Spinal arachnoid cysts in the pediatric population: report of 31 cases and a review of the literature

Clinical article

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Object. The goal of this study was to review all cases of pediatric spinal arachnoid cysts (SACs) surgically treated at the authors’ institution between 1992 and 2008 and to compare these cases to the published literature for the general population.

Methods. The charts of all pediatric patients with SACs were reviewed for demographics, medical history, presenting symptoms, imaging findings, operative procedure(s), complications, and outcomes. Following a complete literature review, the pediatric data were compared with data from the general population and unique findings associated with pediatric patients were identified.

Results. Thirty-one pediatric patients (median age 6.9 years) underwent operative intervention for SACs between 1992 and 2008 (median duration of follow-up 4.2 years). There were 17 female patients (55%) and 14 male patients (45%). Twenty-one patients (68%) presented with symptoms of radiculopathy or myelopathy. The most common presenting symptoms were pain (42%), lower-extremity weakness (39%), gait instability (32%), spasticity (19%), sensory loss (10%), and bladder dysfunction (7%). In 3 patients (10%) SACs were incidental findings. Intradural SACs were more common (18 patients, 58%) than extradural SACs (11 patients, 36%). One patient (3%) had extradural and intradural components. One patient (3%) had a purely intramedullary cyst, and 1 patient (3%) had both an intradural and intramedullary component. Of the 18 intradural SACs, 9 (50%) were located dorsal to the spinal cord and 9 (50%) were ventrally located. One dorsal intradural SAC had an intramedullary component. All extradural SACs were located dorsal to the spinal cord. Intradural SACs were primarily concentrated in the cervical and thoracic regions (67%), whereas extradural cysts were more evenly distributed between the thoracic, lumbar, and sacral regions. Of the 18 patients with intradural SACs, 13 (72%) had significant previous CNS abnormalities, compared with 3 (27%) of 11 patients with extradural SACs. There were 2 operative complications. One patient had a CSF leak treated with a lumbar drain, and the second patient had a pseudomeningocele. No patients had neurological deterioration as a result of surgical intervention. Twenty-one patients (68%) had complete remission of symptoms, 6 (19%) had improvement, 3 (10%) were stable, and 1 (3%) has worsening of symptoms with recurrence that ultimately required cystoperitoneal shunting, despite multiple failed attempts at fenestration.

Conclusions. Spinal arachnoid cysts are rare lesions in the pediatric population. Affected patients present with back pain, weakness, and/or gait instability. In children, SACs predominantly develop in the thoracic region and are more likely to occur intradurally, compared with SACs in the general population. Overall outcomes following surgical fenestration or excision of SACs are excellent, with complete remission or improvement of symptoms achieved in 87% of cases.

Key Words • arachnoid cyst • spinal cyst • intradural • extradural • spine • pediatric neurosurgery

Spinal arachnoid cysts are a rare cause of spinal cord compression, nerve root compression, or both. Patients may present with symptoms of pain, weakness, ataxia, and/or bladder incontinence. In the modern era of neuroimaging, these lesions can also be discovered incidentally. Once the cyst has been diagnosed, typically by MRI, surgery is usually performed to prevent further spinal cord or nerve root compression. Depending on the location of the cyst and the duration and degree of spinal cord compression, postoperative outcomes vary substantially. Some degree of improvement is reported in 45%–70% of cases following surgery, and complete elimination of symptoms is reported in 20%–30% of cases.6,24,42,50,64,69 Most SACs are extradural and arise dorsal to the spinal cord.29,48 Many cases of intradural cysts

Abbreviations used in this paper: CM-I = Chiari malformation Type I; CM-II = CM Type II; SAC = spinal arachnoid cyst.
Spinal arachnoid cysts in the pediatric population

and some rare cases of intramedullary cysts have been reported. Generally speaking, extradural SACs are resected, whereas intradural SACs are usually fenestrated. Although they most commonly develop in the thoracic region, SACs may arise at any spinal level. The mechanism of formation and enlargement of SACs is likely multifactorial, and numerous theories have been proposed.

Owing to the rare nature of SACs, the majority of cases have been reported as single case reports. The largest series was reported in 1988 by Nabors et al., who described their experience with 22 patients treated for SACs. Only 6 pediatric patients, however, were included in that report. In contrast, the present paper is a review of 31 cases involving pediatric patients (median age 6.9 years) treated surgically for SACs from 1992 through 2008 at Children’s Hospital Los Angeles. To our knowledge, this is the largest single series of SACs in pediatric patients.

Methods

Following institutional review board approval, all patients treated surgically over a 17-year period (1992–2008) at Children’s Hospital Los Angeles were identified from a departmental database and reviewed in a retrospective fashion. Pediatric patients with SACs confined to the cervical, thoracic, lumbar, and sacral regions were included; all intracranial arachnoid cysts were excluded from the analysis. Patient demographic characteristics, presenting symptoms, preoperative imaging studies, operative reports, hospital progress notes, postoperative imaging studies, and notes from follow-up clinic visits were reviewed in all cases.

Postoperative outcomes were categorized as complete remission, improvement, stable, or worse. Complete remission was defined as MRI evidence of complete cyst removal or decompression with a corresponding resolution of symptoms. Improvement was defined as MRI evidence of cyst removal or decompression with a corresponding improvement of symptoms. A stable outcome was defined as no change in symptoms and stable cyst size on MRI. A worse outcome was defined as clinical progression of symptoms.

Results

Patient Demographic Characteristics and Presenting Symptoms

Thirty-one pediatric-age patients underwent operative intervention for SACs between 1992 and 2008 and were included in the study. Patient demographic characteristics, presenting symptoms, significant medical history, location of cyst, procedure performed, complications, follow-up time, and outcome are highlighted in Table 1. A summary of the patient demographic characteristics and presenting symptoms is shown in Table 2. There were 17 female patients (55%) and 14 male patients (45%). Twenty-one patients (68%) had a remarkable medical history including the following; scoliosis (7 patients), Prader-Willi syndrome (1 patient), intraventricular hemorrhage at birth (1 patient), chronic hydrocephalus (10 patients), CM-I (3 patients), CM-II (1 patient), open and closed neural tube defects (9 patients), and unilateral (1 patient) or bilateral (1 patient) coronal synostosis. Only 10 patients (32%) presented with a newly diagnosed, isolated SAC.

Twenty-one patients (68%) presented with symptoms of radiculopathy or myelopathy. The most common presenting symptom was pain (42%), followed by lower-extremity weakness (39%), gait instability (32%), spasticity (19%), sensory loss (10%), and bladder dysfunction (7%). In 3 patients (10%) SACs were diagnosed as an incidental finding as a result of MRI performed for other CNS abnormalities or spine deformities. Of the 13 patients presenting with pain, 10 (77%) had back or neck pain, 3 (23%) had bilateral leg pain, and 1 (8%) had unilateral leg pain. Among the 12 patients presenting with lower-extremity weakness, there was representation of cysts in all spinal regions (1 cervical, 1 thoracocervical, 2 thoracic, 4 thoracolumbar, 1 lumbar, 3 lumbosacral, and 1 sacral). Of the 3 patients in whom cysts were found incidentally, one had a sacral cyst that was diagnosed during a workup for a paraspinous tumor. The second patient had a thoracic cyst which was diagnosed during evaluation of asymmetry in the lower thoracic region. The third patient also had a thoracic cyst, which was diagnosed during MRI follow-up for CM-I decompression and shunt-treated hydrocephalus. The cyst was causing significant compression of the cord as demonstrated in Fig. 1. In light of the cyst size and imaging evidence of significant compression of the spinal cord or nerve roots in each case, and the young age of these patients, the lesions were surgically addressed in order to prevent future neurological deficits.

Imaging Findings

In all patients, a preoperative diagnosis of SAC was obtained using MRI. The locations of the SACs are summarized in Table 3. There were more patients with extradural SACs (18 patients, 58%) than intradural (11 patients, 36%) and intramedullary (1 patient, 3%) SACs. Of the 18 extradural SACs, 9 (50%) were located ventral to the spinal cord and 9 (50%) were located dorsal to the spinal cord. One dorsal intradural SAC also had an intramedullary component. All extradural SACs were found dorsal to the spinal cord. Of the total population of 31 patients, 21 (68%) of SACs were dorsal, 9 (29%) were ventral, and 1 (3%) was intramedullary. The distribution along spinal cord segments was different depending upon the type of SAC. Of the 18 extradural SACs, 1 (6%) was cervical, 4 (22%) thoracocervical, 7 (39%) thoracic, 2 (11%) thoracolumbar, 1 (6%) lumbar, 3 (17%) lumbosacral, and 0 sacral. In the 9 patients with ventrally situated intradural SACs, 1 lesion (11%) was located in the cervical region, 3 (33%) were thoracocervical, 2 (22%) thoracic, 1 (11%) lumbar, and 2 (22%) lumbosacral. In the 9 patients with dorsally situated intradural SACs, 1 lesion (11%) was thoracocervical, 5 (56%) were thoracic, 2 (22%) were thoracolumbar, and 1 (11%) was lumbosacral. Of the 11 extradural dorsal SACs, 3 (27%) were thoracic, 4 (36%) thoracolumbar, 1 (9%) lumbosacral, and 3 (27%) sacral. The 1 intradural and extradural dorsal SAC was located in the lumbar region. The 1 purely intramedullary SAC was located in
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<th>Case No.</th>
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<th>Age (yrs), Sex</th>
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* BLE = bilateral lower-extremity; BUE = bilateral upper-extremity; Comp = component; decr = decreased; dev = developmental; fenestr = fenestration; FU = follow-up; HC = hydrocephalus; Intramed = Intramedullary; LS = lumbosacral; MMC = myelomeningocele; NF1 = neurofibromatosis Type 1; NTD = neural tube defect; progr = progressive; Recurr = recurrence; sig = significant; Sz = seizure; S/P = status post; UE = upper-extremity.
the thoracic region. The distribution of SACs by location for the group as a whole was as follows: cervical in 1 case (3%), thoracocervical in 4 cases (13%), thoracic in 11 (36%), thoracolumbar in 6 (19%), lumbar in 2 (6%), lumbosacral in 4 (13%), and sacral in 3 (10%). The mean anterior-to-posterior SAC diameter was 14 mm (range 8–21 mm), and the mean transverse diameter was 18 mm (range 9–36 mm). The mean number of involved spinal cord segments was 4.0, with a range of 1–12 segments.

Operative Complications

There were 2 operative complications in the series. Patient 13 developed a postoperative CSF leak, which was treated via insertion of a lumbar drain. This patient had a history of myelomeningocele repair and significant scarring compromising the remaining dura mater during the operative closure. Patient 26, the only patient with both intradural and extradural components, had a pseudomeningocele, which was repaired later. No patients developed wound infections. No patient had neurological deterioration as a result of operative intervention.

Postoperative Clinical Course

Follow-up times ranged from a minimum of 13 days to a maximum of 12.6 years (median 4.2 years, mean 4.4 years). The summary of postoperative outcomes is shown in Table 4. Analysis of the results in all 31 cases showed that 21 (68%) patients had complete remission of their symptoms, 6 (19%) had symptom improvement, 3 (10%) had stable symptoms, and 1 patient had worsened symptoms. When the outcome in patients with extradural and intradural cysts were analyzed independently, the results were not notably different. Of the 18 patients with intradural SACs, 12 (67%) experienced complete remission of their symptoms, compared with 7 (64%) of the 11 patients with extradural SACs. When remission and improvement were combined, the intradural group had 15 patients (83%) and the extradural group had 10 patients (91%) with improvement or complete remission. The patient with a purely intramedullary cyst had complete remission.

The 3 patients with stable postoperative outcomes included 2 with intradural SACs and 1 with an extradural SAC. The first of these 3 patients (Patient 6) was a 1-year-old girl with no other significant medical history, who presented with bilateral lower-extremity distal motor dysfunction and a dorsal extradural cyst from S-2 to S-4. The cyst was excised without complication. An MRI study performed 3 months postoperatively demonstrated that the cyst had been completely removed and there was no evidence of sacral nerve root compression. However, the patients’ symptoms remained stable and no improvement was noted. The second patient (Patient 27) was a 5.5-year-old girl with a history of myelomeningocele, tethered cord, and hydrocephalus. Fenestration was performed successfully, and no cyst recurrence was noted on follow-up MRI. However, the patient’s gait instability and bladder dysfunction continued as expected owing to her

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**TABLE 2: Summary of demographic and clinical characteristics in 31 patients**

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Value*</th>
</tr>
</thead>
<tbody>
<tr>
<td>sex</td>
<td></td>
</tr>
<tr>
<td>female</td>
<td>17 (55)</td>
</tr>
<tr>
<td>male</td>
<td>14 (45)</td>
</tr>
<tr>
<td>age (yrs)</td>
<td></td>
</tr>
<tr>
<td>median</td>
<td>6.9</td>
</tr>
<tr>
<td>range</td>
<td>1.0–17.5</td>
</tr>
<tr>
<td>history of other sig CNS abnormality</td>
<td></td>
</tr>
<tr>
<td>no</td>
<td>10 (32)</td>
</tr>
<tr>
<td>yes</td>
<td>21 (68)</td>
</tr>
<tr>
<td>presenting symptom</td>
<td></td>
</tr>
<tr>
<td>pain</td>
<td>13 (42)</td>
</tr>
<tr>
<td>lower-extremity weakness</td>
<td>12 (39)</td>
</tr>
<tr>
<td>gait instability</td>
<td>10 (32)</td>
</tr>
<tr>
<td>spasticity</td>
<td>6 (19)</td>
</tr>
<tr>
<td>sensory loss</td>
<td>3 (10)</td>
</tr>
<tr>
<td>bladder dysfunction</td>
<td>2 (7)</td>
</tr>
<tr>
<td>incidental finding of SAC</td>
<td>3 (10)</td>
</tr>
</tbody>
</table>

* Values represent numbers of patients (%) unless otherwise indicated.

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**Fig. 1.** Patient 20. Sagittal T2-weighted MR image obtained in a patient with a history of shunt-treated hydrocephalus and a CM-I decompression. This T4–7 anterior intradural extramedullary cyst was noted on follow-up MRI without corresponding neurological findings. In light of the severe compression of the cord, the cyst was addressed surgically to prevent future neurological deficit. At 5-year follow-up the patient remained active in sports with normal findings on physical examination.
Spinal arachnoid cysts in the pediatric population

TABLE 3: Location of SACs in 31 pediatric patients*

<table>
<thead>
<tr>
<th>Location</th>
<th>Cervical</th>
<th>Thoracocervical</th>
<th>Thoracic</th>
<th>Thoracolumbar</th>
<th>Lumbar</th>
<th>Lumbosacral</th>
<th>Sacral</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>intradural</td>
<td>1 (3)</td>
<td>3 (10)</td>
<td>2 (6)</td>
<td>0 (0)</td>
<td>1 (3)</td>
<td>2 (6)</td>
<td>0 (0)</td>
<td>9 (29)</td>
</tr>
<tr>
<td>ventral</td>
<td>0 (0)</td>
<td>1 (3)</td>
<td>5 (16)</td>
<td>2 (6)</td>
<td>0 (0)</td>
<td>1 (3)</td>
<td>0 (0)</td>
<td>9 (29)</td>
</tr>
<tr>
<td>extradural</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>3 (10)</td>
<td>4 (13)</td>
<td>0 (0)</td>
<td>1 (3)</td>
<td>3 (10)</td>
<td>11 (36)</td>
</tr>
<tr>
<td>dorsal</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (3)</td>
</tr>
<tr>
<td>intradural &amp; extradural</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (3)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (3)</td>
</tr>
</tbody>
</table>

* Values represent numbers of SACs (%).

Discussion

Since the first verified report by Collins and Marks in 1915,20 SACs have been a rare cause of spinal cord or nerve root compression, especially in the pediatric age group. The differential diagnosis for cystic intraspinal lesions includes cystic tumors, ependymal cysts, neuroenteric cysts, teratogenous cysts, and epithelial cysts.45 Campagna et al.12 reported a series of 99 children between the ages of 1 and 6 years who were operated on for intraspinal lesions, and of those cases only 1 was found to be an arachnoid cyst. Most cases of arachnoid cysts in the literature are either reports of single cases,3,5,9,11,15,18,19,25,26,28,29,31,34,35,39,40,43,45,47,49,52–54,56–58,60,61,67,72–74,76–78,81,83,87,88,92 small series of 2–3 cases,1,2,10,16,21,32,36,38,41,46,48,55,62,65,68,71,84,91 or series of 4–10 cases,4,7,22–24,27,33,44,50,51,64 There have been few larger series,6,17,42,63,78 with the largest single-institution experience being a report of 22 cases by Nabors et al.,59 who developed a useful classification system for SACs. The largest pediatric series prior to the present report was by Rabb et al.,69 from our institution, who described 11 pediatric patients with SACs who were treated between 1971 and 1991. Our series of 31 cases of SACs surgically treated at our institution between 1992 and 2008 is the largest reported case series of pediatric SACs to date and provides some insight into the distribution and outcomes of these lesions in pediatric patients.

Although classification has not been standardized, Nabors et al.59 simplified the classification of spinal meningeal cysts by defining 3 major categories: extradural meningeal cysts without nerve root fibers (Type I), ex-
tradural meningeal cysts with nerve root fibers (Type II), and intradural meningeal cysts/intradural arachnoid cysts (Type III). Type I meningeal cysts were further subdivided into IA (extradural arachnoid cysts) and IB (sacral meningoceles or occult sacral meningoceles). Tarlov perineural cysts or spinal nerve root diverticula are the same as Type II meningeal cysts in this classification scheme. In the current series, only Type IA and Type III meningeal cysts were found.

The mechanism for the initial formation of an arachnoid cyst is likely multifactorial and is not completely understood. A few theories have been proposed. Elsberg et al. reported 4 cases of arachnoid cysts in 1934 and proposed an origin from congenital diverticula or a congenital dural defect with herniation of the arachnoid. Support for the congenital theory is further strengthened by a few case reports of familial tendencies and the frequency of association with neural tube defects. Other reports have hypothesized that congenital defects in the distribution of arachnoid trabeculations lead to misplaced cellular remnants resulting in an embryonic malformation, which may act as a nidus for cyst formation. Sixty-eight percent of the patients in our series had a medical history significant for CNS abnormalities, which may lend added support to the theory of congenital malformations in some cases. Unfortunately, we are unable to determine with certainty whether cyst formation in a child with an associated CNS malformation is primary in nature or secondary to the malformation. However, for patients with no significant medical history (32%), one may assume these cysts are primary in nature.

Expansion of an arachnoid cyst after initial formation is also likely multifactorial and may not occur as a result of the same mechanism in all cases. The ball-valve mechanism of enlargement has been described in numerous cases. In the ball-valve theory, the cyst communicates with the subarachnoid space via a small stalk functioning as a one-way valve. McCrum and Williams stressed the importance of having surges in pressure, which fill and expand the cyst, followed by stalk closure, which maintains the higher internal cystic pressure. These surges in CSF pressure may be caused by straining or coughing during normal activities. Without the surges in pressure, the pressure within the cyst and the adjacent subarachnoid space would equilibrate and the cyst would not enlarge. Another proposed mechanism of expansion is via an osmotic gradient formed between the cystic structure and the subarachnoid space, thus facilitating expansion. Yet another proposed mechanism is that the degeneration of cells of the arachnoid trabeculae may lead to an elevated osmotic effect within the cyst. Sandberg et al., however, demonstrated that the chemical composition and the osmolality of cranial arachnoid cysts and the CSF are similar. They found that the protein levels were significantly elevated in the cysts and proposed that the cysts increased in size through colloid osmotic pressure due to the presence of large proteins that cannot pass through semipermeable membranes. Lastly, cyst expansion may be due to active secretion of cells lining the cyst.

The actual mechanism of expansion is likely from a series of events that may include one or more of the aforementioned processes. In our experience, SACs are under pressure, which may even lead to herniation of the spinal cord posteriorly when opening the dura in a patient with a ventrally placed intradural SAC.

In review papers that included intradural and extradural cysts it was found that the majority of cysts (more than 80%) are located posterior to the spinal cord. Wilkins and Odom reviewed 67 cases, including 50 in which lesion location was reported; of these, 47 (94%) were posterior or posterolateral. Perret et al. postulated that SACs develop from dilation of the septum posticum, the thin membranous arachnoid partition that longitudinally divides the posterior subarachnoid space in the midline. Dilation of the septum posticum would explain the frequency with which arachnoid cysts are found in the posterior arachnoid space in the general population. Rabb et al., however, found an association of anteriorly placed cysts in pediatric patients with myelomeningocele, who seemed particularly prone to having anteriorly situated arachnoid cysts that likely developed as a result of altered CSF flow secondary to extensive arachnoid adhesions. Our current pediatric series demonstrates a high proportion of ventrally situated SACs (29%), and of the 9 patients in our series with ventrally situated SACs, 8 (89%) had a history of prior CNS abnormalities.

The most common region for the development of symptomatic SACs is the thoracic canal. performed a literature review of 91 cases of arachnoid cysts from the first case ever discovered at autopsy (1898) to his case report (1968). In his review, 65% cysts were located in the thoracic region: 12% were thoracolumbar, 13% were lumbar or lumbosacral, 6.6% were sacral, and 3.3% were cervical. Another large literature review by Kriss and Kriss in 1997 had similar findings of 80%, 15%, and 5% for the thoracic, cervical, and lumbar regions, respectively. It is worth emphasizing that the thoracic spinal canal is relatively small in diameter and the longest part of the spine, and cysts developing in this region are more likely to become symptomatic than are SACs in other spinal regions, whether in the general population or in pediatric patients. In our series, the distribution of SACs in pediatric patients was as follows: 1 (3%) in the cervical region, 4 (13%) thoracocervical, 11 (36%) thoracic, 6 (19%) thoracolumbar, 2 (7%) lumbar, 4 (13%) lumbosacral, and 3 (10%) sacral. Although pediatric patients have the largest number of SACs found in the thoracic region, as does the general population, they have more SACs located in the lumbar and sacral regions, likely due to the increased prevalence of neural tube defects found in the pediatric population.

In previous reports, SACs were more commonly located extradurally than intradurally. This relationship, however, was not observed in our series. Eighteen (58%) patients in the current series had intradural cysts. Higher prevalence of intradural SACs in the pediatric age group is likely related to congenital CNS abnormalities. Of the 18 patients with intradural SACs, 13 (72%) had significant previous CNS abnormalities. In contrast, of the 11 patients with extradural SACs, only 3 (27%) had significant previous CNS abnormalities. Although arachnoid cysts are usually extra medullary, there have been a few reports of cases of intramedullary arachnoid cysts.
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differentiate an intramedullary cyst from a syrinx, a finding of a normal arachnoid capsule on histopathology is required. Two (9%) of our patients had intramedullary cysts. Patient 21 had a completely intramedullary SAC, whereas Patient 22 had an SAC with both intramedullary and extramedullary components. Both patients had excellent outcomes following surgical fenestration.

Arachnoid cysts usually present as solitary lesions; however, in rare cases multiple septations and multiple independent cysts are found.53,54,58,31,84,91 Arachnoid cysts are sometimes found in association with other anomalies, including spinal cord herniation51,35,60,31 or a syrinx,21,11,33,60,85 and can coexist with nerve root prolapse into the extradural arachnoid cyst with corresponding radiculopathy.77 In our series there was no evidence of spinal cord herniation or nerve root prolapse. However, 6 patients (27%) had complex cysts with either septations, cysts within a cyst, or multiple dural defects. Patient 9 had an extradural cyst with 2 separate dural defects. Patient 7 was noted to have 3 separate cystic structures. In Patients 10, 16, and 20, the surgeon experienced resistance when passing an infant feeding tube in the subarachnoid space during the operation, owing to septations, requiring extension of the laminectomy to ensure complete fenestration. Finally, Patient 21 had an intramedullary cyst within a cyst, which was successfully fenestrated with a myelotomy.

Symptoms of SACs likely develop as a result of pressure on the spinal cord or a spinal nerve root.25,30,51 Often the symptoms can be aggravated by upright posture.25,25,42,63,93 Patients most commonly present with pain, followed by sensory changes (often presenting as gait instability), urinary dysfunction, and/or weakness.24,5,17,24–26,29,33,38–40,43,45,46,45,53,56,62–64,66,67,73,75,77,79,82,83,87,88,92 The presentations in our series of pediatric patients follow the presentations for the population as a whole. Pain was present in 50% of patients, gait instability in 27%, spasticity in 14%, sensory loss in 14%, and bladder dysfunction in 5%.

The treatment of choice for symptomatic SACs is excision or fenestration. When the cyst is located anterior to the spinal cord, fenestration of the cyst is performed to prevent further enlargement. For extradural cysts, excision of the cyst and closure of the cyst/dural communication is performed.51,69,70,80 Effort is made to remove as much of the cyst wall possible; however, recurrence has been reported after total cyst wall removal.64 For refractory cases, insertion of a cystoperitoneal shunt may be required.13,36,37 A cystoperitoneal shunt was used in one of the patients in this series after multiple failed attempts at fenestration. For small extramedullary cysts that are incidental and asymptomatic, conservative management is the preferred option, ensuring that patients are provided with adequate follow-up.58 Although some authors have reported minimally invasive treatment strategies using endoscopic techniques or CT- or MRI-guided aspiration,28,65 resection and wide fenestration remain the most reliable ways to eliminate the mass effect from a SAC. Sacral cysts may be particularly challenging, and less consensus is available regarding the optimal treatment.77 The findings of the current study echo the reported difficulties of treating SACs of the lower spine. Of the 4 patients with lumbosacral cysts and 3 with purely sacral cysts in our series, only 2 (29%) experienced complete remission of symptoms.

Conclusions

Symptomatic SACs in pediatric patients are rare lesions. Patients often present with pain followed by lower extremity weakness and gait instability. Although SACs may be encountered at any spinal level, they are predominately found in the thoracic region, which may be a consequence of the narrower spinal canal in this region and the length of the thoracic cord. According to our series, pediatric patients have a higher percentage of intradural SACs, with half being ventral to the spinal cord. This subtype of arachnoid cyst frequently observed in children may be associated with underlying congenital CNS abnormalities resulting in altered CSF flow dynamics. Overall outcomes following surgical management are excellent, with 68% achieving complete remission of symptoms and 87% achieving improved or remission of symptoms.

Disclosure

The authors report no conflict of interest concerning the materials or 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: Bond, Zada, McComb, Krieger. Acquisition of data: Bond, Bowen. Analysis and interpretation of data: Bond, Zada, McComb, Krieger. Drafting the article: Bond. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Bond. Administrative/technical/material support: Zada, Bowen. McComb, Krieger. Study supervision: Zada, McComb, Krieger.

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