Spinal intramedullary arachnoid cyst: case report and literature review
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Abstract
BACKGROUND CONTEXT: Intramedullary arachnoid cysts are extremely rare; only 14 cases have been reported in the literature so far.
PURPOSE: We report on the case of a 31-year-old woman who presented with back pain and progressive paraparesis secondary to a dorsal intramedullary arachnoid cyst detected on magnetic resonance imaging (MRI): the surgical planning and clinico-radiological outcome are discussed along with a review of the relevant literature.
STUDY DESIGN: Case report and literature review.
PATIENT SAMPLE: One patient affected by intramedullary arachnoid cyst.
OUTCOME MEASURES: Magnetic resonance imaging and pathological findings from operative specimens were used to confirm the diagnosis.
METHODS: A 31-year-old woman presented with a 7-year history of back pain that had worsened 3 months before admission to our department; for this reason, the patient had undergone a spinal MRI revealing the presence of a 1-cm cystic intramedullary lesion at the level T11–T12, with no contrast enhancement. After 2 months, the patient presented with a worsening of clinical symptoms complaining of severe back pain radiating to the lower extremities associated with a progressive paraparesis, urinary incontinence, and abdominal pain. Referred to our department, at the time of admission the patient was bedridden because of the impossibility of maintaining a standing position. The patient underwent a T11–T12 laminectomy with fenestration of the cyst.
RESULTS: She experienced an immediate relief of pain symptoms, and by the seventh postoperative day she was able to stand without help and walk a few meters with assistance. By the sixth postoperative month, the patient had significantly improved, having gained the ability to walk alone without assistance with complete resolution of the bladder dysfunctions, with no cyst recurrence after approximately 2 years of follow-up.
CONCLUSIONS: Intramedullary arachnoid cysts should be considered in the differential diagnosis for intramedullary cystic lesions. A particular consideration deserves their occurrence in asymptomatic patients, who should be adequately informed on the possible natural evolution: when symptomatic, surgical therapy should be promptly offered, considering that a postoperative complete recovery is usually observed, regardless of the surgical technique. © 2014 Elsevier Inc. All rights reserved.

Keywords: Intramedullary arachnoid cyst; Spinal cord; Arachnoidal cysts; Surgical procedure; Spinal cysts; Acute paraparesis

Introduction
Spinal arachnoid cysts are uncommon benign lesions and are often diagnosed as incidental asymptomatic findings; however, symptomatic cases also have been reported, requiring surgical intervention [1–3]. They may be found either in the extradural compartment or, less frequently, in the intradural space [1,4]. The extradural cysts originate
from an arachnoid herniation through dural tearings, whereas the less frequent intradural cysts seem to form mainly due to arachnoidal trabeculae alterations [3]. Intramedullary cysts are extremely rare and only 14 cases have been reported in the literature so far [2,3,5–14]. Both their pathogenetic mechanisms and natural history remain unclear; as well, the choice of the most effective surgical approach in cases of symptomatic intramedullary cysts is still matter of debate.

We report on the case of a 31-year-old woman who presented with back pain and progressive paraparesis secondary to a dorsal intramedullary arachnoid cyst detected on magnetic resonance imaging (MRI): the surgical planning and clinico-radiological outcome are discussed along with a review of the relevant literature.

Case report

A 31-year-old woman presented with back pain that had worsened 3 months before admission to our department; for this reason, the patient had undergone a spinal MRI revealing the presence of a 1-cm cystic intramedullary lesion at the level T11–T12 characterized by hypointensity on T1-weighted images and hyperintensity on T2-weighted images, with no contrast enhancement. The patient had a previous history of occipital cerebral cavernoma for which she had undergone surgical treatment 9 years earlier.

After 2 months, the patient presented a worsening of clinical symptoms complaining severe back pain radiating to the lower extremities, unrelated to posture, associated with a progressive weakness of her lower limbs; she also reported urinary incontinence and abdominal pain.

Therefore the patient was referred to our department. At the time of admission, the patient was bedridden because she was not able to maintain the standing position. The neurologic examination revealed a severe paraparesis with motor power grade 1/5 of the lower extremities; the knee and ankle joints were exaggerated in both lower limbs and the Babinski sign was present bilaterally. Sensory examination detected a partial loss of all sensory modality below D12–L1 territories. Anal sphincter control was normal, but she presented neurogenic bladder with episodes of urinary incontinence and postmiction residue of about 500 mL.

She underwent new spinal MRI, confirming the presence of the cyst, which had slightly increased in size (Fig. 1, Left and Right).

The patient underwent a T11–T12 laminectomy. After dural opening, the spinal cord appeared bulging posteriorly for a 1.7-cm length. The cyst was punctured and about 1.5 mL of clear cerebrospinal fluid (CSF)-like fluid was aspirated; the examination of the fluid confirmed the characteristics of CSF components. The cyst walls collapsed completely, with no evidence of cysts reexpansion after Valsalva maneuvers and fenestration of the cyst was obtained through a small median mielotomy.

The postoperative course was uneventful. The patient experienced an immediate relief of pain symptoms and at the seventh postoperative day she was able to stand without
help and walk a few meters with assistance. She was therefore referred to the physiotherapy program and after 2 months had a progressive improvement of motor deficits; at that time, urinary retention problems were still reported. By the sixth postoperative month, the patient had significantly improved, having gained the ability to walk alone without assistance with complete resolution of the bladder dysfunctions.

Seriated postoperative MRIs (Fig. 2, Left and Right) showed a satisfactory decompression of the spinal cord with no evidence of cyst recurrence at last follow-up (26 months).

Discussion

Intramedullary arachnoid cysts represent an uncommon neurosurgical issue; since the first description by Aithala et al. in 1999 [5], only 14 cases have been reported in the literature (Table) [2,3,5–14].

Because of their rarity, many questions remain unresolved, in particular those concerning the etio-pathogenetic mechanisms, the natural history, and the appropriate surgical procedure.

The development of intradural arachnoid cysts have been explained using different theories that consider at least five main categories, namely congenital, postinflammatory, posthemorrhagic, posttraumatic or secondary to iatrogenic injuries, and idiopathic causes [3]. Fortuna and Mercuri [15] discussed on the possible origin of arachnoid intradural cysts from cells of the arachnoidal trabeculae occurring congenitally at an abnormal site, prevalently at the thoracic level, arising from the septum posticum, which divides the dorsal subarachnoid space in the midline [1]. This does not explain the occurrence of these cysts over the ventral aspect of the spinal cord, as well as in the lumbar region. However, both the theory of the presence of misplaced arachnoid cellular remnants along the spinal axis and the assumption that there may be normal areas of less resistance in the arachnoid at the level of the granulations may explain the formation of arachnoidal diverticula due to variations in CSF hydrodynamics [3,15].

Factors strengthening the congenital theory include the frequent association with central nervous system abnormalities (72% in the Bond et al. series [6]), familial tendencies, and the young age at onset. In particular, according to the literature review, most patients with intramedullary cysts are children (11 of 15 patients) with a mean age at symptom onset of 5.8 years (ranging from 1.5 to 12.0 years); the remaining 5 patients, including the present case, were mainly young adults, with only one exception presenting at 63 years old (respectively 31, 35, 40, 45, and 63 years old). No sex preponderance was detected. None of them reported an association with neurologic congenital anomalies.

History of trauma and infectious disease were reported as precipitating factors in only two cases [5,11], whereas the remaining patients had unremarkable clinical history. Nevertheless, an abrupt clinical onset was described mainly in the pediatric cases, reporting a sudden appearance of severe and progressive neurological deficits with a clinical history length of less than 1 month in most pediatric cases.

On the other hand, adult patients reported initially long-lasting pain symptoms (in one case, even a 10-year history...
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<th>Patients</th>
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<th>Sex/age</th>
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<th>Neurological examination</th>
<th>MRI</th>
<th>Treatment</th>
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<th>Follow-up</th>
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</thead>
</table>
| Pediatric patients | Athala et al. 1999 | M/7 y    | • Severe pain in the abdomen  
• Progressive weakness of acute onset in the lower limbs  
• Inability to walk/stand  
• Bowel dysfunction  
• 5 days  | • Paraparesis with power limbs grade 2/5  
• Truncal ataxia  
• Hyperreflexia with bilateral clonus at lower limbs  
• Bilateral Babinsky sign  
• Romberg’s sign positive  
• Sensory loss around the T5 segment  
• Neck rigidity  
• Quadripareisis  | T1 intramedullary cyst (1 cm dmt) | • Laminectomy T3–T4  
• Median mielotomy  
• Histological examination of the resected cyst wall  
• Clear CSF was aspirated | • On the second postoperative day the pain in the abdomen and neck rigidity had subsided  
• The sensory loss had improved  
• By the third day the ataxia had improved  
• On the fifth postoperative day he was able to walk independently | Not reported |
|            | Sharma et al. 2004 | F/10 y   | NA                | C4–T1 intramedullary cyst | • Laminectomy C3–T2 with partial excision of the cyst  
• Histological examination of the resected cyst wall  
• Laminectomy C4–C6  
• Median mielotomy  
• Marsupialization  
• Histological examination of the resected cyst wall | Marked improvement | 1 mo |
|            | Sharma et al. 2005 | F/4 y    | • Inability to walk/stand and weakness in UL  
• 20-day history  | C4–C6 intramedullary cyst | • Laminectomy T3–T4  
• Median mielotomy  
• Histological examination of the resected cyst wall  
• Clear CSF was aspirated | • Power grade 5/5 in all limbs by postoperative day 3 | 17 mo |
|            | Ghannane et al. 2007 | M/4 y    | • Difficulty walking and weakness lower limbs  
• 15-day history  | T3–T4 intramedullary cyst | • Laminectomy T3–T4  
• Median mielotomy  
• Histological examination of the resected cyst wall  
• Clear CSF was aspirated | Complete recovery | 6 mo |
|            |                | M/8 y    | • Inability to walk  
• Weakness in lower limbs  
• 1-month history  | T3–T4 intramedullary cyst | • Laminectomy T3–T4  
• Median mielotomy  
• Histological examination of the resected cyst wall  
• Clear CSF was aspirated | • Power limbs grade 4/5  
• Complete recovery at 8 months | 8 mo |
|            | Guzel et al. 2007 | F/7 y    | • Inability to walk  
• Weakness lower limbs  
• 1-month history  | C2–C4 intramedullary cyst | • Laminectomy C3–T2 with partial excision of the cyst  
• Histological examination of the resected cyst wall  
• Laminectomy C2–C5  
• Median mielotomy  
• Marsupialization  
• Histological examination of the resected cyst wall | Full recovery except for a slight left upper paresis | 24 mo |
|            | Lmejjati et al. 2008 | M/12 y   | • Inability to walk  
• Weakness lower limbs  
• 15-day history  | T3–T4 intramedullary cyst | • Mielotomy  
• Marsupialization  
• Histological examination of the resected cyst wall | • Power limbs grade 4/5  
• on third postoperative day | 4 mo |
<table>
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<tr>
<th>Authors</th>
<th>Year</th>
<th>Gender/Age</th>
<th>Symptoms</th>
<th>Interventions</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medved et al. 2009</td>
<td>M/1.5 y</td>
<td>Bowel dysfunction (constipation), Inability to walk/stand, 1-week history</td>
<td>Hypotonic paraparesis, Power limbs grade 1/5, Neck rigidity</td>
<td>Hemilaminectomy T5–T6, DREZ mielotomy, Marsupialization, Histological examination of the resected cyst wall</td>
<td>Clear CSF was aspirated, On the seventh postoperative day he was able to walk, Complete recovery at 4 months</td>
</tr>
<tr>
<td>Kataria R et al. 2012</td>
<td>F/9 y</td>
<td>Back pain (1 year), Dribbling of urine, Numbness both LL (6 months)</td>
<td>Weakness both ankles, Saddle anesthesia, Decreased anal tone</td>
<td>Thoraco-lumbar intramedullary cyst (conus)</td>
<td>Gradual improvement of ankle joints, Improved bladder sensation, No bowel dysfunction</td>
</tr>
<tr>
<td>Bond et al. 2012</td>
<td>M/2 y</td>
<td>Back pain, bilateral low extremity pain</td>
<td>Back pain, bilateral low extremity pain, Spastic paraparesis, Power grade 1/5 on the right, and 2/5 on the left lower limb</td>
<td>Laminectomy T9–L2, Median mielotomy, Histological examination of the resected cyst wall</td>
<td>Clear CSF was aspirated, Power limbs 4/5, The patient remains catheterized, Sensation has improved, Power limbs grade 4/5</td>
</tr>
<tr>
<td>Adult patients</td>
<td>Goyal et al. 2002</td>
<td>Low back nonradiating pain (10 y history), Progressive bilateral lower extremity weakness, Bedridden</td>
<td>Power limbs grade 1/5</td>
<td>Laminectomy T9–L2, Median mielotomy, Histological examination of the resected cyst wall</td>
<td>Clear CSF was aspirated, Power limbs 4/5, The patient remains catheterized, Sensation has improved, Power limbs grade 4/5</td>
</tr>
<tr>
<td>Gezici et al. 2008</td>
<td>F/35 y</td>
<td>Weakness of acute onset in the lower limbs, gradually progressed to complete loss of power, Urinary incontinence (2-month history)</td>
<td>T5–T6 intramedullary cyst</td>
<td>T5–T6 DREZ mielotomy bilaterally, Clear CSF was aspirated, Most of the cyst wall was excised and the remainder widely fenestrated bilaterally, Histological examination of the resected cyst wall.</td>
<td>After 3 y she was able to walk independently with little difficulty</td>
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<tr>
<td>Diyora B et al. 2010</td>
<td>F/45 y</td>
<td>2 y history of thoracic back pain Weakness lower limbs-paraplegia Urinary and fecal incontinence</td>
<td>Flaccid paraplegia Absence of sup and deep tendon reflexes Hyposxia below D6</td>
<td>T4–T5 intramedullary cyst</td>
<td>T3–T4 laminectomy Midline myelotomy Cyst wall partially excised</td>
<td>Complete relief of pain</td>
<td>1.5 mo</td>
</tr>
<tr>
<td>Kataria R et al. 2012</td>
<td>F/40 y</td>
<td>6 months Lumbar pain (7 y) with lower limbs irradiation Bowel dysfunction Inability to walk/stand</td>
<td>Paraparesis Urinary symptoms</td>
<td>L1 intramedial cyst</td>
<td>Laminectomy Median myelotomy Cyst wall partially excised</td>
<td>Regained power in the bilateral ankle joints</td>
<td>1 mo</td>
</tr>
<tr>
<td>Present study</td>
<td>F/31 y</td>
<td>Lumbar pain (7 y) with lower limbs irradiation Bowel dysfunction</td>
<td>Progressive spastic paraparesis</td>
<td>T11–T12 intramedullary cyst</td>
<td>Midline myelotomy Cyst wall partially excised</td>
<td>The patient was able to walk with assistance and to stand without help 1 month postoperative; last follow-up completely independent</td>
<td>2 y</td>
</tr>
<tr>
<td></td>
<td></td>
<td>3 months</td>
<td></td>
<td></td>
<td></td>
<td>No bowel dysfunction</td>
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CSF, cerebrospinal fluid; dtm, diameter; DREZ, dorsal root entry zone; extr, extremities; LL, lower limbs; MRI, magnetic resonance imaging; NA, not available; UL, upper limbs.

Neuroepithelial cysts and neuroenteric cysts [15].

Neuroepithelial cysts are intradural, lined with ependymal tissue, arising from the proliferation of ependymal cells. Neuroenteric cysts are lined with ependymal tissue, arising from the proliferation of foregut endoderm. Variations in the pathogenesis of arachnoid cysts include congenital malformations, traumatic cysts, and idiopathic cysts. The differential diagnosis includes mainly neuroepithelial cysts and neuroenteric cysts [15].

On MRI, intramedullary arachnoid cysts appear typically as hypointense on T1-weighted images and hyperintense on T2-weighted images with no contrast enhancement and no peritumoral edema. The cysts are typically round or ovoid, with a thin wall and a smooth contour.

The natural history of spinal arachnoid cysts is still far from being defined. Some patients remain quiescent throughout life, even undergoing spontaneous regression of the cysts. However, this observation concerns the definition of purely intramedullary arachnoid cysts as a dynamic disease. In our case, we observed that the cyst, identified at a first spinal MRI performed for chronic lumbar pain, had become larger at a further radiological examination performed 2 months later for severe worsening of symptoms. Several hypotheses have been proposed to explain the mechanism of enlargement of arachnoid cysts [10].

The question raised by this observation concerns the definition of intramedullary arachnoid cysts as a dynamic disease. In our case, we observed that the cyst, identified at a first spinal MRI performed for chronic lumbar pain, had become larger at a further radiological examination performed 2 months later for severe worsening of symptoms.
of ectopic ependymal cells, are frequently found close to the ependymal canal, in particular over the anterior aspect of the cord, preferring the thoracolumbar region, in particular at the cono-caudal segment. Neuroenteric cysts, also known as endodermal or enterogenous cysts are characterized by a lining of endodermal origin as a consequence of a developmental etiopathogenesis from notochordal remnants; indeed, they are frequently associated with severe developmental neurovertebral abnormalities, namely diastematomyelia, and they involve prevalently the cervical and cono-caudal segments [15].

However, the surgical planning is not significantly modified in the different types of intramedullary cysts, because partial removal of the cyst wall with fenestration to the subarachnoid space, which is often the only possible procedure due to the tight adhesions to the spinal cord, is generally sufficient to prevent recurrence; wall biopsy is generally advocated to exclude a tumor nature [15].

Conclusion

Intramedullary arachnoid cysts have been rarely described in the literature; however, with an increasing frequency during the past decade, probably because of the wide availability of MRI, this should be kept in mind when considering the differential diagnosis for intramedullary cystic lesions. A particular consideration deserves its occurrence in asymptomatic patients, who should be adequately informed on the possible natural evolution. When symptomatic, surgical therapy should be promptly offered, considering that a postoperative complete recovery is usually observed, regardless of the surgical technique.

References