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Resection of giant invasive thoracic schwannoma. Case report

A. Khelifa[1], F. Aichaoui[1], I. Assoumane[2], M. Al-Zekri[1], T. Bennafaa[1], A. Morsli[1]

[1] Neurosurgical Department of BEO University Hospital, Algiers, Algeria
[2] Department of Surgery, National Hospital of Niamey, Niger

Keywords
schwannoma,
nerve sheath tumours,
spinal cord compression

Abstract

Even though spinal nerve sheath tumours, presented especially by schwannomas, are considered to be mostly benign; they can gain a huge size and have an invasive behaviour, causing spinal cord compression, bone destruction, and make the total removal of the tumour a real challenge for the surgeon. This type of tumours is recently described as giant invasive spinal schwannoma (GISS), this type rarely reported in the thoracic region; deserve a special studying vis-a-vis the diagnosis and the management of both the tumour and the bone destruction.

Introduction

Even though spinal nerve sheath tumors, presented especially by schwannomas, are considered to be mostly benign; they can gain a huge size and have an invasive behavior, causing spinal cord compression, bone destruction, and make the total removal of the tumor a real challenge for the surgeon. This type of tumors is recently described as giant invasive spinal schwannoma (GISS), this type rarely reported in the thoracic region (2); deserve a special studying vis-a-vis the diagnosis and the management of both the tumor, and the bone destruction.

Case presentation

The patient is a girl of 30 years old without past medical history, who consulted for a weakness of the lower limbs appeared 5 months before she consults and with a recent worsening; the clinical exam at the admission found a patient who present paraplegia with urinary urgency. Spinal CT then MRI was performed, objectified a spinal cord

Corresponding author:
Khelifa Adel

Neurosurgical Department of BEO University Hospital,
Algiers, Algeria

drkhelifaadel@gmail.com

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compression by an intra and extra spinal giant dumped shape process extending to the left thoracic cavity at the level of Th 8 and Th 9, measuring: 73x47x45 mm; hypointense on T1 weighted images, with heterogeneous signal on T2 weighted images and intense enhancement after gadolinium injection; the process was responsible of a destructive scalloping on the left pedicle and partial destruction of the posterior wall of Th 8, the tumor extends posteriorly to the left paraspinal muscles, and on the thoracic cavity in contact with the aorta (figures 1,2). There were no signs of neurofibromatosis. We operated the patient with the help of the thoracic surgery colleagues, under general anesthesia, and selective intubation of the right lung. Together we performed a total removal of the tumor through a combined approach. First a posterior midline approach was performed through which a spinal cord decompression was obtained, and then through a left posterolateral thoracotomy passing through the sixth interrib space a total removal of the tumor was achieved (figure 3). The thoracic part of the tumor was well encapsulated and easily dissected, but the intra spinal part lacks a capsule and was totally excised with piecemeal removal. A thoracic drain was left. The histological exam found a WHO grade I schwannoma. On post operative the patient was diagnosed with an atelectasis of the left lung which was managed with fibroscopic aspiration and steroids. The patient was oriented to physical medicine where she progressively improved, gaining control on her urinary behavior, then 5 weeks later she was able to walk. Post operative imaging performed 5 months later objectified a total removal of the tumor without residual or recurrence (Figure 4).

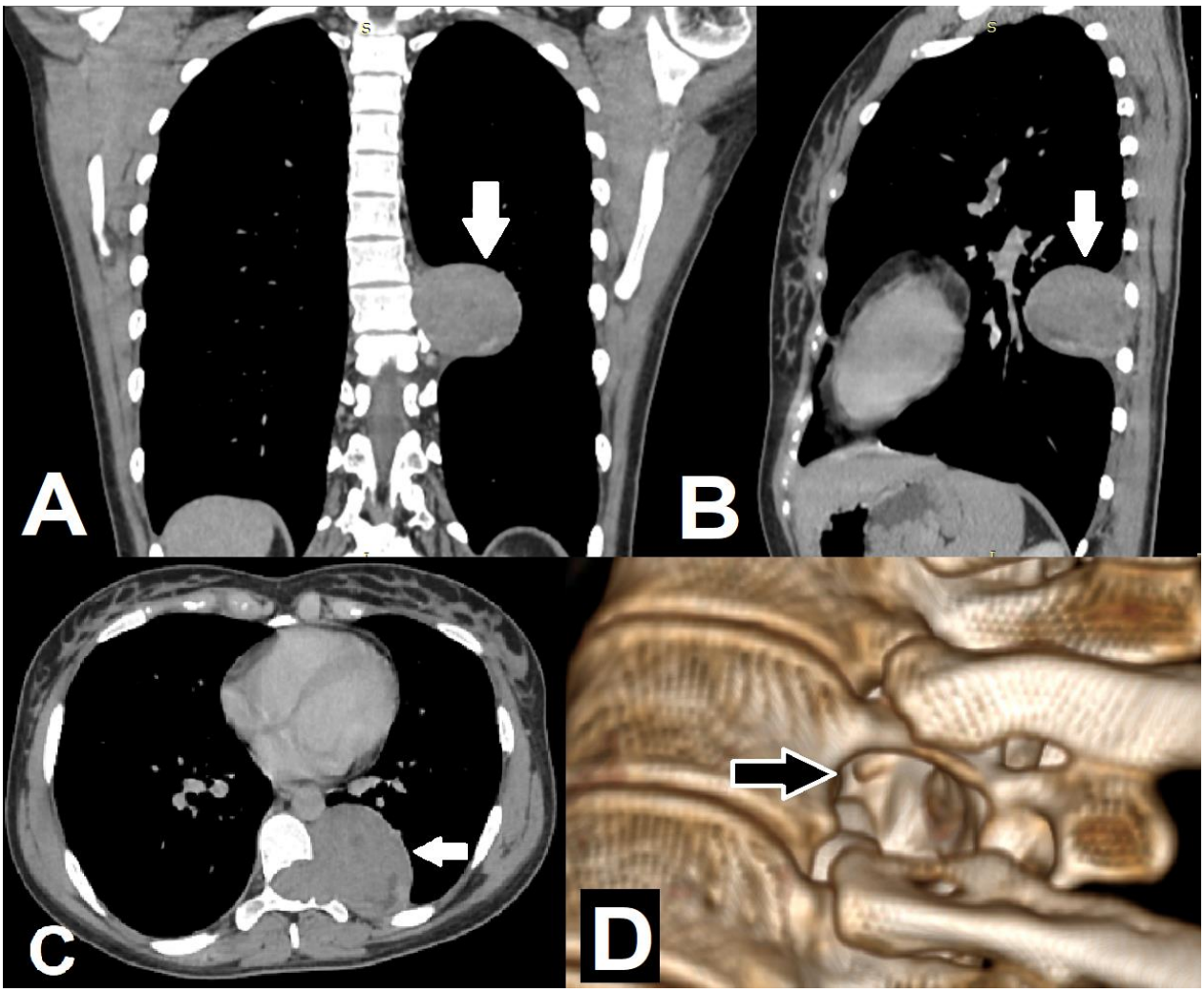

Figure 1. Preoperative thoracic CT; A: coronal reconstruction, B: sagittal reconstruction, C: axial slide; and D: 3D reconstruction. Note the extension of the tumor (white arrow) and the amount of bone destruction on the axial and 3D views.

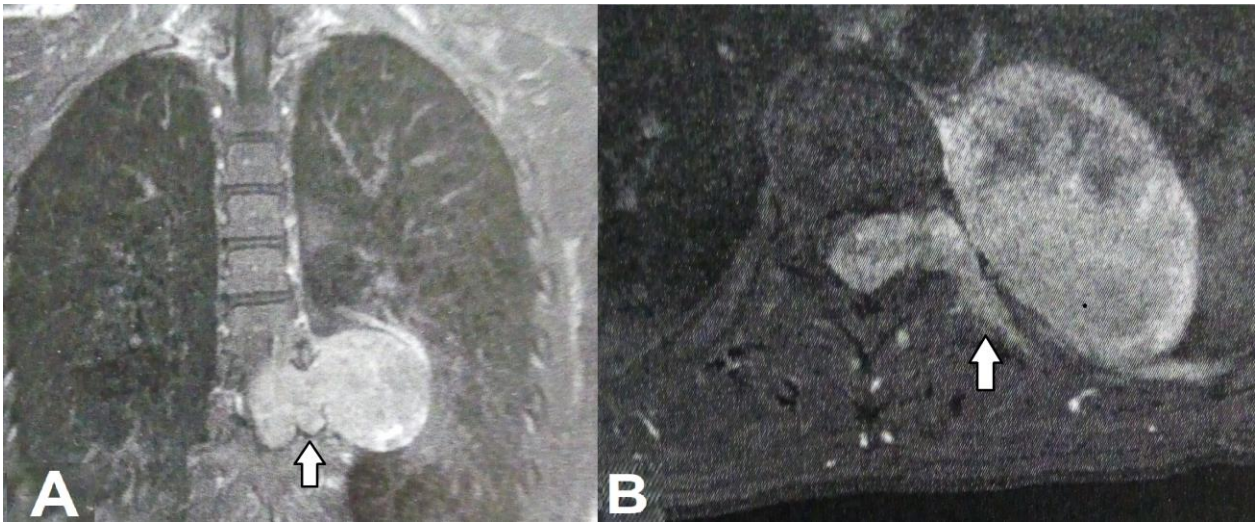

Figure 2. Preoperative T1 injected MRI; A: coronal slide, B: axial slide. Note the soft tissues infiltration (the arrow).

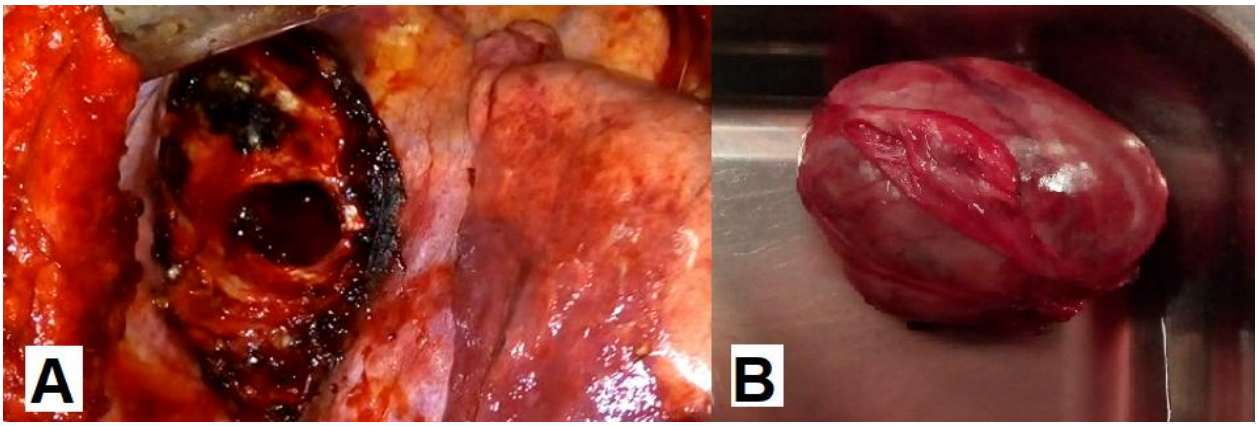

Figure 3. Peroperative views, A: a view through the interrib approach after removing the tumor; B: the intrathoracic part of the tumor. Note on the left image, the widened opening through which the tumor got through to invade the thoracic cavity; the black halo presents the parietal insert of the tumor.

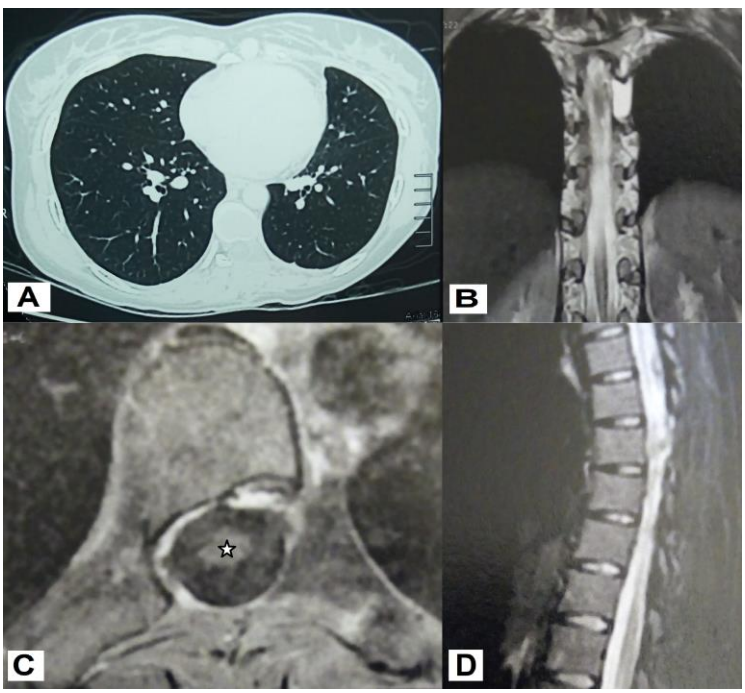

Figure 4. Post-operative images, A: axial slide of a thoracic CT, B: T2 WI MRI coronal slide, C: T1 injected image axial slide, and D: T2 WI sagittal slide. Note the total removal, no residual or recurrence tumor, and the spinal cord decompression (the star).

Discussion

Nerve sheath tumor could be either schwannoma or neurofibroma. Neurofibromas produce a fusiform enlargement of the nerve where schwannomas are more smooth globoid tumors that develop eccentrically (1). Histological exams of schwannomas find usually elongated bipolar cells with fusiform darkly staining nuclei arranged in compact interlacing fascicles that tend to palisade (1). Nerve sheath tumors are mostly intradural but in 10% to 15% they are intra and extra dural in dumbbell shape (1). Nerve sheath tumors are malignant in 2.5% of cases and half of these cases are Neurofibromas (1). In 2001 Sridhar et al proposed a classification for benign nerve sheath tumors with a precise definition of the giant and the invasive spinal schwannomas (table 1); in the same paper the authors reported for the first time, cases of giant thoracic invasive spinal schwannomas (2). Giant invasive spinal schwannoma (GISS) although a benign lesion put the surgeon face to many difficulties related to the size, the infiltration, and the invasive nature. Big lesions need more exposure, though the selection of an adequate approach is mandatory. Midline approach might be sufficient (2) but for more exposure a lateral extracavitary or extracavitary costotrans-versectomies approaches provide access to the extraforaminal extension of the tumor (3); for tumor extending beyond the vertebral body a combined approach is recommended (4). Total removal is not possible in all cases (2,4), and that is mainly because the tumor might lack a capsule (2). The complications related to infiltration are represented especially by bone diffusion, in fact, some amount of bone destruction might jeopardize spinal stability and though the protection of neural structures, motion, and might cause some deformities. The evaluation of the consequences of bone infiltration classes these lesions into stable and unstable. Classically spine surgeons use Denis classification based on the three columns; Kostuik divided those columns on tow, right and left zones and considered a destruction of three or more of these six zones as an unstable lesion (5). Only instable lesions need spinal instrumentation. The lesion of our patient is considered stable, moreover some amount of infiltration is tolerable in the semi rigid spine (from Th3 to Th10) more than in the junctional or on the mobile spine, and that is the first criteria of stability evaluation in "The spinal instability neoplastic score (SINS)" adopted by some spine surgeons (3,6). Total removal of the tumor is the only factor related to long term outcome (2,4), in case where it is not possible, a decompression of neural structures is the priority, but a repeat surgery might be necessary (2).

	Classification
Type I	intraspinal tumor, < 2 vertebral segments in length; a: intradural; b: extradural.
Type II	intraspinal tumor > 2 vertebral segments in length (**giant tumor**)
Type III	intraspinal tumor with extension into nerve root foramen
Type IV	intraspinal tumor with extraspinal extension (dumbbell tumors); a: extraspinal component < 2.5 cm; b: extraspinal component > 2.5 cm (giant tumor)
Type V	tumor with erosion into the vertebral body (**giant invasive tumor**), lateral and posterior extensions into myofascial planes

Table 1. Sridhar et al classification for benign nerve sheath tumors (2).

Conclusion

Giant invasive spinal schwannoma is a benign lesion with high bone destruction potential; its infiltration nature to the surrounding soft tissues could make the total resection a real challenge. Knowing the real instable spinal lesions will orient the indication to spinal instrumentation.

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