of the neurologically injured children with SCIWORA and Chiari 1 malformation was evenly distributed. Two children either underwent surgery or their caregivers strongly recommended surgical intervention despite the absence of direct description.22,23 The remaining children were either treated with external cervical immobilization or surgical intervention was deemed unnecessary. In follow-up evaluation, all of the children regained at least useful motor control, and the child who underwent operative decompression regained function described as normal. Unfortunately, this child suffered the only mentioned treatment complication, postoperative meningitis.23 Progressive neurological injury and deficit remain an indication for operation. With intervention in each case guided by the individual's evaluation and clinical course, overall outcome from such injury seems quite good.

Conclusions

Spinal cord injury without radiographic abnormality in children with otherwise asymptomatic Chiari I malformation is quite an unusual occurrence, but the cases described herein support a less than independent relationship between the two entities. Such injuries tend to occur during the 3rd year of life, to occur in flexion with relatively mild forces, to produce a delayed onset and gradual course of neurological dysfunction, and not to produce early hallmark sensation. Each child's presentation pattern can be explained by either incomplete transverse cervical cord injury or central cervical cord injury. A single unifying theme, however, may emerge with closer evaluation. The pattern of motor and sensory deficit and the intensity changes on MR imaging support the likelihood of anterior, perhaps quite focal, cervicomedullary lesions. Moreover, Chiari I malformation may predispose children to such injury.

The child with otherwise asymptomatic Chiari I malformation may indeed bear increased risk for CNS injury, especially during the immaturely active 3rd year of life. Those discovered before difficulties arise may benefit from careful counseling and close follow-up evaluation. The young child who presents even after the most trivial cervical trauma should be carefully evaluated for suggestion of injury. This becomes more poignant, of course, when neurological deficit, however mild, taints their presentation; subtle, even transient subjective discomfort should not be overlooked. For children with neurological deficit whose initial studies are unrevealing, MR imaging should be strongly considered. Although outcome seems at least functional despite intervention, the implication of ectopic cerebellar tissue in a child with progressive neurological deficit and deficit would support operative decompression of this intradural, extramedullary mass.

References

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