



Giant Schwannoma with Lumbosacral Scalloping in a Teenager: A Case Report and Literature Review

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Abstract

Scalloping describes an exaggeration of the concavity of the posterior surface of vertebral bodies secondary to tumoral growth seen in radiological images and is the result of bone erosion and deformation by tumor. It is associated with some types of tumors like Giant Schwannomas.

These tumors represent type II, IVa and V of classification of spinal Schwannomas by Schridar and define high risk of recurrence. Management is based on the most complete resection possible and the affected spinal segment stability. This entity is rare and unusual in children and teenagers. At these ages it must be suspected that it could be secondary to syndromes as neurofibromatosis type 1 or 2. We present the case of 14 years-old boy with an expansive mass at L5-S1 lumbar segment with radiological scalloping on the bone affected. He was operated by laminectomy, tumoral resection and bilateral and lumbosacral bilateral arthrodesis.

At the procedure intraoperative neurophysiological monitoring was used. Histological diagnostic was typical Schwannoma.

Introduction

Schwannomas are tumors that arise from Schwann cells of peripheral or cranial nerve sheath [1]. They comprise 30% of intradural extramedullary primary spinal lesions and usually benign behavior.

On the other hand, there exists a rare subgroup called Giant invasive spinal Schwannomas that despite being behavior, usually erodes vertebral bodies and can extend at extraspinal space, being described its appearance at any age [2].

Therefore, elaborate a multidisciplinary plan that reaches affected spinal segment stability and most complete resection possible is the main goal of the surgical treatment. In this manuscript we present a child affected by giant invasive spinal Schwannoma at sacral segment which could be resected to safe limits and we describe how affected spinal segment could be stabilized with posterior lumbosacral fusion. In addition postoperative genetic research was realized given the unusual histopathology at this age.

Case Presentation

A 14 year old boy is admitted with lumbar pain irradiating to the left lower limb of a year, associated with muscle weakness and gait disturbance. An examination reveals internal and anterior quadriceps surface hypotrophy of left leg, grade 4/5 muscle strength of quadriceps, hamstrings and anterior tibial muscle; as well as hypoesthesia from knees to toes.

A lumbosacral magnetic resonance is done where a 5.57 cm × 5.57 cm × 5 cm lesions and whose appearance Schwannoma was as can be observed at Figure 1.

Radiological Study was completed by a lumbosacral CT where an erosive sacral lesion is displayed as seen as Figure 2; and an electroneurography and electromyography that show mild signs of chronic neuropathy at left S1 root. During the surgical intervention, we positioned the patient prone and assisted with combined Motor-Evoked Potentials (MEPs) and Somatosensory-Evoked Potentials (SEPs). Middle skin lumbotomy and paravertebral muscle dissection was performed until localization of L5 and S1 lamina by fluoroscopy. At this moment bone erosion at S1 level

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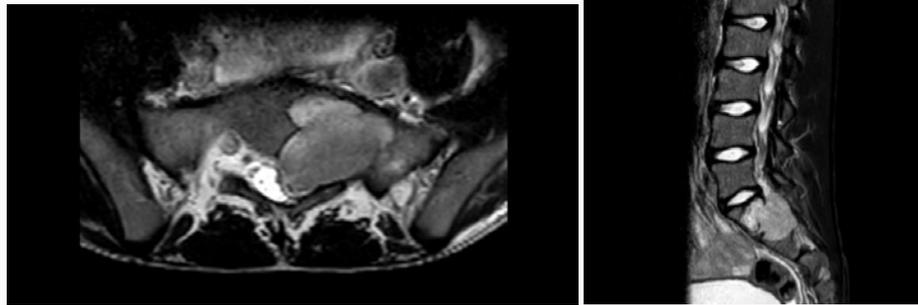


Figure 1: Axial and sagittal slice from preoperative MRI where a lesion had increased signal intensity at STIR sequence and heterogeneous gadolinium-enhanced T1 image, suggestive of Schwannoma.

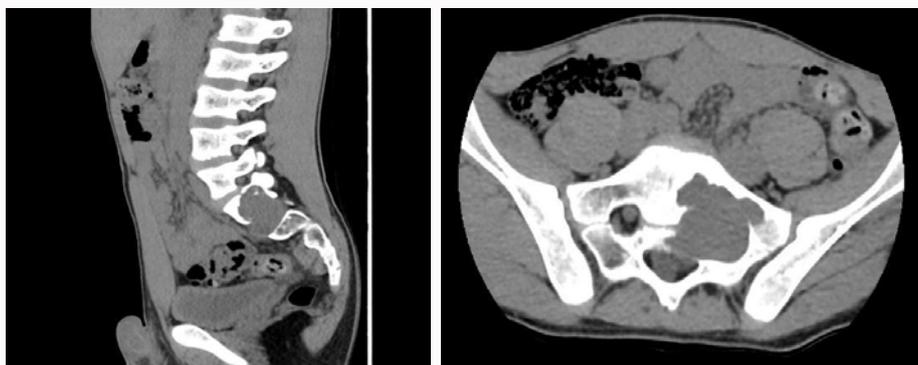


Figure 2: Sagittal and axial slice from lumbosacral CT where an expansive lesion affects S1 and S2 level inducing joint lysis and spreading across sacral foramen near left iliac vessels.



Figure 3: Lumbosacral rx plains where left side pedicle screw L4-L5 and iliac screw fusion is displayed while right side pedicle screw L4-L5-S1 fusion is displayed.

was appreciated. After L5-S2 laminectomy, intratumoral resection by ultrasonic aspiration begins until anterior sacral surface is reached. Given the relationship with iliac vessels and viscera anterior to sacrum, the tumoral fragment attached to the anterior sacral surface must be left.

Biomechanical stabilization of lumbosacral region was made by right side pedicle screw L4-S1 fusion and left side pedicle screw L4-L5 and iliac screw fusion in addition with autologous bone (Figure 3). Postoperative time was unremarkable and the patient was discharged at fifth day with autonomously gait and left inferior limb strength improved.

Histological result was a biphasic aspect tumor where compact fusiform cells area with palisade nuclei; as well as more cellular area without necrosis or mitosis figures. This aspect is typical of Schwannoma. Postoperative MRI displayed a subtotal tumor resection and spinal segment stabilized (Figure 4).

Discussion

Although Spinal Schwannomas are a well studied benign pathology, Giant Schwannomas have very low incidence which management isn't well established. In addition, cases reported aren't always the same age as conventional ones. At the present report, a 14 years-old boy with a Giant Schwannoma from sacral roots is treated,

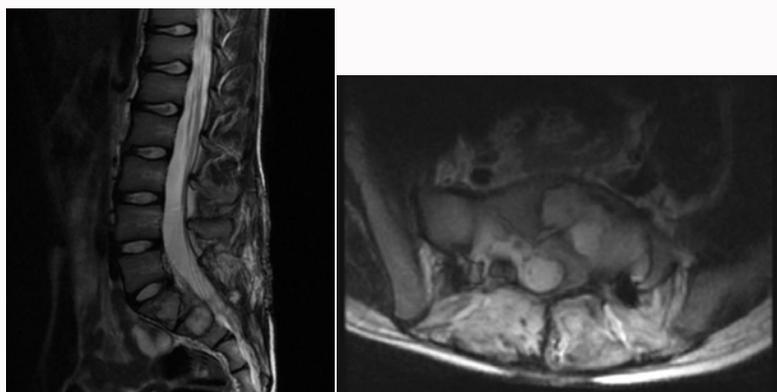


Figure 4: Six-month postoperative study. Lumbar MRI sagittal and Axial Stir sequences slice where subtotal lesion resection is displayed because of fragmentation at anterior sacral surface. Spinal segment does not display instability radiological signs.

an unusual age group given that in literature these cases usually are at adulthood [2-5].

If lesions like Giant Schwannomas are found at the pediatric age, it must be suspected that it could exist as an underlying genetic disorder. Most related to this tumor is type 2 Neurofibromatosis [6]; however, type 1 neurofibromatosis is closely related to scalloping on the vertebral body [7].

Actual patient did not have a family background and the genetic study did not show mutations related to genetic disorders named before.

Scalloping is a radiologic term initially described at ray-X films of spine and it has been subsequently extended to another radiological image test like Computerized tomography or magnetic resonance. It represents an exaggeration of the concavity of the posterior surface of one or more vertebral bodies secondary to bone deformity. At neurofibromatosis it would be secondary to dural ectasia same as other connective tissue disorders like Marfan syndrome or Ehlers-Danlos Syndrome [7,8].

If Schwannoma is not related to genetic disorders, scalloping is secondary to osseous remodeling by raised intraspinal pressure at low growth masses.

With the purpose of establishing best management, it has been tried several classifications for these lesions. Sridhar [9] mentions that Giant Schwannomas fit at Type II (>2 vertebral bodies), IVb (Extradural extension >25 mm) or V (eroding osseous planes and myofascial plane extension). He describes that subtotal resection of this lesion represents high risk recurrency in addition to other factors like youth age, tumoral measure or lumbar segments affection [2]. Present Schwannoma, could be classified like type V, given that anatomical extension required a subtotal resection, patient must keep a close follow-up inasmuch as high risk of recurrency [10-12].

According to the literature, Sacral resection between S1 and S2 vertebrae generates a 30% pelvic instability; rising up to 50% when resection extends 1 cm below sacral promontory [4,13]. These dates show that bone erosion secondary to these tumors in addition to the most complete resection possible, generates great instability to the lumbosacral-iliac segment. Therefore the biggest problem with these surgical interventions is keeping the biomechanical stable spine at the spine segment affected.

Due to the patient characteristics and the damage by the tumor,

a posterior lumbosacral approach was performed with intraoperative neurophysiological monitoring where a transpedicular screws from L4 to S1 were fixed at non-eroded side; and changed to an iliac screw at eroded side given the sacral damage.

The justification for this instrumentation is based on that S1 vertebrae support the most of axial load given that it represents the main link between pelvis and spine. Using an iliac screw, load is redistributed to the pelvis, generating a solid and balanced build [2].

Despite that other authors' purpose is circumferentially arthrodesis and others combined approach for this type of tumors, in this case main principle of this treatment was achieved: one time, on approach and safe, wide resection at the same time that spinal stability keeps [14,15].

Conclusion

Giant Spinal Schwannomas are a rare variety of Schwannomas that make vertebral scalloping characteristically, inducing bone erosion and, therefore, spinal instability. The most complete resection with the solid rebuilding of the affected spinal segment is the main goal at the management of this type of tumor.

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