Cervical Dumbbell Meningioma: case report and literature review

Meningioma Cervical com Haltere: relato de caso e revisão de literatura

Breno Nery¹
Ítalo Pereira Salviano²
Beatriz Paiva Farias²
Eduardo Quaggio³
José Alencar de Sousa Segundo⁴

ABSTRACT

Introduction: “Dumbbell tumor” is a term used to describe tumors that connect and have two or more distinct regions, such as the intradural, epidural and paravertebral space. The incidence of these tumors ranges from 6% to 24% in large series of studies on spinal cord tumors. Dumbbell spinal meningiomas are a rare occurrence, accounting for up to 5% of spinal meningiomas with this feature, and are relevant in the differential diagnosis.

Case report: Female patient, 55 years old, with left cervicobrachialgia and progressive tetraparesis, caused by meningioma in the cervicothoracic transition. After performing tests and using medication for pain relief, the patient underwent laminectomy, facetectomy, tumor resection and correction of the dura mater defect with stabilization and posterior arthrodesis. Histopathological analysis showed a meningioma with markers of better prognosis. The patient had an uncomplicated recovery, with complete improvement of symptoms and no associated motor or sensory deficits.

Conclusion: The case in question is a rare meningioma tumor with peculiar characteristics that caused limiting symptoms. A preoperative investigation is essential to define the diagnosis and plan the surgery, which can result in a significant improvement in the patient’s quality of life and a low chance of recurrence.

Keywords: Cervical meningioma; Meningioma; Dumbbell; Dumbbell tumor; Cervical dumbbell tumors.

RESUMO

Introdução: “Tumor haltere” é um termo utilizado para descrever tumores que se conectam e possuem duas ou mais regiões distintas, como o espaço intradural, epidural e paravertebral. A incidência desses tumores varia de 6% a 24% em grandes séries de estudos sobre tumores da medula espinhal. Os meningiomas espinhais em haltere são uma ocorrência rara, representando até 5% dos meningiomas espinhais com essa característica, e são relevantes no diagnóstico diferencial. Relato de caso: Paciente do sexo feminino, 55 anos, com cervicobraquialgia esquerda e tetraparesia progressiva, causada por meningioma na transição cervicothorácica. Após realização de exames e uso de medicação para alívio da dor, o paciente foi submetido a laminectomia, facetectomia, ressecção tumoral e correção do defeito da dura-máter com estabilização e artrodese posterior. A análise histopatológica evidenciou meningioma com marcadores de melhor prognóstico. A paciente teve recuperação sem complicações, com melhora completa dos sintomas e sem déficits motores ou sensoriais associados. Conclusão: O caso em questão é um tumor meningioma raro com características peculiares que causou sintomas limitantes. A investigação pré-operatória é essencial para definir o diagnóstico e planejar a cirurgia, o que pode resultar em melhora significativa na qualidade de vida do paciente e baixa chance de recidiva.

Palavras-chave: Meningioma cervical; Meningioma; Haltere; Tumor haltere; Tumores cervicais com halter.

¹MD PhD, Neurosurgeon, Neurosurgery Department, Hospital Beneficência Portuguesa de Ribeirão Preto, Ribeirão Preto, SP, Brazil.
²MS, Medical Student, Centro Universitário UNIFACISA, Faculdade de Ciências Médicas, Campina Grande, PB, Brasil.
³MD, Neurosurgeon, Neurosurgery Department, Hospital Beneficência Portuguesa de Ribeirão Preto, Ribeirão Preto, SP, Brazil.
⁴MD, MR, Neurosurgery Department, Sociedade Portuguesa de Beneficência - Hospital Imaculada Conceição de Ribeirão Preto, Ribeirão Preto, SP, Brazil.

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INTRODUCTION

The “dumbbell tumor” was initially described in 1929 by the neurosurgeon George J. Heuer as a way of describing a group of tumors that arise along the entire spine and acquire the shape of dumbbells when they encounter anatomical barriers that modify their structure. Nowadays, however, the term “dumbbell tumors” no longer refers to the shape of the tumor, being used as a term conceptual to describe tumors that connect and have two or more distinct regions, such as the intradural, epidural and paravertebral space.

According to Pojskić and Arnautović the incidence of dumbbell-shaped injuries can vary between 6% and 24%. However, in the cervical spine, this index is significantly higher, reaching up to 44%.

Schwannomas and meningiomas are the two most common types of intradural tumors in the spine. About 90% of spinal tumors are schwannomas, and approximately one-third of them have a dumbbell-shaped configuration. On the other hand, spinal dumbbell meningiomas are a rare occurrence, accounting for up to 5% of spinal meningiomas with this feature, and are relevant in the differential diagnosis.

Meningiomas are, in most cases, benign tumors that originate from non-neuroepithelial progenitor cells present in the arachnoid capsule. It is estimated that meningiomas constitute between 13 and 26% of all intracranial tumors. In addition, involvement of the spinal compartment is relatively rare compared to the intracranial compartment representing approximately 1.2% of all central nervous system meningiomas.

In view of this, we describe the case of a middle-aged woman, complaining of left neck pain radiating to the left upper limb for 6 months, evolving with tetraparesis and the presence of Lhermitte’s sign. After performing a complete physical examination and auxiliary tests, the cause was identified and the patient underwent a surgical procedure, being diagnosed with cervical Dumbbell Meningioma, a rare event in the literature and worthy of being reported.

CASE PRESENTATION

Female patient, MGD, 55 years old, presented a clinical picture of left cervicobrachialgia affecting the region of the C7 dermatome, with progressive worsening for 6 months, progressing to tetraparesis. On physical examination, the presence of Lhermitte’s sign and decreased triceps reflex on the left (mild grade I reflex) were observed. Her sphincter function was intact. After performing cervical and thoracic magnetic resonance imaging with contrast, a lesion was observed in the cervicothoracic transition (C7/T1), with expansion of the intradural and extradural component towards the vertebral foramen of C8 on the left (Figure 1). Later, it was diagnosed that this neural lesion was caused by the presence of a meningioma. The presence of other comorbidities or stigmas of neurofibromatosis was not identified.

Thus, surgery was chosen, previously using Pregabalin 75mg to relieve neuropathic pain. Laminectomy from C6 to T1 was then performed, with facetectomy of C7 on the left for better exposure of the extradural portion of the tumor, opening of the dura mater for resection of the intradural portion of the tumor (Figure 2) and subsequent correction of the dura mater defect with stabilization and posterior arthrodesis with 12 mm lateral mass screws from C6 bilaterally, C7 to the right and T1 bilaterally. Intraoperative electrophysiological monitoring was performed uneventfully. There was anatomical and functional preservation of the ventral root of C8.

Histopathological analysis showed the presence of uniform cells, round and loose chromatic nucleus, pink and abundant...
cytoplasm and imprecise limits (Figure 3). Immunohistochemistry was performed with the EMA marker and positive IH for the Progesterone Receptor marker, common in meningiomas and an indicator of better prognosis (Figure 4).

The postoperative clinical evolution was uneventful, with total improvement of symptoms, without associated motor or sensory deficit. During post-operative follow-up, magnetic resonance imaging was performed, which did not reveal any residual tumor lesion. (Figure 5)

**DISCUSSION**

The “Dumbbell” cervical meningioma is a type of tumor that develops in the meninges of the cervical spine and has a dumbbell shape, as it is composed of two distinct portions, one intradural and the other extradural. This characteristic type of spinal tumor is most commonly seen in schwannoma-type tumors, and more rarely in meningioma-type tumors, which can be difficult to diagnose due to their unusual location.

Few reports were found in the literature about dumbbell-shaped cervical meningiomas, since meningiomas develop from the arachnoid membrane, and rarely penetrate the dura mater, as they usually grow towards the inner side of the dura mater and are unlikely to spread, transform it into a type of dumbbell. However, the similarity of the signs and symptoms presented by the reports found are quite relevant, as well as the consequent surgical intervention and postoperative follow-up, as described in the comparative table (Table 1).

Although this tumor can affect both genders at any age, it was observed that the age with the highest incidence of meningiomas is between the 6th and 7th decade of life. The exact cause of the increased incidence of meningiomas in older people is still not completely understood, but according to the “CBTRUS Statistical Report: Primary brain and other central nervous system tumors diagnosed in the United States in 2011-2015,” factors such as exposure to ionizing radiation, family history of meningiomas, occupational exposure to chemicals and female sex hormones may be related to risk factors for the development of meningiomas.
On the other hand, cases of young patients affected by meningioma as described (Table 1) can be explained by the presence of Neurofibromatosis, an autosomal dominant genetic disease that predisposes to multiple benign tumors of the nervous system. Therefore, combined neurofibromatosis and young age were possibly contributing factors to the development of dumbbell meningiomas, as meningiomas occur in about half of patients with Neurofibroma type 2 and are usually multiple.

In addition, most reports described in the literature show that almost all cases occurred in women (Table 1). This is explained by the role that female sex hormones, such as estrogen, can play in the development of meningiomas. Therefore, although not definitive, the available data suggest an association of the risk of meningiomas in women exposed to endogenous or exogenous sex hormones, being women of reproductive age, postmenopausal with the use of hormone therapy or early menarche. Furthermore, in the adult population, there is a marked female bias with a female: male ratio of 3:1 and increasing to 9:1 for spinal injuries.

In summary, according to the article "Epidemiology and etiology of meningioma", aging, along with genetic, hereditary and environmental factors, can contribute to the development and growth of meningiomas. However, the extent of immunological factors that would likely influence the etiology of meningioma is still very little explored.

Although Meningioma is largely benign, there are challenges regarding to treatment in cases with complex morphology or location, in addition to being able to cause compressive symptoms, depending on the size and space affected. The most common
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Symptoms are local pain and/or root canal, weakness of the limbs and paresthesia. In the literature, the main symptoms observed were progressive complaints of neck pain, weakness in the upper limbs and numbness in the lower limbs. There were two reports of gait disturbance and one case of ankle clonus and the presence of Babinski bilaterally.

The reason why most meningiomas are benign and do not recur is related to the fact that these tumors are composed mainly of normal meningothelial cells, which are the cells that make up the meninges. They tend to grow slowly and can be completely removed by surgery, especially if they are discovered in the early stages. Therefore, surgery is the first-line treatment for this pathology, with low recurrence rates and significant improvement in the patient’s symptoms.

**CONCLUSION**

The case analyzed is considered rare, since the tumor in question is of the meningioma type and was located in the intradural region, with dural invasion, extradural and foraminal extension. It had a peculiar dumbbell shape and caused a variety of symptoms that limited the patient’s daily activities. Therefore, a preoperative investigation is essential to establish the diagnosis and define the best surgical strategy. In addition, it is noteworthy that the complete or partial removal of the tumor can bring a significant improvement in the patient’s quality of life, with low chance of recurrence, due to the histological characteristics of this type of tumor.

**REFERENCES**


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**Table 1. Clinical characteristics and follow-up of patients with dumbbell meningiomas in the literature.**

<table>
<thead>
<tr>
<th>Study</th>
<th>Age/Sex</th>
<th>Location</th>
<th>Signs and symptoms</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chen et al.⁷</td>
<td>16/F</td>
<td>C1 - C2</td>
<td>Grade 4/5 strength and increased tendon reflexes in right upper limb.</td>
<td>After 10 months of follow-up, the muscle strength of the right upper limb was normalized.</td>
</tr>
<tr>
<td>Zhan et al.⁸</td>
<td>47/F</td>
<td>C1 - C2 and foramen magnum</td>
<td>4-month history of gradually worsening neck pain and lower extremity numbness.</td>
<td>There were no postoperative complications and no related clinical symptoms.</td>
</tr>
<tr>
<td>Nguyen et al.⁹</td>
<td>22/F</td>
<td>C5-C8</td>
<td>Grade 2 strength in lower limbs, global hyperreflexia, ankle clonus and positive Babinski.</td>
<td>Surgery without any complications. Showed significant improvements in muscle strength. Six months after surgery, the patient was able to walk with a walker.</td>
</tr>
<tr>
<td>Ozaki et al.¹⁰</td>
<td>49/F</td>
<td>C2 - C3</td>
<td>Gait disturbance, paresthesia and global weakness.</td>
<td>The postoperative period was uneventful, with improvement in preoperative neurological deficits; 2.5 years after the operation, no residual tumor growth is visible by Magnetic Resonance imaging (MRI).</td>
</tr>
<tr>
<td>Oichi et al.¹¹</td>
<td>64/M</td>
<td>C2</td>
<td>5-year of gradually exacerbated left occipital pain.</td>
<td>The postoperative period was uneventful. Six months after surgery, the patient had no clinical symptoms related to the tumor, and MRI showed no tumor recurrence.</td>
</tr>
<tr>
<td>Yoshiura et al.¹²</td>
<td>16/F</td>
<td>C2-C4</td>
<td>3-month of neck pain radiating to left shoulder and weakness in left upper extremity.</td>
<td>After surgery, the patient was discharged without significant neurological deficit. Follow-up studies showed no tumor recurrence at 8 months.</td>
</tr>
<tr>
<td>Sato et al.¹³</td>
<td>76/F</td>
<td>C3-C4</td>
<td>Decreased strength in upper and lower limbs. In addition to paralysis in the C5 region of the left upper limb, coordination disorder, bilateral hyperreflex of the biceps and its distal tendons, and spastic gait.</td>
<td>The postoperative period was uneventful. After the operation and some symptoms were relieved. MRI 2 years after the operation demonstrated a residual tumor around the vertebral artery, but there was no apparent growth.</td>
</tr>
<tr>
<td>Hazuba et al.¹⁴</td>
<td>60/M</td>
<td>C6 - T1</td>
<td>Complete paralysis of upper &amp; both lower limbs.</td>
<td>He was followed up for 5 years and in his postoperative follow-up he presented paralysis of the hand, mild weakness of both lower limbs; able to walk without a cane.</td>
</tr>
</tbody>
</table>

M: male; F: female.


CORRESPONDING AUTHOR

Italo Pereira Salviano, MS
Medical Student
Centro Universitário UNIFACISA
Faculdade de Ciências Médicas
Itarará, Campina Grande, PB, Brasil
E-mail: italopsalviano@gmail.com

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