Spinal pseudomeningoceles and cerebrospinal fluid fistulas

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Spinal pseudomeningoceles and cerebrospinal fluid (CSF) fistulas are uncommon extradural collections of CSF that may result from inadvertent tears in the dural–arachnoid layer, traumatic injury, or may be congenital in origin. Most pseudomeningoceles are iatrogenic and occur in the posterior lumbar region following surgery. The true incidence of iatrogenic pseudomeningoceles following laminectomy or discectomy is unknown; however, the authors of several published reports suggest that the incidence of lumbar pseudomeningoceles following laminectomy or discectomy is between 0.07% and 2%. Pseudomeningoceles are often asymptomatic, but patients may present with recurrence of low-back pain, radiculopathy, subcutaneous swelling, or with symptoms of intracranial hypotension. Very rarely, they present with delayed myelopathy. Although magnetic resonance imaging is the neurodiagnostic study of choice, computerized tomography myelography and radionuclide myelographic study may be helpful diagnostic tools in some cases. Analysis of suspect fluid for β2 transferrin may be a useful adjunctive study. Treatment options include close observation for spontaneous resolution, conservative measures such as bed rest and application of an epidural blood patch, lumbar subarachnoid drainage, and definitive surgical repair.

Key Words • spinal cord • pseudomeningocele • cerebrospinal fluid fistula • magnetic resonance imaging

Epidemiological Characteristics

The causes of pseudomeningoceles may be classified into three categories: iatrogenic, traumatic, and congenital. By far the most common cause is iatrogenic, resulting from inadvertent dural tears during spinal surgery.

Iatrogenic Causes

The true incidence of iatrogenic lumbar pseudomeningoceles is difficult to ascertain because many remain asymptomatic. The reported incidence of unintended durotomy is anywhere from 0.3 to 13% and most frequently occurs as a result of lumbar laminectomy. The higher prevalence of pseudomeningoceles in the lumbar region has been attributed to both the large number of lumbar laminectomies performed in this region and the increased intraspinal CSF pressure in the caudal thecal sac, compared with the cervical and thoracic spine.

In 1947, Swanson and Fincher reported a postoperative incidence of pseudomeningocele of 0.07% in a series in which 1700 lumbar laminectomies had been performed. Mayfield retrospectively reviewed 1408 cases of lumbar laminectomy undertaken between 1971 and 1975 and found a 0.3% incidence of CSF fistula requiring reoperation and an 0.8% incidence of pseudomeningocele in symptomatic patients. Prior to the development of CT scanning and MR imaging, myelography was most commonly used to diagnose pseudomeningoceles and CSF fistulas; thus, the true incidence may have been underestimated. In 1983, Teplick, et al., reported a 2% incidence
syndrome and neurofibromatosis. Pyritz concluded that the prevalence of dural ectasia in patients with Marfan syndrome was as high as 67% but that most are asymptomatic. Dolynechuk, et al., have noted that sacral protrusion of the thoracic meninges has been associated with neurofibromatosis in 64% of 98 cases reported since this phenomenon was first described by Pohl in 1933. The incidence of postsurgical pseudomeningocele, compared with dural ectasia, in either of these syndromes is thought to be much lower. Schneider, et al., have reported a patient with Marfan syndrome who presented with an anterior sacral meningocele. Although postoperative anterior pseudomeningocele has been reported, anterior pseudomeningocele is usually congenital in origin and is associated with defects in the vertebral bodies.

PATHOPHYSIOLOGICAL CHARACTERISTICS

Iatrogenic pseudomeningoceles may or may not be associated with an arachnoid tear, but a dural tear is necessary for one to form. If the arachnoid is not violated, the arachnoid membrane may subsequently herniate through the dural defect and a CSF-filled arachnoid sac forms. Rinaldi and Peach found arachnoidal cells within the capsular membrane of pseudomeningoceles and advocated terming these lesions "true meningoceles." Others have contended that the CSF extravasates through a dural–arachnoid tear and into the surrounding soft tissues, eventually leading to an arachnoid-lined, fibrous capsule. The factors that contribute to a persistent communication between the subarachnoid and the extradural spaces have been debated. Teplick, et al., have suggested that when intact arachnoid herniates into the cyst, the communication is more likely to remain open and form a pseudomeningocele, whereas when an arachnoidal tear occurs, the likelihood of closure of the communication is greater. Tsui, et al., have argued that the volume of the leakage was more important; thus, if a CSF leak is small, the fluid is easily absorbed and the lesion is self-limited. Other authors, however, contend that a smaller communication leads to a higher probability of pseudomeningocele formation, because this acts as a ball valve mechanism, allowing a one-way flow of CSF. Cobb and Ehni have lent support to this theory, describing a pseudomeningocele connected to the subarachnoid space through a small channel. The trapped CSF then collects in the traumatized postoperative bed of the paraspinal musculature and an abnormal connective tissue reaction occurs that leads to poor CSF absorption.

Trauma-induced brachial plexus pseudomeningoceles form by a different mechanism. In brachial plexus nerve root avulsions, the arachnoid and dura that invest the nerve root are torn, and CSF leaks into the perineural soft tissues. A cavity is formed by proliferation and fusion of dura and epineurium. The cyst is then lined by reformed arachnoid. Retraction of severed nerve root by Wallerian degeneration and CSF pulsations may lead to enlargement of the cavity. Barberá, et al., reported on a case of trauma-induced lumbosacral nerve root pseudomeningocele and reviewed 16 other cases. They theorized that the junction between the radicular sheath proximal to the ganglionic root and the dural sac is a "locus minor resistentiae" where traction can...
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TABLE 1
Signs and symptoms associated with pseudomeningoceles and CSF fistulas

<table>
<thead>
<tr>
<th>Sign or Symptom</th>
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<tbody>
<tr>
<td>recurrence of low-back pain</td>
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<tr>
<td>recurrence of radicular pain</td>
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<tr>
<td>posture-related headache</td>
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<tr>
<td>cervical or occipital pain</td>
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<tr>
<td>nausea &amp; vomiting</td>
</tr>
<tr>
<td>photophobia</td>
</tr>
<tr>
<td>cranial nerve palsy</td>
</tr>
<tr>
<td>tinnitus</td>
</tr>
<tr>
<td>palpable mass</td>
</tr>
<tr>
<td>delayed myelopathy</td>
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<td>meningismus or other signs of meningitis</td>
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be centered. Weakness at this junction could explain why congenital cysts appear similar to traumatic lesions. Shahnifar and Slichter have maintained that sectorial defects of the dural sac may occur on a congenital or iatrogenic basis and probably play a role in the cause of cyst formation. As mentioned previously, an increased incidence of pseudomeningocele has been noted in patients with Marfan syndrome, and the cause has been attributed to pulsatile pressure of CSF on a dural membrane that is more elastic than normal.

**CLINICAL FEATURES**

Pseudomeningoceles and CSF fistulas may present with a variety of signs and symptoms (Table 1). Most pseudomeningoceles, however, remain asymptomatic. The time interval between the original surgery to the onset of symptoms may range from months to years. Some pseudomeningoceles may present as a fluctuant mass that enlarges when a patient coughs or sneezes (Fig. 1). Palpation in the region of the cyst may cause pain, as was seen in six of patients with pseudomeningoceles studied by Al-drete and Ghaly. Cutaneous CSF fistulas are often diagnosed by simple inspection of the wound. If there is a watery discharge that produces a clear halo surrounding a central pink stain, then the fluid is assumed to be CSF.

Patients commonly present with recurrence of preoperative symptoms, including periodic low-back and radicular pain aggravated by straining maneuvers. Radicular motor or sensory loss may be present. The signs and symptoms may materialize as a result of nerve roots that become adherent to the fistulous opening into the pseudomeningocele, or from the movement of the spine causing stretching of the anchored nerve root. Nerve roots may also herniate into the pseudomeningocele. Straining can cause wedging of the nerve root with subsequent development of sensorimotor radiculopathy. This concept is supported by the findings of several case reports. Hadani, et al. have reported three cases of nerve root entrapment following laminectomy. Wilkinson has reported a case of nerve root entrapment within a pseudomeningocele that the author attributed to the patient's delayed onset of radicular signs and symptoms. Conversely, Miller and Elder have reported a case of nerve root entrapment within a pseudomeningocele, but they believed that the patient's symptoms were unrelated to this finding. Although uncommon, one must be aware that congenital cysts can present with intermittent back and radicular pain and can be confused with other lesions such as herniated discs.

More rarely, a patient may present with progressive or delayed myelopathy following cervical spine surgery. Goodman and Gregorius have published a case report in which they describe a patient who presented with progressive myelopathy and was found intraoperatively to have spinal cord herniation. They observed the spinal cord periodically bulging into the cyst with each heartbeat and respiration. The authors concluded that myelopathy related to a cervical pseudomeningocele may result from spinal cord herniation (CSF pressure > cyst pressure) or focal cord compression (CSF pressure < cyst pressure). Depending on the location of the lesion, patients with myelopathy may present with a spinal cord syndrome. Horowitz, et al. have reported a patient who presented with a Brown–Séquard and Horner syndromes caused by the development of a cervicothoracic pseudomeningocele following an anterior cervical disectomy. Patients with pseudomeningoceles may also present with symptoms similar to those seen in those with spontaneous intracranial hypotension, including photophobia, cranial nerve palsies, and tinnitus. Very frequently, posture-related headaches, relieved in a recumbent position, may be present. Patients may complain of cervical or occipital pain with or without nausea and vomiting while in a standing position. Lastly, patients with unrecognized CSF fistulas may present with signs and symptoms of acute or chronic meningitis.

**DIAGNOSTIC STUDIES**

Diagnosis by history and physical examination can be augmented by several imaging modalities. Magnetic Resonance Imaging

Today, MR imaging is the diagnostic study of choice.
Prior vertebral elements has been documented. Although imaging can delineate the location, extent, and internal characteristics of the lesion and may demonstrate its level of communication with the thecal sac. It may also elucidate other associated pathological entities such as spinal cord compression or nerve root entrapment, as well as differentiate pseudomeningoceles from syringomyelia, arachnoiditis, or recurrent tumor. Findings noted in spontaneous intracranial hypotension, including intracranial meningeal enhancement, subdural fluid collections, and caudal displacement of the cerebellar tonsils may also be visualized.

Typically it reveals a region of low signal intensity on T1-weighted images and high signal intensity on T2-weighted images consistent with CSF (Fig. 2). Magnetic resonance imaging can delineate the location, extent, and internal characteristics of the lesion and may demonstrate its level of communication with the thecal sac. It may also elucidate other associated pathological entities such as spinal cord compression or nerve root entrapment, as well as differentiate pseudomeningoceles from syringomyelia, arachnoiditis, or recurrent tumor. Findings noted in spontaneous intracranial hypotension, including intracranial meningeal enhancement, subdural fluid collections, and caudal displacement of the cerebellar tonsils may also be visualized.

**Computerized Tomography Myelography**

Myelography combined with CT scanning may better visualize the location of the pseudomeningocele or fistulous communication relative to a surgical site because of the superior bone imaging quality of CT scanning compared with MR imaging (Fig. 3). Calcified pseudomeningoceles have been reported, and erosion into posterior vertebral elements has been documented. Although Barron reported a case study of pseudomeningocele with an enhancing capsule, typically there is no contrast enhancement. Delayed CT myelography, in which the scan itself follows intrathecal contrast administration by several hours, may detect a slow-filling pseudomeningocele. Depending on the location of the pseudomeningocele, the positioning of the patient during the study may need to be modified. In general, however, a supine position allows for adequate delineation of the lesion, because most pseudomeningoceles are posteriorly located.

**Retrograde Radionuclide Myelography**

Retrograde radionuclide myelography has been used to detect chronic dural CSF fistulas. Chronic CSF leaks, as opposed to pseudomeningoceles, may be particularly difficult to detect due to slow leakage and minimal CSF collection. Retrograde radionuclide myelography has been used to detect slow, intermittent leaks after lumbar puncture, CSF leaks after spinal surgery and penetrating traumatic injury, and pleural CSF fistulas.

**Myelographic Study**

Trauma-induced nerve root pseudomeningoceles usually appear on myelography as unilateral lesions that vary in size and have irregular contours. In the cervical region, multiple lesions from C-3 to T-2 are commonly observed. They readily fill with contrast medium unless communication with the thecal sac has ceased. Frequently, flattening of the lateral margin of the thecal sac, absence of the nerve sheath, and pulsatile leakage of contrast into the extraarachnoidal sacs are visualized. Congenital cysts, as opposed to trauma-induced lesions, tend to be bilateral, small, regular in contour, and often have configurations in the sacral region that appear as clusters of grapes.

**Beta 2-transferrin**

Determining the presence of CSF by measuring the glucose level is an unreliable method. However, analysis of fluid for β2 transferrin is highly sensitive in detecting CSF, because, with rare exception, β2 transferrin is found only in the CSF. A small sample size of fluid (<1 ml) is required to detect β2 transferrin. Proteins in the fluid are separated by polyacrylamide gel electrophoresis and then transferred electrophoretically onto a nitrocellulose sheet. Using an antibody reaction (immunoblot), banding patterns are analyzed.

The β2 isoform arises from β1-transferrin through the loss of sialic acid by the action of neuraminidase. Because neuraminidase is only found in the central nervous system, CSF fluid will have two bands, one representing β1 and the other representing β2. Serum and other body fluids normally have only one band, represented by β1.

False positive results can be caused by alteration in the polypeptide sequence or the carbohydrate moiety of transferrin as a result of genetic variation or other causes. This alters the electrophoretic mobility of the molecule and

Fig. 2. Sagittal T1 (left) and T2-weighted MR images obtained of the lumbar spine (same patient as in Fig. 1) demonstrating a large dorsal fluid collection contiguous with the subarachnoid space. Intraoperatively, a pseudomeningocele originating from a small suture hole was observed.

Fig. 3. Computerized tomography myelography demonstrating an anterior cervical pseudomeningocele. This male patient underwent a C-5 corpectomy and anterior cervical arthrodesis in which instrumentation was placed for a C-5 burst fracture and dislocation. Two weeks later he developed a fluctuant mass on the left side of his neck, and axial CT myelography of the cervical spine revealed a large anterior pseudomeningocele. The patient was treated with 7 days of closed lumbar drainage and the lesion resolved.
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results in mistaking these variants for the CSF isoform. The most common allelic variant of the transferrin gene is TfC. Patients that are homozygous for TfC will have a single band of β1. The TIB and TID allele variants result in proteins with slower and faster mobilities, respectively. Heterozygosity for this allele would then lead to a banding doublet. Thus, non-CSF could be confused with CSF. False interpretation can be avoided by simultaneous comparison of CSF fluid and other body fluid obtained in the same patient.88

In individuals who are chronic alcoholics there may be elevated serum concentrations of a partially desialylated transferrin isoform, and an additional intermediate electrophoretic band may appear between β1 and β2.88 Increased concentration of the asialo transferrin isoform has also been reported in patients with cirrhosis, neuropsychiatric disorders, rectal cancer, and a rare disorder of glycoprotein metabolism.88 Careful attention to the clinical information and comparison of patterns of CSF, serum, and other fluids will allow accurate interpretation of unusual transferrin bands.

TREATMENT

Close evaluation of preoperative neuroimaging studies, meticulous surgical technique, and liberal use of microscopic magnification will often avert iatrogenic pseudomeningocele and CSF fistula. All available preoperative neurodiagnostic images should be evaluated for evidence of bone defects caused by possible occult spina bifida or previous surgeries. Each Kerrison rongeur bite should be performed in an ergonomic position and repetitive cisternal punctures for 2 to 3 days.5

Closed Lumbar Subarachnoid Drainage

The first investigators who treated pseudomeningoceles with closed lumbar subarachnoid catheters used teflon or polyethylene catheters.25,26,41,95 These catheters, initially designed for epidural use, were complicated by frequent blockage and kinking and were therefore somewhat difficult to use.87 In 1992, Shapiro and Scully87 reported the use of silicone lumbar subarachnoid catheters in 39 patients with spinal CSF fistulas and pseudomeningoceles. They reported a 92% success rate after 7 days of drainage alone. Complications included a 24% incidence of temporary nerve root irritation that resolved after the drain was removed and a 63% incidence of transient headaches, nausea, and vomiting. In an earlier report Kitchel, et al.,41 noted a similar success rate of 90% in 17 patients treated with 4 days of drainage, but the recurrence rate was higher (18%) when compared with that reported by Shapiro and Scully (8%). The complication rate reported by Kitchel, et al., was similar to that reported by Shapiro and Scully, with a 58% incidence of headaches, nausea, and vomiting. All patients were successfully treated with adjustment in the rate of CSF drainage, intravenous hydration, and analgesic medication. In each study the authors reported one case of meningitis associated with lumbar subarachnoid drainage and both cases were successfully treated with antibiotic therapy.

McCormack, et al.,58 reported using an epidural blood patch combined with a brief course of subarachnoid drainage in one patient with lumbar spinal implants. They speculated that CSF diversion alone in patients with spinal implants may not eliminate the pseudomeningocele because the hardware prevents reapproximation of paraspinal tissues. The blood patch procedure obliterates extradural dead space and provides a substrate for clot formation. Dural healing may thus be optimized because CSF diversion and percutaneous blood patch are complementary in decreasing the CSF pressure differences across the dural breek.
Surgical Treatment

The definitive treatment for CSF fistulas and pseudomeningoceles is reoperation for dural repair. Indications for surgery include failure of conservative measures or progressive radicular or signs and symptoms of myelopathy. For those with a neurological deficit, a delay in surgery may put the patient at risk for further neurological injury.22 The surgery should begin with adequate lighting, and the skin incision should be generous enough to encompass the area of the leak. Once the pseudomeningocele is visualized, it may be followed deep into the durotomy site. Often the pseudomeningocele may need to be resected to identify the region of interest. The durotomy site is protected with a cottonoid, and any necessary bone resection is performed prior to attempting dural closure to provide adequate room for suturing. Under microscopic magnification, the durotomy site is explored to ensure that nerve root or spinal cord strangulation is not present.

In most cases, the dura may be closed primarily, without need of a graft, by using No. 4-0 to 7-0 nonabsorbable sutures on a taper or reverse cutting, half-circle needle. For a large defect, a fascial graft obtained locally or artificial dura may be used to avoid compression of neural elements.25 For durotomies that occur in surgically inaccessible areas, such as far-lateral durotomies, a small plug of muscle or fat may be introduced through an intentional medial durotomy and pulled into the defect66 (Fig. 4).

If a Valsalva maneuver reveals a persistent leak, a fibrin sealant should be placed over the area of the leak. Analysis of the results obtained in animal studies suggests that fibrin sealant alone can withstand high hydrostatic pressures.8,21,72 However, since it remains in situ for only 5 to 7 days, fibrin glue must be supplemented with a dural, muscle, or fat graft placed over the area of the persistent leak. Simply placing Gelfoam or muscle over the durotomy site without also applying fibrin glue is ineffective and has been associated with failure to resolve the leak.59,61,76,86

Paraspinal muscle and overlying fascia should be closed in at least two layers by using No. 0-gauge monofilament with sutures placed 3 to 4 mm apart. Surgically placed drains may lead to the persistence of communication between the intra- and extradural space and may serve as a nidus for infection. Therefore, these drains should not be routinely used in the repair of a dural tear or a pseudomeningocele. Postoperatively, patients in whom lumbar defects have been repaired should stay in bed for at least 2 days, although some authors advocate longer periods of bed rest.18 We routinely provide a corset brace to those patients in whom a water-tight closure was not attained to minimize the pressure difference across the defect.

Some authors argue that all cervical spine pseudomeningoceles should be treated surgically,53 whereas others argue that pseudomeningoceles will resolve with time. Horowitz, et al.,55 reported a case of an anterior cervical pseudomeningocele that resolved after 3 weeks of expectant observation. In contrast to lumbar defects, patients with repaired cervical defects may ameliorate immediately.

The role of an LP shunt in the management of pseudomeningocele and CSF fistulas is not well defined. There are reports of successful management of CSF fistulas27,35,40 and a pseudomeningocele42 with an LP shunt. However, the placement of permanent hardware should be avoided whenever possible and other therapies, including surgical repair, should first be attempted. An LP shunt, in general, should be applied only in those in whom surgical repair failed or in whom surgical repair was not possible due to the location of the defect.

CONCLUSIONS

Pseudomeningoceles and CSF fistulas are extradural CSF collections uncommonly encountered by the spine surgeon. The great majority are the result of iatrogenic durotomy, usually following lumbar surgery. Many of the lesions remain asymptomatic, but some may present with a range of signs and symptoms including posture-related headaches, localized back pain, radiculopathy, and myelopathy. Although MR imaging is the neurodiagnostic modality of choice, CT myelography and radionuclide myelography may provide useful information in difficult cases. A water-tight closure of the dural defect encountered during surgery is the key to avoiding this complication.

When a pseudomeningocele or CSF fistula is encountered, bed rest, an epidural blood patch procedure, or closed lumbar drainage may be attempted. If unsuccessful, direct surgical repair may be necessary, and in rare cases, placement of an LP shunt may be considered.

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