

## Giant invasive spinal schwannomas: definition and surgical management

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*Object.* Confusion exists regarding the term giant spinal schwannoma. There are a variety of nerve sheath tumors that, because of their size and extent, justify the label “giant schwannoma.” The authors propose a classification system for spinal schwannomas as a means to define these giant lesions. The classification is confined to tumors that are essentially intraspinal, with or without extraspinal components. Lesions that erode the vertebral bodies (VBs) and extend posteriorly and laterally into the myofascial planes are classified as giant “invasive” spinal schwannomas.

*Methods.* The records of patients with giant invasive spinal schwannoma were analyzed. The radiological features, operative approaches, and intraoperative findings were noted.

Ten patients with giant invasive tumors were surgically treated over the last 8 years. Six patients were male. Erosion of the posterior surface of the VBs was the diagnostic finding demonstrated on plain x-ray films. Magnetic resonance imaging delineated the extent of the tumors and helped in preoperative planning. Radical excision of the tumors in multiple stages was possible in eight of the 10 patients. Dural reconstruction was required in four patients. All patients required fusion, and an additional stabilization procedure was undertaken in three patients.

*Conclusions.* The authors conclude that giant invasive schwannomas are uncommon lesions and propose a new classification system. Because of their locally “invasive” nature and extension in all directions, careful preoperative planning of the surgical approach is very important. Although radical excision is possible and promises good results, recurrences may occur and multiple surgical procedures may be required.

**KEY WORDS** • giant schwannoma • spine • surgical approach • reconstructive surgery • classification system

**S**PINAL schwannomas comprise approximately 25% of all spinal tumors.<sup>2,5</sup> They are most commonly seen in the thoracic region and are then most commonly found, with almost equal frequency, in the cervical and the lumbar regions. Until this report, no attempt had been made to classify these lesions on the basis of their location and extent. This has resulted in confusion in the use of the term giant spinal schwannoma. By convention, the term refers only to lumbosacral and sacral tumors eroding the VBs and extending into the paraspinal tissues. There are, however, other categories of tumors, that, due to their size and extent, justify the designator giant schwannoma. The authors propose a classification system of spinal schwannomas, in which a giant tumor is defined and discuss the surgical management of patients with giant invasive spinal schwannomas.

### Clinical Material and Methods

The case records of patients with spinal schwannomas surgically treated at our center between 1990 and 1999

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*Abbreviations used in this paper:* MR = magnetic resonance; VB = vertebral body.

were analyzed retrospectively. The lesions were diagnosed by radiography, myelography, and/or MR imaging.

### Classification of Spinal Schwannomas

Based on the radiological findings, the authors have devised a classification system of spinal schwannomas (Table 1 and Fig. 1). Giant spinal schwannomas are defined as those that extend over more than two vertebral levels (Type II), those that have an extraspinal extension of more than 2.5 cm (giant dumbbell Type IVb), and those lesions that erode the VBs and extend posteriorly and laterally into the myofascial planes (giant invasive tumors, Type V). In the present study we focus on patients with the giant invasive, or Type V, spinal schwannomas.

### Results

Ten patients underwent resection of their giant invasive schwannomas in the last 8 years (Table 2). They formed 10.9% of the cases of spinal neuromas surgically treated at our center during the same period. Six of the 10 patients were male. The youngest patient was 13 years of age and the oldest was 60 years of age (mean age 33.8 years). Only one of the patients had neurofibromatosis.

## Giant invasive spinal schwannomas

TABLE 1  
Classification of benign nerve sheath tumors

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 w/ extension into nerve root foramen
Type IV	intraspinal tumor w/ extraspinal extension (dumbbell tumors); a: extraspinal component < 2.5 cm; b: extraspinal component > 2.5 cm (giant tumor)
Type V	tumor w/ erosion into the VBs (giant invasive tumor), lat & posterior extensions into myofascial planes

Findings on plain radiographs included a widened interpedicular distance, erosion of the pedicle, enlargement of the intervertebral foramen, and erosion of the posterior surface of the VBs to a variable extent. The latter finding was diagnostic of a giant invasive schwannoma. In two patients a total block of the contrast column demonstrated on myelography was the definitive indicator of giant schwannoma. On MR imaging the lesions appeared isointense or of varying intensity on T<sub>1</sub>-weighted images, and the entire extent of the lesion, including the extraspinal extensions and erosion of the VBs, was delineated.

Five of the lesions were located in the lumbar region, two each in the thoracic and lumbosacral regions, and one in the cervical region. Three or more operations were performed in three patients, two in four patients, and one in three patients.

Radical microscopic excision of the tumor was achieved in eight patients, and dural reconstruction was necessary in four patients. Fusion in which we used autologous bone was performed in all patients. Three patients required an additional stabilization procedure, as it was believed that the spine was unstable. Bed rest for 3 months was ordered for one patient after which mobilization was allowed after application of an external brace. One patient developed a postoperative cerebrospinal fluid leak through the wound, which subsided following repeated lumbar punctures. Follow-up periods ranged from 6 months to 4 years (mean 20 months). One patient with a lumbar tumor developed an asymptomatic recurrence after 3 years and has been advised to undergo repeated surgery.

### Illustrative Cases

Two patients who presented with problems requiring different approaches and management are described in detail.

#### Case 5

This 25-year-old woman presented with a 5-month history of progressive quadriparesis.

**History.** She had undergone two previous surgeries—one at age 16 years and one at age 19 years—for similar complaints. A myelogram obtained prior to the first operation had demonstrated a total block at the lower border of the C-6 VB. A C3–7 laminectomy had been performed, and radical excision of the intraspinal portion of an intra-

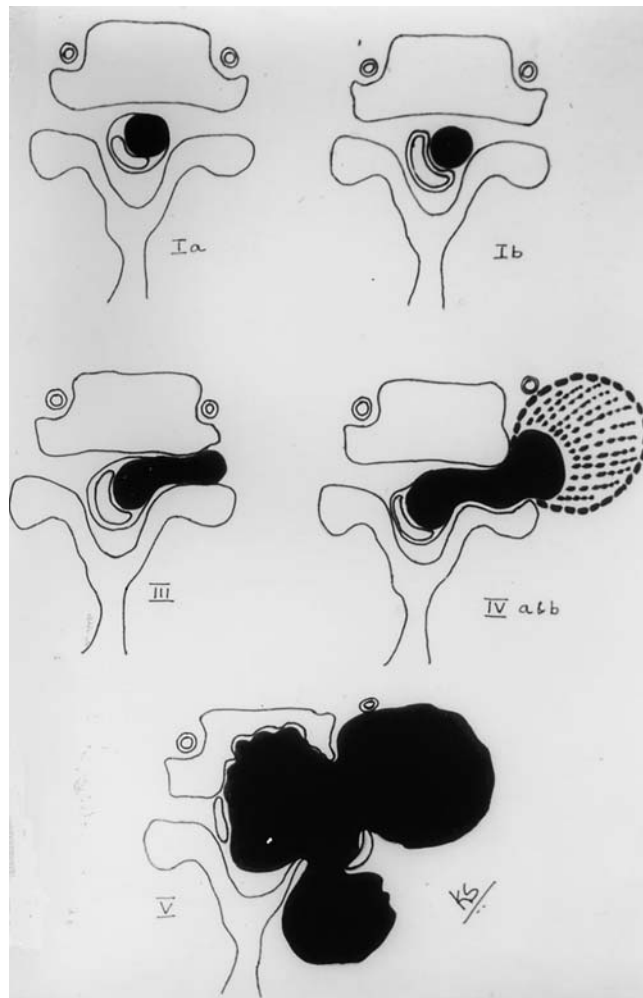


FIG. 1. Diagrammatic representation of the proposed classification of spinal schwannomas Types I to V. Type II (not shown) is an intraspinal schwannoma extending for more than two vertebral segments. See Table 1 for definitions.

extradural schwannoma was accomplished. She regained normal power and sensation within 8 months of surgery. At the age of 19 years she was admitted with a progressively increasing swelling in the neck. The lesion was approached anterolaterally, and a near-total excision of the extradural schwannoma was achieved. A remnant of the tumor was left at the region of the nerve root foramen.

**Examination.** Plain radiography performed at the present admission demonstrated the C3–7 laminectomy, as well as a swan-neck deformity of the cervical spine with localized porosis of the C-6 VB (Fig. 2 upper left). Magnetic resonance imaging of the cervical spine revealed a lobulated lesion isointense in relation to the spinal cord on T<sub>1</sub>-weighted images and of a mixed intensity on T<sub>2</sub>-weighted images, extending from the lower border of the C-4 VB to the middle of the C-7 VB. The lesion had eroded the C-6 VB and was extending through the left intervertebral foramen into the neck with a large extraspinal component. The swan-neck deformity was causing pressure on the cord contralateral to the upper border of C-4, at a level where there was no tumor (Fig. 2 upper right and lower left).

TABLE 2  
 Characteristics of patients with a giant  
 spinal neurofibroma/schwannoma

Case No.	Age (yrs), Sex	Region	No. of Ops	Stabilization Required
1	55, M	lumbar	3	yes
2	25, M	lumbosacral	5	no
3	55, M	thoracic	2	no
4	60, M	lumbar	2	yes
5	25, F	cervical	4	yes
6	15, F	lumbar	2	no
7	30, F	thoracic	1	no
8	19, M	lumbar	1	no
9*	40, F	lumbosacral	2	no
10	13, M	lumbar	1	no

\* This patient had neurofibromatosis.

**Operation.** A posterior approach to the tumor was undertaken as the first step. The previous laminectomy wound was reopened only on the left side where the tumor was exiting from the spinal canal. The bone removal was extended laterally to include the facet joints at C-5 and C-6 to facilitate removal of the extraspinal part of the tumor. Radical excision of the intra- and extraspinal portions of the tumor was performed, leaving only tags of lesion in the eroded C-6 VB. A rib graft was used for fusion across the operated levels. The patient was placed in traction postoperatively for 4 days, after which the anterior approach was used, and a three-level corpectomy (C4-6) was performed to correct the swan-neck deformity and to remove the tumor remnants in the C-6 VB. An anterior C3-7 plate was placed to stabilize the spine (Fig. 2 lower right).

**Follow-Up Course.** The patient made excellent progress in the postoperative period and was mobilized wearing a cervical collar on the 3rd postoperative day. At the last follow-up visit, 30 months after surgery, the patient was asymptomatic and neurologically normal. An x-ray film demonstrated good fusion and implant position.

#### Case 6

**History.** This 15-year-old girl had undergone surgery at another center 1 year prior to presentation to us for back pain, symptoms and signs of a conus medullaris-cauda equina lesion, and urinary retention. The lesion was a giant schwannoma of the lumbar spine. An L2-5 laminectomy had been performed, but the surgery was abandoned following decompression of the tumor because the surgeon had believed that the lesion engulfed the nerve roots.

**Presentation.** The patient presented to us with an increasing lower-limb weakness and had received an indwelling catheter.

**Examination.** Plain radiography of the lumbosacral spine demonstrated the L2-5 laminectomy, posterior scalloping of the L-3 and L-4 VBs of greater than 60% of the body, and a scoliotic curve of the lumbar spine. Magnetic resonance imaging demonstrated the multilobulated lesion extending from the upper border of the L-2 VB to the lower border of L-5 (Fig. 3 left). The tumor was shown on MR imaging to be eroding the L-3 and L-4 VBs and extending to the extraspinal tissues on the left side only

(Fig. 3 center). The lesion extended through the laminectomy up to the subcutaneous plane. A pseudomeningo-coele was observed posterior to the lesion.

**Operation.** The tumor was exposed via a posterolateral approach on the left side where it was exiting from the spinal canal. In addition, the facet joint at that level was also removed. The dura was deficient over most of the lesion, and the tumor was in direct contact with the muscles. Normal dura was exposed at the superior and inferior ends of the lesion as well as the superior and inferior poles of the tumor. Using the operating microscope, the tumor was debulked and space was created to dissect the lesion radically. It was found that the cauda equina was pushed posteriorly and to the right by the lesion. There was no difficulty in dissecting the tumor from the nerve roots, and the tumor was excised radically (Fig. 3 right). The tumor had eroded the VBs up to the anterior vertebral margin, leaving behind a thin shell of bone and intact intervertebral discs. Autologous bone chips were packed into the defect in the VBs. The edges of the normal dura were identified, and using a fascia lata graft, reconstruction of the dural tube was performed.

**Follow-Up Course.** The patient's neurological condition improved in the postoperative period, and by the end of 2 weeks normal power and lower-limb sensation and normal bladder function had returned. The patient was mobilized after 3 months, and she wore a lumbar brace. As of the 18-month follow up, she was mobile and experiencing a minimal residual neurological deficit.

#### Discussion

Spinal nerve sheath tumors are most often single, small, benign lesions that are relatively simple to remove and are associated with a good postoperative result. Giant schwannomas are uncommon. The majority of giant spinal schwannomas are dumbbell shaped, which poses difficulty mainly in the radical removal of the extraspinal part of the lesion. Various approaches have been conducted to help remove these tumors totally.<sup>3,4,6</sup>

Giant invasive (Type V) schwannomas differ from the other giant schwannomas in that they erode the posterior surface of the VBs, infiltrate through the posterior dura, and invade the myofascial planes while still remaining histologically benign. They are reported most often as case reports, and in many large series of spinal schwannomas the authors make no mention of these lesions. Rao<sup>8</sup> and Levy, et al.,<sup>5</sup> while reporting on 80 and 66 spinal schwannomas, respectively, do not refer to these giant tumors. Giant invasive tumors have been described almost exclusively in the lumbosacral spine and as intrasacral lesions.<sup>1,2,7</sup> There have been no reports in the literature of a giant invasive nerve sheath tumor in the thoracic or cervical spine.

Giant invasive (Type V) schwannomas make surgery difficult because of their growth in all directions. They extend 1) commonly over more than three vertebral levels; 2) anterolaterally into the extraspinal space via the foramen, which they erode and widen; 3) posteriorly, thinning and attenuating the dura and the posterior elements and, occasionally, extending into the posterior myofascial planes; and 4) anteriorly, eroding the VBs to varying extents. These extensions cause problems for the surgeon in terms of approach, resectability, and stability of the spine.

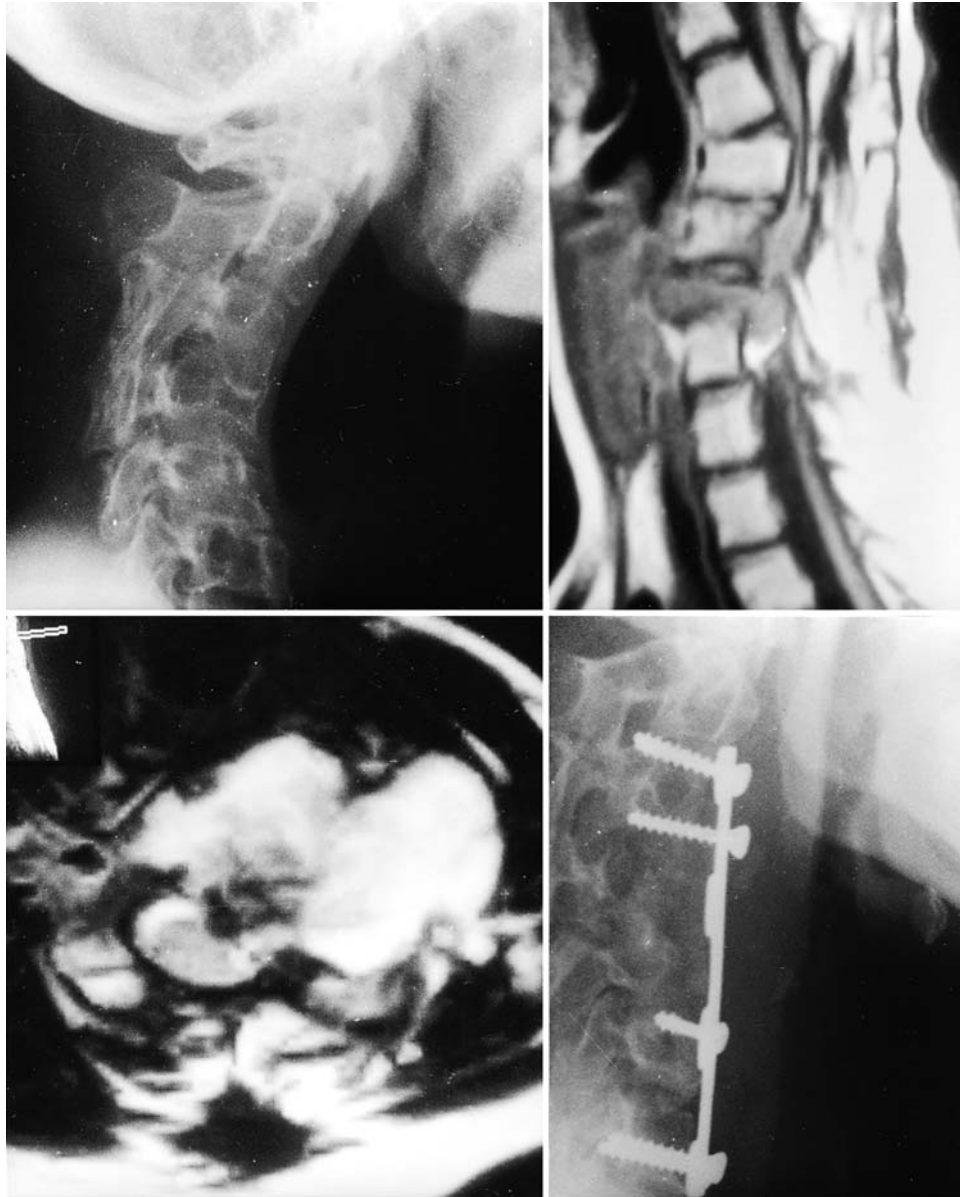


FIG. 2. Case 5. Imaging studies. *Upper Left:* Lateral x-ray film of the cervical spine demonstrating evidence of a previous laminectomy, a kyphotic deformity of the spine, and a localized porosis of the C-6 VB. *Upper Right:* Sagittal T<sub>1</sub>-weighted MR image revealing an intraspinal tumor opposite the C-5 and C-6 VBs and extending anteriorly into the C-6 VB. The kyphotic deformity is causing spinal cord compression opposite the C-4 VB. *Lower Left:* Axial T<sub>2</sub>-weighted MR image demonstrating the hyperintense tumor extending into and eroding the C-6 VB and extending extraspinally through the left intervertebral foramen. *Lower Right:* Postoperative lateral x-ray film obtained following radical excision of the tumor via both the posterior and anterior approaches. Correction of the kyphotic deformity was achieved as well as total excision of the tumor with a median corpectomy from C-3 to C-6. An anterior plate was placed from C-2 to C-7.

#### *Approach and Resectability*

Magnetic resonance imaging has made the preoperative planning of giant tumors easier than when myelography was the most commonly used neurodiagnostic tool. The entire extent of the lesion is seen in all three planes, and the relationship of the tumor to the neural elements, vascular structures, and other organs is clearly delineated. Plain radiographs and/or computerized tomography scans

reveal clearly the bone destruction and are required to evaluate the stability of the spine.

Radical excision is difficult because these tumors lack a capsule that would help in dissecting them from the surrounding structures. It has been the observation of some authors, however, that these tumors often engulf the nerve roots of the cauda equina.<sup>2</sup>

We have found that the relationship between the tumor and the neural structures is constant. Depending on the

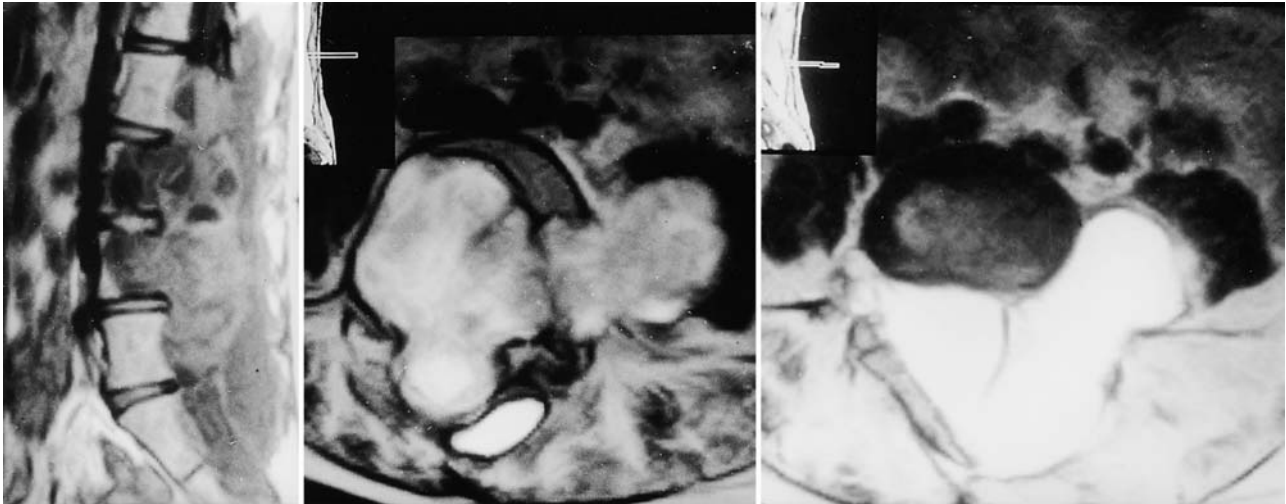


FIG. 3. Case 6. Magnetic resonance imaging studies. *Left:* Sagittal T<sub>1</sub>-weighted image demonstrating a mixed-intensity lesion occupying the entire spinal canal from L-2 to L-5 and eroding and extending into the L-3 and L-4 VBs. *Center:* Axial T<sub>2</sub>-weighted image demonstrating the tumor extending from the spinal canal into the VB, as well as lateral extraspinal extension through the left intervertebral foramen, and posteriorly into the myofascial planes. The tumor is capped posteriorly by a small pseudomeningocele related to previous surgery. *Right:* Axial T<sub>2</sub>-weighted image revealing total excision of the lesion. There is a meningocele extending laterally through the foramen. The reconstructed dura is seen as a thin line.

side of origin of the tumor, the cord/nerve roots are pushed and/or splayed to the opposite direction. On MR imaging it is seen that the tumors extend laterally most often only on one side and that the cord and/or cauda equina is pushed to the opposite side. This relationship is more clearly observed on the axial cuts obtained at either pole of the lesion.

It is, therefore, possible to perform a radical excision of the tumor without causing neurological damage. By approaching the lesion in the midline, the surgeon may encounter the splayed nerve roots first and it may appear as though the nerves are engulfed by the lesion. Using a posterolateral approach on the side of the tumor, initial debulking can be performed to reduce the size, create more space, and then allow the tumor to be separated from the neural elements. We have been able to separate the tumor from the cord and/or nerve roots without any difficulty when using this approach.

Dissecting the tumors from the surrounding soft tissue may be difficult when the tumors do not have a firm capsule and are vascular. The anatomy of the region is confusing when an essentially intradural tumor is seen at the muscle plane without the dural layer to delineate it. In such cases it is always preferable to take the exposure to the normal dura above and below both poles of the tumor and to start the dissection there so that the anatomy of the lesion is understood before the excision is begun.

The tumor may erode into the VBs and may even present as a presacral or retroperitoneal mass.<sup>1,2,7,9-11</sup> The tumor should be followed into the bone and removed. Extraspinal extensions of these tumors are usually multilobulated. Although there is no firm capsule, the surrounding tissues often form a pseudocapsule around the lesion. The bone removal should be extended well lateral into the facet joint and lateral cuts made in the dura to follow the lesion later-

ally. Occasionally, the paraspinal muscles may have to be divided to reach the far end of the extraspinal lesion. It is possible to excise even large extraspinal extensions when using a high-magnification operating microscope and constantly tipping the operating table.

However, even with all these maneuvers, because of the large size, extensive nature, and invasiveness of these tumors, it may not be possible to be certain at surgery that the tumor has been totally removed. The surgeon, therefore, must be aware of the prospect of having to reoperate a number of times.

#### *Reconstruction and Stabilization*

It is important to plan for reconstruction once tumor removal has been achieved. Deficient dura has to be repaired using fascia lata if necessary. Erosion of the VB coupled with removal of the posterior elements with the facet joint at more than one level is likely to leave the spine unstable. Because the VB will slowly reconstitute itself, some surgeons have preferred not to fuse or stabilize the spine.<sup>7</sup> However, if the lesion is in the cervical or upper-lumbar spine, early mobilization may be hazardous if the spinal column has not been stabilized. Erosion of more than 25% of the VB, in our opinion, requires some form of reconstructive procedure. The use of implants, however, is not free of problems, the most important of them being the difficulty of imaging studies during the follow-up period. The surgeon, therefore, has to decide on the use of hardware depending on whether the tumor removal has been radical or not.

Results of radical excision of these large tumors have been encouraging, and most patients do well. Recurrences can be managed with repeat surgeries, and at each attempt an aggressive effort should be made to excise the lesion radically.

## Conclusions

Giant invasive spinal schwannomas are uncommon lesions distinct from other giant spinal schwannomas. Because of their locally invasive nature and extensions in all directions, careful preoperative planning of the surgical approach is very important. Although radical excision of these extensive lesions is possible with good results, they have a tendency to recur, necessitating repeated surgeries.

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