Spinal intramedullary arachnoid cyst in a 4-year-old girl: a rare cause of treatable acute quadriparesis

Case report

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The authors report their experience in successfully treating a 4-year-old girl who presented with sudden onset of quadriparesis that lasted for 20 days. Magnetic resonance (MR) imaging of the spine revealed an intramedullary cystic lesion extending from C-4 to C-6. A C4–6 laminectomy was performed followed by a median myelotomy. The cyst was decompressed and most of the cyst wall was excised. The histopathological findings were consistent with those of an arachnoid cyst. By postoperative Day 3, power had gradually returned to normal in all her limbs. On follow-up reviews at 2 and 17 months, the results of her neurological examinations remained normal. Follow-up MR imaging of the spine at 17 months revealed an intramedullary residual cystic lesion extending from C-5 to C-6, without any mass effect. An intramedullary arachnoid cyst should be considered in the differential diagnosis of an intramedullary cystic lesion.

KEY WORDS • intramedullary arachnoid cyst • magnetic resonance imaging • cervical spine • pediatric neurosurgery

SYMPTOMATIC spinal arachnoid cysts in the pediatric age group are rare. The clinical manifestations range from acute to chronic and gratifying results are often yielded when these entities are diagnosed and treated before the spinal cord becomes irreversibly damaged. The origin of these cysts remains unclear; however, congenital, traumatic, and inflammatory causes have been postulated. They are more often situated extradurally than in an intradural extramedullary location. Extradural cysts are believed to arise from defects in the dura through which the arachnoid herniates, whereas the intradural cysts probably result from an alteration in the arachnoid trabeculae.

Symptomatic spinal intramedullary arachnoid cysts are very rare. To our knowledge, only two case reports of intramedullary arachnoid cysts have been published. We report the third case, involving the youngest patient so far, and our experience in managing this child.

Case Report

Presentation. This 4-year-old girl presented to us with a 20-day history of inability to walk and weakness in her upper limbs. The deficit was sudden in onset and had appeared simultaneously in her four limbs, without any ascending/descending pattern. There was no history of any impaired bladder or bowel function, fever, convulsions, trauma, Koch contact, or any recent immunization. Her developmental milestones were within normal limits.

Examination. The patient was conscious and cooperative. Her vital parameters were normal. There was no respiratory distress or paradoxical respiration. There were no signs of wasting of muscles. Hypertonia was present in her lower limbs, whereas the upper limbs displayed hypotonia. Power in her lower limbs was Grade 0/5. The upper-right limb power was Grade 4/5 and that of the upper-left was Grade 2/5. Superficial abdominal reflexes were equivocal. The knee and ankle jerks were exaggerated in both lower limbs. In both upper limbs, the biceps reflexes were normal but the triceps and supinator jerks were depressed. Bilateral ankle clonus was present, and the plantars were extensor bilaterally. There were no cranial nerve deficits. There was no obvious sensory deficit but because of the child’s young age, pain and temperature sensations could not be tested. The results of funduscopic examination were normal. Neck rigidity was absent. The spine was nontender and no deformity was noted.

Abbreviations used in this paper: CSF = cerebrospinal fluid; MR = magnetic resonance.
The results of laboratory findings were within normal limits. The hemoglobin level was 12 g/dl; white blood cell count was 7200/mm³ with differential count showing 63% polymorphonuclear leukocytes, 34% lymphocytes, and 3% eosinophils. Serum sodium and potassium levels were normal. The results of a Mantoux test were negative. Chest and cervical spine x-ray films revealed normal results. A lumbar puncture revealed normal CSF. Both T₁- and T₂-weighted MR images of the brain and spine were obtained. The MR images demonstrated normal results; however, imaging of the spine revealed a large intramedullary cystic lesion in the cervical cord extending from C-4 to C-6 (Fig. 1).

The etiological diagnosis of the cystic lesion could not be made based on the MR images. In view of the lack of edema in the surrounding spinal cord tissue and also lack of any abnormal vessels adjacent to the lesion, the possibility of the lesion being a hemangioblastoma was considered unlikely, and the absence of abnormal soft tissue within the lesion made the possibility of the lesion being an astrocytoma unlikely. The possibility of the cystic lesion being a teratoid cyst, a syrinx, a cysticercus cyst, or an arachnoid cyst was considered.

Operation. The patient underwent surgery within 5 days of admission to the hospital. A C₄–₆ laminectomy was performed: the dura mater was opened and a median myelotomy was performed. The cyst, which contained clear CSF-like fluid, was decompressed, and most of the cyst wall was excised. The cyst had no extramedullary component or any obvious visible communication with the subarachnoid space.

Histopathological Examination. A portion of the cyst wall was fixed in formalin, embedded in paraffin, and stained with H & E. It showed the presence of cystic structures lined by fibrous walls and meningoepithelial cells (Fig. 2). A histopathological diagnosis of an arachnoid cyst was thus made.

Postoperative Course. Postoperatively the patient had no complications and power gradually returned to Grade 5/5 in all her limbs by postoperative Day 3. The child was discharged on postoperative Day 10. During follow-up visits at 2 and 17 months, the results of her neurological examinations remained normal; she had made a complete recovery. Follow-up MR imaging of the spine at 17 months revealed a smaller intramedullary cyst extending from C-5 to C-6, suggesting a refilling of the cyst. The irregular outline of this residual cyst indicated that it was not tense. The sur-

Fig. 1. Preoperative MR images. Upper Left: A T₁-weighted coronal image of the cervical and upper dorsal spine revealing a well-defined hypointense intramedullary lesion extending from C-4 to C-6 with spinal cord widening. Lower Left: A T₂-weighted coronal image of the cervical and upper dorsal spine demonstrating the hyperintense intramedullary cyst extending from C-4 to C-6 with cord widening. Right: A T₂-weighted sagittal image of the cervical spine revealing the oval hyperintense intramedullary lesion at C₄–₆.
rounding spinal cord was seen and there was no ischemia due to compromise of blood supply, thus the cyst was not causing any significant mass effect (Fig. 3).

Discussion

Spinal arachnoid cysts are a relatively uncommon lesion that may be either intra- or extradural: intradural spinal arachnoid cysts are even less common. Based on a case series of 22 cases, Nabors, et al., proposed a classification of congenital spinal meningeal cysts into three categories: spinal extradural meningeal cysts without spinal nerve root fibers (Type I), spinal extradural meningeal cysts with spinal nerve root fibers (Type II), and spinal intradural meningeal cysts (Type III). Type I meningeal cysts were further classified into Type IA, the extradural arachnoid cyst and Type IB, the sacral meningocele. This classification was based on findings from surgical examinations and histopathological reviews. In their case series the majority of cysts were of the extradural variety (nine each of Types I and II). Only four were of the Type III variety, and none was of the intramedullary type. Based on this classification system the arachnoid cyst in our patient was of the Type III variety.

According to a recent review of intradural spinal arachnoid cysts by Kriss and Kriss, men and women are affected equally and usually in the third to fifth decades of life. The majority of intradural spinal arachnoid cysts occur in the thoracic region (80%), with only 15% in the cervical region, and 5% in the lumbar region. Most are dorsal to the neural elements (80%) in the canal and only 20% are ventrally located. Kriss and Kriss have made no mention of intradural intramedullary arachnoid cysts. Recently, Lee and Cho reviewed 10 symptomatic cervical spinal intradural arachnoid cysts, of which eight were in the pediatric age group, and again none was of the intramedullary type. These two reviews indirectly highlight the extreme rarity of the condition that we report.

Fig. 2. Photomicrograph of the cyst wall showing the flat cuboidal epithelium lining of the cyst along with its collagenous connective tissue wall containing blood vessels. H & E, original magnification × 100.

Fig. 3. Follow-up sagittal MR images. Left: A T₁-weighted image of the cervical and upper dorsal spine revealing a hypointense intramedullary residual cyst extending from C-5 to C-6 with an irregular outline. Right: A T₂-weighted image of the cervical and upper dorsal spine demonstrating a hyperintense intramedullary residual cyst extending from C-5 to C-6 with an irregular outline.
Because of their rarity, the natural history of arachnoid cysts, at all anatomical sites, is still not clearly known. Some of these cysts are quiescent throughout life and some remain dormant for decades before manifesting clinically.\textsuperscript{1,4,6} Arachnoid cysts involving the cervical region, as in our case, are also extremely rare.\textsuperscript{2,10}

Our patient is the third case report of a symptomatic spinal intramedullary arachnoid cyst and the second in the pediatric age group. Aithala, et al.,\textsuperscript{1} were the first to report this condition. They reported on a 7-year-old boy who was admitted for medical treatment with a 5-day history of unexplained neck rigidity and unexplained diffuse constant severe pain in the abdomen, truncal ataxia, and progressive weakness in the lower limbs. He also had hesitancy of micturition and constipation. Decreased sensory perception was evident symmetrically in the lower limbs for all modalities of sensation. Based on MR imaging of the spine a spinal intramedullary cyst was diagnosed at T-4. There was a dramatic and immediate recovery after fenestration of the cyst.\textsuperscript{1} Goyal, et al.,\textsuperscript{4} reported on the second case, a 63-year-old woman who had a history of low-back pain dating back nearly 10 years. For 4 years, she had experienced progressive bilateral lower-extremity weakness, including an increase in the frequency of urination. She had been bedridden for the 3 months prior to presentation due to severe bilateral paraparesis. Spinal MR imaging had revealed an intramedullary cyst at the T9–L2 level. Postoperatively the patient regained power of Grade 4/5 in both her lower limbs. In both these cases, as in our patient, the confirmatory diagnosis of the lesion being an arachnoid cyst was made only postoperatively by histopathological examination.

For all types of spinal arachnoid cysts, MR imaging is the diagnostic procedure of choice because of its potential to demonstrate the exact localization, extent, and relationship of the arachnoid cyst to the spinal cord.\textsuperscript{3} Wide decompression is recommended as the surgical procedure for the treatment of an intramedullary arachnoid cyst as well as removal of as much of the cyst wall as possible, so as to improve the clinical symptoms.\textsuperscript{1,4} Total excision of the intramedullary cyst lining may not be possible due to adhesion of the lesion to the spinal cord.\textsuperscript{2} In our case the cyst was decompressed and most of the cyst wall was excised, following which the patient’s quadriparesis resolved gradually by postoperative Day 3. On follow-up visits at 2 and 17 months the child remained symptom free. In view of the residual cyst detected on the follow-up MR imaging of the spine at 17 months we have asked the parents to continue with long-term follow-up visits for this child. The follow-up period of 17 months being reported by us is the longest period in which successful treatment of a spinal intramedullary arachnoid cyst has been documented. Aithala, et al.,\textsuperscript{1} have not reported any follow up of their patient. Goyal, et al.,\textsuperscript{4} have reported a short follow-up period of 3 months and their patient exhibited gradual improvement. An MR image of the spine obtained 10 days postoperatively had demonstrated a significantly decompressed cyst, but no follow-up MR image was obtained at 3 months.\textsuperscript{4}

**Conclusions**

An intramedullary arachnoid cyst should be considered in the differential diagnosis of an intramedullary cystic lesion. In our patient, urgent MR imaging and prompt surgery to decompress the cyst and remove as much of the cyst wall as possible helped resolve the quadriparesis.

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**References**


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