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Case Report

Pediatric pathology all grown up – An interesting case of adult tethered spinal cord

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ABSTRACT

Background: Cervical myelopathy in an adult is typically the result of degenerative disease or trauma. Dysraphism is rarely the cause.

Case Description: The authors report the case of a 35-year-old male drywall installer who presented with 2 years of progressive left upper extremity weakness, numbness, and hand clumsiness. Only upon detailed questioning did he mention that he had neck surgery just after birth, but he did not know what was done. He then also reported that he routinely shaved a patch of lower back hair, but denied bowel, bladder, or lower extremity dysfunction. Magnetic resonance imaging of the cervical spine demonstrated T2 hyperintensity at C4-C5 with dorsal projection of the neural elements into the subcutaneous tissues concerning for a retethered cervical myelomeningocele. Lumbar imaging revealed a diastematomyelia at L4. He underwent surgical intervention for detethering and repaired of the cervical myelomeningocele. Four months postoperatively, he had almost complete resolution of symptoms, and imaging showed a satisfactory detethering. The diastematomyelia remained asymptomatic and is being observed.

Conclusion: Tethered cervical cord is a rare cause for myelopathy in the adult patient. In the symptomatic patient, surgical repair with detethering is indicated to prevent disease progression and often results in clinical improvement.

Keywords: Cervical myelomeningocele, Cervical myelopathy, Diastematomyelia, Spina bifida, Spinal dysraphism, Tethered spinal cord

INTRODUCTION

Cervical myelopathy in an adult is typically the result of degenerative disease or trauma.[4] Dysraphism is rarely the cause. Cervical myelomeningoceles are a rare form of spinal dysraphism, accounting for <5% of all neural tube defects.[5,11] Patients may present with a myriad of signs and symptoms, including pain, weakness, paresthesia, upper motor neuron signs, muscle wasting, and bowel/bladder dysfunction.[1-3,5,12,13] Unlike myelomeningoceles occurring in the lumbar spine, cervical myelomeningoceles are typically covered by epithelium.[11,15] In the symptomatic patient, surgical repair with detethering is indicated to prevent disease progression and often results in clinical improvement. We present the case of a retethered cervical myelomeningocele in a 35-year-old who also harbored a previously unknown lumbar diastematomyelia.

CASE REPORT

History and examination

A 35-year-old male drywall installer presented with 2 years of progressive left upper extremity weakness, numbness, and hand clumsiness. Neurologic examination demonstrated 4-/5 strength of the left hand intrinsic muscles, decreased sensation to light touch in the left hand, and hyperreflexia of the left upper extremity. Electromyography and nerve conduction testing demonstrated only mild incidental bilateral ulnar neuropathy across the elbow. Magnetic resonance imaging (MRI) of the cervical spine demonstrated cervical spinal cord expansion with T2 hyperintensity at C4-C5. There was an associated defect of the bony posterior elements with projection of the neural elements into the dorsal subcutaneous soft tissue [Figure 1]. At this point, he was referred to our institution where only upon detailed questioning did he mention that he had neck surgery just after birth, but he did not know what was done. This additional history allowed the diagnosis of retethering of a cervical myelomeningocele. A cervical detethering operation was recommended.

He then also reported that he routinely shaved a patch of lower back hair, but denied bowel, bladder, or lower extremity dysfunction. This prompted an MRI of the lumbar spine which demonstrated underlying diastematomyelia with a low-lying conus [Figure 2]. His lower extremity and sacral examination were normal. Because the diastematomyelia was asymptomatic in this adult patient at final height, observation was recommended.

Operation

After satisfactory induction of general endotracheal anesthesia, the patient was positioned prone on gel rolls, and his head was fixed in Mayfield pins in neutral position. Neuromonitoring with somatosensory evoked potentials (SSEP), motor evoked potentials, and electromyography was established. The prior curvilinear midline incision was marked and slightly extended superiorly and inferiorly.

The superior portion of the incision over the last rostral intact lamina was opened sharply and carried down to the fascia using blunt dissection. Progressing along the fascia from superior to inferior, the overlying soft tissue was opened. The myelomeningocele sac was identified and dissected circumferentially as it passed through the fascia. The sac was then separated from the overlying soft tissue [Figure 3a].

The fascia was opened rostral and caudal to the myelomeningocele sac. The rostral (bifid) and caudal (hemi) dysplastic spinal lamina were removed using Kerrison rongeurs. The exposed native dura was contiguous with the myelomeningocele sac [Figure 3b and c]. Intraoperative ultrasound was used to confirm adequate bony exposure and identify dorsal subarachnoid space for dural opening.

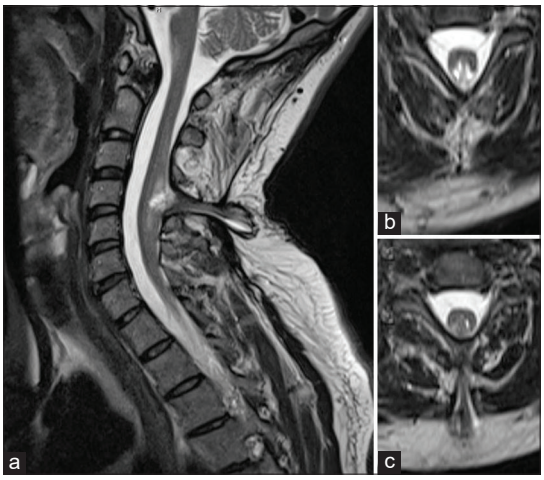

Figure 1: Preoperative sagittal (a) and axial (b and c) T2 MRI of the cervical spine demonstrating intramedullary T2 signal changes and a small syrinx associated with a dorsally projecting exophytic cervical myelomeningocele terminating in the subcutaneous fat.

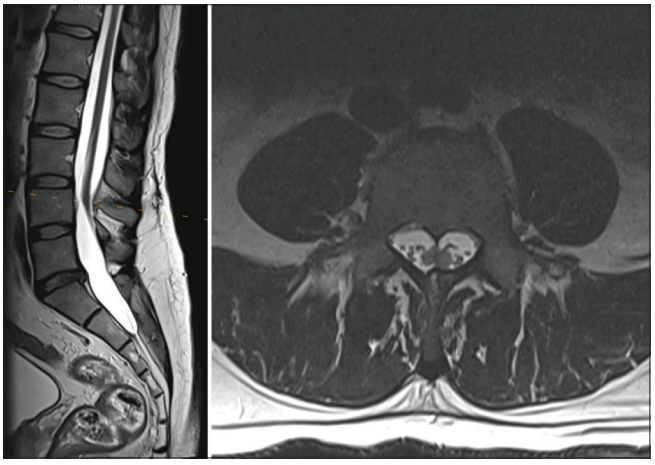

Figure 2: Preoperative sagittal (left) and axial (right) T2 MRI of the lumbar spine demonstrating a low-lying conus and associated diastematomyelia at L4.

Under microscopic magnification, the spinal cord dura was incised rostral to the myelomeningocele. Progressing caudally, the dorsal spinal cord was found to be tethered to the dura by thick arachnoid bands. These were divided as they were encountered. The dural opening eventually extended into the myelomeningocele sac. In the sac was a dorsal projection of the cervical spinal cord approximately the same size as the cervical spinal cord. It was freed circumferentially from its dural attachments. The dorsal projection terminated in dysplastic spinal cord that fused with the overlying dural sac [Figure 3d]. At the transition to dysplastic spinal cord, electrical stimulation did not result in a response in the neuromonitoring. The dorsal projection was sectioned at this

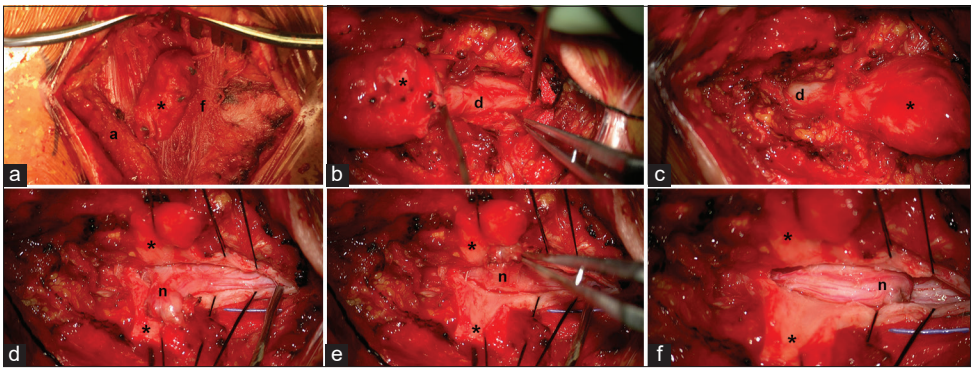

Figure 3: Intraoperative views of the cervical myelomeningocele (cranial at right and caudal at left in each image). (a) Myelomeningocele sac *(asterisk)* extending through the fascia *(f)* and terminating in the subcutaneous adipose tissue *(a)*. (b and c) Myelomeningocele emanating from the normal dura (d) and contiguous with it superiorly and inferiorly. *(d)* Dorsal projection of the neural elements *(n)* after dissection from the overlying myelomeningocele sac. (e) Pial closure of the dysplastic neural stump. (f) The free dysplastic neural stump at rest before dural closure.

location and the pial closed [Figure 3e]. The detethering was then continued inferiorly to the native spinal cord. Eventually the point of last attachment was sectioned, and the cord was free and lax.

The remaining stump of dorsal projection still needed to be addressed. Sectioning the dorsal projection closer to the native spinal cord was avoided due to fear of transiting neural tracts within the projection. Left alone, the stalk came to a resting position along the dorsal surface of the rostral spinal cord [Figure 3f]. In this position, there was sufficient redundant dura to close without compressing the projection or the native spinal cord. This was verified with the ultrasound before and after the dura was closed; there was ample subarachnoid space surrounding the neural elements.

The dura and overlying soft tissue were closed primarily in a watertight fashion. The neuromonitoring signals at the end of the operation were stable compared to the beginning. In fact, the left arm SSEP was mildly improved. There was no significant neuromonitoring signal change throughout the surgery.

Pathological findings

The dysplastic portion of the dorsal projection along with its fused overlying dura was examined [Figure 4]. Hematoxylin and eosin stained sections showed band-like collections of neuroglial tissue and meninges within a background of fibrosis, collagen fiber bundles, and disorganized smooth muscle. Scattered dermal appendages were also identified.

Postoperative course

On postoperative day 1, the patient reported improvement in his left upper extremity weakness and numbness. At 1 month follow-up, he reported continued improvement in his symptoms. He had some residual altered sensorium of the left hand, but improvement in his intrinsic hand muscle strength and no further pain, paresthesia, or clumsiness. He had returned to work without special accommodation. At 4 months follow-up, he had regained full strength in his left hand, and MRI showed a successful detethering without neural compression [Figure 5]. The diastematomyelia remained asymptomatic and is being observed.

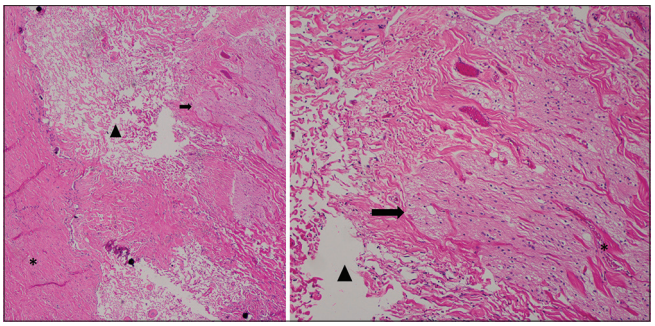

Figure 4: Hematoxylin and eosin staining of the myelomeningocele at the region of the neural-meningeal attachment. Left: low magnification study revealed fibrosis and collagen fiber bundles with prominent interfiber clefts (arrowhead), likely filled with cerebrospinal fluid. Dura is seen at left (asterisk), leptomeninges at center, and bands of neuroglial tissue (arrow) at right. Right: inspection at high magnification showed the clefts (arrowhead) between collagen fibers at left and bundles of neuroglial tissue (arrow) and meninges (asterisk) at right. Neuropil containing scattered oligodendrocytes with a dark nuclei and slight perinuclear halo is readily identified within the neuroglial tissue.

DISCUSSION

Cervical myelomeningoceles are a rare form of spinal dysraphism, accounting for <6% of all neural tube defects.[5,11] As opposed to the classic lumbar myelomeningocele, in which the neural placode is superficially exposed to the environment,

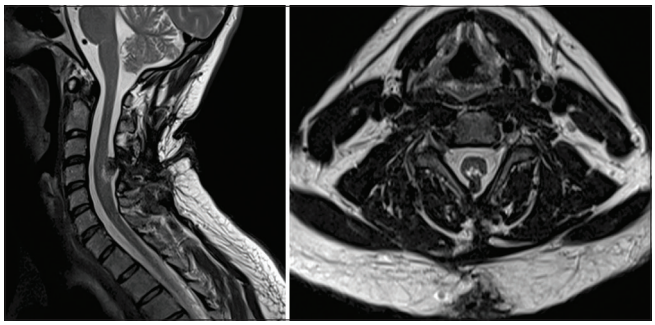

Figure 5: Postoperative sagittal (left) and axial (right) T2 MRI of the cervical spine showing successful detethering of the cervical myelomeningocele without neural compression at 4 months follow-up.

the cervical myelomeningocele typically remains covered by full-thickness epithelium.[11,15] Rossi *et al*. categorized these dysraphisms into two types: an abortive nonterminal myelocystocele in which a meningocele is traversed by a fibrovascular stalk; and, a complete nonterminal myelocystocele, in which a hydromyelia expanded cord is dorsally displaced into the dural sac.[14] In a study of 18 pediatric patients with cervical dysraphisms undergoing operative repair, Salomao *et al.* identified three subtypes: a fibrovascular stalk projecting from the spinal cord to the dural sac, a myelocystocele, and a meningocele.[15] In the present case, the myelomeningocele was comprised of a dorsal exophytic extension of native spinal cord that was tethered to the dural sac. As opposed to lumbar myelomeningoceles, the nervous tissue found in cervical myelomeningoceles is more frequently nonfunctional.[8] Regardless, the use of intraoperative neurologic monitoring may decrease the risk of neurologic injury during detethering. Intraoperative ultrasound was also found to be useful.

Tethered spinal cord in adults most often presents with pain.[9,10] The pathogenesis of delayed symptoms in adult cervical tethered cord is uncertain. It has been postulated that repeated movements in the setting of a fixed, immobile spinal cord results in ischemic insults and alterations in mitochondrial oxidative metabolism with resultant neuronal damage.[16,17] In a retrospective study of 85 adult patients with tethered cord syndrome, Klekamp *et al*. found that 61% of surgical patients demonstrated clinical improvement, while 33% reported no change in symptoms. A significant improvement in pain symptoms was not found.[10] Of 27 adult patients who underwent surgical intervention for tethered cord related to pain symptoms, Iskandar *et al*. found that at 4 years follow-up, 81% of patients reported an improvement.[9] Eller *et al*. observed an immediate increase in the amplitude of SSEP on sectioning the final band of a tethered cervical cord.[6] Smith *et al*. described a 33-year-old female with prior cervical myelomeningocele repair who developed neck pain, quadriparesis, and Lhermitte sign as a result of spinal cord tethering from scar tissue. Following detethering, she had significant improvement in her pain symptoms.[16] It appears that beyond arresting the progression of neurologic deficits, surgical detethering in the adult population often results in symptomatic improvement of pain symptoms.

There have been few reports of cervical myelomeningocele in the adult patient. Presentation may include pain, weakness, paresthesia, upper motor neuron signs, muscle wasting, and bowel/bladder dysfunction.[1-3,5,12,13] Detethering of cervical myelomeningocele has been reported to reduce symptoms and, when present, result in decreased syrinx size.[3,5,12,13] The literature suggests that surgical intervention in the symptomatic adult with tethered cervical myelomeningocele yields beneficial outcomes. In the present case, the patient had significant improvement of his symptoms following surgical detethering.

Interestingly, our adult patient also had a previously undiagnosed lumbar diastematomyelia. Other than a patch of lower back hair that was routinely shaved, the lumbar diastematomyelia was asymptomatic. When these lesions are discovered in childhood, surgery is typically recommended to lessen the risk of developing tethering symptoms, which may be irreversible, overtime.[7] However, when these lesions are discovered in adulthood, observation is typically recommended in the asymptomatic patient.[10] This raises the question of the true incidence of symptom development in lumbar diastematomyelia, however, natural history studies are lacking.

CONCLUSION

Tethered cervical spinal cord is a rare cause for myelopathy in the adult patient. In the symptomatic patient, surgical repair with detethering is indicated to prevent disease progression and often results in clinical improvement. When asymptomatic, such as with this patient's concurrent lumbar diastematomyelia, observation is prudent.

Declaration of patient consent

Patient's consent not obtained as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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