Case report

Spinal extradural arachnoid cyst: A rare cause of thoracic myelopathy

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ARTICLE INFO

Keywords:
Extradural arachnoid cyst
Thoracic myelopathy
Spinal surgery

ABSTRACT

A spinal extradural arachnoid cyst (SEAC) is a rare condition which can lead to back pain, radiculopathy, and compressive myelopathy. It accounts for approximately 1% of spinal tumours. The exact aetiology of SEACs is not well understood; however, this study is supportive of a traumatic aetiology of this disease in addition to supporting a uni-directional valve as a mechanism of CSF accumulation. The purpose of this study is to review the presentation, work-up, treatment and postoperative course of a patient with SEAC. We present the case of a 40-year-old male with a history of trauma who developed a SEAC and was treated surgically at our unit. Although a rare disease, we highlight the importance of early diagnosis and surgical treatment as it is a surgically curable disease with a good postoperative prognosis.

1. Introduction

Spinal extradural arachnoid cysts (SEAC) are rare lesions, accounting for approximately 1% of spinal tumours [1]. They predominantly affect males in the second to fifth decades of life [2,3], and most commonly affect the mid-thoracic spine and thoraco-lumbar spine but may be found at any vertebral level [3–7]. Symptoms may include back pain, radiculopathy, and less commonly, compressive myelopathy. Surgical excision is considered the standard treatment for symptomatic lesions and although several hypotheses exist, the exact aetiology of SEAC formation and expansion is unclear.

We report the case of a 40-year-old male with a clear history of trauma who developed a SEAC and was treated surgically at our institution. This case highlights the importance of early diagnosis and surgical treatment, as well as providing further clarity to the underlying aetiology of these rare lesions. Verbal informed consent from the patient was gained prior to commencing this manuscript.

2. Case study

A 40-year-old male presented with a two-year history of insidious onset mid-thoracic back pain and worsening myelopathy. He described increasing paraesthesia throughout his lower limbs ascending to his trunk and intermittent leg 'spasms'. Bowel and bladder function were unaffected. On examination, he had marked hyperreflexia in his deep tendon reflexes of his lower limb and equivocal plantar responses bilaterally. He had evidence of dorsal column dysfunction with a T10 sensory level to light touch and impaired proprioception of his lower limbs. Pain and temperature sensation were intact, and his gait was unremarkable.

The patient’s past medical history revealed trauma and ankylosing spondylitis. He reported being in a significant motor vehicle accident at 18 years of age, where he sustained significant orthopaedic injuries to his lower limbs. Although unable to recall a spinal column injury, he experienced back and leg pain for several years following the event.

An MRI of the thoracolumbar spine demonstrated a cystic mass at T6–8 with evidence of cord compression (Fig. 1). The cyst measured 54x15mm (SI x AP) and appeared to be extradural. It was hyperintense on T2-weighted imaging, consistent with CSF, and demonstrated no enhancement on contrast sequences. Lateral extension through the T7–8 intervertebral foramina was present without foraminal expansion or erosion. Severe canal stenosis was present at T7 with associated myelomalacia. Given the clinical and radiological findings, he was consented for operative treatment.

A T6–8 laminectomy was performed revealing a large extradural cystic lesion compressing the midthoracic cord (Fig. 2). It was easily separated from the dura except at the left T8 nerve root where it was firmly adhered and extended towards the neural foramen. Here, it was disconnected using two Weck® clips, excised, and submitted for histopathological assessment. Valsalva testing revealed no dural defect.
however a prophylactic repair using a local Duragen® graft and Tisseel®
glue was performed.

Macroscopically, a thin-walled cyst with a semitranslucent fibrous
 capsule was demonstrated. Microscopically, the cyst consisted of dense
fibrous connective tissue, focally showing a flattened layer of
arachnoidal cells (Fig. 3). Scant arachnoidal whorls of epithelial cell
were also present. Immunoperoxidase staining demonstrated the flat-
tened cyst lining cells and whorled arachnoidal cells were EMA positive.
This confirmed the suspected diagnosis of a SEAC.

The patient made an uneventful recovery. When reviewed in clinic at

Fig. 1. MRI thoracic spine.
(A) Axial and (B) sagittal T2-weighted MR images demonstrating a SEAC at T6–8 with evidence of cord compression.

Fig. 2. Intraoperative photographs.
(A) T6–8 laminectomy revealing extradural arachnoid cyst. (B, C) Separation of the superior aspect of the cyst from the underlying dura. (D, E) Tethering of the cyst to left T8 nerve root. (F) Cyst excised revealing intact dura.
Spinal arachnoid cysts can be classified broadly based on anatomical location within the spinal canal and contents, with Nabors et al. publishing the most accepted classification system based on their series of 22 cases [8]. Lesions were classified as extradural cysts without spinal nerve root fibers (Type I), extradural cysts with spinal nerve root fibers (Type II), and intradural meningeal cysts (Type III) [8]. Intramedullary arachnoid cysts have also been reported elsewhere [9]. Type I cysts were further defined as sacral meningoceles (type IB) or extradural meningeal cysts (type IA), as represented in our illustrative case, with the presented histopathological and anatomical findings enhancing the overall understanding of these rare lesions and forming a foundation for future studies.

Despite this, the exact aetiology of SEAC formation and expansion remains incompletely understood. They are thought to be the result of arachnoid membrane protruding through a dural defect with subsequent CSF accumulation [1,3,10]. Both congenital and acquired causes have been described [11,12]. Potential predisposing factors for acquired SEACs include trauma, arachnoiditis, iatrogenic causes as well as underlying structural problems such as Marfan syndrome and dural ectasia [10,13–15]. This case supports the acquired hypothesis, with the patient suffering a significant trauma prior to developing this SEAC.

There are two accepted theories for CSF accumulation and SEAC expansion [10,13]. Firstly, CSF may be actively secreted from residual arachnoid membranes contained within the cyst. Secondly, CSF may passively enter the cyst through a unidirectional valve formed at the dural defect. This case supports the latter hypothesis given the histopathology demonstrating simple flattened and nested arachnoidal cells with no secretory function.

Clinical features of SEACs are often chronic and progressive. Patients may present with back pain, radiculopathy and less commonly, compressive myelopathy [3,10,13,16]. Common neurologic manifestations include gait impairment, lower limb paraesthesia, lower limb weakness, radicular pain as well as bowel and bladder dysfunction [3,4,17]. Symptoms may be exacerbated by Valsalva manoeuvres due to influx of CSF into the SEAC [4,18,19].

Diagnosis of a SEAC is most effectively performed using MRI which is both highly sensitive and specific [3,6,16,17]. As in this case, MRI demonstrates a cystic structure which is isointense to CSF, with high signal on T2-weighted imaging. Identification of a communication site with the intradural cavity may be possible with Cine MRI or CT myelography [3,6,10,11,16,17]; however, in our practice both were felt unlikely to aid or significantly alter the surgical approach.

For asymptomatic patients, SEACs may be managed conservatively with routine observation [20]. As neurologic features are due to chronic expansion of the SEAC and compression of neural elements, non-operative management is unlikely to be effective once a patient becomes symptomatic [11,21]. Hence, surgical intervention is the first-line treatment for those presenting with significant pain or myelopathy, and prognosis is excellent [3,6,10,21]. Definitive surgical treatment includes laminectomy or laminoplasty with complete excision of the cyst and obliteration of the communicating pedicle. To decrease risk of recurrence, watertight dural repair should be performed where a defect is observed [11], and we advocate for prophylactic repair where one is not clearly visualised. Simple aspiration is not recommended due to the risk of recurrence [20].

4. Conclusion

Our patient ultimately made an excellent recovery from his operation with early improvement of myelopathic symptoms. This case supports the hypotheses regarding SEAC formation and expansion. It also highlights the importance of early diagnosis and surgical treatment as, although rare, SEACs are essentially surgically curable with a good post-operative prognosis. The authors encourage further research with retrospective multi-institutional cohort studies to define the epidemiology and predisposing factors of this rare disease.

Funding

No funding was received by any author for the completion of this study.

Declaration of Competing Interest

None.

Acknowledgments

No acknowledgements.
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