Huge Spinal Extradural Meningeal Cyst in the Thoracolumbar Spine: A Case Report of a Rare Cause of Low Back Pain

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Symptomatic intraspinal extradural meningeal cysts are rare. We present the case of a 17-year-old female with low back pain, progressive numbness, and radiation pain to the flank and lower limbs. Magnetic resonance imaging and computed tomographic myelography revealed a huge intraspinal extradural meningeal cyst extending from T12 to L3 with cord and dural sac compression. The patient underwent surgery to open the cyst and close the connecting dural defect. Pathologic examination of the cyst disclosed non-specific fibrous connective tissue without an inner arachnoid single-cell lining. She achieved complete recovery after the operation. There was no recurrence of the cyst at a 2-year follow-up.

Key Words: spinal cyst, extradural cyst, meningeal cyst, arachnoid cyst, spinal cord compression

Intraspinal extradural meningeal cysts (EMCs) are rare causes of spinal cord or nerve root compression. Previous investigators have referred to spinal EMCs as extradural arachnoid cysts [1], pouches [2], diverticula [3], perineural cysts [4], and occult intrasacral meningoceles [5]. The classification of these lesions in the literature is indistinct, confusing and, in certain categories, histologically misleading. Therefore, Nabors et al simplified the classification of spinal EMC into three major categories [6]: spinal EMC without nerve root fibers (Type I), spinal EMC with nerve root fibers (Type II), and spinal intradural meningeal cyst (Type III). Type I is further divided into two subgroups: Type IA is an EMC (intraspinal extradural arachnoid cyst) and Type IB is a sacral meningocele (occult sacral meningocele). Type II is a Tarlov’s perineural cyst or a spinal nerve root diverticulum and Type III is a spinal intradural arachnoid cyst.

It is difficult to make a diagnosis of a spinal EMC based solely on histopathologic examination; in fact, the lining of the cyst cavity usually lacks arachnoid tissue. Together with modern imaging techniques, including magnetic resonance imaging (MRI) and computed tomography (CT) myelography, surgical and histopathologic findings can differentiate spinal EMC and give the correct diagnosis.

We describe a rare case of a symptomatic Type IA thoracolumbar spinal EMC that was identified by MRI and CT myelography that was successfully treated with surgery.

CASE PRESENTATION

A 17-year-old female had a congenital ventricular septal defect that was repaired surgically during childhood. She suffered from low back pain, with radiation to the right flank and progressive numbness in bilateral lower limbs for 12 months. Physical examination showed numbness in right L1 and bilateral L2 to S1 dermatome. There was no
motor deficit in the lower limbs. Deep tendon reflexes were intact. MRI of the thoracolumbar spine demonstrated a dorsally located intraspinal extradural cystic lesion extending from T12 to L3 with anterior compression of the thecal sac (Figure 1A and 1B). The signal intensity of the lesion was similar to that of cerebrospinal fluid (CSF). The mass also appeared to enlarge and slightly protrude through the neuroforamen. CT myelography revealed an intraspinal extradural cyst filled with contrast medium and anterior displacement of the dural sac (Figure 1C).

The patient underwent thoracolumbar laminectomy, which disclosed a large posterior extradural cystic mass compressing the dural sac. The cyst was opened and yielded CSF under high pressure. An ostium about $3 \times 5$ mm in size was identified at the dorsal lateral aspect of the dural sac near the junction of the theca and right L1 root sleeve. The CSF passed through this dural defect between the intraspinal subarachnoid space and the cystic cavity. After resection of the posterior cystic wall, the dural defect was closed with 6-O silk and reinforced with a free muscle flap.

Histopathologic examination of the cyst wall disclosed nonspecific fibrous connective tissue without a single-cell layer of inner arachnoid lining (Figure 2).

The patient was in total remission after surgical treatment at 6-month and 2-year follow-up. MRI also revealed no evidence of recurrence of the spinal EMC (Figure 3).

Figure 1. (A): Preoperative T2-weighted magnetic resonance imaging (MRI) sagittal view: a huge extradural meningeal cyst (arrows) extends from T12 to L3 with anterior compression of the dural sac. (B) T2-weighted MRI axial view: the cyst (arrow) protrudes through the bilateral L1 neuroforamen. (C) Computed tomography myelography reveals an intraspinal extradural cyst (arrow) filled with contrast medium and anterior displacement of the dural sac.
DISCUSSION

Spinal meningeal cysts are uncommon, accounting for 1% of all spinal tumors [7]. Spinal EMCs occur most frequently in the mid to low thoracic spine (67%), followed by the lumbosacral spine (20%), the thoracolumbar region (9%), and the cervical region (4%) [2,8,9]. Thoracic EMCs are more frequent in adolescents and sacral EMCs are more often found in adults. Most lesions like the one in this case are dorsally located in the spinal canal, with half protruding through the neural foramen. The incidence is higher in males, and most symptoms and signs develop during the second decade of life [8].

Thoracic and cervical EMCs usually present with myelopathy, lumbar EMCs with low back pain and radiculopathy, and sacral EMCs with radicular symptoms, bowel or bladder dysfunction [6,8]. The duration of symptoms is shorter for thoracic lesions due to the smaller diameter of the spinal canal. Intermittent exacerbation of symptoms may occur with postural changes and Valsalva maneuvers. More than 30% of patients have intermittent episodes of remission, although most progress in severity over months.

The etiologies of spinal EMCs remain uncertain. Most investigators believe that the cysts arise from herniation of the arachnoid through a congenital dural defect near or at the nerve root or dorsal midline of the dural sac [6,8,10–12]. Few patients have a familial tendency or associated congenital anomalies [6,7,13–15]. Some authors speculate that the proliferation and dilatation of the spinal arachnoid obstruct the CSF pathway, thus forming EMCs [12,16]. We prefer the congenital and developmental etiologies because our patient had a congenital anomaly (ventricular septal defect) and no arachnoid proliferation was found in her cyst. Progressive enlargement of spinal EMCs is presumably due to active and passive fluid transport [2,16,17]. However, the ball-valve mechanism is widely accepted [2,11,18].

Imaging diagnostic tools include plain films of the spine, CT myelography, and MRI. Plain radiographs may reveal widening of the spinal canal with scalloping of the vertebral bodies. MRI is a powerful tool in diagnosing EMCs, and does not require intrathecal injection of contrast medium. It can clearly delineate the cyst and the content characteristics of CSF on both T1- and T2-weighted sequences, and define its anatomic relation to the surrounding structures [19–24]. Although some authors think that MRI should completely replace myelography in imaging diagnosis, CT myelography still plays a role in

Figure 3. T2-weighted magnetic resonance imaging sagittal view 2 years after surgery shows no evidence of cyst recurrence.

Figure 2. Photomicrograph of the cyst wall shows a fibrocollagenous layer without an inner arachnoid layer (hematoxylin & eosin, original magnification × 50).
demonstrating the connection between the cyst and the subarachnoid space [6,25,26].

Type I spinal EMC was previously called spinal extradural arachnoid cyst. However, histopathologic examination seldom shows a single-cell arachnoid layer in the cyst wall [6]. The lesions usually consist only of nonspecific fibrous tissue [25]. Pathologic examination in our case also revealed a nonspecific fibrous cyst wall without arachnoid lining. Therefore, the diagnosis of Type I spinal EMC is difficult based solely on histopathologic findings. Only combined imaging studies, intraoperative findings, and histologic examinations give a correct diagnosis [6,25].

The differential diagnosis includes disseminated sclerosis, ependymal cysts, epidermoid cysts or dermoid cysts, enterogenous cysts, teratogenous cysts, inflammatory diseases, and intervertebral disc lesions [26,27]. MRI is the imaging modality of choice to differentiate these lesions [28]. Epidermoid and dermoid cysts are frequently located in the lumbosacral region and have inhomogeneous intensity, well-defined contrast-enhanced limits, and absence of surrounding tissue edema on MRI. The cysts are lined with keratinizing squamous epithelium, with or without sebaceous glands and hair follicles [27,29]. An ependymal cyst is typically lined with cuboidal-to-columnar, nonstratified, ciliated epithelium and presents with an intramedullary cyst with focal and significant expansion of the spinal cord for a few segments on MRI [30]. The enterogenous cyst is rare. MRI commonly shows anterior intradural-extradural cysts with isointensity to hyperintensity on both T1- and T2-weighted images [28]. The cyst wall is only simple cuboidal or columnar epithelium, with or without cilia. A teratogenous cyst occurs at any level of the intradural compartment and is variably composed of a neuroecto-, ecto-, endo-, and mesodermal derivative [27].

Surgery is recommended in associated symptomatic cases to avoid permanent damage to neural elements. The methods of surgical therapy for Type I spinal EMCs vary in the literature. They include total removal of the cyst [6,8,18,31], opening the cyst and closure of the fistulous communication [6,11,24–26,32], and alternative treatment with ventriculoperitoneal or lumboperitoneal shunting [6,33]. Total excision of the cyst is usually difficult due to adhesion and is unnecessary because the residual cystic wall does not promote recurrence. In our opinion, opening the posterior cystic wall and closing the connecting dural defect to eradicate the valve-like mechanism is sufficient. Ventriculoperitoneal or lumboperitoneal shunting to lower CSF pressure is reserved for recurrent cases.

In most reports, surgery provides some improvement of symptoms or stabilization of neurologic deficits without further deterioration. Only a few patients continue to deteriorate despite adequate spinal decompression. This may be related to permanent vascular insufficiency or damage to neural elements induced by long-term compression. One study revealed that only those with a short pain history and clear neurologic deficit profited from surgery [23]. In our patient, neurologic symptoms progressed within 1 year before surgery and she had good surgical results. This implied that the duration of associated symptoms might be the prognostic factor. Recurrence of the cyst after surgical intervention has not been seen in our patient and is very rarely reported in the literature.

Although a spinal EMC is rare, it should be kept in mind for patients with low back pain and associated neurologic symptoms. MRI is a very effective diagnostic tool. The final diagnosis should be made based on intraoperative findings, imaging studies, and histopathologic results. Surgical management by unroofing the cystic wall and closure of the connecting dural defect can cure the disease.

References

胸腰脊椎內之巨大硬脊膜外腦脊膜囊腫：
罕見的下背痛原因 ─ 病例報告

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有症狀之脊椎內硬脊膜外腦脊膜囊腫相當罕見。本文描述一位 17 歲的女性，主訴持續一年的下背痛，伴隨雙側下肢及右腰漸進性的酸麻。核磁共振影像檢查及電腦斷層脊髓攝影顯現一巨大脊椎內硬脊膜外腦脊膜囊腫，範圍分佈自胸椎第十二節至腰椎第三節，造成前側脊髓及脊膜腔嚴重的壓迫。病患接受胸腰椎手術，發現硬脊膜有一處缺損與囊腫內交通，於是將囊腫後壁切除並修補脊膜腔與囊腫之交通缺損處。組織病理顯示該囊腫壁為無特異性之纖維結締組織，與脊椎內硬脊膜外腦脊膜囊腫之診斷吻合。患者之症狀在手術後完全緩解，於門診持續追蹤兩年後之核磁共振影像檢查亦無發現復發跡象。我們回顧相關文獻並且探討其臨床表徵、可能的致病機轉、影像檢查、鑑別診斷及病理組織之特性與治療方式及預後。

關鍵詞：脊椎囊腫，硬脊膜外囊腫，腦脊膜囊腫，蜘蛛網膜囊腫，脊髓壓迫
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