СЛУЧАЙ ИЗ ПРАКТИКИ

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MULTIPLE SPINAL EXTRADURAL MENINGEAL CYSTS PARTIALLY ASSOCIATED WITH A DURAL DEFECT

Introduction. *Spinal extradural meningeal cysts, extradural outpouchings of the arachnoid that communicate with the intraspinal subarachnoid space through a small dural defect, are relatively uncommon.*
Case Report. *We report a case of 35-year-old man with multiple spinal extradural meningeal cysts in the thoracolumbar region. The operative findings revealed a dural defect in one of the cysts that allowed communication between the extradural cyst cavity and the subarachnoid space. Application of the Valsalva maneuver allowed cerebrospinal fluid to flow into the cyst's cavity; however, reverse flow did not occur.*
Result. *These findings indicate that a valve-like mechanism developed in the enlarging cyst, but there was no communication between the extradural cyst cavity and the subarachnoid space in the other cysts.*
Conclusion. *The number of independent cysts makes this case unique and suggests an underlying defect in the dura of the spinal canal in a limited region. Surgical resection of the cyst wall and closure of the dural defect obtained a favorable result.*
Key Words: *Spinal extradural cyst, Neurological deficits*

Introduction

Spinal extradural meningeal cysts are rare and are seldom a cause of spinal cord compression. They are thought to arise from congenital defects in the dura mater, they almost always communicate with the intrathecal subarachnoid space through the small defect in the dura [1-3], and they have been described as arachnoid cysts, pouches, and diverticula [4, 5]. These meningeal cysts are rare and may occur at all levels of the thecal sac. Spinal meningeal cysts are classified into three major categories: extradural cysts without nerve root fibers (Type I), extradural cysts with nerve root fibers (Type II) and intradural cysts (Type III) [6].

Spinal meningeal cysts are most common in the thoracic spine. When the spinal cord develops enlarging cystic cavities and spaces, the treatment is exclusively surgical [7-9]. These cysts are found predominantly in males [1-3]. The clinical presentation ranges from asymptomatic to pain, weakness, numbness, paralysis, and paralysis. The symptoms tend to occur during the second decade of life [1-3]. Patients usually present with progressive spastic or flaccid para- or quadriparesis [1-3]. Approximately 10% of patients present with monoparesis [1]. Sensory deficits are less prominent [10]. The clinical symptoms develop over months [2], although partial relief may occur in more than one-third of the patients [3]. Some cases are associated with long-term remission that extends for years [11].

Reciprocal obstruction and recanalization are thought to mediate remission and relapse [11, 12]. Nontraumatic spinal extradural meningeal cysts are thought to have congenital, iatrogenic and inflammatory etiologies [1-3, 4, 6]. Cyst expansion is thought to be due to active secretion of the internal cell lining [2], an osmotic spinal gradient between the subarachnoid space and cyst [3, 12], pulsatile cerebrospinal fluid (CSF) dynamics [4, 6], active fluid secretion, hydrostatic forces and valve-like mechanisms [1, 3, 4, 5, 13]. Active secretion by the inner cell lining has never been proven, and this hypothesis has been discredited [1, 3].

Case Report

A 35-year-old man was referred to our hospital with a six month history of progressive bilateral leg weakness. Five years prior to admission, he occasionally experienced intermittent urinary incontinence, and three years prior to admission, he complained of problems with defecation. He had no history of infection or trauma.

Examination

On examination, we found no cutaneous stigmata of neurological disease and no dysmorphic features. The patient had no evidence of phacomatosis. Neurologically, the muscle strength of his right leg reduced to Grade 3+/3+. Bilateral upgoing plantar responses were present. Clonus was present at the both the patella and the ankles. There was hypoesthesia in small mid-thigh areas bilaterally and on the dorsal side of the left foot.

Laboratory and Radiological Examinations

Routine laboratory tests yielded normal results. Thoracolumbar vertebral roentgenograms (Th7-L2) revealed an enlarged interpedicular space. Magnetic

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resonance imaging (MRI) revealed the presence of multiple extradural cystic lesions posterior to the cord that flattened and shifted the spinal cord anteriorly from Th-8 to L1 (Figure 1). This lesion was best visualized as a high signal intensity relative to the CSF on T2-weighted MRI (Figure 2). The lesion ventrally compressed the thecal sac and spinal cord. The cysts contained fluid that was demonstrated to be the same as CSF. Electromyography indicated the presence of myelopathy.

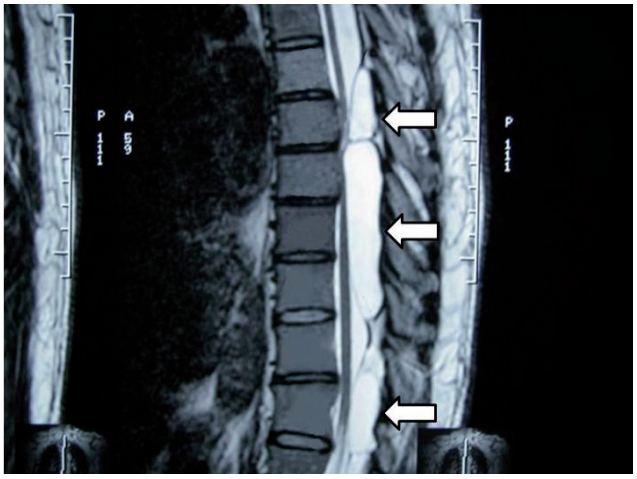

Figure 1. A magnetic resonance image (MRI) in a 35-year-old man with lower extremity weakness, occasionally experienced intermittent urinary incontinence and problems with defecation; the presence of multiple extradural cystic lesions (arrow) posterior to the cord that flattened and shifted the spinal cord anteriorly from Th-8 to L1

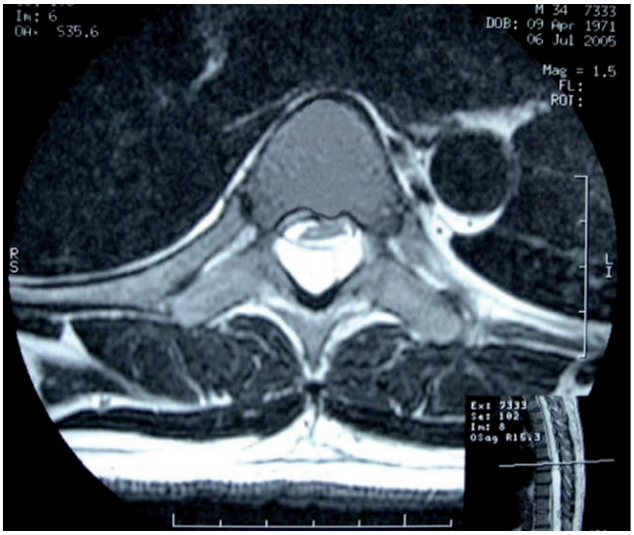

Figure 2. Cystic lesion was best visualized as a high signal intensity relative to the CSF on T2-weighted MRI. The lesion ventrally compressed the thecal sac and spinal cord. The cysts contained fluid that was demonstrated to be the same as CSF

Operation

The cystic lesion was exposed via laminectomy extending from Th7–Th9, and a partial laminectomy was performed at the L-1 laminae. On reflecting the bone, the canal was found to be occupied by multiple large translucent cysts. The cyst wall was white, fibrous, and tense. Following the removal of CSF-like fluid by puncture, the cyst re-expanded within a few minutes. The dorsal wall was opened vertically to explore the intracystic cavity. There was a small dural defect adjacent to the right Th-7, bilateral Th-8 and left Th-9 nerve roots. During forced inflation of the patient's lungs by the anesthesiologist using the Valsalva maneuver, CSF flowed into the cyst cavity via the dural defects, without subsequent reflux. No other defects were noted. The dural defect was closed using No. 4-0 nylon. The ventral wall of the cyst was easily separated from the theca. The L-1 cyst was dissected in a similar manner, but no dural defects were found. The wound was closed in a watertight fashion.

Postoperative Course

During the postoperative course along with neurorehabilitative care, there was an increase in motor strength and improvement in the vegetative symptoms. The clonus remained.

Pathological Findings

A histopathological study of the cyst wall revealed a thin layer of arachnoid lined by discrete nests of meningothelial cells. In some places immediately adjacent to the arachnoid, there was a thin layer of refractile collagenous connective tissue resembling attenuated dura. The final pathological diagnosis was an arachnoid cyst (Figure 3).

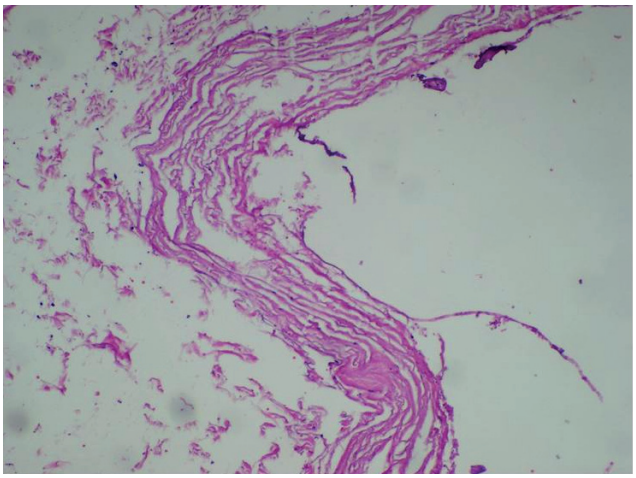

Figure 3. A histopathological study of the cyst wall revealed a thin layer of arachnoid lined by discrete nests of meningothelial cells, there was a thin layer of refractile collagenous connective tissue resembling attenuated dura. The pathological diagnosis was an arachnoid cyst

Discussions

Extradural arachnoid cysts of the spine are an uncommon cause of myelopathy secondary to spinal cord compression. Although they are most commonly found in the thoracic spine, they may occur in other parts of the spine [9, 14-16]. They are commonly found posterior to the spinal cord, but there have been several reports of cysts in the posterolateral and anterior positions [14]. They are more common in males, and their peak incidence is in the second decade of life [9]. The precise causes and natural history of arachnoid cysts are unclear. They are known to be associated with trauma, surgery, arachnoiditis, and neural tube defects [15, 17, 18]. Familial cysts have been reported, and there is an association with lymphedema and distichia [7, 8].

According to the literature, there is no known underlying cause, and these lesions are thought to have congenital, iatrogenic and inflammatory etiologies. In our case, there was no history of trauma or surgery and no evidence of arachnoiditis. The history of progressive myelopathy for many years and the secondary bone changes observed on radiographs and at the time of surgery suggest a long-standing lesion with a congenital origin. Both active and passive fluid-transport mechanisms have been hypothesized to explain the formation and enlargement of extradural meningeal cysts [5]. Two cases of extradural arachnoid outpouchings have been reported, although in each case, there was only one diverticulum [9]. These cases suggest that the initial arachnoid herniation enlarges due to pressure pulses within the CSF that are caused by straining or coughing and by a valve-like mechanism at the cyst neck that prevents emptying. In other reports, intermittent relief of symptoms has been obtained in conjunction with changes in posture, suggesting that in certain positions, the diverticulum may empty [1, 14].

Histopathologically, the cyst wall has been reported to consist of fibrous connective tissue with or without an inner single-cell lining, which may represent an arachnoid membrane [1, 6]. Cells with secretory capability are frequently absent [12]. Microscopy of the cavities mostly shows gliosis of the lining with an annular fiber arrangement. The cavities appear to develop by fluid cutting through sites of structural weakness. The location and lining of the cavities has provoked discussion relating to whether the central canal is involved. Tumors may be associated with cyst cavities. Most investigators now prefer the passive fluid-transport theory to explain the etiology of cyst expansion via pulsatile CSF dynamics and an osmotic gradient with or without valve-like mechanisms [4, 12, 13, 19]. In the early stages, pulsatile CSF dynamics may promote cyst enlargement [4, 5], and an osmotic gradient can facilitate further expansion [3, 12]. It is possible that free fluid exchange occurs across the cyst wall in the spinal tumors. Both operative findings and post-myelographic CT scan suggest communication between extradural meningeal cysts and the subarachnoid space, as observed when intrathecal contrast material enters the cystic cavities, in approximately half of reported cases [1]. A recent study suggests that such communication exists in nearly all cases of meningeal cysts [6].

Complete resection is ideal treatment [6]; surgery is most likely succeed if symptoms match findings. Unfortunately, not all isolated spinal meningeal cysts can be fully resected. In such case, percutaneous drainage, or shunting the cysts into the peritoneal cavity might relieve symptoms [6, 20]. Minimal invasive surgery have also met with some success. Neo et al., reported treated a giant spinal extradural arachnoid cyst by selectively closing, or occluded the dural defect with clips [21].

In the case presented here, the thoracic cyst was deflated immediately after intraoperative puncture, but it expanded within a few minutes, except in the lumbar region. The intraoperative findings confirmed that CSF influx through the dural defect followed forced inflation of the lungs. Although an osmotic pressure gradient may be partly responsible for cyst enlargement, the intraoperative findings strongly supported a valve-like mechanism [13].

MRI is a useful tool for diagnosing intraspinal meningeal cysts [13]. In the case presented here, MRI provided clear anatomical information and demonstratedpathological change. The hyperintense cystic lesion located in the spinal cord caused long-standing spinal cord compression. The prognosis of extradural meningeal cysts is favorable, and one-third of patients achieve complete recovery [13]. The persistence of a high intensity signal in the spinal cordon T2-weighted MRI may indicate the presence of a permanent subclinical lesion in our case. Resection of the cyst wall from the posterior aspect of the thecal sac and closure of the dural defect were performed to eradicate the valve-like mechanism [10, 13, 22].

Conclusion

In our report, there were multiple cysts, but only a single cyst with a cavity that demonstrated multiple separate necks extending through a dural opening. These necks allowed the cyst (or, more precisely, the diverticulum) to communicate with the spinal subarachnoid space.

Conflict of interest

None

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ТҮЙІНДЕМЕ

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ЖҰЛЫННЫҢ ҚАТТЫ ҚАБЫҒЫНЫҢ АҚАУЫМЕН ІШІНАРА БАЙЛАНЫСТЫ ЖҰЛЫННЫҢ КӨПТҮРЛІ ЭКСТРАДУРАЛЬДЫ МЕНИНГЕАЛЬДЫ ЖЫЛАУЫҚТАРЫ

Кіріспе. Омыртқадағы менингеальды жылауықтар, үлкен емес дуральды ақау арқылы субарахноидальды кеңістікпен байланысқан арахноидальды қабықтың экстрадуральды томпаюы салыстырмалы түрде сирек кездеседі.

Клиникалық жағдай. Кеуде және бел аумақтарында жұлынның көптүрлі экстрадуральды – менингеальды жылауықтары бар 35 жастағы ер кісінің жағдайы қарастырылады. Интраоперациялық жолмен жылауықтардың бірінен жұлынның қатты қабығының ақауы – субарахноидальды кеңістіктен эпидуральды жылауық бар екендігі анықталды. Вальсальва құрылғысы жұлын сұйықтығының жылауық қуысына ағуына мүмкіндік берді, алайда кері ағу байқалмады.

Нәтиже. Нәтижелер қақпақшалы механизм жылауық қуысының ұлғаюына себепкер болғандығын көрсетеді, бірақ экстрадуральды қуыстар мен басқа жылауықтардың субарахноидальды кеңістігі арасында байланыс бар екендігі туралы деректер анықталмады.

Тұжырым. Оқшауланған жылауықтардың саны бұл жағдайды бірегейлендіреді және шектелген аймақта жұлынның қатты қабығында ақаудың бар екендігін болжамдайды. Жылауық қабырғасының хирургиялық резекциясы және дуральды ақауды жою оң нәтиже беруде.

Негізгі сөздер: Омыртқаның экстрадуральды жылауығы, неврологиялық тапшылық.

РЕЗЮМЕ

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МНОЖЕСТВЕННЫЕ СПИННОМОЗГОВЫЕ ЭКСТРАДУРАЛЬНЫЕ МЕНИНГЕАЛЬНЫЕ КИСТЫ ЧАСТИЧНО СВЯЗАННЫЕ С ДЕФЕКТОМ ТВЕРДОЙ МОЗГОВОЙ ОБОЛОЧКИ

Введение. Позвоночные экстрадуральные менингеальные кисты, экстрадуральное выпячивание арахноидальной оболочки связаное с субарахноидальным пространством через небольшой дуральный дефект, относительно редки.

Клинический случай. Рассматривается случай 35-летнего мужчины со множественными позвоночными экстрадурально-менингеальными кистами в области грудного и поясничного отделов. Интраоперационно был обнаружен дефект твердой мозговой оболочки в одной из кист, что обеспечило сообщение эпидуральной кисты с субарахноидальным пространством. Маневр Вальсальвы привел к поступлению спинномозговой жидкости в полость кисты, однако обратного оттока не наблюдалось.

Результат. Результаты показывают, что клапанный механизм способствовал увеличению полости кисты, но данных о наличии сообщения между экстрадуральными полостями и субарахноидальным пространством других кист не было обнаружено.

Заключение. Количество изолированных кист делает этот случай уникальным и предполагает наличие дефекта в твердой мозговой оболочке спинного мозга в ограниченной на ограниченном участке. Хирургическая резекция стенки кисты и закрытие дурального дефекта дает положительный результат.

Ключевые слова: Позвоночная экстрадуральная киста, неврологический дефицит.