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Review Article

Extradural arachnoid cyst – Case report and literature review

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ABSTRACT

Extradural arachnoid cysts (EAC) are a rare entity that may cause spinal cord or radicular nerve compression, usually in young adults. The most common locations are the posterior or posterolateral region of the thoracic spinal canal or the thoracolumbar junction.

The most accepted theory is that they are formed by the protrusion of the arachnoid through a defect in the dura, they progressively increase in size through physiological Valsalva maneuvers performed dailyand then they can become symptomatic by direct neural compression.

Surgery is the treatment of choice for these cases, with complete removal of the cyst and closure of the dural defect. Extradural arachnoid cysts have generally a good prognosis.

This article aims to present a case of an extradural arachnoid cyst in a 13 year old patient with spinal cord compression syndrome (paraparesis with hyperreflexia, clonus and alteration of deep sensitivity in the lower limbs), without previous history of trauma. Dorsal spine magnetic resonance imaging revealed an extradural cystic lesion with significant spinal cord compression similar to the CSF in T2 weighted images. The lesion extended from T6 to T9.

1. Introduction

Spinal arachnoid cysts (SAC), also named meningeal cysts, are a rare condition first described by Spiller in 1903, although the first reported case as an autopsy finding can be traced back to 1898 by Nonne [7]. They are diverticula of the dura mater, arachnoid or nerve root sheath that causes the accumulation of cerebrospinal fluid (CSF) within extradural space. They present as collagenous fibers and membrane with flat lining cells and foci of calcification may be seen within the fibers. Histopathological confirmation of the nature of the cyst wall (arachnoid) is not always possible [9].

Two studies analyzing a total of 35 patients with SACs demonstrated that intradural arachnoid cyst are more common than extradural arachnoid cysts [14,15], finding intradural cysts in 50–54% of the patients and extradural cysts in only 27 to 38%. Extradural arachnoid cysts actually account for less than 1% of all spinal epidural lesions [6], most commonly seen in the thoracic spinal canal, followed by lumbar and cervical [7,14]. They are usually present in the second decade of life and seem to be more frequent in males but sex distribution in published series varied significantly [14].

The cause of these cysts remains unclear, they most probably have a congenital origin but some may be acquired from trauma, infection, or inflammation [16]. They result from a protrusion of the arachnoid that communicates with the subarachnoid space through one or many defects in the dura mater [3]. A few have been reported as being non communicating with the subarachnoid space [1].

The size of spinal extradural cysts varies dramatically. One study selected 51 patients and found the mean length of the EACs to be of 5.04 vertebral bodies (range 2–13) [17]. In our report, the length of the cyst was about 2 vertebral bodies which is inside the range of size found in the study.

As it expands, they may cause compression on the spinal cord and/or nerve roots causing long tracts dysfunction and/or neuropathic pain.

2. Case description

A 13-year-old female patient presented with paraparesis grade 2, hyperreflexia, clonus and deep sensitivity alterations in lower limbs. These alterations were progressive for 3 months until she arrived in our institution. There was neither history of trauma nor other comorbidities. After general serum exams were made, we performed radiological examination of the patient with dorsal spine magnetic resonance imaging that revealed an extradural cystic lesion with significant spinal cord compression similar to the CSF signal. The lesion extended from T6 to T9

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(Fig. 1).

A laminectomy was made through T6 to T9 and a pouch of membrane resembling to dura-mater was found lying over the dorsal aspect of the dural sac. It contained CSF and after the cyst had been perforated and drained it refilled completely with CSF within seconds. Afterwards, a communication between the cyst and the arachnoid space was found and ligated. Then, the cyst was microsurgically resected and the specimen was sent for pathology analysis. A complete removal was performed (Fig. 2).

The patient had no complications after the surgery and was discharged from the hospital two days later. Follow- up MRI scans taken 2 months after the surgery shows the absence of medullar compression and no residual cyst was encountered (Fig. 3). The patient has completely recovered from her motor symptoms and has normal strength on both legs after 13 months of surgery.

The postoperative histological findings were compatible with the presence of an arachnoid cyst with few foci of calcification.

3. Discussion

EACs can develop intradurally or extradurally, most are located on the posterolateral aspect of the spinal canal, compressing the spinal cord toward the anterior aspect of the canal [13].

The dural defect is usually found near the nerve root sleeve. Some authors believe that stretching forces between the movable thecal sac and the relatively fixed roots are the major contribution factor [13].

Congenital malformations associated witha neural tube defect, inflammatory process of any kind and trauma are possible causes for arachnoid cysts, but the majority of the AC's are idiopathic. In these cases, the dural defect is considered congenital. We believe this is the case of our patient as we do not have any history of trauma, infection/inflammation or any other neural tube congenital malformation. According to Ogura et al, there are two types of idiopathic AC's: sporadic (isolated) and syndromic (familial) [9]. Ac's in familial form appears as part of Milroy's syndrome (spinal extradural arachnoid cyst, familial lymphedema, history of bronchial asthma, and a double row of eyelashes) [1] and is one of the phenotypes of lymphedema-distichiasis syndrome (LDS) caused by a mutation in the FOXC2 (Forkhead box protein C2) gene [9].

In the syndromic form it is frequent the finding of multiple cysts and tend to occur out of the thoracolumbar area. So, in the presence of multiple AC's we must consider that they are in the context of a syndrome [9].

It is also proposed that arachnoid cysts are developed by mechanical stress in the spine, such as seen in diffuse idiopathic skeletal hyperostosis (DISH), wedge deformity of the vertebral body and disc herniation [8].

Perret et al. postulated that arachnoid cysts are the result of widening of the septum posticum, which is a membranous partition in the dorsal thoracic spinal canal [2]. There are, still, some cysts that cannot be explained for any of these conditions.

There are three theories to explain the refilling of AC's:

By active secretion of fluid from the cyst walls made though active transportation. The liquid inside the cyst has a lesser concentration of proteins than the cerebrospinal fluid which supports this theory.
Elevation in oncotic pressure: proliferation and abnormal distribution of the arachnoid cells during the embrionary phase. These cells degenerate posteriorly causing an elevation in oncotic pressure inside the cyst.
Valve mechanism: anatomic communication between the cyst and the arachnoid space allowing CSF to flow and enlarge the AC with physiologic Valsalva maneuvers [1]

AC's are frequently asymptomatic but, may cause myelopathy, radiculopathy, and focal pain, depending on the size and location. They may present either with a rapid or a gradually progressive deficit. The symptoms are due to alterations in the pressure inside the cysts. Extradural arachnoid cysts can be confused with other cyst-like lesions, such asperineural cysts (Tarlov's cyst), neurenteric cysts, meningoceles, cystic neoplasms (dermoid, epidermoid and teratoid cysts), inflammatory cysts and cysticercosis [12,4].

Magnetic resonance imaging (MRI) is the standard for diagnostic workout. AC's are hypointense in T1 and hyperintense in T2 sequence, so is the CSF. There is no gadolinium enhancement of the cysts. MRI flow studies are ideal to localize the site of cerebrospinal fluid leakage. CT myelography with delayed scanning can be used in some cases where a MRI is contraindicated.

Surgery is the treatment of choice for this pathology, however, there are reported cases with long term follow-up in the literature for asymptomatic patients. Surgical indications are pain and neurologic deficits. There is no consensus on the best surgical technique, but we believe that complete resection of the cyst through laminoplasty or laminectomy must be the preferred strategy because it offers the relief of the pressure inside the spinal canal and do not cause spine instability. Other surgical methods includes shunting procedures, percutaneous image-guided aspiration, and minimally invasive endoscopic

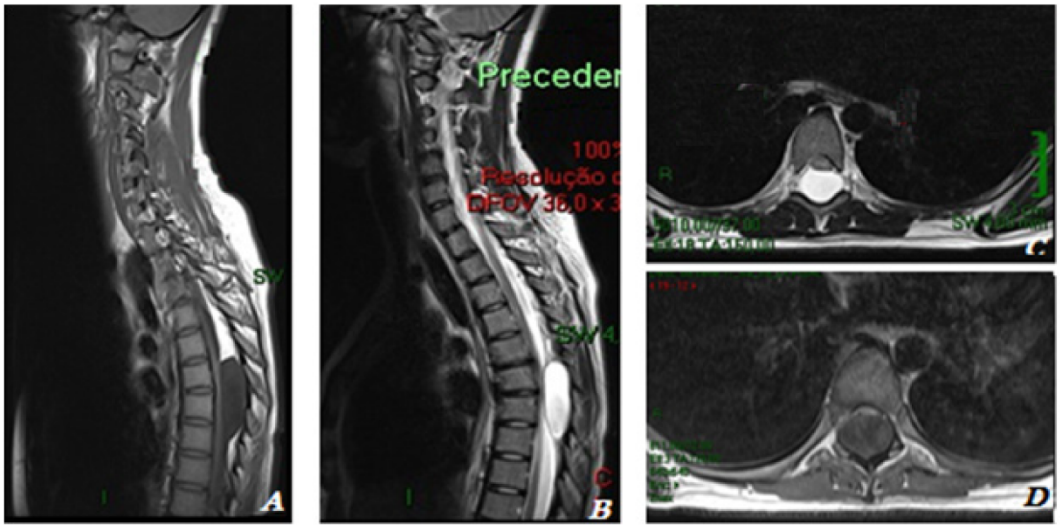

Fig. 1. Pre operative T1-weighted sagittal MRI (A), T2-weighted sagital MRI (B), T2-weighted axial MRI (C) and T1-weighted axial MRI (D). Observe how the lesion seems to be extradural on the posterior aspect of the dural sac and it has the same signal than the CSF.

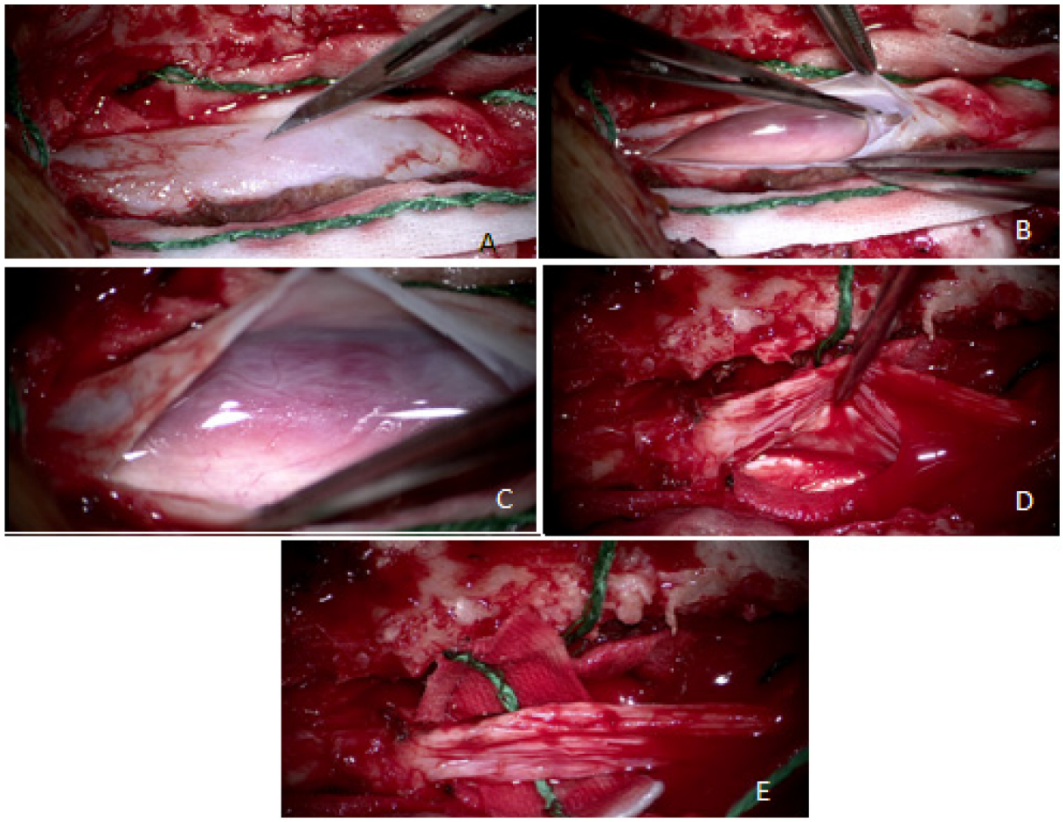

Fig. 2. After laminectomy a pouch of membrane resembling to dura-mater was found lying over the dorsal aspect of the dural sac (A). This pouch was opened and dissected revealing that its ventral wall was separated of the dorsal aspect of the dural sac (B–E).

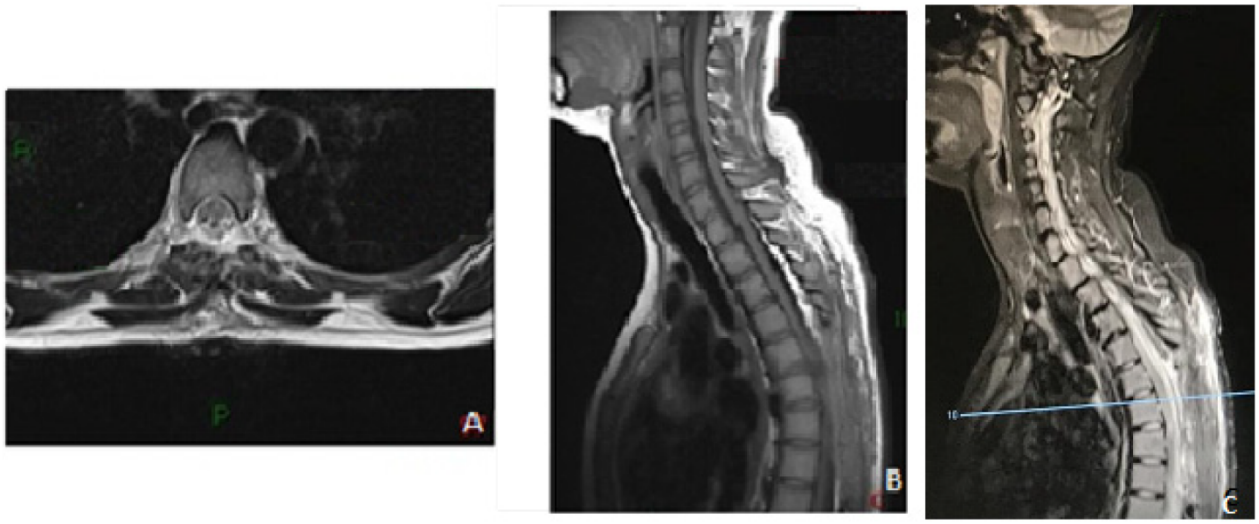

Fig. 3. Two month post operative T2-weighted axial MRI (A), T1-weighted sagital MRI (B) T2-weighet sagital MRI. Note the complete removal of the cyst.

approaches are rarely used [18], because It is believed that cyst excision with complete cyst wall resection, provides the least chances of recurrence [14]. We opted for a laminectomy in our case because it was mid thoracic and we have preserved the facets bilaterally which minimizes the odds of instability. The patient hasn't had any deformity after 1 year of follow up. However, we are following her closely for any sign of kyphosis.

Lee et al. analyzed 44 operations in a literature review of spinal extradural arachnoid cysts and observed that complete resection followed by the obliteration of the dural defect improves symptomatology in the majority of cases (recurrence in 2% of cases). However, if the dural defect was not addressed, the recurrence rate was high as 66.7% [3,1].

Other option includes fenestration and correction of the dural defect but these have worse results (3.6% of recurrence) [3]. Percutaneous cyst puncture using CT or MRI guidance can be performed but the cyst, frequently, reccurs following its puncture.

Long lasting symptoms are less responsive to treatment because of the spinal ischemic injury caused by the compression. Thus, neurologic recuperation depends on the size of the cyst and the time from symptoms

beginning to treatment.

Familial forms of AC's have poorer surgical outcome than sporadic forms [9].

4. Conclusion

In our clinical case, the patient had no history of trauma or arachnoiditis. There was also no evidence of trapping of the nerve roots (type 1A). The extradural cyst was over the posterior aspect of the dural sac and the communication between the cyst and the subarachnoid space was ligated and the cyst wall posteriorly resected. Postoperative histologic finding was consistent with arachnoid cyst showing calcification foci within collagenous fibers and membrane with flat lining cells.

The EAC is a rare but a rather important pathology to be known by all neurosurgeons since if it's addressed correctly and on time the patient will probably recover fully. Our patient has had an /excellent clinical and radiological outcome, with the recovery of all her deficits in the first and 6th month follow up consultation, she was ambulating and with normal strength on both legs.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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