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Case Report

Marsupialization and distal obliteration of a lumbosacral dural ectasia in a nonsyndromic, adult patient

Ha Son Nguyen, Andrew Lozen, Ninh Doan, Michael Gelsomin, Saman Shabani, Dennis Maiman

Department of Neurosurgery, Medical College of Wisconsin, Milwaukee, WI, USA

Corresponding author: Dr. Ha Son Nguyen, 9200 West Wisconsin Avenue, Milwaukee, WI 53226, USA. E-mail: hsnguyen@mcw.edu

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Abstract

Dural ectasia is frequently associated with connective tissue disorders or inflammatory conditions. Presentation in a patient without known risk factors is rare. Moreover, the literature regarding the treatment options for symptomatic dural ectasia is controversial, variable, and limited. A 62-year-old female presents with intractable, postural headaches for years. A lumbar puncture revealed opening pressure 3 cm of water. A computed tomography myelogram of the spine demonstrated erosion of her sacrum due to a large lumbosacral dural ectasia. An initial surgery was attempted to reduce the size of the expansile dura, and reconstruct the dorsal sacrum with a titanium plate (Depuy Synthes, Westchester, PA, USA) to prevent recurrence of thecal sac dilatation. Her symptoms initially improved, but shortly thereafter recurred. A second surgery was then undertaken to obliterate the thecal sac distal to the S2 nerve roots. This could not be accomplished through simple ligation of the thecal sac circumferentially as the ventral dura was noted to be incompetent and attempts to develop an extradural tissue plane were unsuccessful. Consequently, an abundance of fibrin glue was injected into the thecal sac distal to S2, and the dural ectasia was marsupialized rostrally, effectively obliterating the distal thecal sac while further reducing the size of the expansile dura. This approach significantly improved her symptoms at 5 months follow-up. Treatment of dural ectasia is not well-defined and has been variable based on the underlying manifestations. We report a rare patient without risk factors who presented with significant lumbosacral dural ectasia. Moreover, we present a novel method to treat postural headaches secondary to dural ectasia, where the thecal sac is obliterated distal to the S2 nerve roots using an abundance of fibrin glue followed by marsupialization of the thecal sac rostally. This method may offer an effective therapy option as it serves to limit the expansile dura, reducing the cerebrospinal fluid sump and the potential for intracranial hypotension.

Key words: Dural ectasia, intracranial hypotension, marsupialization

INTRODUCTION

Dural ectasia involves the enlargement of the spinal canal, concavity of the posterior vertebral body, reduction of cortical

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bone thickness of the pedicles and laminae, expansion of the neural foramina, formation of a meningocele, scoliosis, spondylolithesis, or vertebral fractures.[1] Lumbosacral dural ectasia is commonly associated with connective tissue disorders, such as Marfan syndrome,[2-9] osteogenesis imperfecta, Loeys-Dietz syndrome,[10-14] ankylosing spondylitis (AS),[15,16] neurofibromatosis,[17,18] fibromuscular dysplasia,[19] and Larsen syndrome;[20] however, the pathology has also been linked to vertebral fracture, spine surgery,[21] trisomy 13,[22] scoliosis,[23] and trauma.

The pathology may be asymptomatic,[24] but can cause postural headaches due to a "cerebrospinal fluid sink" mechanism, as well as back pain, leg pain, or rectal pain.[25,21] If significant arachnoid cysts are present, the compression of lumbosacral nerve roots may lead to neurological deficits, including cauda equina syndrome (CES).[26,27] Moreover, dural ectasia may induce structural spinal instability.[28-30] Conservative management has been successful, including flat bed rest, hydration, and blood patch.[1,31,32] Surgery has been performed for persistent symptoms or CES. General methods have revolved around surgical bony decompression, followed by thecal sac reconstruction[33,34] with possible lumboperitoneal shunt placement.[26]

To our knowledge, no report exists regarding dural ectasia in a patient without known risk factors. We report such a patient, and describe a novel technique to obliterate the dural ectasia through marsupialization of the thecal sac followed by Tisseel glue (Baxter Healthcare Corp., Deerfield, IL, USA) isolation of the estasia from the spinal canal. We also review the surgical techniques employed in the literature.

CASE REPORT

The patient is a 62-year-old female who complains of headaches for her entire life but recently became progressively severe over the last couple of years. Headaches become progressively severe after 20-30 min of being upright. After lying down, the headaches slowly subside. Magnetic resonance imaging brain and magnetic resonance angiogram neck without significant findings. A lumbar puncture revealed opening pressure 3 cm of water, concerning for low spinal pressure. A computed tomography (CT) myelogram of the spine demonstrated the erosion of her sacrum due to a large dural ectasia [Figure 1], which was felt to be the source of her postural headaches.

Surgery was recommended, entailing a sacral laminectomy and reduction of the dural ectasia.

An incision was made over the sacrum, followed by deep tissue dissection and subperiosteal dissection on the residual sacrum. The dura was immediately identified. Approximately 5 cm of bone circumferentially around the ectasia was removed with Kerrison rongeurs. Subsequently, the dura was incised, and large amounts of cerebrospinal fluid (CSF) drained. An ellipse within the dorsal dura was then excised, and over-sewed with 5-0 nylon. Tisseel and Duragen (Integra Life Sciences, Plaisnboro, NJ, USA) were applied. Valsalva maneuvers showed

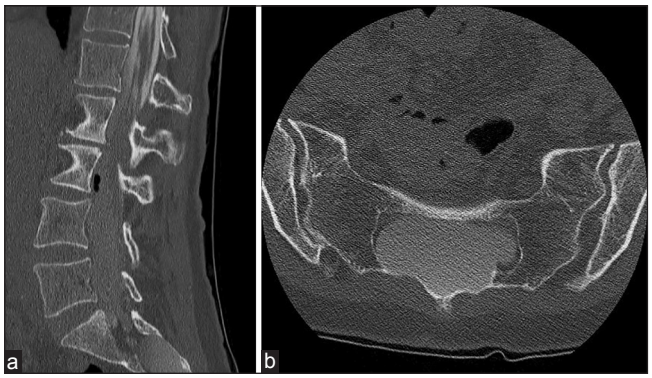

Figure 1: (a) Computed tomography myelogram (sagittal plane) demonstrates large lumbosacral dural ectasia. (b) Computed tomography myelogram (axial plane) demonstrates large lumbosacral dural ectasia

adequate closure. Subsequently, a titanium mesh fitted to the bony defect and fastened with screws. The wound was closed in layers. Evoked potentials, electromyography, and motor response did not change during the procedure.

Positional headaches initially improved but recurred at 1 month. A repeat CT myelogram of the L-spine demonstrated recurrence dural ectasia, with a size comparable to preoperative imaging. Repeat surgery was recommended to obliterate the thecal sac distal to the sacral nerve roots. Dissection occurred down the prior surgical cavity, as well as more distally into the sacrum. The metal mesh was removed. The dura was opened from L5 to S3. A dural stay stitch was placed. Meticulous inspection revealed no arachnoid cyst. The nerve roots were dissected free from the lateral walls of the dura and positioned anteriorly using cottonoids. Additional dural tissue was resected to narrow the size of the thecal sac. An attempt was made to dissect the ventral dura free in order to ligate the thecal sac at approximately S2, coming below the lowest nerve roots. However, this was unsuccessful as the tissue was notably tenuous with clearly defined erosions. Subsequently, the surgical cavity distal to the S2 roots were filled with Tisseel and Surgicel (Ethicon, Somerville, NJ, USA), forming a solid mass and filling the distal extasia in its entirety. The thecal sac was then marsupialized using 5-0 prolene, reducing the size of the thecal sac by approximately 30-35%. Tisseel and Surgicel were applied along the dural suture line. Valsalva maneuvers showed no evidence of CSF leak. The wound was then closed in layers. The patient woke up well. At 5-month follow-up, patient notes minimal headaches, which responds to caffeine.

DISCUSSION

The pathogenesis leading to dural ectasia is unclear. Given its frequent association with connective tissue disorders or inflammatory conditions, the prevailing theory implicates CSF pulsations as the primary cause of progressive distention of a weakened dural sac. Associated features include the enlargement of the spinal canal, concavity of the posterior vertebral body, reduction of cortical bone thickness of the pedicles and

laminae, expansion of the neural foramina, the formation of meningoceles, fracture of posterior elements, spondylolithesis, and scoliosis.[1] The most caudal levels of the spine tend to be most affected since CSF pressures are greatest at these levels based on Pascal's law.[21,35] On the other hand, dural ectasia has been reported after trauma and after spinal surgery[21,36] as well. In additional, the pathology can also occur at cervical[37] or thoracic levels[38] without lumbar involvement. Curiously, our patient had no history of connective tissue disorder, and no signs/symptoms to suggest an undiagnosed disorder. Moreover, she had no prior spine trauma or spinal surgery. The literature is scarce regarding dural ectasia in patients without such risk factors.

Treatment of dural ectasia is not well-defined and has been variable based on the underlying manifestations. Where the pathology has been associated with spinal instability/deformity, surgery has been directed to stabilize the spine through reconstruction, instrumentation, and fusion.[29,33,39-41] Where the pathology has been associated with postural headaches, conservative management has had some success, including flat bed rest, hydration, and blood patch.[1,31,32] Treatment assumes that the postural headaches due to a "cerebrospinal fluid sink" mechanism, which causes traction on pain-sensitive structures such as cranial nerves, dura, and meninges; however, imaging to seek the site of a CSF leak associated with dural estasia has predominantly been negative.[24,31,42] However, if a site if discovered, percutaneous injection of fibrin glue and surgical repair of the leak have had success.[43] For patients who developed dural ectasia after posterior fossa decompression for Chiari malformation, the use of a titanium plate to support dura over the cerebellar hemispheres has been effective.[36] Where the pathology has been associated with leg pain/rectal pain, procedures have been directed at the dural ectasia. Several investigators have felt that reduction of the expansile dura would decrease pressure and improve symptoms. Wera *et al.*[35] reported a patient received a percutaneous injection of fibrin glue for treatment of dural ectasia for back and leg pain; the goal was to use fibrin glue to decrease the expanded dural space and strengthening the attenuated dura; unfortunately, the patient suffered CES after the procedure.

If significant arachnoid cysts are present, the compression of lumbosacral nerve roots may lead to neurological deficits, including CES.[26,27] However, CES may also occur without arachnoid cysts; the pathophysiology of this presentation remains unclear. Dural ectasia and CES have predominantly been reported among AS patients. According to Ahn *et al.*,[26] conservative management have had limited success for patients with AS and CES Dinichert *et al.*[44] and Ahn *et al.*[26] placed a lumboperitoneal shunt in seven patients with AS with CES; the groups believed that CES was caused by injury to the spinal cord and nerves secondary to an arterial pulse wave transmitted to the CSF, in a dural sac with reduced elasticity due to chronic inflammation and residual adhesions; 6 of 7 patients had symptoms that improved. Ahn *et al.*[26] reported decompressive laminectomy for 11 patients with AS with CES (3 improved, 6 stable, 2 worsened). Ha *et al.*[45] reported a limited laminotomy followed by filum detethering for a patient with AS-CES, where symptoms did not improve. Similarly, Liu *et al.*[27] performed lysis of adhesions along a tethered conus, without clinical improvement. For patients with concurrent sacral cyst along with dural ectasia, the cyst has been treated with marsupialization[34] or resection.[24]

Our patient had a prolonged history of postural headaches. Imaging suggested a gradual erosion of her sacrum due to the dural ectasia. An intact sacral wall limited the size of expansion of the cyst and the sump of the CSF. However, once the cyst eroded through the sacrum completely, leading to an enlargement of the potential line of the cyst, the spinal headaches worsened. The initial surgery attempted to reduce the size of the expansile dura while buttressing the reconstruction with a titanium plate to prevent recurrence of thecal sac dilatation. This logic mirrors the use of titanium plate for those that developed dural ectasia after posterior fossa decompression for Chiari malformation. Unfortunately, this approach did not prevent the recurrence of debilitating postural headaches. The second surgery was designed to obliterate the thecal sac distal to the S2 nerve roots. This could not be accomplished through ligation of the entire thecal sac circumferentially since the ventral dura of the thecal sac could not be elevated successfully. Consequently, an abundance of fibrin glue was injected into the thecal sac distal to S2, and the thecal sac was marsupialized rostrally, effectively obliterating the thecal sac while further reducing the size of the expansile dura. This approach significantly improved her symptoms. Percutaneous injection of fibrin glue was not considered since there was no obvious site of CSF leak on her imaging. Lumboperitoneal shunting or laminectomy alone would not be effective since that would increase the intracranial hypotension from the CSF sump.

CONCLUSION

Treatment of dural ectasia is not well-defined and has been variable based on the underlying manifestations. We report a rare patient without risk factors who presented with significant lumbosacral dural ectasia. This pathology should remain on the differential for persisting postural headaches, even for patients without risk factors. Moreover, we present a novel method to treatment of postural headaches secondary to dural ectasia, where the thecal sac is obliterated distal to the S2 nerve roots using an abundance of fibrin glue followed by marsupialization of the thecal sac rostally. This method can be effective since the expansile dura is significantly reduced, reducing the CSF sump and the potential for intracranial hypotension.

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Conflicts of interest
There are no conflicts of interest.

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