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Extradural Arachnoid Cyst: An Uncommon Cause of Spinal Cord Compression in Children: A Case Report

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Abstract **Case Report**

The objective of this work is to highlight and discuss a rare cause of curable spinal cord compression with a favourable evolution, through the case of a posterior thoracic epidural arachnoid cyst for which we will discuss its clinical, paraclinical, therapeutic and evolutionary characteristics, with review of the literature. The patient was a six and a half year old boy with no previous pathological history. He was admitted for progressive gait problems with myalgia. MRI showed an extradural cystic lesion in the thoracic spinal cord opposite D5-D10 with a signal similar to that of the CSF. A laminectomy followed by evacuation of the cyst with partial removal of the wall was performed. Pathological examination confirmed the diagnosis. The evolution was marked by a total disappearance of the initial clinical symptomatology in the patient. Although rare, the extradural arachnoid cyst is a potential cause of reversible spinal cord compression for which surgical resection is the treatment of choice.

Keywords: curable spinal cord, extradural arachnoid cyst, diagnosis, myalgias.

1. INTRODUCTION

Arachnoid cysts are a very rare cause of spinal cord compression [1]. They are unusual in children. Their origin is unknown, although theories suggesting congenital, traumatic and inflammatory causes have been proposed [2]. Their location is variable and can be extradural, intradural or extra medullary [3]. Most often, they are asymptomatic and are discovered incidentally on routine radiological examination. Intramedullary arachnoid cysts with neurological expression are a very rare entity with few cases described in the literature [4]. We report a case of a symptomatic thoracic extradural arachnoid cyst in a boy revealed by signs of neurological deficit indicative of spinal cord compression. Surgical treatment was successful.

2. CASE REPORT

This is a male child, aged 6 and a half years, with no particular pathological history, who consulted for progressive gait problems that appeared 5 months ago, with myalgias and low back pain. On admission, the patient was apyretic, hemodynamically and respiratorily stable with dorsolumbar spinal stiffness. Walking was impossible, with paresis rated at 2/5 on the right side and 3/5 on the left side by muscle strength testing. The osteotendinous reflexes were preserved. There were no sensory or sphincter disorders.

Magnetic resonance imaging (MRI) revealed a cystic intraductal and posterior extradural spindle-shaped lesion, opposite the vertebrae from D5 to D10, in T1 hyposignal, T2 hypersignal, with a signal identical to that of the cerebrospinal fluid (CSF), not enhanced after injection of contrast medium (PDC) and without communication with the subarachnoid spaces, measuring 3cm in transverse diameter and 9cm in height. The latter enlarged the medullary canal at this level and placed the dorsal medulla against the posterior wall of the facing vertebrae.

A laminectomy of T3 and T4 exposed the cystic lesion, the contents of which were similar to those of the CSF. After opening the cyst and evacuating its contents, a partial excision of its wall waś performed.

The postoperative course was straightforward with a clear neurological improvement and a gain in muscle strength. A spinal MRI check-up showed a total regression of the arachnoid cyst with restitution of the normal volume of the initially compressed marrow.

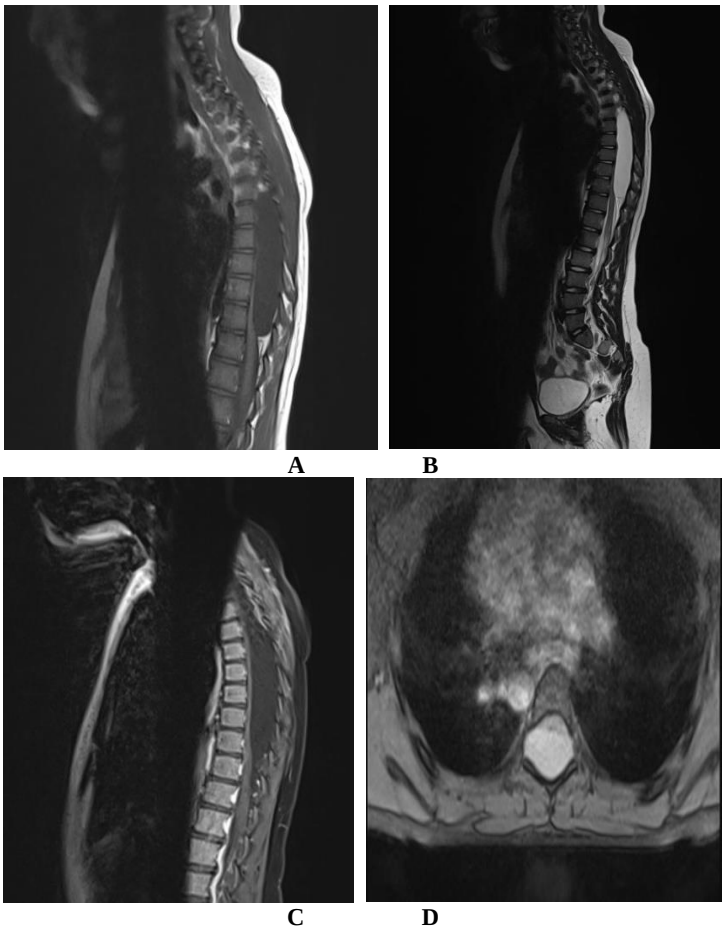

Figure 1: Spinal cord MRI in sagittal T1 (a), T2 (b), FATSAT (c) and axial T2 (d) slices showing a thoracic cystic formation, signal identical to that of the CSF, hypointense in T1, hyperintense in T2, unenhanced after injection of PDC, posteriorly localised and pressing the cord against the posterior wall of the vertebral bodies

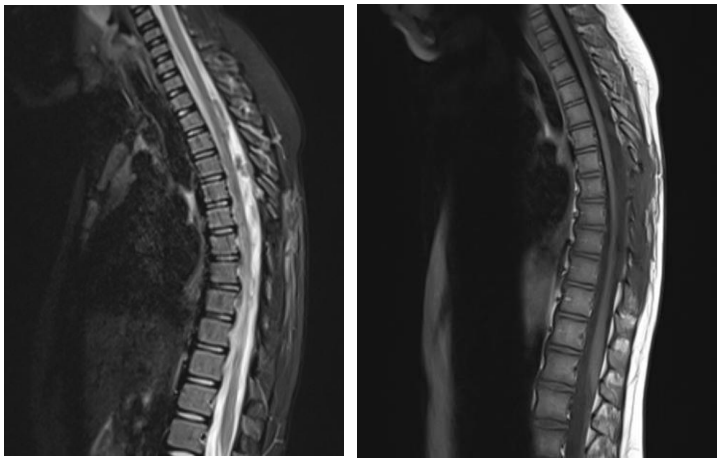

Figure 2: Spinal cord MRI in T1 and T2 sagittal slices showing total regression of the arachnoid cyst with evidence of musculoskeletal changes at the surgical site

3. DISCUSSION

Spinal epidural arachnoid cyst (SEAC) is a rare entity [5] that affects both sexes with a slight male predominance. Only a few cases have been described in the literature. Their origin is not clearly established, some authors suggest a congenital source, while others propose a traumatic or post-surgical origin secondary to an acquired dural lesion [6].

The mechanism of growth is also a subject of dispute; while some suggest secretion of CSF by the cyst wall, free osmosis of water and hydrostatic pressure, others have proposed a valve mechanism at the origin of expansion- regression of the cyst [6]. This theory states that intermittently the flow of CSF into the cyst promotes its expansion to a certain level where the cyst becomes large and would block the flow of CSF. By obeying La Place's law, the accumulation of blocked CSF will lead to an emptying of the cyst contents and vice versa. This valve mechanism was observed intraoperatively by Rohrer *et al.*, [6].

Histologically, there is a fibrous connective tissue made up of an arachnoid monocellular alignment which is sometimes absent on histological study.

Proposed in 1988 by Nahors MW *et al.*, [7], a classification divides cysts according to their location, site and nerve compression: Type I: extradural arachnoid cyst without nerve compression; Type II: extradural arachnoid cyst with nerve compression; Type III: intradural arachnoid cyst. In a few cases, the KAER may have an intradural extension [8]. For type I, a communication of the CSF between the cyst and the arachnoid space has almost always been found [9]. Non-communicating forms remain rare [10].

As for their ethiopathogenesis, hypotheses have been put forward. KAER is thought to be the result of arachnoid herniation through congenital aplasia of the dura mater [11] or due to congenital diverticula of the dura mater. A collagen anomaly of congenital origin would be at the origin of the structural failure of the dural framework which, by weakening, becomes loose, elongated and ectatic. The association of arachnoid cysts with certain pathologies such as multiple sclerosis, Marfan's syndrome, spinal dysraphia and syringomyelia would support the genetic origin [12].

Topographically, thoracic location is predominant (65%), followed by lumbar and lumbosacral location (13%), thoracolumbar location (12%). Cervical location represents only 3% of cases [13, 14]. The usual location is posterior or posterolateral. Clinically, the discovery is often fortuitous, with no notifiable signs.

An inaugural picture of spinal cord compression is rare and almost never described [15], revealing itself mainly by a progressive spastic paraparesis in the lower limbs with paresthesias. There may also be a rapidly progressive motor deficit. In sum, the presentation depends mainly on the level of compression with fluctuating symptomatology. However, rapid decompensation can occur and there is no correlation between the date of onset of symptoms and their severity because, for example, in the thoracic region, unlike the lumbar region, the size of the canal is smaller and therefore the expression will be more obvious. Similarly, the lumbar region is more vulnerable than the thoracic region because of the cauda equina roots [5].

Magnetic resonance imaging (MRI) is the reference examination for the diagnosis of spinal epidural arachnoid cysts. It allows a three-dimensional morphological study of the marrow, sub-arachnoid and epidural spaces. It specifies the location, size, partitioned, single or multiple cysts and the degree of spinal cord compression [15].

KAER is seen as a formation behind the spinal cord following the CSF signal on T1 and T2 weighted sequences. Gradient echo sequences with their excellent spatial resolution visualise the communication between the KAER opening and the subarachnoid space. Intravenous injection of Gadolinium allows the elimination of possible differential diagnoses such as neurenteric cysts, synovial cysts, dermoid cysts, epidermoid cysts, hydatid cysts or purely cystic tumours such as schwannomas and hemangioblastomas in their cystic form.

For asymptomatic forms, conservative treatment is recommended with regular clinical and radiological monitoring. For symptomatic forms, most authors agree on the surgical indication of complete excision of the cyst, followed by ligation of the pedicle communicating the cyst with the subarachnoid space and repair of the dural defect. This technique prevents recurrence of the cyst. The prognosis depends on the diagnostic and therapeutic precocity but also on the patient's condition.

4. CONCLUSION

KAER is a pathology to be known as a differential diagnosis of epidural cystic lesions. In our patient, early diagnosis by MRI followed by surgical decompression with total removal of the cyst allowed a resumption of physical activity with regaining of deficits.

Consent

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

Ethical Approval

"All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki."

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Authors' Contributions

All authors read and approved the final manuscript.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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