Thoracic intramedullary arachnoid cyst in an infant

Case report

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Symptomatic intramedullary arachnoid cysts are rare, especially in children; these lesions are rarely described as a cause of spinal cord compression in this age group. The authors report on an 18-month-old boy who experienced a sudden loss of his ability to stand and walk due to a paraparesis. Magnetic resonance imaging of the spine exhibited a cystic intramedullary lesion at the level of T5–6. A hemilaminectomy was performed, and after myelotomy the cystic lesion was decompressed by fenestration to the subarachnoid space. The histopathological examination verified the diagnosis of an arachnoid cyst. In the postoperative course the boy experienced complete resolution of the initial paraparesis. (DOI: 10.3171/2008.10.PEDS08202)

Key words: • arachnoid cyst • infant • intramedullary cyst

Spinal arachnoid cysts, also called arachnoid diverticula, represent benign lesions that are commonly asymptomatic clinically. They become clinically relevant due to a compression of the spinal cord or nerve roots and consecutive appearance of neurological symptoms.10 The extra- or intradurally located arachnoid cysts can be subdivided into 3 main categories according to the classification of Nabors et al.,15 as follows: extradural cysts without spinal nerve root fibers (Type I); extradural cysts with spinal nerve root fibers (Type II); and intradural cysts (Type III). Spinal arachnoid cysts are considered to be rare lesions and are commonly located at the thoracic level.9,12 The more frequent extradural arachnoid cysts predominantly result from a herniation of the arachnoid mater caused by flaws of the dura mater, whereas the less frequent intradural cysts are most likely to develop because of a modification of the arachnoid trabeculae. However, the pathogenesis of these lesions is to a great extent unexplained, and several origins, such as congenital anomalies or traumatic or inflammatory causes, are discussed.9,13,16 The appearance of neurological symptoms during infancy caused by a spinal, intradural, and intramedullary thoracic arachnoid cyst has only been described 4 times in the literature and can therefore be considered to be extremely rare.

Case report

History. We describe the case of an 18-month-old boy who had been having bowel dysfunction (constipation) for several days. According to his parents, he had been able to walk since the age of 8 months, but had suddenly lost his ability to stand or walk for ~1 week prior to the medical examination. There was no evidence of a trauma in the anamnesis of this young boy.

Examination. The boy was in good general health, with normal vital parameters. The neurological investigation detailed a hypotonic paraparesis of the lower extremities, motor power Grade 1/5, and neck rigidity. The monosynaptic reflexes examined in the lower and upper limb (patella reflex, ankle jerk reflex, biceps reflex, and triceps reflex) were bilaterally equal and of normal intensity. No clinical signs for corticospinal tract lesions (like Babinski and Hoffmann signs) or clonus were seen. The superficial abdominal reflexes, the anocutaneous reflex, and also the anal sphincter strength were found to be normal. In terms of spinal dysraphism, no superficial cele or myeloschisis for a spina bifida aperta were detectable. For occult spinal dysraphism, neither cutaneous lesions like hairy patches, vascular nevi, aretic meningocoele, or midline mass such as subcutaneous lipomas were visible, nor was limb asymmetry or foot deformity detected. The examination of the abdomen revealed no pathological findings. Admission MR imaging of the total spine revealed a cystic intramedullary lesion at the level of T5–6, with no enhancement following administration of Gd contrast (Fig. 1). Additionally, the MR imaging studies exhibited conus medullaris at the level of L-1. There was no evi-
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dence for bifid laminae or tethered cord syndrome. The MR imaging of the brain and the skull base revealed no intracranial pathological entities and no evidence for an associated Chiari malformation. Differential diagnosis for a cystic intramedullary lesion at the T5–6 level included a focal syringomyelia, epidermoid cyst, ependymoma, or other medullary neoplastic lesion.

**Operation.** Following hemilaminectomy at the level of T-5 and T-6, and dorsal root entry zone myelotomy, the cystic lesion was identified just 1 mm beneath the medullary tissue. The cyst was fenestrated and its wall was partially resected. Macroscopically, the cyst had neither communication with the subarachnoid space nor an extramedullary component.

**Histopathological Findings.** For the histopathological examination, a fraction of the cyst wall was fixed in formalin, embedded in paraffin, and stained with H & E. The microscopy findings revealed an intramedullary arachnoid cyst with thick fibrous tissue covered by arachnoid cells (Fig. 2).

**Postoperative Course.** The postoperative course was uneventful, and the MR imaging study performed immediately postoperatively confirmed the removal of the cystic lesion (Fig. 3). The preoperatively existing paraparesis resolved, and the patient was able to walk a few meters with assistance and to stand without help at the 1-month follow-up examination. There was no evidence for bowel or urinary bladder dysfunction.

**Discussion**

Arachnoid cysts are very seldom the cause of an intraspinal lesion in children. Campagna et al.\(^3\) reported on a series of 99 children between the ages of 1 and 6 years in whom surgery was performed because of intraspinal lesions. In only 1 patient was an arachnoid cyst the cause of spinal cord compression. The extra- or intradurally located spinal arachnoid cysts are primarily benign lesions, which become clinically relevant due to compression of the spinal cord or nerve roots. The arachnoid cysts are composed of a normal or slightly thickened arachnoid mater, a kind of mesothelium, and are filled with a clear, anemic liquid that corresponds to the CSF.\(^8\) The exact mechanism for the development of arachnoid cysts is not yet known, but several possible causes are discussed.

Extradural arachnoid cysts are thought to be caused by congenital defects of the dura mater or the arachnoid diverticulum, from which the cysts can gradually deve-
Concerning the development of intradural arachnoid cysts, the different theories are divided into 5 main categories: 1) congenital; 2) arachnoid adhesions secondary to an inflammatory process; 3) arachnoiditis secondary to subarachnoid hemorrhage, contrast medium, spinal anesthetics, fibrin glue, and bone dust; 4) traumatic or iatrogenic injuries to the vertebral column; and 5) idiopathic.\(^\text{12}\) Agnoli et al.\(^\text{1}\) state that the degeneration of cells of the arachnoid trabeculae leads to an elevated osmotic effect within the cyst, causing subsequent enlargement. According to Catala and Poirier,\(^\text{4}\) a valve mechanism based on a unidirectional valve as well as an increased secretion from the cells of the cystic wall could be further reasons for the growth of the cyst. The origin of intramedullary arachnoid cysts is rarely described in the literature. Fortuna and Mercuri\(^\text{6}\) postulate the atypical presence of intramedullary arachnoid granulations leading to cystic development on different levels of the spine. This hypothesis is supported by Goyal et al.\(^\text{8}\), who reported a case of an intramedullary arachnoid cyst in a 63-year-old woman. They believed that the intramedullary formation of cysts in misplaced cellular remnants had resulted from embryonic abnormalities.

In general, arachnoid cysts can be associated with other defects of the neural tubes. In 1937, Voss\(^\text{19}\) was the first to describe the possible correlation between congenital cysts and dysraphic anomalies such as spina bifida. Rabb et al.\(^\text{16}\) strengthened this observation by examining 11 patients between the ages of 19 months and 18 years who had spinal arachnoid cysts (and a coexisting myelomeningocele in 6 of the patients). The reported spinal arachnoid cysts were predominantly found in the thoracic spine (80%); 15% were in the cervical spine; and cysts were least frequently found in the lumbar spine (5%).\(^\text{9,12}\) Especially in children, arachnoid cysts are commonly located in the thoracic spine.\(^\text{16}\) The clinical pathological features further depend on the size and the level of the arachnoid cyst. The clinical symptoms are most frequently caused by compression of the spinal cord or nerve roots; this is associated with pain, gait disturbances, or weakness of the extremities.\(^\text{1,2}\) Spinal intradural arachnoid cysts are commonly detected during adolescence or in young adulthood, which further emphasizes the rarity of the present case.\(^\text{12}\) The relatively small diameter of the thoracic medulla in combination with a slowly growing
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TABLE 1: Literature review of patients with spinal intramedullary arachnoid cysts*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age, Sex</th>
<th>Clinical Findings</th>
<th>Cyst Location</th>
<th>Outcome</th>
<th>FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aithala et al., 1999</td>
<td>7 yrs, M</td>
<td>progressive paraparesis, severe abdominal disturbance, neck rigidity</td>
<td>T-4</td>
<td>complete recovery</td>
<td>5 days</td>
</tr>
<tr>
<td>Sharma et al., 2004</td>
<td>10 yrs, F</td>
<td>progressive quadriaparesis</td>
<td>C4–T2</td>
<td>good recovery</td>
<td>1 mo</td>
</tr>
<tr>
<td>Sharma et al., 2005</td>
<td>4 yrs, F</td>
<td>progressive quadriaparesis</td>
<td>C4–6</td>
<td>complete recovery</td>
<td>17 mos</td>
</tr>
<tr>
<td>Ghannane et al., 2007</td>
<td>4 &amp; 6 yrs, UK</td>
<td>progressive paraparesis</td>
<td>T3–4 (both patients)</td>
<td>complete recovery</td>
<td>UK</td>
</tr>
<tr>
<td>Guzel et al., 2007</td>
<td>7 yrs, F</td>
<td>progressive quadriaparesis</td>
<td>C2–4</td>
<td>slight lt upper paresis</td>
<td>24 mos</td>
</tr>
<tr>
<td>Lmejjati et al., 2008</td>
<td>12 yrs, M</td>
<td>progressive paraparesis</td>
<td>T3–4</td>
<td>complete recovery</td>
<td>4 mos</td>
</tr>
<tr>
<td>Present study</td>
<td>18 mos, M</td>
<td>progressive paraparesis, constipation, neck rigidity</td>
<td>T5–6</td>
<td>complete recovery</td>
<td>1 mo</td>
</tr>
</tbody>
</table>

* FU = follow-up; UK = unknown.

Cyst may have led to the cessation of medullary compliance. This could be a reason for the sudden onset of the symptoms in our patient.

Aithala et al.² were the first to publish a study positing spinal intramedullary arachnoid cysts as the cause of neurologic symptoms in children. In a pediatric case report of a 7-year-old boy, they report the sudden appearance of diffuse, very strong abdominal pain and a progressive weakness of the lower extremity caused by a thoracic intramedullary arachnoid cyst. In 2007, Ghannane et al.⁷ described 2 more pediatric cases in patients who were 4 and 8 years of age, who experienced a sudden progressive spastic paraparesis. In both cases, diagnosis of a thoracic arachnoid cyst with spinal cord compression was confirmed. Along with those 3, the case report by Lmejjati et al.¹⁴ from 2008 described a fourth pediatric case of a 12-year-old boy with a symptomatic arachnoid cyst in the thoracic spine. In addition, there are 3 more reported cases of symptomatic cervical intramedullary arachnoid cysts in children between 4 and 10 years of age.⁹,¹⁷,¹⁸

The rarity of intradural intramedullary arachnoid cysts as shown in the present case is further confirmed by the investigations reported by Kriss and Kriss¹¹ (1 case) and Lee and Cho¹³ (3 cases). In their analyses of intradural spinal arachnoid cysts, an exclusively extramedullary localization was observed. To the best of our knowledge, a symptomatic intradural arachnoid cyst has not yet been reported in such a young patient (Table 1).

Use of MR imaging is of crucial importance in the preoperative workup of patients with these lesions. Because of the widespread use of this imaging technique, an increasing number of asymptomatic cystic lesions in children and adults will be reported. The differential diagnosis of cystic intramedullary lesions includes neuroepithelial cysts, epidermoid cysts, astrocytoma, ependymoma, teratoma, ganglioglioma, cysticercosis, hydatid cyst, syringomyelia, dermoid tumor, neurenteric cyst, and myelomeningocele.⁹ Whereas in asymptomatic patients a “wait and see” strategy seems to be justified, in symptomatic patients surgery is the treatment of choice.

In all reported cases of symptomatic intramedullary arachnoid cysts, surgery was performed as the treatment of first choice. Surgical options are laminotomy or laminectomy, followed by dorsal root entry zone or medially myelotomy, followed by decompression of the cyst and either resection of its wall or fenestration to the subarachnoid space. The patients recovered in most cases during the postoperative course.²,⁷,⁹,¹⁴,¹⁷,¹⁸

Conclusions

Thoracic intramedullary arachnoid cysts in children have been rarely described in the existing literature, but should be kept in mind in the differential diagnosis of intramedullary cystic lesions. The MR imaging of the vertebral column is crucial when diagnosing such lesions, and surgery is the treatment of choice in clinically symptomatic patients. After successful surgical therapy, a complete recovery of the neurological symptoms is the usual course. This circumstance emphasizes the importance of the differential diagnosis of intramedullary arachnoid cysts in infants, especially in the absence of contrast enhancement.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References


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