Management of complex giant spinal arachnoid cysts presenting with myelopathy

Technical note

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A giant spinal arachnoid cyst is an unusual cause of progressive epidural compressive syndrome. The authors describe 4 cases of a “complex” subtype of this lesion and discuss aspects of surgical management. The patients presented with progressive spastic paraparesis and were found to harbor extensive spinal extradural arachnoid cysts with multiple septations and significant paraspinal extensions. Extensive laminotomy and excision of the cyst along with its extensions were performed in all cases.

Compared with previously indexed cases of surgically managed extensive spinal extradural arachnoid cysts, the cases reported here are unique because of their complex nature. Curative treatment consists of radical excision inclusive of the paraspinal extensions as well as closure of a dural defect, if found. A laminotomy or laminoplasty should be performed to avoid postoperative instability related to the extensive exposure. Extended follow-up and instrumentation may be required in select cases. (DOI: 10.3171/2011.3.SPINE10672)

Key Words • arachnoid cyst • extradural lesion • myelopathy • spine • surgical treatment

Illustrative Cases

Case 1. This 13-year-old girl presented with progressive lower limb spasticity of 2 years’ duration and symptoms suggestive of a neurogenic bladder of 3 months’ duration. At presentation, she was wheelchair bound. Tone was spastic and power was Grade 4 in all muscle groups in the lower limbs. Touch and pain sensations were impaired from T-8 downward, with gross impairment of vibration and proprioception in both legs. Magnetic resonance imaging of the lumbosacral spine showed a giant extradural arachnoid cyst from the T-4 level extending inferiorly up to L-2 (Fig. 1A and B). The cyst had displaced the spinal cord ventrally and had extensions through the neural foramina in the lower dorsal region, close to the pleural cavity and retroperitoneum (Fig. 1C and D). Histopathological examination showed a single-cell layer of inner arachnoid lining. Postoperative MR imaging showed total excision of the lesion. A chest radiograph on the 1st postoperative day documented a minimal pleural effusion (Fig. 2) that spontaneously resolved within 5 days. At discharge, the patient had functionally improved to Nurick Grade 3, although her bladder symptoms persisted. Six months later, she was asymptomatic and had no spinal deformity.

Case 2. This 30-year-old man presented with spastic paraparesis of 1 year’s duration and was Nurick Grade 3 at admission. He demonstrated Grade 4 power in all muscle groups in the lower limbs and had a graded sensory loss below the L-2 dermatome. Deep tendon reflexes were exaggerated in the lower limbs, and the plantar reflex was bilaterally extensor. An MR image showed a giant arachnoid cyst extending from the T-11 to S-3 vertebral level, pushing the cord anteriorly, invaginating through the interlaminar spaces and into the paraspinal area through the intervertebral foramina (Fig. 3A–C). The lumbar laminae were thinned out on spinal CT (Fig. 3D). Histopathological examination showed a single-cell layer of inner arachnoid lining. Postoperative MR imaging showed total excision of the lesion. Although not significantly better, the patient had functionally improved to Nurick Grade 2. Six months later, he was asymptomatic and had no spinal deformity.

Abbreviations used in this paper: GSAC = giant spinal arachnoid cyst; SAC = spinal arachnoid cyst.

This article contains some figures that are displayed in color online but in black and white in the print edition.
at discharge, this patient functionally improved to Nurick Grade 2 at a follow-up visit 2 years later, and imaging showed no evidence of recurrence or kyphoscoliosis (Fig. 4).

**Case 3.** This 45-year-old man presented with a history of paresthesias in both lower limbs for 3 years and spasticity-related progressive difficulty in walking for 2 years. On examination, he had normal power in all muscle groups. There was a graded sensory loss below the T-6 level and impaired proprioception in the lower limbs. His gait was spastic. Magnetic resonance imaging showed a well-defined, dorsally located extradural arachnoid cyst extending from the T-5 to T-11 vertebral levels and extending laterally into multiple neural foramina. Another small similar extradural lesion was noted at the T3–4 level extending into the left-sided foramina. Histopathological examination showed a single-cell layer of inner arachnoid lining. Postoperative MR imaging showed total excision of the lesion. The patient was asymptomatic 6 months after surgery, and there was no radiological evidence of any deformity (Fig. 5).

**Surgical Procedure**

The laminae, though thinned out, were intact in all the cases. Laminotomy was performed using the craniotome attachment of a pneumatic drill (Midas Rex) pedicled either superiorly or inferiorly to expose the underlying extradural cyst (Fig. 7). Breaks in the laminae were performed medial to their junction with the facets to secure a base to reattach the laminae during closure. While raising the laminae as a flap, outpouchings of the cyst wall in areas of ligamentum flavum deficiency were maneuvered into the spinal canal by using a dissector and cottonoids, which were advanced under the laminae from a lateral direction. A thick arachnoid wall enabled easy dissection of the ex-
tractal cyst off the laminae and pedicles without inducing a tear. The ventrally pushed dural sac was peeled off the cyst wall with cottonoid dissection (Fig. 8A). The extraforaminal extensions of the cyst along the roots were delivered into the operating field by gentle traction on the neck of the cyst at the foramina followed by coordinated inward traction on progressively distal parts of the extensions (Fig. 8B and C). Nerve roots were seen ventral to these extensions of the arachnoid cyst. Bleeding at all stages was well controlled with Gelfoam and/or Surgicel. After excision of the cyst, the dura was carefully inspected for any defect(s) that would have led into the arachnoid cyst. Such a rent, identified only in the patient in Case 1, was sutured with 5-0 Prolene. Anchoring of the laminotomy flap was done with nonabsorbable suture material (2-0 nylon), which was passed between the free end of the laminae on the flap and corresponding points on the base of the laminae at all levels. A suction wound drain, placed over the laminae, was left in situ for 2 days, after which the patients were mobilized with a Taylor-Knight brace that they were advised to wear for 6 months.

Discussion

Spinal arachnoid cysts are unusual causes of spinal cord or root compression, constituting < 1% of all spinal tumors.1 Among the less common extradural subtype of these lesions are the rare GSACs. Spinal arachnoid cysts have been reported to range from < 1 to > 10 vertebral levels, and those at the high end of this spectrum have loosely acquired the “giant” descriptor.9,10,18,19 It would
be useful, albeit arbitrary, to define this term for future reporting as a threshold extent that spans 6 vertebral levels. A review of surgically managed GSACs (PubMed) revealed that the largest of 4 previously described lesions spanned about 12 levels (Table 1). The extensive lesions described in our series, constituting 1% of all surgically managed SACs in our institute over a 10-year period, are of added interest because of their complex nature rendered by septated loculations, exuberant paraspinal extensions, and the associated arduous surgical extirpation.

The pathogenesis of extradural SACs remains controversial. Classified by Nabors et al. as Type 1A, most of these lesions are hypothesized to be congenital. Their development is hypothetically related to dural diverticula of arachnoid membrane herniations through dural defects in the midline or at or near a nerve root. They can also be acquired, with the implicated pathogenesis being dural defects developing in relation to surgery, trauma, or arachnoiditis. A widely accepted mechanism to explain their enlargement is the ball-valve hypothesis, which postulates the operation of an anatomical communication as a 1-way valve between the subarachnoid space and the cyst. Such a communication may get sealed off at a later stage of their development or may not exist at all, constituting the so-called noncommunicating, or closed, arachnoid cyst. In such closed cysts, other proposed mechanisms for cyst expansion, such as active fluid secretion from the cyst wall, passive osmosis of water, and CSF hydrostatic pressure, probably hold true.

The pathogenesis of the complex anatomical features of GSACs is probably multifactorial. While a dural defect might have initiated their development, intrathoracic pressure-related fluctuations in their volume would have caused areas of the cysts to get sealed off during periods of relative collapse. Septations arising in sealed-off regions would have become apparent during reexpansion of the cysts at a later stage. Passive osmosis or active fluid secretion by the cyst wall would have facilitated expansion of these independent septated areas. Paraspinal extensions would have occurred due to a strong ball-valve mechanism in place, causing the cyst to expand through the foramina and indented areas of relative ligamentum flavum deficiency, both regions of comparatively low resistance.

Findings on plain radiography and CT, although nonspecific, are indirect pointers to the wall effect of these large cysts. On MR imaging, the standard diagnostic examination, these lesions are typically hypointense on T1- and hyperintense on T2-weighted sequences and do not enhance with contrast. Myelography, myelo-CT, and cine MR imaging, by virtue of their ability to detect pulsating flow, are useful tools to visualize the communication(s) between the cyst and the subarachnoid space, and thereby aid in intraoperative localization of the dural defect. Such information is of considerable value in complex GSACs in which the presence of such communication(s), as cited earlier, may have been essential in their pathogenesis. It may, however, turn out that a dural defect remains elusive at surgery, as in 3 of our cases.

A radiological mimic of an SAC and often an interchangeable used term, although wrongly, is a “spinal meningeal cyst,” which can be differentiated intraoperatively by its dural wall and histopathologically by the presence of a thick, collagenous outer wall and the absence of a thin inner layer of arachnoid. Careful radiological analysis should enable differentiation of an SAC from other lesions, such as dermoids and epidermoids (Fig. 9A), perineural cysts, hydatid cysts (Fig. 9B), neural cysts (Fig. 9C and D), meningeal diverticula, and meningoceles, some of which have been reported to acquire gigantic proportions.

Surgery for complex GSACs should address the complete release of multicompartmental fluid contents and total excision of the wall inclusive of its extraforaminal components. With such a surgical end in sight, procedures like marsupialization, partial excision with a cystoperitoneal shunt, or a cystotheostomy should be considered lesser procedures because of the risks of recurrence rendered by potential remnants in the extraforaminal region or in relation to unruptured loculations. We thus advocate total excision even when it could result in a large dural tear, as in cases in which the cyst wall is densely adherent to the dura. The dura can be repaired with fascia or an artificial dural substitute. An expansile duraplasty, a described alternative in such cases, may not be applicable.
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to GSACs, as it may not relieve fluid-related compression in some septated areas or in the paraspinal extensions.

A factor that would help with relatively easy total resection of the cyst is the prevention of its rupture as far as possible. The use of ultrasonography to confirm normal CSF pulsatility in the thecal sac at the polar ends of the excision site will aid in ascertaining the completeness of excision in the craniocaudal extent of the lesion. The radicality of excision of the paraspinal extensions can be assessed using a flexible spinal endoscope, which can also be used to aid breakage of the septations. A careful search for a dural defect and its closure should follow cyst excision. Although the benefit of intradural exploration in such cases to look for possible intradural components has yet to be determined, there have been a few reports of similar lesions coexisting in dual compartments. A dural defect, though expected in such cases, may not always be found at surgery, and hence, the surgeon’s inability to find one should not be considered a surgical failure to treat this pathogenetic factor.

Among the intraoperative complications, blood loss is foremost given the sheer extent of the required exposure. Injury to intercostal vessels while delivering the paraspinal extensions should not be a concern as they run in an extrapleural course. Complications unique to complex GSACs are effusions or hematomas related to their dissection off the closely associated retroperitoneal and juxtapleural structures. Effusions could also be sympathetic in nature, showing spontaneous resolution within a few days, as in our Case 1. Obtaining serial radiographs to detect possible complications in the immediate postoperative period would thus be prudent.

Delayed complications that could affect functional outcome in surgery for such an extensive lesion with extraforaminal extensions are instability, misalignment, and worsening kyphosis. Some factors proven to be conducive of postoperative spinal deformity are an age younger than 3 years, preexisting spinal deformity, decompression spanning the axial cervical spine or cervicothoracic region, removal of 3 or more laminae, and aggressive facetectomy. Procedures such as laminoplasty, skip laminectomies, and preservation of the posterior osteoligamentous tension band have not proved to reduce postlaminectomy kyphosis. Based on these facts, we would advocate instrumentation for GSACs occurring in young children, in cases spanning the cervical or cervicothoracic regions, and in patients with a preexisting deformity. Our preference for a nonexpansible laminotomy for exposure is based on the proper apposition and reconstruction of the posterior elements of the spinal column that it affords. Besides, we have not yet encountered any postoperative deformities in other thoracic and lumbar intradural or extradural lesions using this procedure. A longer follow-up in 2 of our pediatric patients would be prudent to check for the development of deformity during the latter part of spinal maturation.

**Conclusions**

In this first-of-its-kind report, we defined and dis-

**TABLE 1: Literature review of cases of surgically managed nontraumatic giant extradural SACs**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Extent of Lesion</th>
<th>Radiology</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Papo et al., 1977</td>
<td>Midthoracic to sacral canal</td>
<td>NA</td>
<td>Partial resection, cystoperitoneal shunt</td>
</tr>
<tr>
<td>Kahraman et al., 2008</td>
<td>T4–L3</td>
<td>yes</td>
<td>Cystoperitoneal shunt, total resection &amp; repair of dural defect</td>
</tr>
<tr>
<td>Rashid et al., 2008</td>
<td>T8–L3</td>
<td>no</td>
<td>Total resection</td>
</tr>
<tr>
<td>Joaquim et al., 2009</td>
<td>C2–T1</td>
<td>no</td>
<td>Total resection &amp; repair of dural defect</td>
</tr>
<tr>
<td>Present study</td>
<td>T4–L2, T11–S3, T5–11, T9–S3</td>
<td>yes</td>
<td>Total resection</td>
</tr>
</tbody>
</table>
| * NA = not available.*
discussed a complex giant subtype of spinal extradural arachnoid cyst causing progressive myelopathy. Surgery in such cases should aim at total excision of the cyst with its extraspinal ramifications. A dural defect should be carefully sought and repaired. Its absence could be indicative of a closed variety of the arachnoid cyst with a different pathogenesis.

Disclosure

The authors do not report any conflict of interest concerning the materials and 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: Furtado. Acquisition of data: Furtado, Thakar, Murthy, Dadlani. Analysis and interpretation of data: Furtado, Thakar, Murthy, Dadlani. Drafting the article: Furtado, Thakar, Dadlani, Hegde. Critically revising the article: Hegde. Reviewed final version of the manuscript and approved it for submission: all authors. Administrative/technical/material support: Furtado, Thakar, Murthy, Dadlani. Study supervision: Hegde.

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