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Case Report

Thoracic Intradural Arachnoid Cyst: When to operate? Regarding a series of cases

Cisto Aracnóide Intradural Torácico: Quando operar? A propósito de um relato de dois casos

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ABSTRACT: Introduction: Historically, Schlesinger detected the first arachnoid cyst in 1893, and Spiller reported this in 1903. Arachnoid cysts (AC) are benign tumors that develop in the cerebrospinal axis of the arachnoid membrane common in people in the second decade of life, and predominantly in the middle and lower thoracic region (65%), in the lumbar and lumbosacral spine (13%), thoracolumbar (12%), sacral (7%) and cervical (3%). The intramedullary location of the AC is rare, with only a few cases reported in the literature. **Objective:** Describe a series of 2 cases of intradural thoracic AC, explaining and further elaborating on its treatment. **Materials and Methods**: Description of 2 cases of patients with thoracic intradural AC and including a literature review. **Discussion/Results**: Most ACs are uncovered incidentally and, therefore, asymptomatic. They remain constant in size, leading many physicians to recommend conservative treatment and periodic monitoring. On the other hand, the standard treatment for symptomatic thoracic intradural ACs involves surgical exploration and relief of neural tissue compression. Preferably before causing significant neurological deficits, as there is a greater chance of a satisfactory postoperative result. **Final considerations**: Thoracic intradural ACs, despite being rare, when symptomatic, require surgical treatment and should not be delayed due to the risk of progressive neurological motor deficit.

KEYWORDS: Arachnoid; Cyst; Intradural; Thoracic; Treatment.

RESUMO: Introdução: Historicamente, o primeiro cisto aracnoide (CA) foi detectado por Schlesinger em 1893, e o primeiro relatório foi de Spiller em 1903. Os CAs são tumores benignos que se desenvolvem no eixo cerebrospinal da membrana aracnóide sendo comuns em homens, na segunda década de vida, e predominantemente na região torácica média e baixa (65%), na coluna lombar e lombossacra (13%), toracolombar (12%), sacral (7%) e cervical (3%). A localização intramedular do CA é rara, com apenas alguns casos relatados na literatura. **Objetivo**: Descrever uma série de 2 casos sobre CA intradural torácico, explanando com maior enfoque sobre seu tratamento. **Materiais e Métodos**: Descrição de 2 casos de pacientes com CA intradural torácico e revisão de literatura. **Discussão/Resultados**: A maioria dos CAs são encontrados incidentalmente, sendo, portanto, assintomáticos, e permanecem constantes em tamanho, fazendo com que muitos médicos recomendem tratamento conservador e com monitoramento periódico. Já o tratamento padrão, para CA intradural torácico que se apresenta com sintomatologia, envolve exploração cirúrgica e alívio da compressão do tecido neural, de preferência, antes de causar déficits neurológicos importantes, pois assim há maior chance de um resultado pós-operatório satisfatório. **Considerações Finais**: CAs intradurais torácicos apesar de raros, quando sintomáticos, precisam de tratamento cirúrgico, não devendo ser protelado devido ao risco de déficit motor neurológico progressivo.

PALAVRAS–CHAVE: Cisto; Aracnoide; Intradural; Torácico; Tratamento.

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INTRODUCTION

Historically, Schlesinger detected the first arachnoid cyst (AC) in 1893, and Spiller did the first report in 1903[1]. ACs are benign tumors developing along the cerebrospinal axis in the arachnoid membrane, common among men in their second decade of life[2]. The intracranial is the most commonly affected region; however, when they are predominantly in the middle and lower thoracic spinal regions (65%), in the lumbar and lumbosacral parts of the backbone (13%), thoracolumbar (12%), sacral (7%), and cervical (3%)[3]. Furthermore, spinal CAs are present in most cases in the intradural/extramedullary location, yet they are extremely rare, as only a few cases are reported in the literature[4].

The intradural cyst generally consists of diverticula, full of fluid, which can be found incidentally or from clinical presentation from radicular or medullar compression[5]. The etiologies include inflammatory and infectious processes, tumors, and traumatic and congenital events[6]. Its formation occurs due to the accumulation of cerebrospinal fluid in the arachnoid layer, as the cyst wall is composed of flattened arachnoid cells made up of a thin translucent membrane that does not bring about a solid component or epithelial covering[7].

The intradural thoracic ACs are frequently asymptomatic and multiple. They are painful or myelopathic due to medullar compression when symptomatic[8] related to progressive weakness of the legs, paresthesia, neuropathic pain, scoliosis, gait disorders, and spasticity. These cysts can cause permanent motor deficits in rare cases; however, the postoperative prognosis is good if the operation is performed before neurological deficits occur[9].

The clinical and imaging studies perform the diagnosis, such as magnetic resonance imaging (MRI), the gold standard, as it is a non-invasive method and provides a high degree of sensitivity and accuracy. It can determine the location of communication between the subarachnoid space of the spine and the cyst[10]. The differential diagnostics of the intramedullary cystic lesions include intradural/extramedullary lesions, such as squamous cells, synovial cysts, schwannomas, and mainly the arachnoid membrane) a squamous cell cyst and synovial cyst[11].

This article aims to describe two cases of thoracic intradural ACs, focusing further on their treatment.

METHOD

The two cases of patients with AC were described, and the literature revision was searched in PubMed/MEDLINE, SciELO, and Bireme/LILACS. In MEDLINE, the authors utilized ("arachnoid cyst" AND "thoracic intradural") as descriptors, searched in "Descritores em Ciências da Saúde" (DeCS) (Health Science Descriptors). In SciELO and Bireme, they used ("arachnoid cyst" AND "thoracic intradural") searched in DeCS. The researched articles were selected based on the criteria as systematic literature reviews, epidemiological studies, and a series of case reports in English, Spanish, and Portuguese languages published in the last ten years. The abstracts were read in all the articles and selected based on the best approaches to thoracic intradural arachnoid cysts. Sixteen articles were found in the PubMed database search, three articles in Scielo, and forty-one in Bireme. After the inclusion criteria were applied and duplicated, unrelated and unavailable articles were excluded, and those that did not address the theme satisfactorily; a total of thirteen articles adhered to the acceptable article constraints.

In April 2020, the patients went to "Hospital Santa Marta" (Santa Marta Hospital) (HSM) in Brasília for evaluation. After clinical analysis, the exams were diagnosed as thoracic intradural arachnoid cysts, accompanied by treatment and case monitoring. The cases and subject were discussed in January and February 2021, then the "Termo de Consentimento e Livre Esclarecimento" (Informed Consent Form) (TCLE) was agreed to by collecting the patients' signatures in April of that same year. After that, the following study began to be performed and written.

RESULTS

Case 1

A male 38-year-old patient, who was a stock clerk without comorbidities, complained of back pain and fatigue in his lower limbs at his appointment that had worsened in the past twelve months. Those symptoms were accompanied by progressively evolving lower limb paraparesis in the last few weeks. He also suffered from gait instability and proprioceptive alterations compatible with the impairment of the posterior funiculus and the Babinski sign observed in the physical exam. An MRI was performed to detect the presence of an expansive intradural/extramedullary lesion caused by the compressive effect on the spinal cord and signs of myelopathy. The initial diagnosis was based on a cystic spinal tumor of congenital origin. Laminectomy was the recommended surgical treatment on T7-T8, as a cystic lesion was observed after opening the dura-mater extending to two levels, the transparent wall, and the hyalin content. Thus, it was necessary to perform a medullar decompression by fenestration of the cystic tumor throughout its full extension and then to remove fragments from the wall and submit them for anatomopathological evaluation. At the end of the procedure, the patient did not display any motor or intraoperative neurophysiological alterations.

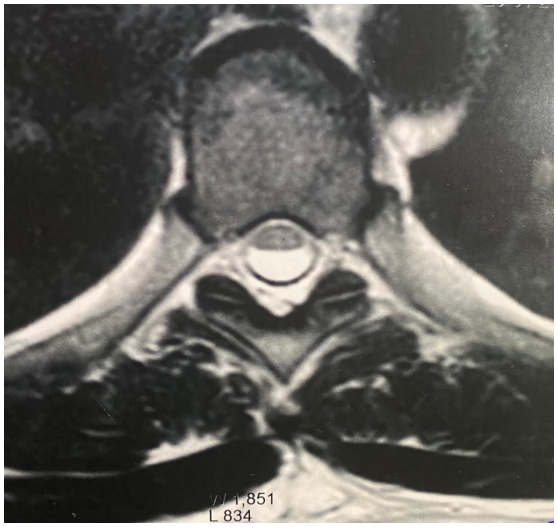


Image 1 - The Preoperative Magnetic Resonance Image: One can observe a widened area of the dorsal subarachnoid space of the vertebral channel at the height of the distal portion of the T7 vertebral body, compressing the posterior surface of the marrow. It measures about 2 cm in caudal cranium diameter. There is even a signal alteration in the marrow because of probable compression.

Case 2

A female 37-year-old patient, presented cervical brachial and chronic pain arising from breast cancer treatment in 2017 when she underwent chemotherapy and radiotherapy. Her cervical and thoracic spine MRI was performed in October 2018, which displayed discal protrusions from C3 to C7, hemangioma in T9, and an intradural and extramedullary cyst measuring T4 13x7x10 mm in size touching the spinal cord, but without any signs of myelopathy. Although the cystic tumor had been uncovered incidentally, based on this context, a conservative treatment approach was selected for outpatient follow-up, as it showed no signs of myelopathy or symptomatologic characteristics. Another MRI was performed in April 2019 without any relevant changes compared to the previous exam; thus, the conservative approach was continued. In August 2019, another MRI was performed that uncovered a hemangioma at T9 and a ventral thoracic spinal cord dislocation from T4 to T6, previously almost touching the dura mater, without signs of medullary herniation or dural defects, with conservation of the fluid flow, and besides there was also a posterior indentation of the spinal cord in T4 like the "scalpel sign" but without any suggestive signs of myelopathy. Afterward, the patient returned complaining about pain in the interscapular region, however, it was more intense on the right side, as she suffered approximately two crises daily, lasting from two to three minutes. In March 2020, the patient complained about posterior thorax pain irradiating interscapularly throughout the entire innervation territory compromised by the intradural cyst. Thus, another MRI was performed that did not display any relevant alterations related to the previous exam on April 5, 2019; however, as the patient presented a more serious condition and evident symptomatology, surgical treatment was indicated for decompressing the cystic tumor by fenestration to achieve improved pain relief and provide an enhanced prognosis.

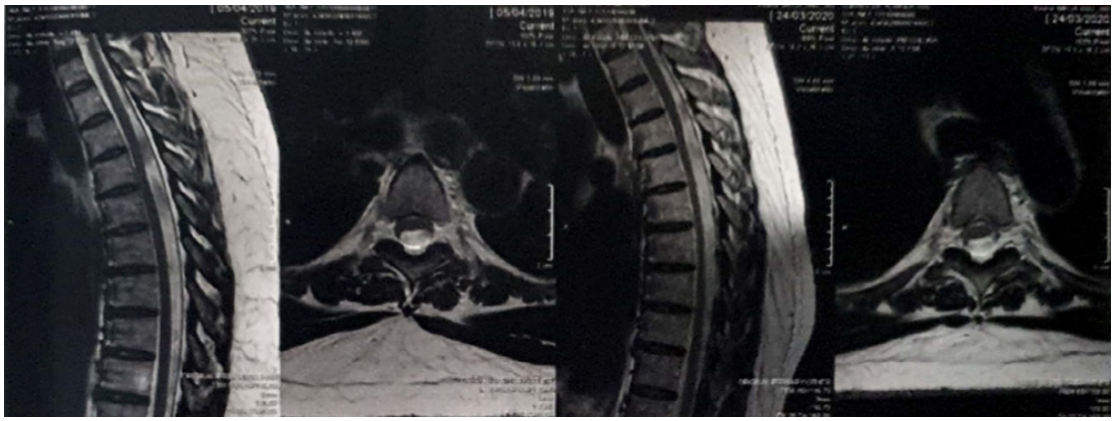


Image 2 - Preoperative Magnetic Resonance Imaging: Focal ventral shifting of the thoracic spinal cord was observed from T4 to T6, almost touching the dura mater anteriorly, without signs of modular herniation or dural defects. The posterior fluid space is focally widened to the T4 level. There is an indentation in the posterior spinal cord at the T4 level, like the "scalpel sign," which suggests the possibility of a dorsal arachnoid membrane. Furthermore, a hemangioma was confirmed in the vertebral body at T9. There were no relevant changes in the findings from the MRI performed on April 5, 2019; however, the patient presented a worsened condition and characteristic symptomology.

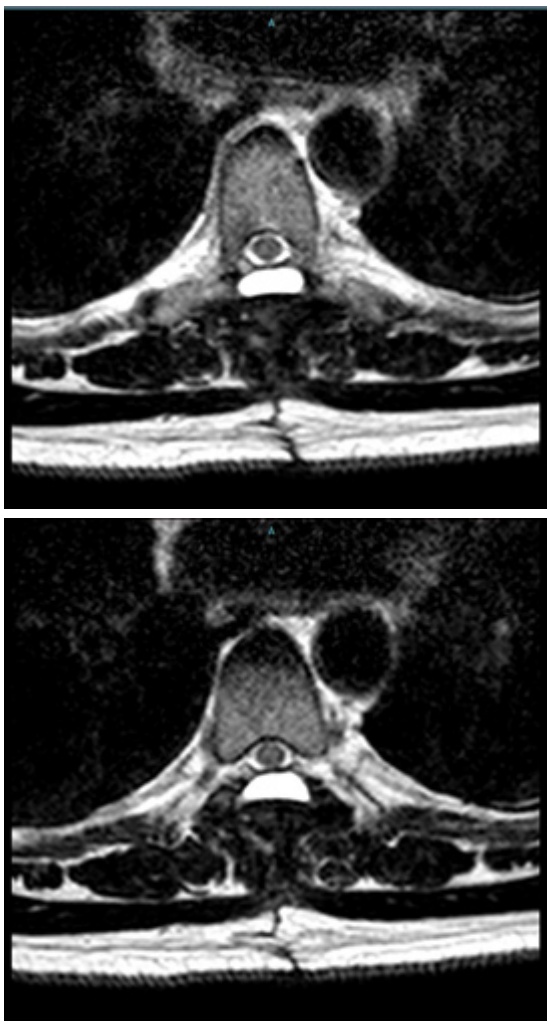
DISCUSSION

Most ACs are found incidentally by backbone imaging exams and remain constant in size. The MRI is the gold standard exam for this type of pathology, making many physicians recommend conservative treatment

as period monitoring, as was initially performed for the case 2 patient[12]. However, in case 1, as there was a progressive neurological deficit, gait instability, and the risk of irreversible, progressive spinal cord lesions, he was submitted to an urgent surgical treatment regime that consisted of laminectomy by dural opening and cyst fenestration[13] to resolve the neurological symptoms.

The standard treatment for thoracic intradural AC, present with symptomatology, involves surgical exploration and compression relief of the neural tissue, preferably before causing significant neurological deficits, as there is a greater chance of achieving satisfactory surgical results[5].

Case 1



Images 3 and 4: Postoperative Magnetic Resonance Imaging: evolutive control was proven after surgical handling by resectioning the posterior elements of T6 and T7 and the partial spinal process of T5. Oval-shaped collection in the posterior epidural space at the T6 and T7 levels (surgical site) that compresses the posterior surface of the dural sac. The focal tapering with the anterior shifting of the thoracic spinal cord at the T7 level and the signal change at the T8 level, these findings are probably related to the previous compression event.

In case 1, the cyst was not an incidental finding, as the patient had already presented at the appointment from neurological symptoms that suggested spinal cord compression, as the MRI confirmed that finding. Afterward, to avoid more serious complications and improve the patient's prognosis, a laminectomy was performed for spinal cord decompression that achieved the desired effect and the patient improved, and the presented neurological symptomatology ceased.

Case 2

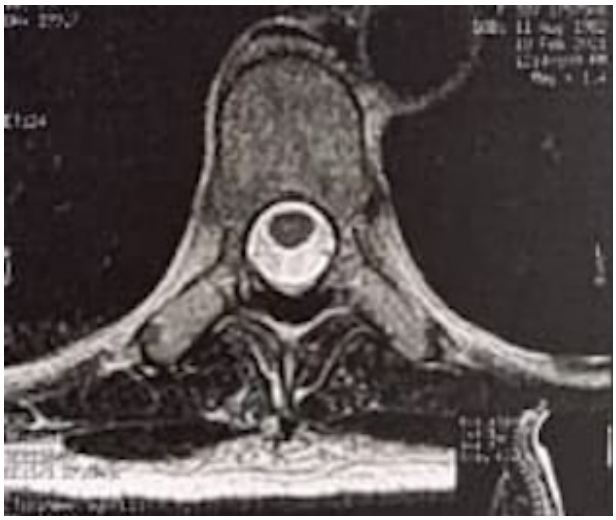


Image 5 - Postoperative Magnetic Resonance Imaging: a laminectomy was performed on T4 and T5 to collect extradural fluid on the laminectomy level corresponding to the seroma. There was a slight degenerative change in the thoracic backbone and slight edema in the paravertebral muscular at the surgical site.

In case 2, the cyst was found incidentally and initially followed up with conservative measures for almost two years, as the patient's cyst did not increase in size, nor was there neurological symptomatology. However, after that period, the patient started to present neurological deficits, which changed the medical conduct, as it was necessary to perform surgery for cystic tumor decompression by fenestration. That resulted in improved prognostics for the condition, as the MRI showed post-surgery that the procedure was performed for spinal cord decompression, causing the symptoms.

In both cases, the post-surgical MRI proved the presence of epidermal seromas, which are normal findings from surgical manipulation of the anatomic field utilized for recession/fenestration.

FINAL CONSIDERATIONS

Thoracic intradural ACs, although they are rare when symptomatic, they require surgical treatment. That cannot be delayed due to the risk of imminent progressive

neurological deficit. Eventually, urgent surgical treatment must be recommended to avoid increased motor loss and consequently improved the quality of life. The reported cases demonstrate surgical recommendations effectivelyt for patients who have already showed symptoms and those initially followed up as outpatients.

Conflict of interest: The authors do not declare any conflicts of interest.
Authors' participation: Sara Araújo de Medeiros Mendes – drafted the introduction, methodology, and results; Hyale Melo Lima – drafted the discussion, final considerations, and references; Ronaldo Borges Tonaco– collected and analyzed the clinical cases; William Antônio Quirino – analyzed and interpreted the clinical cases; Jobe Petter – research study advisor and final text revisor.

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