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Case Report

Cervical Intramedullary Schwannoma: Case Report and Review of the Literature

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Abstract
Cervical intramedullary schwannomas are extraordinarily rare. Gross total resection is the best therapeutic option for these types of tumors. Although rare, intramedullary schwannomas should be considered as a differential diagnosis of intramedullary lesions since a good prognosis can be guaranteed to the majority of these patients. We present a case of a cervical intramedullary schwannoma surgically treated in a 19-year-old male patient who initially presented with motor neuron disease.

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Introduction

Schwannomas represent 30% of all spinal tumors [1]. They are usually located in the intradural-extramedullary space. Intraparenchymal location of schwannomas in the central nervous system (CNS) is rare, because brain and spinal cord do not have Schwann cells. Intramedullary schwannomas (IS) are extraordinarily rare, accounting for 1.1% of spinal schwannomas and 0.3% of intraspinal tumors [2–4]. Misdiagnosis of IS as glioma may lead to inappropriate treatment. Comprehensive analysis of clinical and radiological features, including a high index of suspicious, is the best way to preoperatively diagnose IS. Gross total resection (GTR) is usually possible and represents the most beneficial treatment. On the other hand, the role of adjuvant radiotherapy is not clear.

In the present study, we report a case of a cervical intramedullary schwannoma surgically treated and discuss clinical and radiological findings related to these types of tumors.

Case Presentation

A 19-year-old male with a 3-year history of pain and sensitive alterations (dysesthesias and numbness) predominantly in his right arm and hand with progressive spastic quadriparesis. Patient functional pre- and postoperative status was assessed with the modified McCormick scale (Table 1) [5, 6].

Neurological Examination

The patient presented with muscle wasting, bilateral atrophy of the thenar and hypothenar eminences, quadriparesis (3 on the MRC scale), hyperreflexia (4+), bilateral Hoffmann and Trömner reflexes, bilateral Babinski sign and right hypoesthesia caudal to C5–C6 dermatome. Discrimination and propioception were also altered. Sphincters were not affected. There were no neurofibromatosis stigmas. The preoperative modified McCormick grade was IV.

Neuroradiology

The preoperative MRI on T2WI showed a 33 × 13 mm intramedullary cystic tumor located on C6–C7, regular and well delimitated with minimum perilesional spinal cord edema. No syrinx was noted. The tumor was isointense on T1WI and T1-gadolinium showed annular enhancement of the lesion (Fig. 1).

Operation

Given the clinical and radiological findings, the patient first underwent open biopsy of the lesion. The histological specimen was sent to the Department of Pathology for histological diagnosis. Definitive surgery was performed in the National Cancer Institute of Mexico (INCan) through posterior cervical approach with C6–C7 laminoplasty. When the dura mater was opened, the spinal cord was expanded in that region. Incision at the pia mater disclosed a yellow-grayish tumor, firm and moderately vascular with a good dissection plane. Total excision of the lesion with bipolar coagulation, ultrasonic aspirator and intraoperative neurophysiological monitoring (somatosensory and motor-evoked potentials) was performed.

Pathology Findings

Tumor histology showed a biphasic neoplasia consisting of hypercellular and hypocellular alternating areas. The cells were spindle, elongated, often wavy with ill-defined eosinophil cytoplasm and oval nucleus with little apparent nucleolus. In the more compact zones, the cells were arranged in a fascicular pattern with thick-walled stromal vessels. Immunohistochemical studies of S-100 Protein, Glial Fibrillary Acidic Protein (GFAP), Epithelial Membrane Antigen (EMA), Progesterone Receptors (PR), and Ki-67 were tested. Altogether, these findings confirmed schwannoma and excluded glioma and meningioma (Fig. 2).

Postoperative Course

The patient had an uneventful recovery. At the immediate postoperative period the sensitivity and strength remain unchanged. After discharge, the patient was sent to physical therapy. At follow-up review 4 weeks later, the patient had recovery strength (4– on the MRC scale), improved sensitivity and the modified McCormick score was III. The postoperative MRI showed complete removal of the tumor (Fig. 3).

Discussion

James Kernohan is recognized as the first pathologist to report an IS in 1931 [7]. However, until these dates approximately 70 cases of IS have been reported [2, 8, 9]. The most common location of IS in order of frequency is the cervical spine (63%), and the thoracic (26%) and lumbar (11%) levels. Men are more affected than female in a 3:1 ratio [8]. The mean age of presentation is around the fourth decade of life, but congenital IS cases have been reported [8, 10]. Although vestibular schwannomas are distinctive of Neurofibromatosis type 2, emergence of IS has been reported in this hereditary disease [9, 11].

Regarding the clinical spectrum, Wu et al. compared different features of patients with intramedullary tumors (173 ependymomas, 70 astrocytomas and 7 schwannomas) admitted during a 7-year period (2003–2010) [12]. They found somatic pain and nerve root pain were the statistically significant initial symptoms of IS compared with ependymomas ($p = 0.005$) and astrocytoma ($p = 0.019$), but these clinical differences were not seen between ependymomas and astrocytomas ($p = 0.175$). In addition, the mean age for intramedullary ependymoma, astrocytoma and schwannomas was 35, 33 and 44 years, respectively. IS had a longer mean duration of illness (4.4 years vs. 2.3 years for astrocytomas and 1.7 years for ependymomas) [12]. The most common initial symptoms are pain and/or dysesthesias that last for several months or years, and motivate the patients to consult a physician rather than the motor deficit, diminished sensitivity, and sphincter dysfunction [2, 13].

The origin of the schwannoma are the Schwann cells, which are not normally found within the parenchyma of the brain and spinal cord. This has raised speculation about the pathogenesis and has led to several theories to explain the presence of this tumor in the CNS, although none has gained acceptance. Hypotheses include 1) migration of Schwann cells during embryonic development, 2) Schwann cells ensheathing aberrant intramedullary nerve fibers, 3) neoplastic growth of Schwann cells in the dorsal root entering zone where posterior nerve roots lose their sheaths on entering the pia mater, 4) extension of Schwann

cells along the intramedullary perivascular nervous plexus and 5) transformation of multipotential cells of neuroectodermal origin into Schwann cells [2, 13, 14].

MRI is the preferred imaging study to visualize intraspinal tumors. IS are most commonly reported to be iso- or hypointense on T1WI, and moderately hyperintense on T2WI images [15]. However, signal intensity depends on macroscopic features (solid or cystic) and histological composition (Antoni A or Antoni B pattern) [2]. Preoperative radiological diagnosis of IS is difficult and differentiation from other intramedullary gliomas is practically impossible. For this reason, the role of MRI in the diagnosis of IS is limited and the histopathological analysis is crucial to establish a diagnosis.

GTR of the lesion must be attempted without damaging the surrounding neural tissue. The majority of lesions reported in the literature had a well-demarcated dissection plane with no adhesions to spinal cord and GTR was achieved. However, if the lesion is adherent to the neural tissue, a subtotal resection is likely to improve neurological function. In addition, a second surgery can be performed to remove the remaining tumor [16].

Conclusions

IS is a slow growing and histologically benign tumor and complete functional recovery can be achieved after total excision. Although MRI enables a good pre-surgical evaluation of IS, there are no pathognomonic clinical signs that allow differentiation from other intramedullary tumors. Therefore, IS should be considered in the differential diagnosis of an intramedullary lesion in the cervical or thoracic spine of a middle-aged male patient with chronic somatic or root pain as the initial symptom, without signs of severe neural damage.

Statement of Ethics

Since this a retrospective case report, ethical approval was waived. However, written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Disclosure Statement

No potential conflicts of interest were disclosed from the authors.

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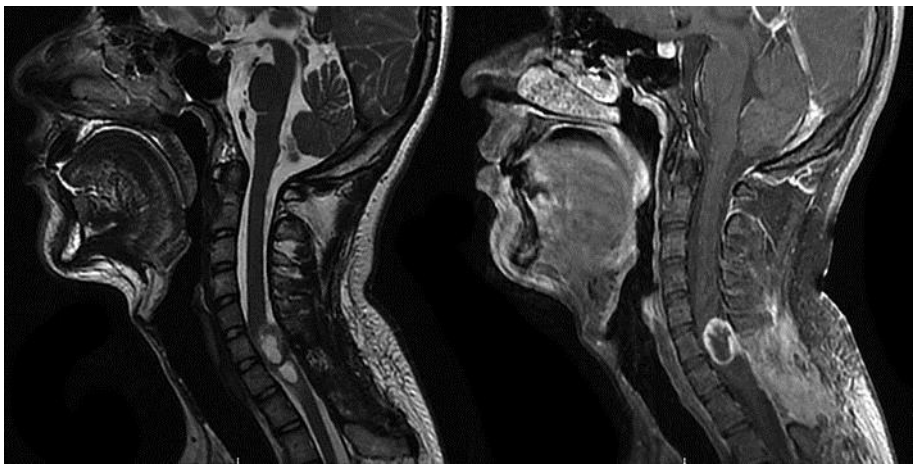

Fig. 1. T2 (left) and T1-gadolinium (right) MRI showing a cervical intramedullary tumor.

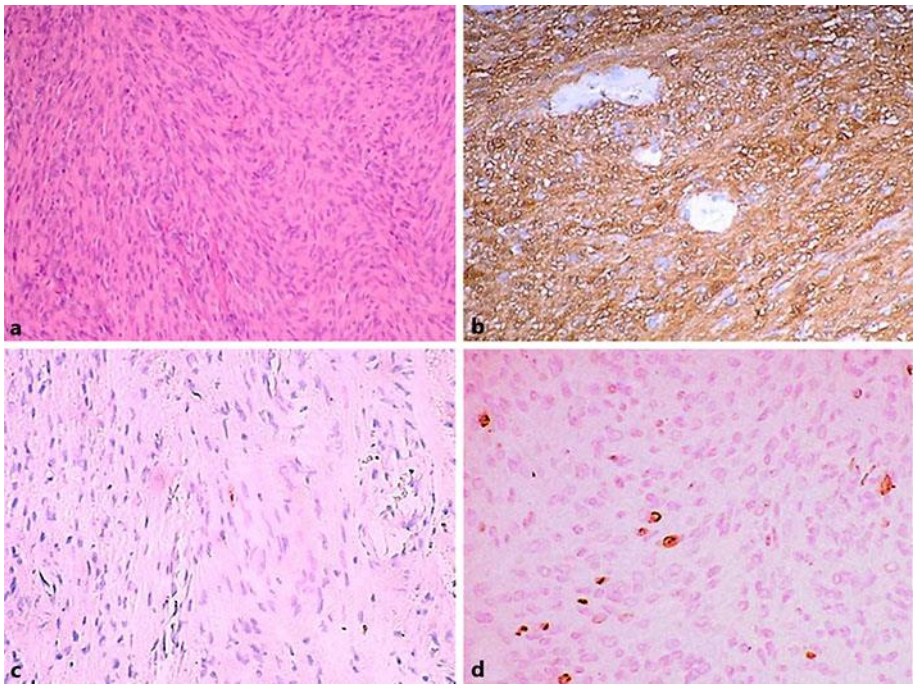

Fig. 2. Histopathological study showed a schwannoma with biphasic components (hypercellular and hypocellular areas) (a). Immunohistochemical studies showed strong reactivity for S-100 Protein (b). Glial Fibrillary Acidic Protein (GFAP), Epithelial Membrane Antigen (EMA) and Progesterone Receptors (PR) were negative (c). A low rate proliferative index was demonstrated by Ki-67 expression (d).

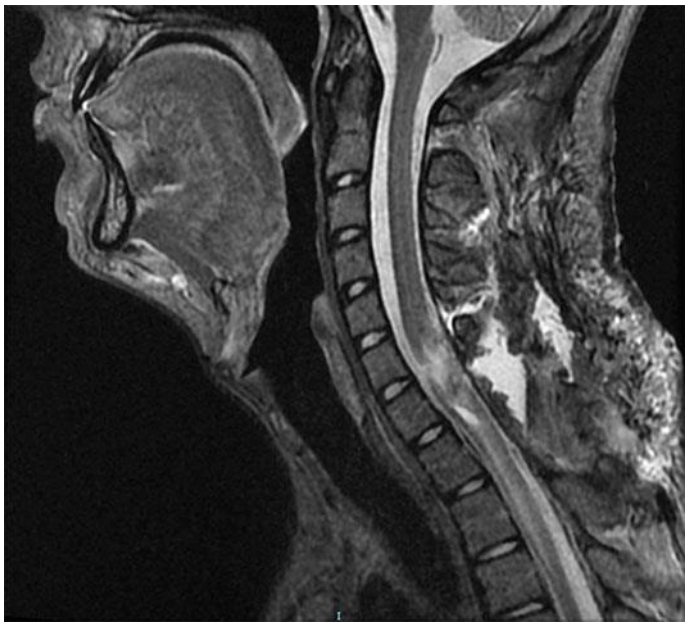

Fig. 3. Postoperative T1-gadolinium MRI showing removal of IS.

Table 1. Modified McCormick scale

Grade	Modified McCormick scale
I	Intact neurologically, normal ambulation, minimal dysesthesia
II	Mild motor or sensory deficit, functional independence
III	Moderate deficit, limitation of function, independent with external aid
IV	Severe motor or sensory deficit, limited function, dependent
V	Paraplegia or quadriplegia, even with flickering movement