Article published online: 2023-10-10

Multiple Medullary Schwannoma Associated with Lumbar Disc Herniation and Cauda Equina Syndrome: Case Report

Schwannoma medular múltiplo associado a hérnia de disco lombar e síndrome da cauda equina: Relato de caso

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Arq Bras Neurocir

Abstract

Introduction Schwannoma is a Schwann cells neoplasm that can occur in various parts of the nervous system, including the medullar region of the cauda equina. Bone involvement is uncommon.

Objective To report a multiple schwannoma case that was initially diagnosed as cauda equina syndrome.

Clinical Case We report the case of a 44-year-old male who presented with pain irradiating to the lower limbs, urinary retention, and saddle paresthesia. Imaging exams revealed tumors in the cervical spine and in the T12/L1, L2/L3, and L3/L4 levels, the last three causing nervous compression. Histopathological analyses confirmed a Schwannoma pattern.

Results The case was solved by surgical resection.

Conclusion The case was solved and, in the end, the patient was stable and with improved pain and waits for a cervical surgery.

Keywords
- schwannoma
- neoplasm
- lumbalgia

Resumo

Introdução Schwannoma é uma neoplasia de células de Schwann que pode ocorrer em diversas partes do sistema nervoso, incluindo a região medular da cauda equina. O acometimento ósseo é incomum.

Objetivo Relatar um caso de schwannoma múltiplo que foi diagnosticado inicialmente como síndrome da cauda equina.

Relato de caso Relatamos o caso de um paciente masculino de 44 anos que apresentou quadro álgico com irradiação para os membros inferiores, retenção urinária

Palavras-chave
- schwannoma
- neoplasia
- lombalgia

received
September 16, 2021
accepted
January 18, 2022

DOI https://doi.org/10.1055/s-0042-1744111.
ISSN 0103-5355.

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Thieme Revinter Publicações Ltda., Rua do Matoso 170, Rio de Janeiro, RJ, CEP 20270-135, Brazil

e parestesia em cela. Exames de imagem revelaram tumores na cervical e em níveis de T12/L1, L2/L3 e L3/L4, com os três últimos causando compressão nervosa. A análise histopatológica comprovou padrão de schwannoma.

Resultados A correção do quadro se deu através da ressecção cirúrgica.

Conclusão O quadro foi resolvido com o paciente apresentando-se estável e com melhora do quadro álgico. O paciente aguarda cirurgia a nível cervical.

Introduction

Schwannoma, also known as neurilemoma or neurimoma,[1,2] is a neoplasm that occurs in Schwann cells, which are responsible by the formation of the myelin sheath of the peripheral nerves. In most cases, schwannomas are benign,[3] affect more women than men,[4] and are more frequently seen in the 40th and 50th decades of life. Its incidence is of 0.5 per 100.000 individuals per year.[3] Their most common location is the vestibulocochlear nerve (NC VIII), but they can also affect other nerves.[5] Multiple schwannomas are usually be associated with neurofibromatosis (type I or II) or with schwannomatosis,[3] which are rare conditions. The clinical manifestations are late, due to their slow growth, and occur due to neurological compression.[6]

The aim of the present article is reporting a multiple schwannoma case that was initially diagnosed as cauda equina syndrome.

Material and Methods

Information was collected from medical records, patient interview, the diagnostic methods to which the patient was submitted, surgical findings, and a literature review.

Case Report

Patient, C.L., male, 44 years old, white, bricklayer, presented to the hospital due to a 2-month-long backache.

The backache irradiated to the lower limbs and was associated with urinary retention, saddle paresthesia, reduced sensitivity, and paraparesis. At the first evaluation, a diagnosis of cauda equina syndrome was made. Magnetic Ressonance (MRI) (►**Fig. 1** and ►**Fig. 2**) demonstrated lesions corresponding to C6/C7, T12/L1, L2/L3, and L3/L4 discal herniation. The patient was referred to urgent surgical medullar decompression (L3-L4 laminectomy). During the intraoperative evaluation, the lesion was suggestive of tumoral etiology. The collected tissue was sent to biopsy. The pathological report confirmed an intradural lesion with a suggestive Schwannoma pattern. A subsequent magnetic resonance imaging (MRI) exam showed that the discal herniation was associated with intradural tumoral lesions.

A subsequent surgical approach was made to resect the T12/L1 and L2/L3 lesions and revise the L3/L4. level After the surgery, the patient presented pain improvement.

Currently, the patient is stable, walking normally, without new deficits and with normal neurological examination. The cervical schwannoma is under strict monitoring for the best moment for surgery.

Discussion

Schwannoma is a benign tumor of the nervous sheath[3] that usually appears isolated, but it can also occur more rarely in multiple form with slow growth,[7] with consequent delay to present symptoms,[6] which happens when it reaches 4 cm in size.[8] The classic symptom is pain, which worsens at night,[9] associated with paresthesia due to compression of sensitive roots.[10] Moreover, the patient can also present with other sensitive and motor changes.[7]

Usually, the roots of the back are the most affected location. Generally, the tumors are intradural,[10] extramedullary ($\sim$ 99%), and most are found in the cervical and lumbar regions. In computed tomography (CT), schwannomas are not distinguishable from neurofibromatosis or discal herniation. However, in MRI, schwannoma have characteristic signs, which are hypointensity in T1, intense improvement in T1 + C, and heterogeneous hyperintensity in T2; generally, large tumors appear with hemosiderin areas in T2 sequences.[11] Normally, schwannomas appear as solid, well-defined rounded lesions[10] and are frequently associated with bone remodeling.[11] As they grow, they can extend to several levels of the spine or protude to the neural foramen. Macroscopically, these tumors are encapsulated and arise eccentrically to the nerve, with nerve fibers along their surface.

Fig.1 C7 tumor showed in MRI T2-weighted.

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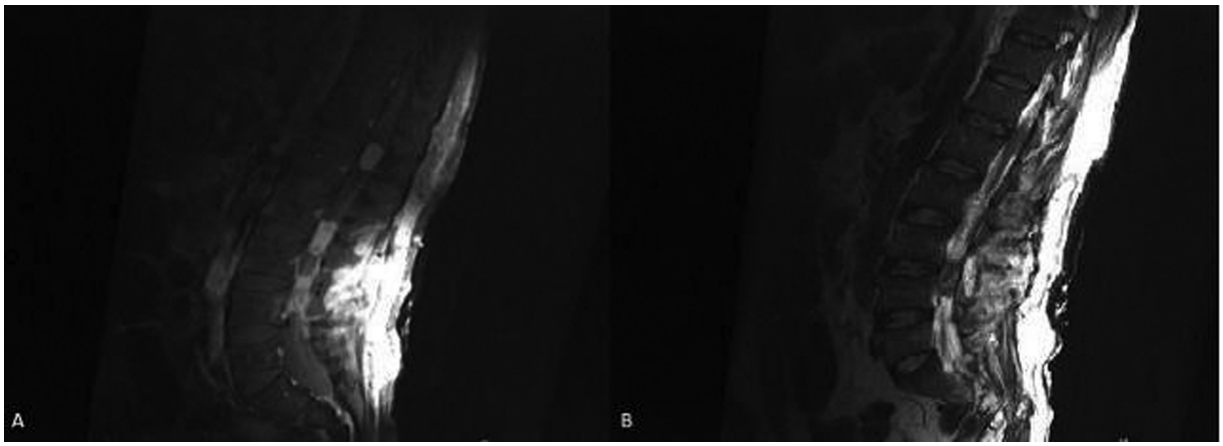

Fig. 2 . Massas em T12/L1, L2/L3 e L3/L4 reveladas por ressonância magnética (A, B).

Histopathological analyses enable confirmation of the diagnosis trough fusiform cells, which can be type Antoni A or Antoni B. The pattern Antoni A consists of elongated cells that are thickly involved and organized into fascicles, and alternating palisades with Verocay bodies can occur. Type Antoni B is less compact and tends to cystic degeneration.[12]

Among the differential diagnoses, the most common are neurofibromatosis type 2 (NF2) and neurofibromatosis type 1 (NF1). Neurofibromatosis type 2 is an autosomal dominant disease, progressive and rare,[13] that usually affects the vestibular nerves and originates cranial and multiple spinal tumors.[14] The schwannoma phenotype is very similar to NF2; nevertheless, the former has a good prognosis and the latter does not. Therefore, differentiation between these tumors is necessary in order to determine the right treatment.[15] Neurofibromatosis type 1 is also an autosomal dominant neurocutaneous disorder linked to the formation of benign and malignant tumors; however, it has a different phenotype from NF2. Neurofibromatosis type 1 can present coffee latte stains, neurofibromas in any type, axillary and inguinal freckles, optical nerve glioma, Lisch nodules (pigmented iris hamartomas), typical bone disease lesions, such as small wing sphenoidal dysplasia, thinning of the cortex of long bones with or without pseudarthrosis, and family history.[14] Neurofibromatosis type 1 is linked to the *NF1* gene, which is located in the long arm of chromosome 17 (17q12).[10]

Schwannoma can also be confused with disc herniation. Both can lead to neurological compression, originating the symptoms.[16] The most common locations for herniation are the C6/C7 level in the cervical spine, the L4/L5, and L5/S1 levels in the lumbar spine.[17] The latter causes cauda equina syndrome,[18] just as a medullary tumor in this region would.[19]

Disc protrusion is more common than schwannomas, being the disease that most leads to spinal surgery today. It occurs more in males in the 40th decade of life.[20]

The first line of treatment for schwannomas is surgical. Since the tumor does not infiltrate the originating nerve, it can be totally separated, without nerve lesion.[12]

Conclusion

Spinal schwannoma is a rare neoplasm with unspecific clinical presentation for which the most used imaging exam is MRI. Confirmation of the diagnosis is obtained by histological and immunohistochemical studies. The treatment is surgical and its aim is complete resection. Incomplete resection can lead to future recurrence.[3] The present case report describes a rare concomitant lumbar and cervical multiple schwannoma associated with symptoms of cauda equina syndrome. Surgical approach for the resection of lumbar schwannoma with arthrodesis leads the patient to improvement of symptoms.

Ethics Statement

In the present case report, neither the patient nor any of his data have been identified.

Conflict of Interests

The authors have no conflict of interests to declare.

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