Cervical intramedullary schwannoma: a case report and review of the literature

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Abstract

Intramedullary schwannomas unrelated with neurofibromatosis are uncommon tumors, but if correctly diagnosed and properly treated they may have a good prognosis.

They have a wide range of clinical presentations, commonly presenting as a slowly progressive motor or sensory syndrome. We present a case report of a patient without neurofibromatosis with a surgically treated cervical intramedullary schwannoma.

Introduction

Spinal schwannomas are tumors originating from the Schwann cells[1] and correspond to 30% of spinal tumors, most of which have an intradural extramedullary location.[2]

They are generally associated with neurofibromatosis types 1 and 2.[3]

Intraparenchimal schwannomas of the central nervous system (CNS) are extremely rare when no relationship with neurofibromatosis is present and several parts of CNS can be affected, such as the spinal cord, cerebellum and brain stem.[4-6]

Intramedullary lesions represent 0.3% of all medullary tumors and 1.1% of spinal schwannomas.[7] This article reports a case of intramedullary schwannoma and presents a review of literature.

Case Report

Our patient is a 40-year old Caucasian male. He was admitted to the department of Neurology and Neurosurgery of the Heliopolis Hospital, São Paulo, Brazil, presenting spastic tetraparesis and sphincterian disturbances. The symptoms had developed progressively with onset two years before admission.

Initially the patient complained of left inferior limb weakness that affected the right inferior limb after five months together with urinary urgency , high thoracic column pain without irradiation and arms and shoulders muscular fasciculations. After ten months the paresthesia and motor weakness achieved the superior limbs. The neurologic examination showed an asymmetric progressive spastic tetraparesia especially in the left side whith impaired deambulation. Diffuse pyramidal signs and normal deep sensitivity were also described. The first diagnostic hypothesis was amyotrophic lateral sclerosis and the patient was treated with rilusole for 14 months. After this, and given his physical weakness and sphincterian dysfunction progression, he sought assistance from another medical center where a cervical spinal magnetic resonance (MRI) was performed. This demonstrated a C4-C6 intramedullary lesion (hypointense in T1-weighted, hyperintense in T2-weighted,

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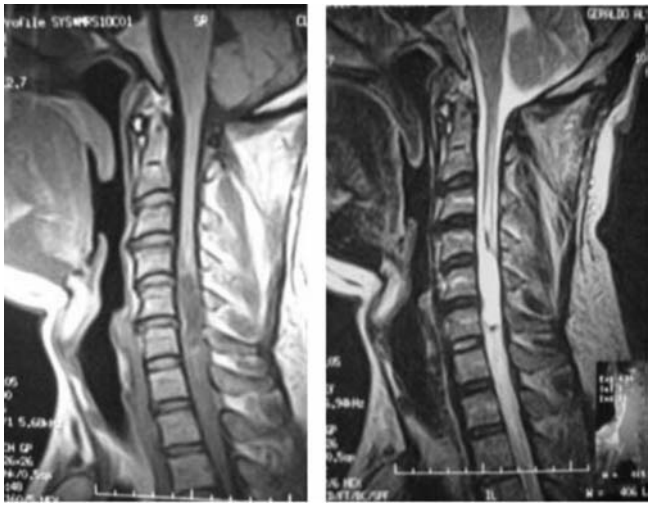

Figure 1. Preoperative cervical spine magnetic resonance (Sagittal T1- and T2-weighted images).

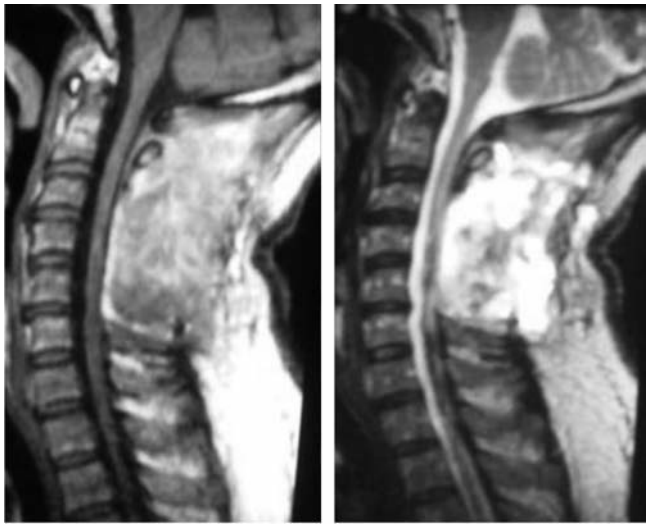

Figure 2. Postoperative magnetic resonance. Gross total ressection.

heterogeneous contrast impregnation and syrin-gomyelia) (Figure 1). Intravenous steroids were started in the pre-operative period (30 mg/kg in bolus) followed by a maintenance 5.4 mg/kg dosage over the following 23 hours. Surgical treatment was made through a C3-C5 laminotomy with a careful duramater microscopic opening, one centimeter mielotomy and tumor subtotal resection (Figure 2).

The intraoperative aspect of the lesion was of a grayish and infiltrative mass making total resection impossible. The nerve roots were not involved by the tumor.

Microscopically examination demonstrated compact palisade cells with lined nucleus mixed with some enucleated areas called Verocay bodies (Figure 3).

During the immediate post-operative period a worsening of tetraparesis was noted with recovery in the first 48 hours. After 24 months, a significant regression of the motor and sphincterian dysfunction was observed and this allowed the patient to hold objects with his hands and walk with help again.

Discussion

The first surgical description of a spinal tumor was made in 1888 by Sir Victor Horsley[8] who reported an extramedullary intradural meningioma resection. However, it was just in 1907 that Von Eiselberg published the successful resection of an intramedullary neurofibrosarcoma. Though Kernohan has been recognised as the first neurosurgeon to report an intramedullary schwannoma case in 1952, Penfield had already described an intramedullary lesion with schwannoma characteristics in 1932.[9]

Up until today, approximately 50 cases of intramedullary schwannomas not related to neurofibromatosis have been described, some of them are shown in Table 1. The melanotic schwannomas, although not the scope of this discussion, are even rare; from 39 cases reported, just 5 were intramedullary lesions and 10% of those malignized,[10] as shown in Table 2.

The male:female ratio for intramedullary schwannomas is 3:1 with a mean age of 40-years old. They are usually single lesions affecting the cervical spinal cord (63%), the thoracic spinal cord (26%) and the lumbar spinal cord (11%). They have a slow growth pattern and because of this the average interval between first symptoms and diagnosis is 28.2 months (from six months to 20 years).[11]

The most described clinical manifestation is the pyramidal syndrome followed by sensitivity complaints and sphincterian dysfunction. There are reports of muscular fasciculations as the first symptom. Another complaint is the

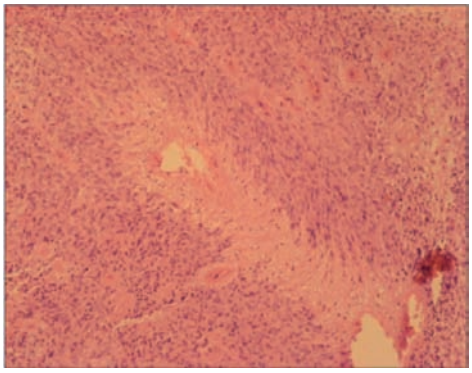

Figure 3. Electronic microscopy of a schwannoma with palisade cells and Verocay bodies.

Table 1. Intramedullary schwannoma cases without Von Recklinghausen disease.

Author/year	Sex	Topography		Duration of symptoms	Treatment
Penfield, 1932[9]	M	C5	8 years	Subtotal resection	-
Rasmussen, 1940[19]	M	C4 – C7	4 years	Subtotal resection	PR
Roka, 1951[20]	M	Cervical	3 years	Subtotal resection	Worsening
Riggs & Clary,1957[16]	M	C4 – C5	3 years	Autopsy finding	-
Ramamurthi, 1958[8]	M	T2	9 months	Total resection	PR
Lang & Bridge, 1959[21]	M	Cervical	1 year	Total resection	PR
	M	Thoracic	3 years	Total resection	Worsening
Scott & Bentz, 1962[22]	F	T3 – T4	12 years	Subtotal resection	Stable
Lu, *et al.*, 1963[23]	M	C4 – C5	3 months	Total resection + RDT	PR
	M	C2 – C5	18 months	Subtotal resection	PR
McCormick, 1964[17]	M	L2	6 weeks	Autopsy finding	-
Slooff, *et al.*, 1964[24]	M	C4 – C7	4 years	-	-
Guidetti, 1967[25]	-	Connus	-	Total resection	TR
Mason & Keigher, 1968[26]	M	T8 – T10	3 months	Total resection	PR
Chigasaki, 1968[27]	F	T3	-	Subtotal resection	-
Van Duinen, 1971[28]	M	C3	-	Total resection	-
Cambier, *et al.*, 1974[29]	M	C2 – C4	16 months	Total resection	Worsening
Wood, *et al.*, 1975[2]	M	C1 – C3	3 months	RDT	Death -PE
Schmitt, 1975[30]	M	Connus	6 months	Autopsy finding	-
Isu, *et al.*, 1976[31]	F	C1	6 months	Subtotal resection	-
Pardatscher, *et al.*, 1979[32]	M	C4 – T9	9 months	Decompressive	Death
Vailati, *et al.*, 1979[33]	F	T8 – T9	1 year	Total resection	PR
Shalit & Sandbank, 1981[34]	F	C2 – T2	6 months	Total resection + RDT	TR
Cantore, *et al.*, 1982[35]	M	T12 – L1	-	Total resection	TR
	F	C3 – C5	2 years	Total resection	PR
Lesoin, *et al.*, 1983[36]	F	C3 – C7	6 months	Total resection	TR
	M	Conus	5 years	Total resection	TR
Rout, *et al.*, 1983[37]	F	C2 – C6	5 years	Total resection	TR
	F	C2 – C5	5 years	Subtotal resection	PR
Sharma, *et al.*, 1984[3]	M	C2 – C6	18 months	Total resection	TR
Ross, *et al.*, 1986[7]	M	C4 – C5	4 months	Total resection	TR
	F	C2 – T1	4 years	Subtotal resection	PR
Gorman, *et al.*, 19893[8]	F	C2 – C5	8 months	Total resection	PR
Herregodts, *et al.*, 1991[1]	F	T3 – T4	5 years	Total resection	PR
Nicácio, *et al.*, 2007	M	C4 – C6	2 years	Subtotal resection	PR

RDT, radiotherapy; PR, partial recovery; TR, total recovery; PE, pulmonary embolism.

Table 2. Intramedullary melanotic schwannoma cases in literature.

Author/year	Age /Sex	Signs and symptoms	Duration of symptoms	Location	Treatment	Results
Solomon, *et al.*[39]	69/M	Brown-Séquard	4 y	C3	Total resection	
Marchese &McDonald[40]	72/F	Tetraparesis	20 y	C4-C6	Total resection	PR
Sola-Perez, et al.[41]	63/F	Radicular pain		C7 - T1	Partial resection	PR
Acciarri, *et al.*,[42]	44/F	Tetraparesis	10 y	T2 - T3	Total resection	PR
Santaguida, *et al.*[10]	35/M 39 M	Hemip./Parap.	10 m	C4-C5/C4-C6	Total resection + CMT+RDT	

RDT, radiotherapy; CMT, chemotherapy; PR, partial recovery .

motor-sensitive alternal deficit associated with amiotrophy in patients with predominantly one-sized located medullary tumors.[12] The X-ray findings are correlated to tumoral growth characteristics.

Mielography denotes precisely the tumor location and the relationship with dura mater and the spinal cord. However, MRI is the gold standard to study intramedullary tumors.

In 1988, Takemoto stated that MRI allows pre-operative diagnosis of schwannomas, neurofibromas, meningiomas and hemangioblastomas.[13] On the other hand, according to Nicoletti in 1994, neither the MRI nor CT scan can differentiate the intramedullary tumor histological type.[14] Sagittal and axial images demonstrate a widening of the spinal cord.

Perilesional edema and cystic cavities can be observed. These tumors are hypointense or isointense on T1-weighted sequences and generally hyperintense on T2-weighted sequences. When gadolinium is injected there is a heterogeneous enhancement.

According to Demachi, there is no correlation between the classification of Antoni and the MRI findings.[15] The Antoni A-type is characterized by the presence of compact wave-shaped cells rounded by a reticular net. The Antoni B-type has large and loose cells surrounded by a collagenous web.[12]

The infiltrative pattern of some intramedullary schwannomas make total gross resection impossible and some authors suggest in these cases the use of radiotherapy for residual lesions.[2] According to the new WHO classification of tumors there are three types of schwannomas: cellular, plexiform and melanotic.

However, the controversial question about this pathology emerges from the unknown pathogenesis. Previous studies have claimed that the central nervous system cells have no Schwann cells, thus making the presence of intramedullary schwannomas a paradox.

In the pursuit of an answer to that question, several theories have been suggested over the last fifty years. In 1957, Kernohan, McCarty, Riggs and Clary proposed that the origin of such lesions could be from Schwann cells' proliferation derived from nerve fibers of the spinal arteries.[16]

Ramamurthi et al. suggested that a few ectopic Schwann cells of the embrionary neural tube (during the fourth gestational week) could be the origin of these schwannomas.[8]

In 1964, MacCormick and Wood stated that intramedullary schwannomas came from some Schwann cells found in aberrant intramedullary nervous fibers arising through the posterior roots.[17] But the most acceptable theory was reported by Rusell and Rubenstein in 1971. According to them, these tumors emerge from the transformation of neuroectodermal pial cells into Schwann cells, leading to a possible fast neoplastic growth of Schwann cells located in a "critical area" in the dorsal roots.[18]

Conclusions

Although rare, the intramedullary schwannomas should be considered as a possible diagnosis for young adults presenting with an intramedullary lesion. Once suspected, surgical treatment is recommended. Gross total resection is the goal but sometimes this cannot be accomplished due to the infiltrative characteristic of the tumor. Finally, a better understanding of the etiology and physiopathology will certainly contribute to the treatment of these patients.

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