Rapidly progressive paraplegia due to an extradural lumbar meningocele mimicking a cyst

Case report

INO FISS, PH.D., MARCO DANNE, M.D., CHRISTIAN HARTMANN, PH.D., MARIO BROCK, PH.D., AND RUEDIGER STENDEL, PH.D.

1Department of Neurosurgery, Charité–University Medicine Berlin, Campus Benjamin Franklin, Berlin; and 2Clinical Cooperation Unit Neuropathology, German Cancer Research Center, Heidelberg, Germany

Unlike arachnoid meningoceles, arachnoid cysts frequently cause local pressure effects probably because there is no free communication between the cyst and the subarachnoid space. Following the first detailed description of cystic lesions of spinal nerve roots by Tarlov in 1938, a simplified classification of spinal meningeal cysts was developed in 1988, containing three major categories. The authors report on a lumbar intraspinal extradural meningocele that caused incomplete paraplegia in an otherwise healthy 31-year-old man in whom magnetic resonance imaging revealed stigmata of Scheuermann disease. Intraoperatively, the lesion was classified as a transitional-type lesion, in accordance with Type IA of the Nabors classification, because a communication with the subarachnoid space was observed. After complete removal of the meningocele, the patient’s recovery was prompt and complete.

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KEY WORDS • arachnoid cyst • lumbar spine • meningocele • Scheuermann disease • spinal cord compression

Most anomalies of the spinal subarachnoid space and the meningeal membranes are not uncommon, and many are congenital. In most cases, these anomalies are innocuous and do not require surgical treatment, but arachnoid cysts are known to frequently cause local pressure effects, likely due to an absence of a free communication between the cyst and the subarachnoid space. Interestingly, there are transitions between meningoceles and cysts in which a residual communication between the subarachnoid space and the diverticulum still exists, but local pressure effects have already emerged. We present the case of a patient with an extradural lumbar meningocele mimicking a cyst that caused rapidly progressive paraplegia due to local pressure effects. The surgical procedure, a schematic overview of the relevant types of extradural cysts and meningoceles, and a description of the entities that should be considered in a differential diagnosis are presented.

Abbreviations used in this paper: CSF = cerebrospinal fluid; MR = magnetic resonance.
Plain lumbar radiography revealed erosion of the right lamina of the vertebral body of L-1 more than L-2, as well as a widened interpedicular distance and an enlarged neural foramen at this level.

Operation. Before surgery, the hypothetical diagnosis was arachnoid rather than perineurial cyst because of the lesion’s location proximal to the dorsal root ganglion. Intraoperatively the lesion was classified as an extradural arachnoid meningocele of the transitional type in accordance with Type IA of the classification proposed by Nabors and colleagues because we observed a communication with the subarachnoid space.

We performed an L1–2 laminectomy rather than a laminoplasty because the lamina was extremely thinned out. Intraoperatively, we noted a grayish epidural pouch free of neural structures compressing the dura posteriorly (Fig. 3A). The pouch did not collapse when compression was applied, nor did compressing the thecal sac increase the size or tension of the pouch. After puncturing the pouch, which released a large amount of clear fluid, an opening into the subarachnoid space was suspected (Fig. 3B). Because of dense fibrous adhesions, the posterior wall was excised. Subsequently, a tiny opening, measuring about 1 mm, was identified at the L-2 level medial to the right nerve root (Fig. 3C). Furthermore, a pulsatile, breath-synchronous outflow of CSF was seen. Inspection with the operating microscope identified a valvelike mechanism resulting from two overlapping arachnoid layers. The opening was closed with a suture and the sac was removed (Fig. 3D).

Histological Examination. Histologically we observed a thin layer of arachnoid cells, collagen fibers, singular fibroblasts, and discrete perivascular lymphocytes as signs of minimal, chronic unspecific inflammation (Fig. 4 left). The thin layer of arachnoid cells was labeled by epithelial membrane protein–specific antibodies (Fig. 4 right). There were no signs of malignancy.

Postoperative Course. Within 2 days of surgery the patient recovered completely and was free of pain. He was discharged on the 7th postoperative day. His neurological status at discharge and at 6-month follow-up was completely normal.

Discussion

Tarlov published the first detailed description of cystic lesions of the spinal nerve roots in 1938 and introduced the term “perineurial cysts.” In 1970 he published a communication in an attempt to eliminate the confusion about the
Different types of abnormalities that had arisen over time. \(^{12}\) Perineurial cysts occur along the nerve roots, at or distal to the junction of the posterior root and the dorsal ganglion. Unlike meningeal cysts, at least part of the lining of perineurial cysts contains nerve fibers and, at times, ganglion cells. These perineurial cysts are usually seen on the sacral nerve roots, but they may occur at other levels. \(^{11}\) In comparison, arachnoid meningoceles are initially formed by herniation of arachnoid through a dural defect, and they communicate with the subarachnoid space by a narrow neck. \(^{5,7}\) When the communication becomes occluded, the term arachnoid cyst seems more appropriate. \(^{4,5}\) In contrast to asymptomatic arachnoid meningoceles, arachnoid cysts frequently cause local pressure effects, which is likely caused by an absence of a free communication between the cyst and the subarachnoid space. \(^{9}\) A schematic overview of relevant types of extradural cysts and meningoceles is provided in Fig. 5.

Interestingly, it has been proposed that spinal extradural arachnoid cysts can cause Scheuermann disease by cystic compression and occlusion of the venous channels draining the vertebral bodies. \(^{2}\) Because our patient had local signs of Scheuermann disease (kyphosis with the apex at the L1–2 level and Schmorl nodules at T11–L2 [Fig. 1]), a similar mechanism might be suspected in his case.

A hint for a congenital origin was found by Bergland, \(^{1}\) who described extradural meningeal cysts in three members of one family. Other authors have reported finding an extradural meningeal cyst in a 20-month-old baby and thought a failure of mesenchymal structure fusion surrounding the neural tube was a possible cause. \(^{8}\)

According to the approach proposed by other authors, \(^{3,5,6}\)
surgical treatment of the cyst was straightforward with excision of the cyst and closure of the communication by a suture. In contrast, the cyst walls of perineurial cysts (Tarlov cysts or Nabors Type II cysts) should be handled more carefully to preserve nerve root function. Table 1 shows the entities that should be considered in the differential diagnosis in cases of intraspinal cysts and meningoceles.

We believe, in accordance with Tarlov,\textsuperscript{12} that to refer to meningoceles or diverticula as cysts is inaccurate because, unless the necks of these lesions become occluded, they are not cysts by definition. As in the presented case, transitions between “meningoceles” and “cysts” occur, in which there is residual communication between the subarachnoid space and the diverticulum. Only when pressure was relieved after excision of the posterior wall was a pulsatile outflow of CSF clearly visible.

This observation might be explained by the intermittent opening of a valvelike communication between the pouch and the subarachnoid space composed of two overlapping layers of arachnoid. The pressure changes that occur within the CSF are most influenced by pressure transmitted via the venous system from the body cavities. Pressure peaks

![Diagram of the spine and cysts](image)

**TABLE 1**

Differential diagnosis of meningoceles*

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Spinal Location</th>
<th>Incidence</th>
<th>Pathological Features</th>
<th>Imaging Features</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>perineurial cyst</td>
<td>lateral sacral</td>
<td>common</td>
<td>lining contains neural elements</td>
<td>appears as nerve root mass; sacral erosion</td>
<td>can be excised</td>
</tr>
<tr>
<td>arachnoid diverticulum</td>
<td>usually lumbar</td>
<td>common</td>
<td>lined w/ arachnoid</td>
<td>enlarged nerve root sleeve</td>
<td>asymptomatic, of no clinical significance</td>
</tr>
<tr>
<td>intradural arachnoid cyst</td>
<td>usually thoracic</td>
<td>rare</td>
<td>lined w/ arachnoid</td>
<td>may cause spinal cord compression</td>
<td>can be excised or fenestrated</td>
</tr>
<tr>
<td>extradural arachnoid cyst</td>
<td>usually thoracic</td>
<td>uncommon</td>
<td>lined w/ arachnoid; does not contain neural elements</td>
<td>associated w/ kyphosis dorsalis juvenilis</td>
<td>complete excision after ligation of neck/</td>
</tr>
<tr>
<td>intrasacral meningocele</td>
<td>sacral, midline</td>
<td>uncommon</td>
<td>lined w/ arachnoid; does not contain neural elements</td>
<td>sacral erosion</td>
<td>can be excised, usually asymptomatic</td>
</tr>
<tr>
<td>extradural pseudomeningocele</td>
<td>usually lumbar</td>
<td>common</td>
<td>not lined w/ meninges</td>
<td>extradural CSF root sleeve</td>
<td>of no clinical significance</td>
</tr>
<tr>
<td>nerve avulsion</td>
<td>usually cervical</td>
<td>uncommon</td>
<td>not lined w/ meninges</td>
<td>empty root sleeve</td>
<td>usually not reparable</td>
</tr>
</tbody>
</table>

* ant = anterior; pst = posterior.
of up to 180 mm Hg have been recorded during coughing.\textsuperscript{13} Intermittent elevation of abdominal pressure probably facilitates early enlargement. Inevitably, the confines of the spinal canal will limit spherical enlargement and the growing pouch, as in the present case, will come to resemble a cylinder. Subsequently, the development of very large pouches is promoted by intermittent surges of pressure facilitating pulsatile inflation, whereas outflow of CSF is prevented by a valvelike mechanism. This mechanism may be supported by sufficient force exerted by the body of the pouch on the neck to close the communication, as described by the law of Laplace.\textsuperscript{5} These large pouches are likely to produce pressure-related symptoms. Because a true neck was missing in the present case, we assume that a true valvelike mechanism consisting of two overlapping arachnoid layers enabled the development of such a large meningocele, causing pressure symptoms.

Acknowledgment

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References

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Address reprint requests to: Ingo Fiss, Ph.D., Department of Neurosurgery, Charité–University Medicine Berlin, Campus Benjamin Franklin, Hindenburgdamm 30, 12203 Berlin, Germany. email: ingo.fiss@charite.de.