Multiple Spinal Extradural Arachnoid Cyst : A Case Report

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Abstract

Introduction: Spinal extradural arachnoid cysts(SEAC) are a rare cause of spinal cord compression, nerve root compression, or both, accounting for approximately 1-3% of all primary spinal space-occupying lesions. Multiple SEACs are rarely reported in the literature. Aim of this article is to illustrate our experience of surgical treatment of this rare but curable disease.

Case Report: We present a case report of 15-year-old boy who presented with progressive lower extremity weakness, pain and dysaesthesia. Magnetic resonance (MR) of the spine revealed two extradural arachnoid cysts. The patient underwent a thoracic laminoplasty for en bloc resection of the spinal extradural arachnoid cyst. Postoperatively, the patient's motor strength and ambulation improved immediately.

Conclusion: We have described a rare case of back pain and leg weakness in patient with multiple thoracolumbar spinal extradural cysts. Clinical outcome after Laminoplasty and surgical excision of cyst was excellent and there has not been any evidence of cyst recurrence and symptomatic worsening till now(three years post surgical enbloc excision).

Keywords: spinal extradural arachnoid cyst, laminoplasty, excision

Introduction

Spinal arachnoid cysts are a rare cause of spinal cord compression, nerve root compression, or both, accounting for approximately 1-3% of all primary spinal space-occupying lesions[1]. Spinal extradural arachnoid cyst (SEAC) is more commonly found among male patients and during the second decade of their life. SEACs can be found in any location, although mostly reported to be located at mid thoracic to the thoraco-lumbar junction, commonly in a posterior position[2].

SEAC is an out-pouching herniation of arachnoid membrane through a dural defect that may communicate to intradural subarachnoid space. The aetiology of this herniation is still unclear and can be either congenital or acquired. These cysts can result in fluctuating symptoms associated with cord or root compression. It is assumed that they can be enlarged by subsequent pressure change in the cerebrospinal fluid (CSF) during exercise and Valsalva manoeuver as there is micro-communication between the cysts and subarachnoid space[2].

Patients may present with symptoms of pain, weakness, ataxia, and/or bladder incontinence. In the modern era of neuroimaging, these lesions can also be discovered incidentally. Once the cyst has been diagnosed, typically by MRI, surgery is usually performed to prevent further spinal cord or nerve root compression.

Despite the rarity of SEACs, it is important and worthy to keep it as differential diagnosis as they are surgically curable disease. Diverse surgical techniques have been introduced and many reports reviewed favourable outcome of SEACs with surgical treatment. Aim of this article is to illustrate our experience of surgical treatment of this rare but curable disease.

Case Report:

15-year-old boy presented with progressive lower extremity weakness, pain and dysaesthesia, aggravated by upright posture and partly relieved with lying down, myelopathy, and severe gait ataxia. Patient had no penetrating trauma, contrast media application, inflammation, haemorrhage, or surgical history.

Radiography of the thoracolumbar spine revealed thinned pedicles and increased interpedicular distance. Magnetic resonance (MR) of the spine demonstrated two extradural arachnoid cysts extending from T-5 to T-9 and T-10 to T-11, causing anterior displacement of the dura. The cyst was isointense with the cerebrospinal fluid

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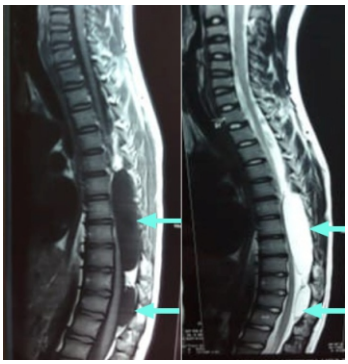

Figure 1: Sagittal Cuts On MRI: T1W and T2W showing two lobulated arachnoid cysts (arrows shows compression in spinal cord)

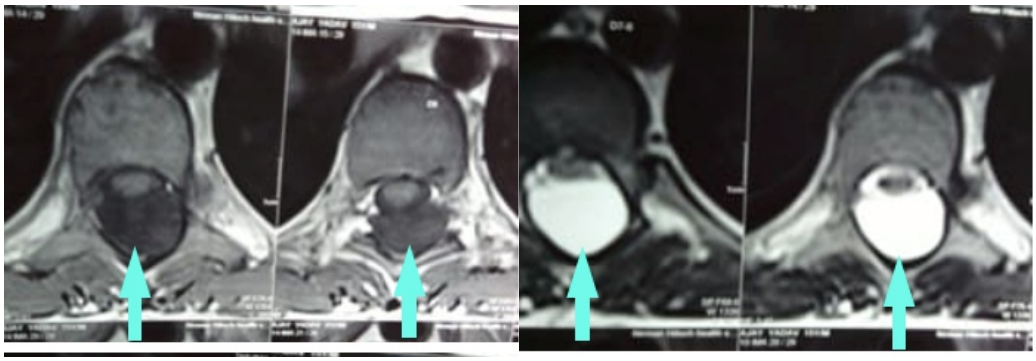

Figure 2: Axial cuts on MRI: T1W and T2W showing arachnoid cysts (Arrows shows compression in spinal cord)

(CSF) on all sequences and was adjacent to the posterior epidural fat (Figure 1 and Figure 2).

The patient underwent a thoracic laminoplasty for en bloc resection of the spinal extradural arachnoid cysts(Figure 3, Figure 4 and Figure 5). A small communications with the subarachnoid space were seen at the level of T-7 and T-10. They were sealed with tissue fibrinogen after repair with primary suture. Postoperatively, the patient's motor strength and ambulation improved immediately.

Discussion:

SEAC is a rare disease entity accounting for 1% of all spinal tumors[3]. The mechanism for the initial formation of an arachnoid cyst is likely mutifactorial and remains still unclear and can be congenital or acquired[2]. A few theories have been proposed. Elsberg et al[4] reported 4 cases of arachnoid cysts in 1934 and proposed an origin from congenital diverticula or a congenital dural defect with herniation of the arachnoid. Support for the congenital theory is further strengthened by a few case reports of familial tendencies[5,6], and the frequency of association with neural tube defects. Some reports demonstrated an association with dural ectasia or Marfan syndrome[7]. In this condition, a primary defect in the organisation of collagen with decreased tensile strength weakens the ligamentous structures and other supporting tissues. Dural stretching can lead to dural thinning to such an extent that it becomes ectatic and even deficient in areas[8]. Other reports have hypothesised that congenital defects in the distribution of arachnoid trabeculations lead to misplaced cellular remnants resulting in an embryonic malformation, which may act as a nidus for cyst formation[9]. Dural defect is often found around the nerve root sleeves. One possible explanation is that tension across the movable dural sac and relatively fixed roots can predispose such dural tears. If patients have underlying structural abnormality such as Marfan syndrome, the probability of such tears may be further increased[2].

The cause of dural defect can be acquired. Trauma, arachnoiditis or iatrogenic cause can result in small dural tear and subsequent CSF accumulation to develop SEACs[2]. In our experience, patient had no history of trauma, arachnoiditis or previous spine operation. No specific underlying disease was documented.

Expansion of an arachnoid cyst after initial formation is also likely multifactorial and may not occur as a result of the same mechanism in all cases. The ball-valve mechanism of enlargement has been described in numerous cases. In the ball-valve theory, the cyst communicates with the subarachnoid space via a small stalk functioning as a one-way valve[10].

McCrum and Williams[11] stressed the importance of having surge in pressure, which fill and expand the cyst, followed by stalk closure, which maintains the higher internal cystic pressure. These surges in CSF pressure may be caused by straining or coughing during normal activities. Without the surges in pressure, the pressure within the cyst and the adjacent subarachnoid space would equilibrate and the cyst would not enlarge. Another proposed mechanism of expansion is via an osmotic gradient formed between the cystic structure and the subarachnoid space, thus facilitating expansion. Yet another proposed mechanism is that the degeneration of cells of the arachnoid trabeculae may lead to an elevated osmotic effect within the cyst[10]. Sandberg et al. [12], however, demonstrated that the chemical composition and the osmolality of cranial arachnoid cysts and the CSF are similar. They found that the protein levels were significantly elevated in the cysts and proposed that the cysts increased in size through colloid osmotic pressure due to the presence of large proteins that cannot pass through semipermeable membranes. Lastly, cyst expansion may be due to active secretion of cells lining the cyst. The actual mechanism of expansion is likely from a series of events that may include one or more of the fore mentioned processes[10].

Nabors et al. categorized SEACs in three major groups of meningeal cyst, non-meningeal epidural cysts, and neurenteric cysts. Meningeal cysts are further classified in 3 subgroups : 1) type 1 : extradural meningeal cyst that contains no neural tissue, 2) type 2 : extradural meningeal cyst that contains neural tissue, 3) type 3 : intradural meningeal cyst. Type 1 meningeal cysts consist of extradural arachnoid cysts (type Ia) and sacral meningoceles (type Ib)[13].

The majority of cysts (more than 80%) are located posterior to the spinal cord. Wilkins and Odom, reviewed 67 cases, including 50 in which lesion location was reported; of these, 47 (94%) were posterior or posterolateral. Perret et al[14]. postulated that SACs develop from dilation of the septum-posticum, the thin membranous arachnoid partition that longitudinally divides the posterior subarachnoid space in the midline. Dilation of the septum-posticum would explain the frequency with which arachnoid cysts are found in the posterior arachnoid space in the general population. Rabb et al[15]., however, found an

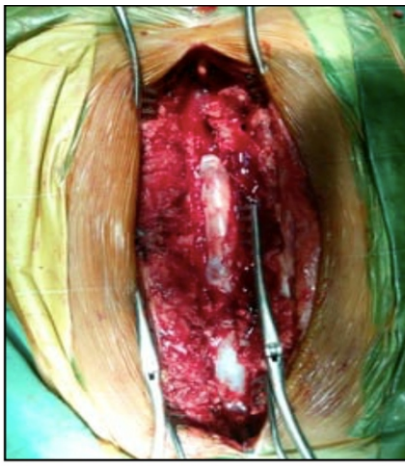

Figure 3: intraoperative picture Showing two arachnoid cysts.

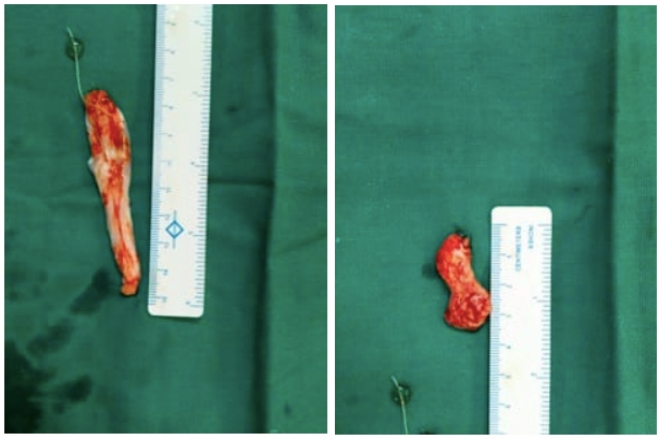

Figure 4: Showing excised arachnoid cysts

association of anteriorly placed cysts in paediatric patients with myelomeningocele, who seemed particularly prone to having anteriorly situated arachnoid cysts that likely developed as a result of altered CSF flow secondary to extensive arachnoid adhesions.

The most common region for the development of symptomatic SACs is the thoracic canal. Cloward[16] performed a literature review of 91 cases of arachnoid cysts from the first case ever discovered at autopsy (1898) to his case report (1968). In his review, 65% cysts were located in the thoracic region: 12% were thoracolumbar, 13% were lumbar or lumbosacral, 6.6% were sacral, and 3.3% were cervical. Another large literature review by Kriss and Kriss[17] in 1997 had similar findings of 80%, 15%, and 5% for the thoracic, cervical, and lumbar regions, respectively. It is worth emphasising that the thoracic spinal canal is relatively small in diameter and the longest part of the spine, and cysts developing in this region are more likely to become symptomatic than are SACs in other spinal regions, whether in the general population or in pediatric patients[10].

In General, SACs are more commonly located extradurally than intradurally. But higher prevalence of intradural SACs in the paediatric age group is likely related to congenital CNS abnormalities. Arachnoid cysts are usually extramedullary.

Arachnoid cysts usually present as solitary lesions; however, in rare cases multiple septations and multiple independent cysts are found. Also multiple SACs are reported rarely in literature[18].

Symptoms of SACs likely develop as a result of pressure on the spinal cord or a spinal nerve root. Often the symptoms can be aggravated by upright posture. Patients most commonly present with pain, followed by sensory changes (often presenting as gait instability), urinary dysfunction, and/or weakness.

These lesions are twice as common in males and usually present in the second decade. Lumbar and sacral cysts are usually present in the third and fifth decades[19]. The most common presenting symptoms are pain, paresthesia, intermittent claudication, and variable degrees of spastic weakness. Bowel or bladder dysfunction may occur with sacral cysts[19].

Radiographs of the spine usually show bone erosion with widening of the canal, erosion of pedicles, foraminal enlargement, and scalloping of the vertebral bodies or the sacrum. The valve-like mechanism with intermittent surges in CSF pressure is thought to explain expansion of the cyst and bone erosion. The diagnosis can be established by myelography, which demonstrates an extradural defect with smooth displacement of the margin of the thecal sac. CT myelograms may demonstrate the communication between cysts and the subarachnoid membrane by Pooling of contrast medium in the lesion indicating communication with the subarachnoid space[19,20]. But it is an invasive procedure. MRI is the diagnostic procedure of choice as it is noninvasive and can demonstrate the cyst nature, exact size, and anatomic relationship with the spinal cord[21]. In a previous study, cine-MRI was used to detect this communication site, enabling the treatment of a giant extradural arachnoid cyst with selective closure of the dural defect[22]. Extradural arachnoid cysts must be differentiated from neuroepitelial, neuroenteric, dermoid, epidermoid and teratoid cysts[23].

The management of these cysts has not been standardized. Treatment methods include complete surgical excision of the cyst wall with closure of the communication site between the cyst and the subarachnoid space after laminectomy, partial resection of the cyst wall and closure of the communication site between the cyst and the subarachnoid space, only closure of the communication site between cyst and the subarachnoid space or percutaneous aspiration of arachnoid cyst[24]. The treatment of choice for symptomatic SACs is excision or fenestration. When the cyst is located anterior to the spinal cord, fenestration of the cyst is performed to prevent further enlargement. For extradural cysts, excision of the cyst and closure of the cyst/dural communication is performed. Effort is made to remove as much of the cyst wall possible; however, recurrence has been reported after total cyst wall removal. For refractory cases, insertion of a cystoperitoneal shunt may be required. Extensive laminectomy can result in worsening of scoliosis[19]. To avoid this, Laminectomy can be performed, preserving the spinous processes and the facet joints[20]. For small extramedullary cysts that are incidental and asymptomatic, conservative management is the preferred option, ensuring that patients are provided with adequate follow-up. Although some authors have reported minimally invasive treatment strategies using endoscopic techniques or CT- or MRI-guided aspiration, resection and wide fenestration remain the most reliable ways to eliminate the mass effect from a SAC.

Conclusion:

We have described a rare case of back pain and leg weakness in patient with multiple thoracolumbar spinal extradural cysts. Multiple SEACs are rarely reported in the literature. Excision of the cysts at the spinal cord level leads to a favorable outcome. Clinical outcome after Laminoplasty and surgical excision of cyst was excellent and there has not been any evidence of cyst recurrence and symptomatic worsening till now(three years post surgical enbloc excision).

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