Expanding occult intrasacral meningocele associated with diastematomyelia and multiple vertebral anomalies

Case report

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The authors report the case of a 7-year-old girl who presented with persistent low-back and left leg pain and was diagnosed with expanding occult intrasacral meningocele (OIM), diastematomyelia, tethered cord, and multiple vertebral anomalies. She was followed for 27 months. Removal of a bone spur and sectioning of the tight terminal filum did not relieve her left leg and back pain. Serial magnetic resonance (MR) imaging after initial detection of the OIM revealed gradual but significant enlargement of the lesion. Fifteen months after terminal filum release and bone spur resection, the OIM was excised. No case of OIM reported to date contains documented serial MR imaging confirmation of enlargement of OIM. Furthermore, this is the first report of OIM associated with diastematomyelia. Because most OIMs expand, the authors recommend that, in addition to surgery for associated congenital anomalies, the OIM be totally excised to resolve symptoms completely.

Key words • meningocele • sacrum • tethered cord • pediatric neurosurgery

OCCULT intrasacral meningocele was first described by Enderle in 1932.1 The walls of this rare cystic lesion are composed of fibrous tissue, and the cavity is lined with arachnoid membrane. These meningoceles contain no neural elements, and the spinal canal is always enlarged at the site of occurrence.5,7,11 In most cases the OIM is attached to the distal end of the dural sac by a narrow pedicle, and CSF usually flows freely between the cyst and the dural sac.

In this report, we describe a case in which OIM was associated with diastematomyelia, tethered cord, and multiple vertebral anomalies. The follow-up period was 27 months, during which time cyst enlargement was demonstrated on serial lumbosacral MR imaging. To the best of our knowledge, this is the first reported case of OIM to be associated with diastematomyelia and MR imaging–confirmed enlargement of an intrasacral meningocele.

Case Report

Presentation. This 7-year-old girl presented to our center in April 1998 with the complaint of left leg, left buttock, and lower-back pain. Spinal column deformity and foot asymmetry were also present.

Examination. Neurological and physical examinations showed hypalgesia below L-2, rotoscoliosis, left pes planus, and a smaller left foot than right. Careful inspection revealed no somatic stigmata in the lumbosacral area. Plain whole-spine radiography revealed an expanded spinal canal in the lumbosacral region, rotoscoliosis of the thoracic spine, and multiple thoracic vertebral anomalies. The latter included hemivertebra, butterfly vertebra, block vertebra, and sagittal clefts. Multiple rib anomalies were also noted on these x-ray films. The spinous processes and laminae of the vertebrae below T-10 were incompletely developed.

Lumbar MR imaging during the initial workup in May 1998 demonstrated expansion of the spinal canal, in the sacral region, and an intrasacral cyst. The cyst was filled with fluid that was isointense to CSF, and its cranio-caudal length was 31 mm (Fig. 1 left). Thoracic MR imaging demonstrated duplication of the spinal cord from T-10 to L-3 due to a bone spur at L-2. The conus medullaris was low lying and terminated at L-4. The initial diagnoses were diastematomyelia, tethered cord, and intrasacral meningocele with multiple spinal column anomalies.

Operation. We recommended sectioning of the terminal filum and removal of the bone spur, but the patient’s family refused this operation at first. One year later, in May 1999, they decided to proceed with surgical intervention. During the procedure, we removed the bone spur and sectioned a portion of the tight terminal filum but did not explore the intrasacral cyst. The postoperative period was

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computerized tomography; MR = magnetic resonance; OIM = occult intrasacral meningocele.
uneventful, but the patient’s low-back and left leg pain remained unchanged. Early postoperative control lumbar MR imaging revealed the expected postoperative changes, and measurements indicated that the intrasacral cyst had enlarged to 38 mm in length (Fig. 1 center). Because the patient’s pain was unaltered by surgery, we suspected that the intrasacral lesion was the cause; however, the family refused a second operation.

Postoperative Course. At 15 months postoperatively, repeated MR imaging of the lumbar spine revealed that the OIM had increased in length to 42 mm, indicating an 11-mm expansion since its detection (Fig. 1 right). The patient continued to suffer the same pain, and we believed that steady enlargement of the cystic lesion was a strong indication for excision. Fortunately, we were able to convince the family that a second surgery was essential.

Second Operation. A sacral laminectomy was performed to expose a thin-walled cyst filled with fluid under tension. The lesion displaced the thecal sac laterally. Once the cyst wall was opened, a fistulous tract was observed between the thecal sac and the lesion. Gross inspection showed no neural elements or nerve roots within the cyst. We ligated the tract between the cyst and thecal sac and removed the entire lesion.

Second Postoperative Course. Analysis of the cyst fluid identified it as CSF, and histopathological examination of the wall of the lesion showed fibrous tissue containing no neural elements. The patient’s back and leg pain disappeared immediately after the second operation, and she was referred to a pediatric orthopedic surgeon for management of her scoliosis.

Discussion

The literature contains three types of sacral cysts: anterior sacral meningoceles, perineural cysts, and OIMs. Occult intrasacral meningocele is a rare lesion that must be differentiated from other sacral cystic lesions. Fewer than 90 cases of OIM have been documented in the literature. Our patient was symptomatic at a very young age. Furthermore, OIM in her case was unique because of its associated diastematomyelia with multiple spinal column anomalies and its MR imaging–confirmed enlargement of the intrasacral cyst during a 2-year period.

In addition to the fact that OIMs are uncommon, their origin and pathogenesis are unclear. Their association with other congenital spinal anomalies, such as spinal lipoma, sacral hyperkeratosis, sacral hyperpigmentation, sacrococcygeal dimples, neurofibromatosis, and tethered cord syndrome, indicates a congenital origin. Various theories of their pathogenesis have been proposed; they include the following.

Dysraphic Theory

In most cases of OIM, the lesion has been attributed to failure of neural tube closure or failed closure of the surrounding mesoderm because of developmental arrest during various stages of embryogenesis. Reports in which
authors have noted large open connections between the cyst and the dura mater, as well as cases with associated spina bifida, support this theory.4,12,17

**Acquired Dural Disruption Theory**

Proponents of this theory contend that the OIM forms as a result of a dural defect caused by direct traumatic injury or operative intervention, with resultant herniation of the arachnoid through the defect.4,11,12

**Proliferative Theory**

Based on analysis of the developmental process in myeloschisis, Patten19 suggested that the open neural tube in OIM may be the result of local overgrowth that interferes with closure, rather than a consequence of developmental arrest.

**Failure of Meningeal Sac Ascension**

In this hypothesis it is believed that an OIM develops when the meningeal sac fails to ascend from its embryonic coccygeal position to its normal adult position at S-2.7

**General Comments**

Except for the theory of acquired dural disruption, the aforementioned hypotheses suggest that pathogenesis involves failure of normal spinal development and its neural elements.4 Although three of the four theories propose a congenital origin, the diagnosis in most reported cases of OIM has been established during adulthood (Table 1). The more typical late onset of symptoms reflects the nature of these lesions, because gradual enlargement of the OIM compresses the sacral nerve roots more intensely over time.

As with many other conditions, more widespread use of MR imaging for evaluation of low-back pain has resulted in higher diagnostic rates of OIM in the pediatric population. Increasingly, asymptomatic cases are being diagnosed incidentally during MR imaging sessions in patients of all ages. The signs of OIM, however, are sometimes overlooked or mistakenly linked to other conditions. In a literature review, we found five adult cases in which the diagnosis was established late even though the patients had exhibited symptoms since adolescence (Cases 2–5 and 9 in Table 1). In these instances, the problems were initially attributed to other pathological entities.

The walls of OIM lesions are composed of collagenous connective tissue without neural components. Most of these cysts communicate with the subarachnoid space and are filled with CSF. Intraoperative confirmation of the absence of neural elements is important for distinguishing between an intrasacral meningocele and a perineural cyst.15 Magnetic resonance imaging is noninvasive and is the diagnostic modality of choice for investigating sacral cysts. If the meningocele contains CSF, MR imaging will

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**TABLE 1**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author(s) &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Method of Diagnosis</th>
<th>Clinical Findings</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Wilson &amp; Walkey, 1932</td>
<td>16, M</td>
<td>plain radiography</td>
<td>lumbalgia, sciatica</td>
<td>complete resection</td>
<td>symptomatic relief</td>
</tr>
<tr>
<td>2*</td>
<td>Baker &amp; Webb, 1952</td>
<td>28, F (10 yrs’ duration)</td>
<td>plain radiography</td>
<td>sciatic pain &amp; stiffness</td>
<td>complete resection</td>
<td>symptomatic relief</td>
</tr>
<tr>
<td>3*</td>
<td>Abbott, et al., 1957</td>
<td>28, F (12 yrs’ duration)</td>
<td>plain radiography, myelography</td>
<td>urinary &amp; rectal incontinence, weakness of lower extremities</td>
<td>complete resection</td>
<td>symptomatic relief</td>
</tr>
<tr>
<td>4*</td>
<td>Palazzoli, 1963</td>
<td>21, F (5 yrs’ duration)</td>
<td>plain radiography, myelography</td>
<td>low-back &amp; sciatic pain</td>
<td>complete resection</td>
<td>symptomatic relief</td>
</tr>
<tr>
<td>5*</td>
<td>Kak, et al., 1972</td>
<td>30, M (15 yrs’ duration)</td>
<td>plain radiography, myelography</td>
<td>urinary incontinence</td>
<td>complete resection</td>
<td>symptomatic relief</td>
</tr>
<tr>
<td>6</td>
<td>Florez &amp; Ucar, 1976</td>
<td>7, F</td>
<td>plain radiography, myelography</td>
<td>low-back pain, paresthesia, nocturnal enuresis, neurofibromatosis Type 1, sacral neurofibromas</td>
<td>complete resection</td>
<td>pain relieved, persistent difficulty micturating</td>
</tr>
<tr>
<td>7</td>
<td>Atabay, et al., 1994</td>
<td>12, M</td>
<td>plain radiography, MRI</td>
<td>urinary &amp; rectal incontinence, weakness of lower extremities, foot deformities</td>
<td>complete resection</td>
<td>asymptomatic except minimal sensory deficit</td>
</tr>
<tr>
<td>8</td>
<td>Okada, et al., 1996</td>
<td>13, F</td>
<td>plain radiography, myelography, CT, CT myelography, MRI</td>
<td>numbness of lower extremity, decreased pinprick sensation</td>
<td>complete resection</td>
<td>muscle wasting persisted</td>
</tr>
<tr>
<td>9*</td>
<td>Mishra, et al., 2000</td>
<td>19, M (2 yrs’ duration)</td>
<td>plain radiography, MRI</td>
<td>low-back pain, leg atrophy</td>
<td>complete resection</td>
<td>muscle wasting persisted</td>
</tr>
<tr>
<td>10</td>
<td>Franco, et al., 2001</td>
<td>18, F</td>
<td>plain radiography, MRI</td>
<td>urinary incontinence</td>
<td>complete resection</td>
<td>symptomatic relief</td>
</tr>
<tr>
<td>11</td>
<td>present case</td>
<td>7, F</td>
<td>plain radiography, MRI</td>
<td>low-back &amp; leg pain, diastematomyelia, tethered cord</td>
<td>complete resection</td>
<td>symptomatic relief</td>
</tr>
</tbody>
</table>

* Cases symptomatic since childhood but diagnosed and treated in adulthood.
demonstrate characteristic CSF intensity within the cyst cavity; however, surgical findings and histopathological evaluation are essential for definitive diagnosis of OIM.

In our case, we initially attributed the young girl’s left leg and back pain to her tight terminal filum. The first operation, which involved sectioning of the tight terminal filum and removal of the septum, had no effect on the symptoms. The fact that the craniocaudal length of the intrasacral cyst increased by 11 mm during a 27-month period indicated to us that the OIM might be the primary cause of the persistent left leg and back pain. Indeed, our patient’s symptoms resolved immediately after the OIM was totally excised.

The reports to date indicate that OIMs usually enlarge and that pain increases as the cyst grows and compresses the nerve fibers. Because of this, we believe that any patient in whom this condition is diagnosed should undergo excision of the lesion. In most cases in the literature, total resection of the OIM resulted in complete resolution of lower-extremity pain and back pain. Other authors have described “slow enlargement” of these cysts, but none has offered serial CT or MR imaging confirmation of lesion enlargement. Ours is the first case of OIM in which this type of consecutive imaging-based investigation has clearly documented the enlargement of such a lesion.

The literature contains two asymptomatic cases in which the OIM was incidentally detected. One involved an elderly woman who harbored an OIM associated with tethered cord syndrome complex, and the other involved a 41-year-old woman with bilateral anterior sacral and intrasacral meningeal and perineural cysts. These intrasacral meningeoceles have also been described in patients with Marfan syndrome. Our patient harbored OIM associated with diastematomyelia and multiple vertebral anomalies. According to Grivegnee, et al., OIM is rarely associated with spinal column anomalies. Occult intrasacral meningocele in conjunction with diastematomyelia has not been reported.

In conclusion, OIMs are rare cystic lesions of the sacral spine that tend to increase in size and cause pain over time. In a small number of patients with OIM spinal abnormalities are also present, but the clinician should been cautioned about immediately attributing all pain to coexisting lesions. Many OIMs expand and cause symptoms. We recommend that all such cysts be excised to ensure complete neurological recovery and symptomatic relief. Magnetic resonance imaging is the tool of choice for investigating sacral cysts, but definitive diagnosis of OIM requires histopathological examination of a surgical specimen.

References
