Sacral intraspinal meningocele in a patient presenting with abdominal pain

Case report

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Sacral meningoceles are typically asymptomatic. When they are symptomatic, patients commonly present with signs and symptoms of nerve root compression and back pain. The authors report the case of a 10-year-old girl with an intraspinal sacral meningocele who presented initially only with severe right lower quadrant pain. The patient underwent successful surgical treatment of the meningocele and experienced subsequent resolution of the abdominal pain.

This is the first reported case of an intraspinal sacral meningocele in which the only presenting symptom was abdominal pain and which was successfully treated with surgery. It is postulated that the sacral meningocele caused severe abdominal pain secondary to compression of the sacral parasympathetic fibers that pass through the sacral plexus on each side of the cord corresponding to the S-2 and S-3 levels. (DOI: 10.3171/PED-07/07/053)

KEY WORDS • abdominal pain • meningeal cyst • myelography • pediatric neurosurgery • sacral meningocele

Spinal meningeal cysts are uncommon, accounting for 1 to 3% of all spinal tumors.4,9,13 Spinal meningeal cysts occur most frequently in the thoracic spine (65%), followed by the lumbar and lumbosacral spine (13%), the thoracolumbar spine (12%), the sacral spine (6.6%), and the cervical spine (3.3%).3,13 Thoracic cysts typically occur in adolescents, and sacral cysts are more often found in adults. Sacral cysts are sometimes referred to as sacral meningoceles.

Symptoms referable to spinal cysts are dependent on the level of involvement, and the proximity of the cyst to the spinal cord and nerve roots. Sacral cysts are rarely symptomatic and are usually found while investigating complaints of low back or sacrococcygeal pain, sensory and motor disturbance in the lower extremities, and bowel or urinary dysfunction.7,17 We present the case of a patient with an intraspinal sacral meningocele who presented initially only with atypical abdominal pain, without signs and symptoms usually ascribed to nerve root compression in the lumbosacral spine.

Case Report

History and Presentation. This 10-year-old girl was referred to our neurosurgical office with complaints of severe right lower quadrant abdominal pain and an inability to sit. Sitting precipitated the right abdominal pain. Standing was fairly comfortable but caused subjective right-sided leg weakness requiring the use of a cane for ambulation. She otherwise had no radicular complaints. A week prior to her visit, she began to experience rectal pain while performing Valsalva maneuvers. There was no bowel or bladder incontinence. She had been unable to attend school for over 6 weeks due to her symptoms. She was hospitalized 1 month prior to her neurosurgical evaluation for a complete gastrointestinal and general surgical workup. The results of ultrasound and CT examinations of the abdomen and pelvis were unremarkable except for a suggestion of a sacral cyst. A MR imaging study of the lumbar spine with and without gadolinium enhancement revealed an intraspinal cyst in the lower sacral area (Fig. 1). The cyst was relatively large and filled the lower canal at the S1–2 area. The cyst was isointense with CSF, suggesting a spinal meningocele. The patient was treated with narcotic agents and discharged with a prescription for continued medication.

Examination. During the physical examination, sitting was noted to cause the patient moderate distress due to pain in the right lower quadrant. The results of lower back palpation were unremarkable. There was tenderness to palpation in the right lower quadrant of the abdomen. She was slightly weaker on the right side and ambulated antalgical-

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computed tomography; MR = magnetic resonance.
ly, favoring the right side and using a cane. However, no appre-
ciable weakness was identified on objective motor ex-
am. The results of a sensory examination were unremark-
able, and the patient had normal perineal sensation. A rectal
examination was deferred.

A lumbar myelogram and postmyelogram CT study (CT
myelography, Fig. 2) showed no communication between
the sacral cyst and the thecal sac. Since all abdominal caus-
es of the patient’s symptoms had been ruled out, it was rea-
sonable to postulate that the cyst was compressing the lower
sacral nerves, thereby creating abdominal visceral pain via
the parasympathetic nerves.

Operation and Postoperative Course. The patient under-
went a sacral laminectomy with closure of the ostium and
subarachnoid space, which obliterated the meningeal cyst. Intraoperatively, the cyst was found not to include any nerve
fibers, and a communication was observed between the sub-
arachnoid space and the cyst that was not demonstrated by

Discussion

Spinal meningeal cysts are actually diverticula of the spi-
nal meningeal sac, nerve root sheath, or arachnoid. Nomen-
clature has been confusing, and a useful classification has
been described by Nabors et al.10 On the basis of imaging,
operative, and histological examination, the cysts can be
classified into three major categories as follows: Type I, ex-
tradural meningeal cysts without compression of the spinal
nerve root fiber; Type II, extradural meningeal cysts with
compression of the spinal nerve fibers (Tarlov perineural
cysts or spinal nerve root diverticula); and Type III, intra-
dural meningeal cysts (intradural arachnoid cysts).16 Extra-
dural cysts can be further divided into Type IA, extradural
meningeal/arachnoid cysts, and Type IB, (occult) sacral

FIG. 1. Sagittal T1-weighted (A) and T2-weighted (B) and axial
T2-weighted (C) MR images showing the sacral cyst with CSF in-
tensity.

FIG. 2. Computed tomography myelographic images. Upper:
Axial image showing nonopacification of the sacral cyst. Lower:
Sagittal reconstruction showing noncommunication of the sacral
cyst.

CT myelography. There was complete postoperative resolu-
tion of her pain syndrome. After a short convalescence, the
patient was able resume her normal activities including at-
tending school.
Sacral intraspinal meningocele

Meningoceles (Table 1). The radiographic and intraoperative findings in our case were consistent with an extradural cyst, specifically a Type IB, (occult) sacral meningocele cyst.

Type IA cysts arise above the sacrum, and their pedicles are usually adjacent to the entrance of the dorsal nerve root. Type IB sacral meningoceles contain a pedicle at the caudal tip of the dural sac adjacent to dorsal sacral or coccygeal nerve roots. These lesions have been sometimes identified as extradural arachnoid cysts. In some cases, the pedicle is quite broad with a large communication; the thecal sac does not taper and instead broadens within the sacral canal. This type of cyst is sometimes called an “occult” meningocele, but Nabors et al. simplified the sometimes confusing nomenclature by designating all sacral meningoceles that are extradural without nerve root fibers as sacral meningoceles.

Type II cysts or Tarlov perineural cysts arise from nerve root sleeves rather than from the dural sac. These cysts are synonymous with cystic diverticula or ectasias. Tarlov cysts are often multiple and bilateral.

Type III cysts or intradural cysts usually occur anywhere along the posterior spinal subarachnoid space. Like Tarlov cysts, they are often multiple and asymptomatic.

In all spinal cysts, the mass effect and CSF pulsations can erode the sacrum both anteriorly and posteriorly. Osseous erosion implies the presence of a valve mechanism causing intermittent surges of CSF pressure. In some cases, an enlarged caudal sac can balloon into the pelvis as an anterior sacral meningocele.

Spinal cysts are an uncommon cause of spinal cord or nerve root compression. When they are symptomatic, a patient may present with weakness, sensory loss, incontinence, or myelopathy, depending on the level of compression. Sacral cysts may present with radicular symptoms with bowel or bladder dysfunction. Our patient was unusual in that she did not present with any of these symptoms. We postulate that the mechanism for this patient’s unusual presentation was compression of the S-2 and S-3 nerve roots, which eventually contribute to the parasympathetic fibers that pass through the sacral plexus. The spinal visceral afferents transmit conscious sensation (gut distention, cardiac ischemia) and unconscious visceral sensations (blood pressure, chemical composition of the blood).

They are carried by the thoracolumbar sympathetic and sacral parasympathetic nerve trunks, receive convergent visceral and somatic inputs, and are the substrate for referred pain (visceral pain referred to overlying or nearby somatic structures). These fibers distribute to the descending colon, rectum, urinary bladder, and lower portions of the ureters, explaining our patient’s abdominal pain and, later, the rectal pain. Valsalva maneuvers and postural changes such as sitting can either directly or indirectly increase the pressure effects of spinal meningeal cysts resulting in increased symptoms.

In general, the various cystic spinal masses are not distinguishable by their imaging characteristics alone. A spinal meningeal cyst is best demonstrated on MR images. The cysts may be isointense with CSF, although Type I cysts may be more proteinaceous because of a lack of a broad connection with the subarachnoid space. As a result, the signal intensity may be higher than normal CSF due to elevated protein content and lack of pulsation-related signal loss.

Myelography and/or CT myelography may be helpful adjunctive modalities in revealing the connection between the cyst and the subarachnoid space. In this case report, the results of the MR imaging study were sufficient to make a diagnosis of a meningocele cyst. The CT myelography was not absolutely necessary and obviously did not influence the treatment plan. Demonstration of a communication allows for definitive diagnosis of spinal meningeal cyst; however, some cysts may lack a broad communication with the subarachnoid space. Nabors et al. demonstrated in their series that CT myelography may not always demonstrate a communication between the cyst and the subarachnoid space and should not preclude the diagnosis of a meningeal cyst.

In this particular case, the MR imaging cross-sectional studies demonstrated an intrasacral cyst measuring 3.6 cm in width, located 1.6 cm anteroposteriorly, and 3 cm in length with mild associated scalloping and broadening of the sacral canal (indicating that the lesion was longstanding). The signal was similar to that of CSF on all MR imaging pulse sequences. The cyst did not opacify with contrast from a myelographic intrathecal injection at the L2–3 level. Its central location suggests a sacral cyst of the meningocele variety.

Histologically, sacral meningoceles do not contain nerve fibers and ganglion cells in the cyst wall, whereas perineural (Tarlov) cysts do. Both congenital and developmental origins have been proposed for each type of spinal meningeal cyst.

Because sacral cysts are frequently asymptomatic, surgical intervention is indicated only if there are clinical manifestations. In certain instances it may be difficult to definitively determine if a cyst is symptomatic or not. The use of percutaneous fine-needle cyst drainage and lumbar drains to lower the hydrostatic and pulsatile CSF pressures within the meningocele have been used to help determine the value of surgery. If there is symptomatic relief, placement of a lumbo-peritoneal shunt may be considered as a treatment option. We believe, however, that the best treatment is surgical closure of the ostium or pedicle.

Most cases of sacral cysts reported in the literature involve Type II Tarlov cysts, but this may be due to former difficulties in categorizing spinal cysts. There have been relatively few case reports in which a sacral cyst was felt to have been the etiology of abdominal pain.

Slipman and colleagues described a large Tarlov cyst that eroded through the sacrum and extended into the retroperitoneum. The patient chose not to undergo surgical intervention so it is unknown whether treatment of the cyst would have indeed effected a cure.

The authors postulated that the abdominal pain was secondary to direct irritation of the peritoneum rather than sa-

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<th>Type</th>
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<tr>
<td>I</td>
<td>extradural meningeal cyst w/out compression of spinal nerve root fibers</td>
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<tr>
<td>IA</td>
<td>extradural meningeal/arachnoid cyst</td>
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<tr>
<td>IB</td>
<td>(occult) sacral meningocele</td>
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<tr>
<td>II</td>
<td>extradural meningeal cyst w/ compression of spinal nerve root fibers (Tarlov perineural cyst, spinal nerve root diverticulum)</td>
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<td>III</td>
<td>spinal intradural meningeal cyst (intradural arachnoid cyst)</td>
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eral root compression, which is what we postulate in the current case report.

In the series of Nabors et al., a patient with a sacral meningocele diagnosed by MR imaging presented with low back pain and abdominal pain. This Type I spinal meningeal cyst arose from the midline and extended from S-1 to S-3. The same series also included a boy who presented with similar symptoms but was diagnosed with a Tarlov cyst involving bilateral S-2 nerve roots. Neither of these patients underwent surgery.

Conclusions

Although sacral meningoceles are usually asymptomatic, patients with these lesions may present with symptoms of nerve root compression, usually manifesting as perineal or lower extremity signs and symptoms. We have described a case in which a patient with sacral nerve root compression due to a sacral meningocele presented with isolated right lower quadrant pain, mimicking a gastrointestinal disorder.

References
