Clinical Case Report

Dorsal intramedullary giant dermoid tumor: Case Report

Bruna Mendes Lopes Meira[1], Carla Larissa Cunha Sottomaior[1], João Gabriel de Melo Silva[1], Luís Henrique Rossignolli Almeida Prado de Oliveira[1], Sophia Rodrigues Molina[1], Ricardo de Amoreira Gepp[2]

[1] School of Health Sciences - ESCS, Brasília, Brazil

[2]Coordinator - Sarah Network of Rehabilitation Hospitals, Brasília, Brazil

To whom correspondence should be addressed: Bruna Mendes, MD

e-mail:

brunamendesl@outlook.com

Available at:

http://www.archpedneurosurg.com.br/

Intramedullary dermoid tumors are rare benign neoplasms that correspond from 1 to 2% of all intramedullary tumors, affecting specially the lumbosacral region. They are composed by remnants of embryonic tissue derived from the ectoderm. Despite its benign character, neurological injury occurs as a consequence of its expansive process and the collateral damage derived from the complete resection of the cyst wall, which can cause hypoesthesia and radicular pain, in addition to other effects such as neuromuscular scoliosis. The primary treatment for intramedullary tumor is resection. The case reported involves an 8 years old child with an extensive dorsal intramedullary dermoid tumor, presenting clinically complete paraplegia, lower limbs atrophy, hypoesthesia below the T4 level and urinary incontinence. The physical examination revealed upper motor neuron syndrome in lower limbs, mild cognitive delay and significant scoliosis. Image test showed a severe scoliosis and an expansive intramedullary extending from C5 to the T9 plane. The patient underwent extensive cervical and thoracic laminotomy followed by median myelotomy and the resection of the lesion. An arthrodesis treated the secondary vertebral deformity. Postoperative period showed good surgical recovery and control exams revealed ample resection of the lesion and adequate control of scoliosis with partial maintenance of rotational deformity. The purpose of this report is to describe a rare condition for a better understanding of the clinical features, to highlight the importance of an early diagnosis and approach and to serve as clinical evidence for future studies.

Keyworks: Vertebral deformity; Spinal cord neoplasms; Intramedullary dermoid tumors; Dermoid cysts

INTRODUCTION

Intramedullary dermoid tumors are rare neoplasms, corresponding to about 1 to 2% of all intramedullary tumors at all ages.[1] The most frequently affected area in dermoid tumors is the lumbosacral region, involving the cauda equina and the medullary cone, corresponding to about 60% of the cases.[2] These lesions are benign, but with a growth potential similar to a true neoplasms and, thus, can cause severe neurological damage, especially when not treated at the right time and in the correct way. [3]

The dermoid cyst is essentially composed of remnants of embryonic tissue, derived mainly from the ectoderm, during the closure of the neural tube in the third to fifth week of development [4]. The vast majority of these changes are associated with spinal dysraphisms, but there are less frequent forms associated only with the dermal sinus (occult spina bifida). The clinical presentation is characterized by the presence of neurological deficit. Secondary neuromuscular scoliosis can also occur, which causes a great impact on the quality of life of those affected.

Received 11 August 2020 / Published 24 September 2021

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The authors describe a rare case of a child with an extensive dorsal intramedullary dermoid tumor in association with spinal dysraphism, who underwent surgical resection of the lesion and arthrodesis to treat secondary vertebral deformity.

CASE REPORT

Male patient, 8 years old, had complete paraplegia for 3 years, with atrophy in the lower limbs, hypoesthesia from the T4 level and urinary incontinence. The development of the paraplegia from the beginning is unknown due to judicial secrecy, since the previous history of the patient includes violence and loss of child custody.

On physical examination, presented spastic paraplegia with lower limbs hyperreflexia, a positive Babinski sign bilaterally, spasticity in the lower limbs, significant scoliosis and a small surgical scar resulting from a surgery performed five years ago to remove a skin lesion in the dorsal region, which could be suggestive of dermoid spinal sinus, but there are no information or previous diagnostic of the patient. In addition, a mild cognitive delay was noted.

Column radiography for scoliosis evaluation revealed a Cobb angle from T3 to L2 with 115 ° sitting and 68 ° with traction. Magnetic resonance imaging (MRI) of the total spine showed massive expansive intramedullary lesion extending from C5 to the T9 plane, widening the spinal cord and causing distension on the posterior edge of vertebral bodies, characterized by hypointense T1 with intervening septa, heterogeneous hyperintense T2 and predominantly

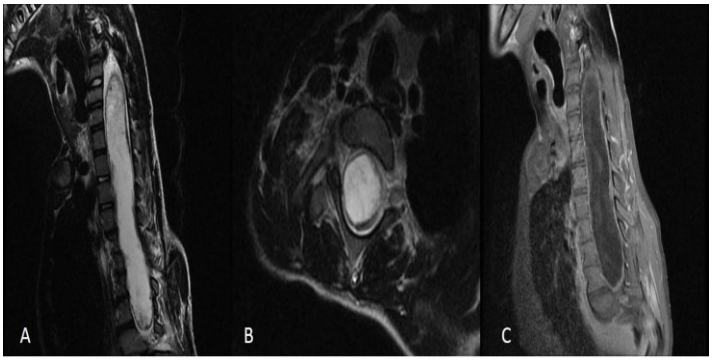

Figure 1- MRI showing a large intramedullary lesion. (A) Sagital view showing a large tumor inside spinal cord in T2 picture. (B) Axial section in

T2 peripheral by contrast medium. A small hyperintense in T2 was also observed in the medullary portions immediately above and below the expansive formation of the spinal cord, respectively in the plane of C4-C5 and T9-T10 (Figure 1). He did not present characteristic perpetuity of dermal sinus when the tumor surgery was performed. He presented a small skin scar, but as the patient lived in a support institution he had no report of what had been done previously.

The patient underwent extensive cervical and thoracic laminotomy. After the removal of the laminae, an opening of the dura mater that communicated the intradural space with the extradural space was observed, being suggestive of a remnant of dermal sinus. Neurophysiological monitoring was not performed due to the fact that the patient had no active movement of the lower limbs (paraplegia). The bleeding from the surgery of the tumor was 180ml, and 600ml from spinal arthrodesis, with 380ml of concentrated red blood cells replaced.

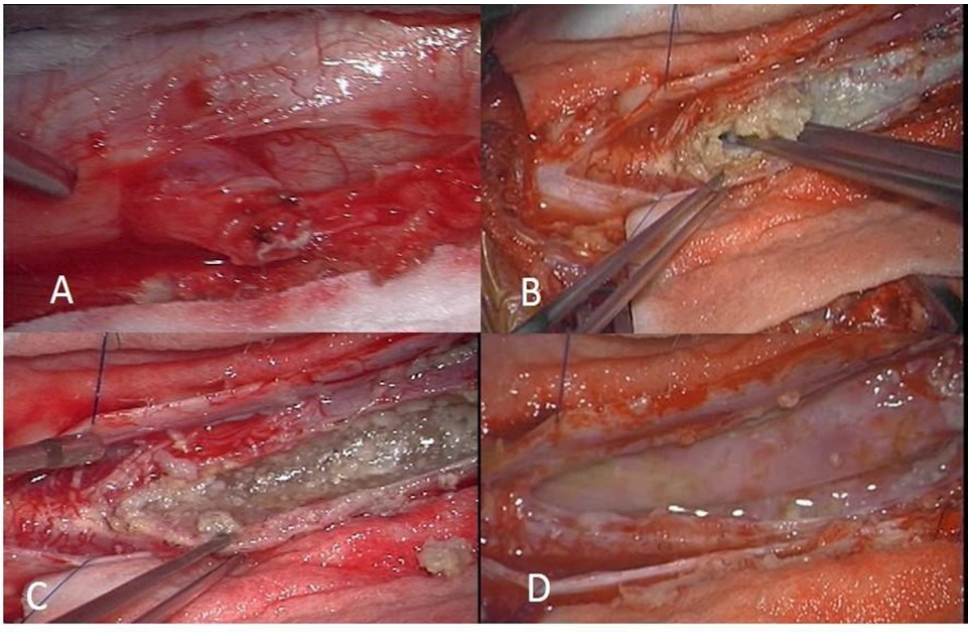

Figure 1: Surgical pictures showing the myelotomy and the resection of the dermoid tumor. After the resection the final aspect of spinal cord without tumor.

DOI: 10.46900/apn.v3i3(September-December).53

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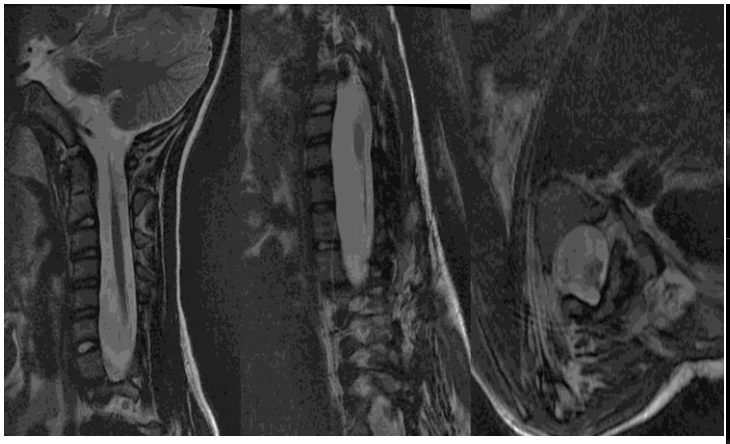

Figure 3 - Postoperative MRI in both sagital (A and B) and axial (C) views showing no residual dermoid cyst.

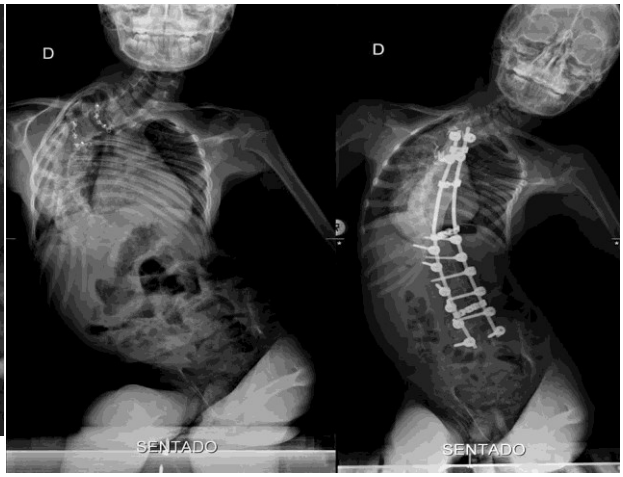

Figure 4 - X-ray pictures comparing the scoliosis before and after the tumor resection and spine fusion

Median myelotomy was performed and the presence of an intramedullary tumor with classic features of a dermoid lesion was confirmed, later corroborated by histopathological examination. Complete resection of the lesion was performed with removal of the cyst-forming epithelium and the perimeter that communicated the spinal cord lesion with the extradural space was resected (Figure 2). The laminae were repositioned using titanium miniplates and wires. After resection, scoliosis surgery with fixation was performed, using transpedicular hooks and screws. Partial correction of the patient's coronal deformity was possible.

In the postoperative, the patient evolved well clinically and showed good surgical recovery but maintained spastic paraplegia. The patient then underwent a spinal cord rehabilitation program.

Control exams showed ample resection of the lesion and adequate control of scoliosis with partial maintenance of rotational deformity (Figures 3 and 4).

DISCUSSION

Dermoid tumors are a rare condition. According to the Central Brain Tumor Registry of the United States, this primary spinal cord tumor appeared in the frequency of 0.19 per 100000 person-year from 1997 to 2001 [16]. This incidence has a bimodal behavior, with peaks between 0-4 years old and 15-19 years old. In addition, in the analysis by Arseni et al., which included 12 cases between 1935 and 1976, no difference of gender was observed, 59% were associated with congenital malformations [12,13]. The predilection for the lumbosacral region is explained by the later closure of the caudal neuropore, being the last to close in the formation of the neural tube [14].Dermoid tumors are due to an incomplete separation between the ectoderm and the neuroectoderm, causing the presence of ectodermal inclusions between these two layers, which may be congenital or acquired [5,14]. This error usually occurs between the third and fifth weeks of embryonic development [4], and subsequent differentiation of this fragment would originate the dermoid tumor [5]. The insertion of cutaneous residues during surgical procedures in patients with dysraphism has also been suggested as a possible explanation for the appearance of this type of pathology [5,6]. It has a thin wall with stratified squamous epithelium, having a content composed of viscous fluid material, keratin, epithelial remains and some epidermal appendages, such as sebaceous and sweat glands, and may also have teeth [2,7]. The leakage of the cyst in the subarachnoid space can install non-infectious meningitis [3]. The wall of the Dermoid tumor secretes contents that increase its volume, explaining its slow growth. Although this content can be easily removed, complete resection of the cyst wall has led to severe nerve injury, given the proximity and adherence to the spinal cord. Thus, remnants of the cyst wall after resection may explain its high recurrence, although these mechanisms are still not well understood [10].

The clinical picture varies in relation to the location and extent of the tumor, resulting from the injury caused by the compression of adjacent structures [4]. Symptoms are usually nonspecific and of late onset. Children may experience pain, motor regression and weakness [4], in addition to developing other conditions such as sensory disorders and urinary dysfunction [8,9]. It is presented more frequently in their chronic form, with radicular pain or neurological deficits over a long period. When it ruptures and its contents leak, there is a more acute presentation,

DOI: 10.46900/apn.v3i3(September-December).53

such as meningitis, which can be represented by intractable pain or symptoms of degenerative lumbosacral stenosis (DLSS) [14,15]. The diagnosis is made by means of histopathological analyzes, with magnetic resonance imaging being the radiological test with the greatest capacity to detect findings suggestive of a dermoid tumor [4].

The later the tumor is diagnosed, the greater the compressive condition, and the most likely the symptoms are to appear [4]. Furthermore, its uncontrolled growth increases the odds of leaking and presenting the acute presentation [14,15] and also the likelihood of leaving remains after the surgery due to its large volume [11].

The primary treatment for all kinds of intramedullary tumors is surgery and a crucial factor for the recurrence rate is the volume of resection. [11] Since most patients are symptomatic in view of the mass effect, symptoms tend to decrease after surgery, although they could recur if the tumor grows back[1]. This case is one uncommon intramedullary spinal cord disease, and the authors described all the evolution and the deformity correction.

CONCLUSION

Although being a rare condition, the intramedullary dermoid tumors can be presented with symptomatology of other diseases that affect the nervous system, including clinical manifestations such as hypoesthesia, sphincter deficits and upper neuron motor syndrome, emphasizing the importance to include this disease as a differential diagnosis.

The relevance of the surgery approach description is in the proximity and adherence to the spinal cord, raising the chance of secondary injuries despite the benign character of the dermoid tumor. Thereby, if the tumor is not totally removed, the recurrence rate is high. As well, its recrudescence is one of the biggest challenges when managing these lesions, making the evolution description as important.

It is important to highlight that had the patient been diagnosed and had the correct management of the dermoid spinal sinus been offered, he would probably not present the severe neurological deficit and scoliosis. Hence, these consequences could be avoided if the approach had been made earlier.

Therefore, since it's a rare condition which causes a great impact on the quality of life, this article aims to be a clinical evidence for future studies to better understand and approach this disease.

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DOI: 10.46900/apn.v3i3(September-December).53