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Giant Spinal Schwannomas. Presentation of Two Cases with Interdisciplinary Surgical Resection

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Abstract

Purpose: To report two quite rare cases of giant schwannomas of the cervical and thoracic spine. Surgical resection is usually the treatment of choice for these neoplasms. **Methods:** The first patient is a 48-year-old male that came to our department, complaining about weakness and paresthesia of both lower extremities and upper left extremity, and a considerably big dorsal palpable lump. MRI and CT scan were used to diagnose giant cervical schwannoma. Because of the intensity of symptoms, he was subjected to surgery. Multidisciplinary resection involving neurosurgical and oncosurgicalteamn was performed for intravertebral and extravertebral approaches. The second patient is a 55-year-old female that came to the hospital complaining about incapacitating pain and dysesthesia involving T6, T7, and T8 dermatomes, with no other deficits. MRI and CT scan were used to diagnose giant thoracic schwannoma. Because of the intensity of the pain, surgery was decided as the treatment of choice. Video assisted thoracoscopic surgery was performed and total resection was achieved, with improvement of the symptoms. **Results:** Both patients showed complete remission of the symptoms, with no major deficits after surgery. **Conclusion:** Although infrequent, giant spinal schwannomas may be the cause of visible lumps in the back with radicular symptoms. When asymptomatic, conservative treatment could be viable, but when they present symptoms, surgery offers good results.

Keywords

Spinal Schwannomas, Giant Schwannomas, Interdisciplinary Surgery

1. Background

Schwannomas originate from Schwann cells in the neural sheath. Approximately 95% are benign, encapsulated and slow growing. Spinal schwannomas may be extradural or intradural. In extradural growth they can present an extraspinal pattern or stay inside the spine. In intradural growth, they can get intramedullary and cause spinal cord and nerve root compression [1].

2. Introduction

For a spinal schwannoma to be defined as giant, it must have a length of at least 2 vertebral bodies or extraspinal extension of 2.5 cm [2] [3]. Approximately 90% of schwannomas are sporadic. Schwannomas may be part genetic syndromes (NF-2, Schwannomatosis and Carney complex) or be sporadic cases. NF-2 accounts for 3% of syndromicschwannomas, schwannomatosis accounts for 2%, and meningiomatosis with or without NF-2, for 5% [4]. Incidence of giant spinal schwannomas is unknown, since it's an infrequent presentation. For non-giant spinal schwannomas incidence is 0.24 cases per 100,000/year [5]. One of the largest reported series of giant spinal schwannomas included 32 patients. A mean age of 47 years was found, with male (71.9%) predominance, none of the patients presented NF-2 or schwannomatosis.

Clinical presentation of these tumors can range from asymptomatic to severe motor, sensitive and autonomic deficits. When asymptomatic, conservative treatment is of choice. For patients with severe deficits, surgical resection is performed. Gross-total resection (GTR) is usually curative and safe in most cases. Suboptimal resections usually occur when the tumor adjacent to deep neural or visceral structures [6]-[11]. Resection of giant schwannomas involves extensive intra and extraspinal corridors, and can be performed by anterior, posterior, combined or even posterior retro pleural with Thoracoscopic Guidance approaches to the spine [12] [13] [14] [15] [16]. Several classifications have been used to improve precision in diagnostic communication and treatment planning. Some of the most widely used are Eden and Shridhar, Toayam and Asazuma's Classification [3] [17] [18].

These classifications take into consideration the imaging studies performed, the anatomical relationship of the tumor with the bony structures, the tumor volume, the relationship with the dura mater and the spinal canal, and also attempt to discuss surgical management according to the previous characteristics [19] [20] [21] [22].

The use of the Davinci robot has been reported with good outcomes in the resection of giant Schwannomas and spine stabilization [23] [24].

Dumbbell tumors can not only cause the compression of cervical cord and nerve root, but also invade the important structures and the surrounding organs, causing great harm to the patient [21] [22] [25] [26]. However, although useful in clinical practice, Toyama's classification system shows its low reliability and reproducibility in the study published by Mengchen *et al.* (2017) [27].

In the publication by Xin *et al*. 2020, the current classifications were reviewed and compared, identifying the advantages and disadvantages of the use of each one, which were summarized in one list, see table 1 for a better understanding [28].

Despite the efforts made, there is still no validated and standardized algorithm of action for the treatment of giant spinal schwannomas, we share the report of two cases managed at the Centro Medico Nacional 20 de Noviembre.

The objective of this article is to report two cases of giant scwannomas managed by non-conventional techniques with excellent post-surgical results, thus exemplifying that each similar case should be studied in depth and individualized in the search for the best possible results.

Giant Schwannomas are infrequent, however, given that most of their life they are asymptomatic when they establish clinically they have involved important structures, the direction in which they grow as well as the structures they involve and the vertebral levels they occupy will determine the complexity of the procedure for their extirpation, other surgeons and anatomists have tried to establish scales or classifications to facilitate decision making when facing them, however according to the literature review none of them has proven to be completely useful given the limitations they have demonstrated, it's not the purpose of this article to discuss the limitations of one or the other, however, **Table 1** has been attached that briefly discusses them [28].

3. Clinical Case 1

This 48-year-old male patient, with no relevant history, presented to the department complaining of growing, painless, dorsal lump. He also presented bilateral weakness, and paresthesia of lower extremities, that increased during physical activity and caused claudication when walking for long periods, and he recently developed left arm weakness and gait disturbance.

During PE, multiple *café au lait* spots on trunk and extremities, and axillary freckles were detected. Cutaneous fibromas on the thorax and scapular region. Neurological examination showed weakness 4/5 in the left C5 and T1 myotomes, and bilateral weakness of L4-S1 myotomes. Reflexes were diminished in both lower extremities, and he presented slow and uncoordinated gait. Neurophysiological exam showed a pathway dysfunction suggestive of structural lesion in mentioned level. Electromyography of upper limbs diagnosed left C5-C6 sensorimotor radiculopathy. The patient presented a palpable lesion at cervical level that correlated with a heterogeneous image that enhanced in a discrete way with the application of contrast medium (in Dumbell). The MRI showed in the axial section a Dumbell image with an intravertebral extradural portion and an extravertebral portion running anteriorly (see image 1). We had a multidisciplinary evaluation, that involved a oncological surgical plan for the cervical extraspinal component, and they suggested a posterolateral neck approach, with a combined team of neurosurgeons and oncological surgeons. Genetic evaluation confirmed the patient met more than 2 criteria of NF-1. Cervical myelopathy secondary to

C1-C2 intraspinal extradural tumor with paravertebral extension C2 to C5 associated with neurofibromatosis type 1 was diagnosed.

Surgery was performed with a posterolateral neck approach, the extraspinal

Table 1. The advantage and disadvantage of Published classification Xin *et al*. 2020.

Classification	Advantage	Disadvantage
Eden's classification	1) It is the "gold standard" when neither CT nor MRI has been developed 2) The basic classification	1) It is insufficient to a guide the choice of surgical strategy 2) It is not based on modern imaging techniques 3) The membranous hierarchy of spinal schwannomas has been neglected
Asazuma's classification	1) It is the first classification based on CT or MRI 2) It is highly useful for devising surgical approaches	1) It is not comprehensive 2) It cannot be used for thoracic or lumbar schwannomas 3) The guiding significance of the surgical approach is not fully applicable today 4) The membranous hierarchy of spinal schwannomas has been neglected
Peking university third hospital classification	1) It is comprehensive 2) It further rationalizes the surgical approach for cervical dumbbell tumors	1) It cannot be used for thoracic or lumbar schwannomas 2) The membranous hierarchy of spinal schwannomas has been neglected
Sun's classification	It takes into consideration tumor volume and tumor localization relative to the dura and spinal canal	1) The relationship between the tumor and surrounding structure is not clear 2) The surgical guidance of this classification is not described clearly 3) It is not comprehensive
SRIDHAR's classification	1) It aims to define giant spinal schwannomas and discuss their surgical management 2) The focus of this classification is on the relationship between the tumor and the vertebral body and foramen	The membranous hierarchy of spinal schwannomas has been neglected
Jinnai's classification	1) It describes the relationship between the tumor and the dura mater and/or intervertebral foramen 2) It improves the GTR rates of tumors to some extent 3. It tries to use the growth patterns of schwannomas to explain the clinical symptoms	1) It does not indicate the surgical guidance based on the classification 2) It does not pay attention to the arachnoid 3) It does not systematically explain the growth model of schwannomas

portion of the tumor was resected by the oncological surgeons, and the intraspinal part by the neurosurgical team verse imagen No. 2. The outcome was good, with no major deficit after surgery, blood loss of 300 mL, surgical time 220 minutes. No incidences during electrophysiological monitoring, and discharge 72

hours after. Histopathological diagnosis: Classic Schwannomaver imagen 3.

4. Clinical Case 2

Female patient, 55-year-old, presented to neurosurgical consultation, complaining of paravertebral numbness and burning sensation in the dorsal region, T6 to T8 levels. She didn't refer weakness or any motor deficit. MRI images were obtained, they showed a paravertebral tumor with intra and extra spinal growth, it was isointense in T1, hyperintense in T2, and presented enhancement to gadolinium, see image 4 [A) - B)]. The mass occupied more than two vertebrae in length and grew into foraminal structures but didn't compress the spinal cord.

The case involved mediastinal structures; an interdisciplinary session was called. Video Assisted Thoracoscopic approach was decided. Selective intubation and VATS approach was performed by the thoracic surgery team. Our neurosurgical team performed a microscopical approach, with Gross Total Resection achieved, please see image 4 [C), D) and E)]. No deficits appared after surgery, with good surgical outcomes. surgical time 150 minutes, blood loss 120 cc, no complications. The patient was discharged 4 days later, and follow up was done 1 month later, with good improvement of the symptoms, en la image 4 [F), G), H), I)]. The control tomography in axial and axial section can be observed, as well as the post-surgical images with visualization of the post-surgical scar. The histopathology images reported thoracic schwannoma, see image 5.

5. Discussion

As mentioned in the body of the text, giant schwannomas are infrequent, however, most of their life they are asymptomatic, when they show sintomatolgy probably involve important structures, the direction in which they grow as well as the structures they involve and the vertebral levels they occupy will determine the complexity of the procedure for their extirpation, other surgeons and anatomists have tried to establish scales or classifications to facilitate decision making when facing them, however according to the literature review none of them has proven to be completely useful given the limitations they have shown, it is not the purpose of this article to discuss the limitations of one or the other, however, Table 1 has been attached to briefly discuss them. It is important during the study of each particular case to have adequate imaging studies CT, MRI, angiography, and or those who agree, electromiography to diagnose the established damage and as a prognosis, anatomical knowledge of the surrounding structures and to have an interdisciplinary team, considering that there is no surgeon who dominates all body areas or known all the different techniques, each action taken based on scientific knowledge can make a difference in the outcome of the patient.

6. Conclusion

Although infrequent, it's convenient to understand the clinical behavior and treat-

ment of giant spinal schwannomas. They can present a wide range of signs and symptoms, with a variable asymptomatic period. Available classifications try to set a guideline to facilitate therapeutic choices and surgical planning. Optimal management may involve multidisciplinary teams, and a variety of surgical techniques depending on the location and structures damaged by the tumor. Thorough planning is important to improve postoperative outcomes and quality of life in giant spinal schwannomas.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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