Delayed postoperative tethering of the cervical spinal cord

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Tethering of the spinal cord in the lumbar and sacral regions of children with congenital anomalies is a well-recognized problem; however, tethering in the cervical region has rarely been reported. A search of the literature revealed no reports of symptomatic postoperative cervical spinal cord tethering. The authors present five cases of delayed postoperative cervical spinal cord tethering and discuss the benefit of detethering in these patients. All five patients were young (16 to 42 years of age) at presentation. All had done well after an initial surgical procedure but returned between 1 and 31 years postoperatively with symptoms including severe headache, upper-extremity pain, and progressive neurological deficits. In each case, magnetic resonance imaging indicated dorsal tethering of the cord in the cervical region. Surgical exploration with microscopic sharp detethering of the cervical cord was performed on each patient with favorable results. To avoid re-tethering, wide Tutoplast duraplasty is recommended.

Key Words: tethered spinal cord • Chiari malformation • cavernous malformation • syringomyelia

Symptomatic tethering of the spinal cord is well known in the lumbar and sacral region and occurs primarily in children with congenital anomalies, including spina bifida, myelomeningocele, lipoma of the cord, diastematomyelia, and thickened short filum. These patients are typically stable with non-progressive neurological deficits for many years following surgical treatment for the underlying disorder, but later often present with pain or loss of neurological function or worsening scoliosis. It is postulated that, during childhood, growth of the spinal column with a fixed cord terminus produces stretching and tension on the delicate neural tissues of the spinal cord, which causes neurological deterioration by mechanical distortion and/or cord ischemia.

Tethered spinal cord has also been described in adults related to occult congenital anomalies, fibrous adhesions, and trauma. In these cases magnetic resonance (MR) imaging has allowed visualization of the spinal cord with a typically tethered appearance. Many surgical series have included favorable results with respect to microsurgical detethering, with arrest of neurological deterioration and relief of pain.

To our knowledge, there have been no reports of postoperative tethering in the cervical spine, although Eller, et al., described congenital cervical cord tethering in a previously unoperated patient found to have spina bifida occulta at C-1. This 23-year-old woman presented with headaches, positional neck pain, and progressive sensory deficits with mild weakness. She responded well to operative detethering. Berrington reported a case of posttraumatic cervical spinal cord tethering in a 53-year-old woman who exhibited progressive neurological deficits including bulbar symptoms 3 years after being rendered quadriplegic at the C-7 functional level. Her symptoms responded favorably to detethering by cord transection and lysis of the adhesions.

We present five patients with delayed postoperative tethering of the cervical spinal cord who responded favorably to further surgical intervention. These patients were young (aged 16, 19, 27, 33, and 42 years), each presenting with pain and progressive neurological complaints 1 to 31 years after intradural intramedullary surgical procedures in the cervical region. Magnetic resonance images were consistent with dorsal tethering of the spinal cord in each case; however, these studies were not read as such by the neuroradiologists. All patients experienced resolution of both pain and neurological complaints after undergoing surgical detethering and duraplasty.
Case Reports

Case 1

This 16-year-old girl had a history of Chiari I malformation and syringomyelia for 1 year prior to the current presentation. She had been treated with suboccipital craniectomy, C1-2 laminectomy, syringosubarachnoid shunt, and duraplasty. She had done well after the surgery and had no neurological deficits; however, 1 year later she presented with a 2-month history of severe suboccipital headache, clumsiness, and progressive left-hand weakness and numbness.

Examination. The patient exhibited a 4/5 weakness in the flexor and intrinsic muscles of the left hand. Sensory examination disclosed decreased pinprick and light-touch sensation in the left forearm and hand. Mild hyperreflexia was noted, greater on the left than the right. Her gait was normal. Cervical MR imaging revealed dorsal tethering at the operative site but no recurrence of the syrinx.

Operation. At surgery, dense fibrous scarring was found, tethering the cord to the dura at C-2; the adhesions were lysed with sharp microdissection. A C-3 laminectomy was required to establish a normal plane of dissection. The cord was completely released, after which it became relaxed and ventral in position. The syringosubarachnoid shunt was also removed. A wide cadaveric dura (Tutoplast) duraplasty was then performed.

Postoperative Course. The postoperative course was favorable, with complete cessation of pain. Left-hand strength returned to 5/5 over several months, and sensory and reflex findings returned to normal. The patient continues to be asymptomatic 3 years after detethering.

Case 2

This 33-year-old woman had been born with a myelomeningocele at C3–4, which was closed surgically at birth but required three repairs for cerebrospinal fluid (CSF) leak (the last at 18 months of age). She also had meningitis which was successfully treated. At presentation she was normal neurologically, including bowel and bladder function, and was attending college. She had moderate hydrocephalus but never required shunting. She reported an 18-month history of progressive neck pain and weakness in all four extremities, greater on the right than the left. Painful dysesthesias increased with neck flexion. She also complained of an unsteady gait and clumsiness.

Examination. The neurological examination was significant for 4/5 weakness of the right deltoid, biceps, triceps, and wrist flexor, extensor, and intrinsic muscles. Right-foot dorsiflexion was also weak at 4/5. Sensation was grossly normal. Diffuse hyperreflexia and a bilateral Babinski sign were present. Gait was spastic and broad-based. Tethering of the spinal cord at C3–5 was seen on MR imaging.

Operation. At surgery, C1–5 laminectomy was performed for exposure. Scar tissue was found extending from the dorsal aspect of the cord through the dura and attached to the paraspinal musculature at the C4 level. Arachnoid bands were also found tethering the cord to the dura from C3 to C5. The scar was removed and all adhesions were lysed with sharp microdissection. The cord was relaxed and ventral in position after detethering. Wide Tutoplast duraplasty was used for closure.

Postoperative Course. The patient improved postoperatively in motor strength with minimal residual deficit graded at 5–5 because the right arm and hand muscles were slightly weaker to resistance testing than the left. Right-foot dorsiflexion returned to normal at 5/5 within several weeks of surgery. Her spasticity, hyperreflexia, and gait disturbance resolved nearly completely, and she remains asymptomatic 3 years after detethering.

Case 3

This 27-year-old man had a Chiari I malformation and syringomyelia at 17 years of age, which were treated with suboccipital decompression and a syringosubarachnoid shunt. He was asymptomatic for 10 years but then began to have headaches and noticed bilateral decreased fingertip sensation as well as shock-like sensations in the legs, exacerbated by neck flexion.

Examination. Examination disclosed normal motor strength throughout, but decreased sharp-dull discrimination in both hands and feet, worse on the right side than the left. Hyperreflexia was not apparent. Coordination and gait were normal. Tethering of the cord was demonstrated on MR imaging at the site of the shunt at C-2, and at the C-6 level (Fig. 1).
Operation. A cervical laminectomy was performed extending down to C-6. Dense arachnoid scarring was found at C2–3 and C-6 and lysed with sharp microdissection. The scar at the C-6 level was at the terminus of the syringosubarachnoid shunt, which was removed. After detethering, the cord was relaxed and ventral in position. Wide Tutoplast duraplasty was performed.

Postoperative Course. The patient’s pain was reduced postoperatively, but he had a mild residual sensory deficit in the right hand which has improved slightly. Strength remains at 5/5 in all muscle groups bilaterally. He has continued well for the 16 months since surgery.

Case 4

This 19-year-old woman, who had undergone cervical laminectomy (C1–2) for removal of a cavernous malformation of the cord at 14 years of age, had fascial lata duraplasty performed at the original surgery but required a revision for CSF leak. Nevertheless, she did well after surgery with a mild residual left hemiparesis. At 19 years of age, she began to complain of intermittent severe headaches, increasing numbness, weakness of both lower extremities, and electric shooting pains in both legs, made worse by neck flexion.

Examination. Examination revealed a stable mild spastic left hemiparesis at 4+/5, but decreased sensation and increased bilateral hyperreflexia compared with her baseline state. Dorsal tethering of the spinal cord at C2–3 was revealed on MR imaging (Fig. 2).

Operation. A C-3 laminectomy was performed to develop a safe plane of dissection. Thick scarring of the cord to the dura was present, particularly on the left dorsolateral aspect of the cord as suggested on the MR image (Fig. 2B). Multiple arachnoid bands to the dura in the region of the previous operative site were divided with sharp microscopic dissection; the cord became relaxed and ventral in position directly following lysis.

Fig. 2. Case 4. A: Sagittal T1-weighted magnetic resonance (MR) images showing the cervical spine. The abnormal cystic changes at the cervicomедullary junction are stable postoperative changes following removal of a cavernous malformation. B: Sagittal T2-weighted MR images showing the dorsolateral malpositioning of the cervical spinal cord at the C2–3 level. The cerebrospinal fluid (CSF) space ventral to the cord is excessive compared with the complete absence of any CSF signal between the cord and the dura on the left dorsolateral aspect of the cord. C: This axial T1-weighted MR sequence was particularly helpful in this case. Thick scarring of the cord causing it to adhere to the dura on the left dorsal surface of the cord was found at surgical exploration and was successfully lysed with sharp microneurosurgical technique.
of the adhesions. Wide Tutoplast duraplasty was utilized for closure.

Postoperative Course. The patient did well postoperatively, with complete relief of headaches and shooting dysesthetic pains. She returned to her baseline neurological function with a mild 4+/5 spastic hemiparesis, and has nearly normal sensory perception. She has remained asymptomatic for the 14 months since surgery.

Case 5

This 43-year-old woman was treated at another institution for syringomyelia by placement of a cervical syringosubarachnoid shunt at 26 years of age. The shunt was revised for recurrent symptoms 3 years later. At 32 years of age she exhibited an inferior extension of her syrinx, which was treated with a low thoracic shunt. At age 35 years, her Chiari I malformation was decompressed and the cervical shunt was again revised with good resolution of symptoms until about 1 year prior to being referred to our institution. She then complained of progressive sensory loss in both hands and increased left-hand weakness. As an electromyographic technician she was no longer able to type or place needles. She described severe tingling and burning dysesthesias, shooting pains into all four extremities exacerbated by neck flexion, and frequent falls due to decreased positional awareness. Neck muscle spasms were frequent and debilitating. Physical therapy, ultrasound, massage, and analgesic and antispasmodic medications were of little or no benefit.

Examination. Examination was remarkable for asymmetrical hyperreflexia of the left biceps and triceps muscles with bilaterally diminished sharp-dull discrimination in both hands. Weakness (4/5) of the left biceps, triceps, wrist flexor, and intrinsic hand muscles was present. Magnetic resonance imaging of the cervical spine (Fig. 3A) showed dorsal tethering from C-5

![Fig. 3. Case 5. A: Preoperative sagittal T₁-weighted magnetic resonance (MR) images of the cervical spine showing the dorsally displaced segment of spinal cord with a rough, scarred surface at the previous laminectomy site from C-5 to C-7. Note the large ventral cerebrospinal fluid (CSF) space directly opposite the dorsally tethered segment. Small syringal cavities are present at and below the tethered region. B and C: Sagittal T₁-weighted (B) and T₂-weighted (C) MR images obtained 3 months after cervical spinal cord detethering. Note the large CSF space dorsal to the cord afforded by wide duraplasty. The ventral CSF space is present but decreased from the preoperative images. The syrinx both at and below the detethering site appears collapsed, though neither a shunt revision nor a myelotomy was performed. There may be a small site of residual tethering present at the most rostral aspect of the duraplasty with slight residual kinking of the cord at this level; however, there is a marked improvement in the degree of cord deformation.](image-url)
to C-7 and small syringal cavities at and below the site of tethering.

Operation. Reopening the previous cervical laminectomy from C-5 to C-7 revealed the spinal cord to be densely scarred and adherent to the overlying dura; the nerve roots appeared to be stretched and under tension. Multiple arachnoid bands were sharply lysed with microscopic dissection, resulting in relaxation of the cord and nerve roots. The previously placed shunt tube was not identified and myelotomy was not performed; no attempt was made to drain the syrinx. A wide Tu-toplast duraplast was used for closure.

Postoperative Course. The patient showed marked improvement in her symptoms postoperatively. Strength in her left hand and arm has returned to near normal at 5/5, and she no longer has shooting dysesthetic sensations with neck movement or neck pain, which once required medication. At 7 months post-surgery, she no longer complains of falling or loss of balance but states that her sensation was subjectively worsened in the immediate postoperative period; she has since returned to baseline. Follow-up sagittal T2- and T2-weighted MR images obtained 3 months postoperatively (Fig. 3B and C) showed an expansive CSF space dorsal to the cord at the operative site, from C-5 to C-7 with complete collapse of the small syrinx. A small area of residual tethering may be present at the rostral end of the surgical site because there is a slight kinking of the cord remaining at this level. However, there has clearly been remarkable improvement in the degree of spinal cord deformation compared with the preoperative MR image.

Discussion

Patients with cervical cord tethering present with a delayed onset of neurological symptoms referable to the cervical region, including myelopathy, upper-extremity pain, weakness, and numbness. This syndrome may appear after various intradural procedures that involve intramedullary exploration in the cervical region. Symptoms may not occur for many years after the initial procedure. Apparently, any violation of the pial surface may lead to scar formation between neural tissue and the dura. It is unclear when scar tissue forms and why symptoms are so delayed (31 years in Case 2). All of these patients had reached their adult height prior to becoming symptomatic; therefore growth could not have caused the neurological deterioration, as is proposed for lumbar tethering in children.

We postulate that repeated movements of the head and neck with a relatively fixed spinal cord results in stretching and tension on delicate neural tissues. This mechanical distortion, along with intermittent or progressive cord ischemia as suggested by others, must play a role in causing neuronal damage. Progressive neurological deficits would then arise when the ability of the spinal cord to compensate for these repeated insults is exceeded. However, these deficits are reversible by detethering. Pain was the chief complaint from all of our patients. Electric or shock-like sensations associated with neck flexion (Lhermitte's sign) were present in all five and should alert the clinician to the possibility of cervical tethering in postoperative patients presenting with delayed onset of symptoms.

Magnetic resonance imaging proved invaluable in the diagnosis of a tethered cervical spinal cord in our patients. It must be emphasized that the neuroradiologist's initial reading of the MR images in each case described no interval change; however, after careful review by the senior neuroradiologist, the studies clearly revealed dorsal malpositioning of the cord and the appearance of tethering in the regions of the previous operative sites (Figs. 1 and 2). Specifically, in these studies the presence of excess CSF ventral to the cord (with typical signal characteristics of low intensity on T1-weighted sequences and high intensity on T2-weighted sequences) and the absence of CSF signal dorsal to the cord were diagnostic of tethered cord. This was apparent on both the sagittal and the axial images (Fig. 2). In nontethered segments, the cord returns to its typical central location in the spinal canal surrounded by CSF. The proper interpretation of the MR images led to corrective neurosurgical intervention in each of these patients.

Cases 1, 3, and 5 were treated with suboccipital decompression, syringosubarachnoid shunt, and duroplasty for a Chiari I malformation with syrinx (Table 1). One patient (Case 3) developed arachnoid scarring at both the entrance and the terminal portion of the shunt tube and required the most extensive laminectomy in our series for adequate detethering (Fig. 1). As Batzdorf advocates, we no longer use the syringosubarachnoid shunt in this procedure, and have not yet experienced symptomatic tethering in any Chiari I patient treated without a shunt. If the pial surface is not violated by introduction of the shunt, scar formation and subsequent tethering should not be possible. This is clearly only anecdotal evidence, but it supports utilizing decompression alone and avoiding the use of syringosubarachnoid shunts for syringomyelia associated with Chiari I malformations. The small syrinx present preoperatively in Case 5 (Fig. 3) completely resolved after detethering, even though no attempt was made to drain the syrinx or to revise the shunt. In some cases syrinx formation may be secondary to tethering of the cord.

Progressive neurological deficits are associated with cervical spinal cord tethering. Neurological deficits and symptomatic complaints appear to be reversible and respond well to surgical microscopic sharp detethering. All five of our patients are doing well without signs of recurrence with a mean follow-up period of 22 months. Although recurrence remains possible, with a time period from operation to delayed tethering of 31 years in Case 2, we advocate wide Tusoplastic duroplasty, as performed in our patients, to give the best chance of avoiding retethering.

Conclusions

Postoperative cervical spinal cord tethering, delayed from 1 to many years after intradural intramedullary
surgery in the cervical region, is newly described, probably underdiagnosed, and presents with a new onset of neurological complaints and progressive deficits referable to cervical spinal cord pathology. Dorsal malpositioning of the spinal cord, visible on MR imaging, in symptomatic patients is diagnostic. Microsurgical sharp detethering of the cervical spinal cord appears to be safe and allows for resolution of symptoms and return of normal neurological function. To avoid re-tethering, wide Tutoplast duraplasty is recommended. In our series there was no morbidity or mortality.

References

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Delayed postoperative cervical spinal cord tethering

TABLE 1

Profiles of five patients with cervical spinal cord tethering

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Original Presentation</th>
<th>Surgical Procedure*</th>
<th>Interval</th>
<th>Signs &amp; Symptoms</th>
<th>Surgical Procedure</th>
<th>Follow-Up Period</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>16, F</td>
<td>Chiari I, syrinx</td>
<td>suboccipital craniectomy, C1-2 laminectomy, syringosubarachnoid shunt</td>
<td>1 yr</td>
<td>headache, neck pain, spastic hemiparesis, sensory deficit, Lhermitte's sign</td>
<td>C-3 laminectomy, spinal cord detethering, shunt removal, duraplasty</td>
<td>3 yrs</td>
</tr>
<tr>
<td>2</td>
<td>33, F</td>
<td>C3-4 myelomeningocele</td>
<td>closure of myelomeningocele, CSF leak repair × 3</td>
<td>31 yrs</td>
<td>spastic hemiparesis, gait ataxia, neck pain, sensory deficit, Lhermitte's sign</td>
<td>C-1-5 laminectomy, spinal cord detethering, duraplasty</td>
<td>3 yrs</td>
</tr>
<tr>
<td>3</td>
<td>27, M</td>
<td>Chiari I, syrinx</td>
<td>suboccipital craniectomy, C1-2 laminectomy, syringosubarachnoid shunt</td>
<td>10 yrs</td>
<td>headache, neck pain, sensory deficit, Lhermitte's sign</td>
<td>C-3 laminectomy, spinal cord detethering, shunt removal, duraplasty</td>
<td>16 mos</td>
</tr>
<tr>
<td>4</td>
<td>19, F</td>
<td>Cavernous angioma C1-2</td>
<td>C1-2 laminectomy, excision of cavernous angioma, duraplasty; re-exploration for CSF leak</td>
<td>5 yrs</td>
<td>headache, hemiparesis, sensory deficit, Lhermitte's sign</td>
<td>C-3 laminectomy, spinal cord detethering, shunt removal, duraplasty</td>
<td>14 mos</td>
</tr>
<tr>
<td>5</td>
<td>43, F</td>
<td>Chiari I, syrinx</td>
<td>syringosubarachnoid shunt, revision of shunt, Chiari I decompression</td>
<td>8 yrs</td>
<td>neck pain, hemiparesis, sensory deficit, dysesthesias, Lhermitte's sign</td>
<td>C5-7 laminectomy, spinal cord detethering, duraplasty</td>
<td>7 mos</td>
</tr>
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* CSF = cerebrospinal fluid.