Management of Intradural and Extradural Spinal Schwannomas

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11.1 Introduction

Spinal schwannomas (also called neurilemmomas or neurinomas) are typically benign round encapsulated neoplasms of Schwann cell progenitors in the spine. Schwannomas grow as appendages to the parent dorsal nerve root and contain neoplastic Schwann cells [1, 2]. Schwannomas form two patterns that are classified as Antoni A (ie, highly cellular regions with closely packed elongated nuclei) or Antoni B (ie, poorly cellular and loosely packed regions) [3]. Spinal schwannomas are frequently intradural-extramedullary or extradural [1, 4], although there are rare examples in the literature of intramedullary tumors [5–7]. They are the most common nerve sheath tumors, which comprise roughly one-fourth of primary spinal tumors, and usually present in the fourth through sixth decades of life [4, 8]. Ninety-five percent of Schwannoma cases are believed to be sporadic [9], although multiple tumors can imply a syndrome [10]. The incidence is reported to be 0.3–0.5 per 100,000 individuals [11].

Schwannomas can also be categorized into variants, such as cellular, plexiform, or melanotic. Cellular schwannomas have higher cellularity and mitotic rates than other tumors, which may lead to more erosive localized damage without malignant potential [12]. Older data suggested that these tumors typically arise paravertebrally or from the pelvis [13]. Another form is described as plexiform, which are rarely found in the spine [14]. These tumors usually have Antoni A-type tissue, and are often dermal in younger patients [12]. Melanotic schwannomas are less common and pigmented with spindle and epithelioid cell morphology [12].

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Occasionally, patients will present with multiple schwannomas, which is typically syndromic (eg, neurofibromatosis type II [NF2] or schwannomatosis) [10]. NF2 and schwannomatosis can overlap in terms of phenotype or presentation but they are genetically and molecularly distinct [15]. NF2 is an autosomal dominant multiple neoplasia syndrome that is derived from a mutation to a tumor suppressor gene on chromosome 22 [16]. Patients develop tumors of the central nervous system, including bilateral vestibular schwannomas, as well as ophthalmic and cutaneous lesions [16]. Approximately two-thirds of NF2 patients have spinal tumors, which tend to be more aggressive than other forms of schwannoma [17, 18]. Conversely, schwannomatosis is described as a third form of neurofibromatosis and is defined by the development of multiple schwannomas without vestibular nerve involvement [19, 20]. It can be a sporadic entity, but it can also involve a familial genetic component, such as a mutation on the *INI1/SMARCB1* tumor suppressor gene [21].

We review the current management recommendations for treating patients with intradural or extradural spinal schwannomas with the aim to compile a resource of current evidence for these common tumors. Surgical approaches and strategies for intradural and extradural tumors are discussed, as well as other considerations related to management. Surgical pearls are also provided to help avoid common pitfalls.

11.2 Work–Up & Presentation

Diagnosing spinal schwannomas starts with taking a thorough history, conducting a thorough neurological exam, and electromyogram and nerve conduction velocity test data, if necessary. Furthermore, a neurogram can provide additional anatomical details on the relationship of the tumor, nerve root, and potentially the dura. The most common preoperative symptom for spinal schwannomas is localized pain with or without radiation [1, 11, 22, 23]. Other less frequent symptoms include sensory deficits in a dermatomal distribution, bowel or bladder incontinence, motor weakness, gait disturbances, or muscle atrophy [22]. Rarely, patients have presented with elevated intracranial pressure and associated bilateral papilledema [24].

Schwannomas are often asymptomatic and found incidentally on imaging. Their presentation can be diverse and unusual, but they typically have characteristic features. Schwannomas tend to be hyperintense on T2-weighted imaging and range from hypointense to isointense on T1-weighted imaging [2, 25]. The tumors often have heterogeneous enhancement. A study of 92 patients with schwannomas found that 55.4% of tumors showed fluid signal intensity on T2-weighted magnetic resonance imaging (MRI) and 58.7% showed rim enhancement on contrast T1-weighted imaging [26]. Imaging on computer topography (CT) often exhibits hypo-to-mild hyperdensity but may not be distinguishable from the spinal cord without intrathecal contrast [2]. Furthermore, spinal schwannomas can present with pedicle erosion and remodeling with a widened foraminal diameter [2]. Rarely, schwannomas will present with subarachnoid or subdural hemorrhaging [27, 28], which can be

Table 11.1 Summary of different potential surgical approaches and procedures depending on the spinal region and tumor location

Region	Tumor location	Approach and procedure
Cervical	Ventral (midline)	Anterior approach (eg, corpectomy) with or without posterior fusion
	Ventral (lateral)	Posterior approach (eg, laminectomy) or posterolateral approach (eg, facetectomy) with or without fusion
	Lateral recess	
	Dumbbell	
	Dorsal	
Thoracic	Ventral	Anterior approach (eg, thoracotomy with corpectomy and fusion)
	Lateral	Posterior-based approaches (eg, transpedicular, costotransversectomy and fusion)
	Dumbbell	
	Dorsal	Laminectomy or laminotomy with or without facetectomy and fusion
Lumbar/sacrum	Ventral	Laminectomy or laminotomy with or without facetectomy and fusion
	Lateral	
	Dumbbell	
	Dorsal	

challenging to diagnose [29]. The apparent diffusion coefficient (ADC) can also be useful in certain situations for assessing pre-operatively the benign nature of the tumor.

Different surgical approaches are summarized in Table 11.1.

11.3 Intradural Tumor Management

11.3.1 Intradural Extramedullary Tumors

Schwannomas are encapsulated tumors that splay rather than invade the dorsal sensory root. Intradural extramedullary tumors are the most common presentation, accounting for approximately two-thirds of all spinal schwannomas (Fig. 11.1) [11, 30, 31].

There are many surgical strategies that can be utilized, depending on tumor size and location [32]. In many cases, conventional posterior laminectomy or hemilaminotomy with tumor debulking may be sufficient to achieve gross total resection [22, 33]. Unilateral laminectomy has been shown to result in improved postoperative pain and adequate tumor debulking while maintaining regional stability [34]. In certain patients, minimally invasive options—like interlaminar approaches—may be employed [35, 36].

The location and size of the tumor may dictate the type of surgery indicated. For example, a retrospective review of 110 patients compared the outcomes (blood loss, hospital length of stay, pain scores) for patients who underwent a microsurgical laminectomy, hemilaminectomy, or laminectomy with pedicle screw fixation [37]. The results showed that hemilaminectomy was best suited for removal of cervical

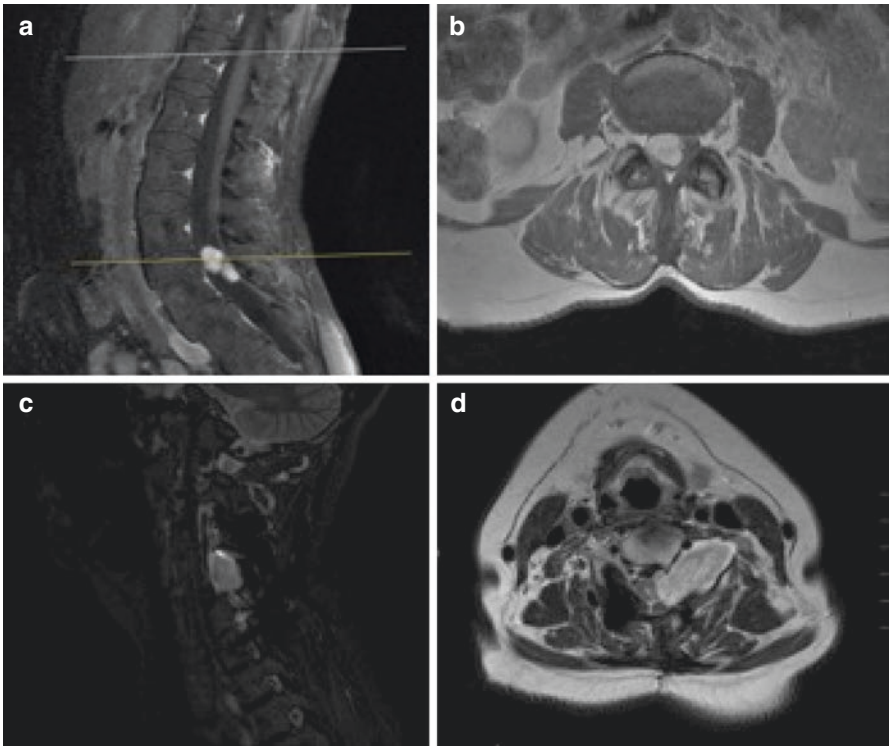

Fig. 11.1 Images showing two patients with intradural tumors. T1-weighted sagittal (**a**) and axial (**b**) images of a patient with an exclusively intradural tumor. Sagittal (**c**) and axial (**d**) images of a patient with both intradural and extradural schwannoma

tumors, laminectomy was best suited for removal of thoracic tumors, and laminectomy with pedicle screw fixation was best suited for removal of lumbar tumors. However, the benefits were minimal and thus appropriate judgment must be used when determining the correct approach and technique.

In some instances, an intradural schwannoma may be ventral to the cervical or thoracic spinal cord and cause compressive myelopathy. In the lumbar region and in select instances in the thoracic region, a posterior approach with only a laminectomy or a posterolateral approach that is medial to the paraspinal muscles is typically adequate for dorsally-located tumor debulking [38, 39]. For tumors in the cervical spine, the anterior approach may be more favorable for ventral canal tumors, given the inability to manipulate the cord [40]. An anterior cervical corpectomy with spinal arthrodesis and fixation has been used to treat a midline ventral intradural schwannoma [41]. Alternatively, a posterior facetectomy may allow for a posterolateral approach to a lateral recess tumor in the cervical spine (Fig. 11.2).

Gross total resection of schwannomas without severe neurological deficits is often achievable. For example, a study of 128 patients who underwent surgery for spinal schwannomas demonstrated that gross total resection was accomplished in

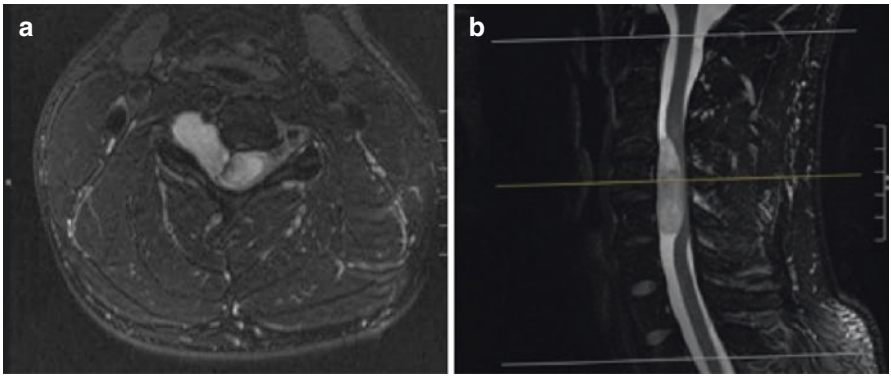

Fig. 11.2 T2 axial (**a**) and T2 sagittal (**b**) MRI images of a 27-year-old man with a dumbbell type schwannoma that extended through the C4–5 foramen. The tumor is amenable to a surgery utilizing a posterolateral approach and posterior facetectomy

97% of tumors [22]. Preservation of the spinal nerve root is feasible, but is sometimes sacrifice of the parent root is necessary to completely resect the tumor. Even when the nerve root is sacrificed, the neurological deficit may be minimal if the affected sensory nerve root only is transected [42]. Although the exact mechanism is unclear, the hypothesis is that as the tumor progresses, the nerve root is damaged and becomes dysfunctional, allowing muscles to be reinnervated by other nerve roots [43]. In support of this hypothesis, a study of 31 patients who required nerve root sacrifice for tumor resection at spinal levels critical for function (ie, C5–T1 and L3–S1) showed that postoperative disabling deficits were minimal, and that the spinal roots that produce the tumors were frequently nonfunctional at the time of surgery [22, 44]. Additionally, intraoperative neural monitoring or direct nerve root stimulation can be used as an adjunct to help assess whether sacrificing the nerve root will result in any postoperative deficit [23].

There may be specific risk factors that predict whether patients will have neurological deficits postoperatively. A study of 64 patients with solitary spinal intradural schwannomas (between T11 and S1) demonstrated that the absence of preoperative lower extremity pain, presence of either preoperative sensory or motor disturbance, or a tumor between T11 and L2 were more likely to have postoperative deficits after concurrent nerve root sacrifice than those without any of these risk factors [45]. Moreover, the authors found that the age, sex, duration of disease, the presence of diabetes, and the tumor length were not predictive of postoperative deficits.

11.3.2 Intramedullary Tumors

Intramedullary schwannomas are rare and represent approximately 1% of cases [1, 2, 11], which is not surprising considering their origin from Schwann cells located

outside the spinal cord. The exact pathogenesis of intramedullary tumors is unclear. A number of hypotheses have been proposed, including derivation from ectopic Schwann cells [46]. Complete resection is not always possible because these tumors can be infiltrative.

Due to the rarity of these tumors, there is no consensus on surgical approach or technique. A posterior approach with a laminectomy or laminotomy can be sufficient for adequate tumor debulking. However, it is crucial to determine the location of tumor origin to ensure a safe surgical corridor [47]. Lee et al. described a case series of ten patients with intramedullary tumors and argued that if the tumor originates from the dorsal root entry zone and not from within the spinal cord, then a myelotomy can be avoided [47]. If the tumor originates from a purely intramedullary location, typically a myelotomy must be performed.

11.4 Extradural Tumor Management

Extradural schwannomas are found outside the thecal sac and occasionally away from the nerve root (Fig. 11.3) [30]. They are relatively uncommon in comparison to intradural extramedullary tumors. Celli et al. sought to establish the clinical presentation, tumor characteristics, and surgical outcomes for extradural schwannomas [30]. They retrospectively reviewed 24 cases at their institution and found that the patients were predominantly women (71%) and tumors were more likely to develop in the cervical and thoracic spine [30]. On rare occasions, extradural schwannomas can develop within the vertebrae and grow into the spinal canal, causing cord compression [48].

Extradural schwannomas have traditionally been removed via an open midline posterior approach with a laminectomy or hemilaminectomy and potential fusion, depending on whether facetectomy is required to debulk the tumor [31, 49, 50]. This strategy is similar to intradural surgical techniques without durotomy and typically is adequate for gross total resection and alleviation of symptoms [11]. Conversely, recent studies have noted that a minimally invasive approach is possible for appropriate tumors with expandable tubular retractors (Fig. 11.4) [50, 51]. Standard microsurgical techniques similar to minimally invasive lumbar microdiscectomy are used [51]. The advantages of a minimally invasive approach include the following: (1) the avoidance of fusion due to lack of facetectomy and iatrogenic spinal instability; (2) less tissue destruction; and (3) less blood loss [51]. Figure 11.5 demonstrates a tumor that is amenable to gross total resection via a minimally invasive spine surgery technique. A potential disadvantage to minimally invasive techniques is there is minimal potential for tumor mapping. Therefore, not all tumors are amendable to minimally invasive techniques, and surgical judgment should be utilized when determining whether an open or minimally invasive approach is appropriate.

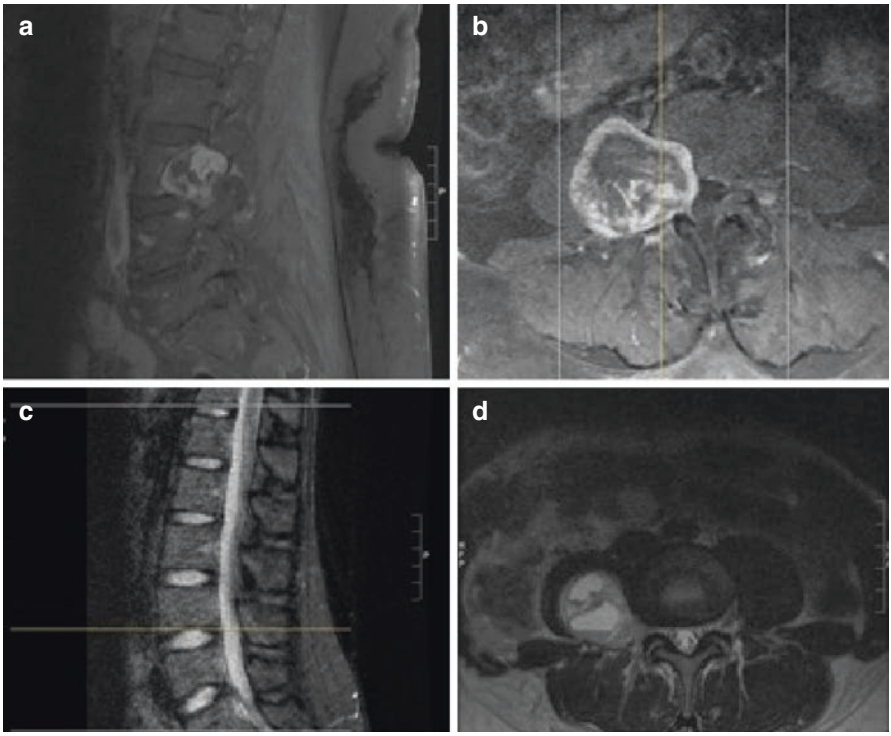

Fig. 11.3 Images showing two patients with extradural schwannomas. Sagittal (**a**) and axial (**b**) MRI sequences of a patient with lumbar right-sided extra-dural schwannoma. Sagittal (**c**) and axial (**d**) views of a right-sided schwannoma invading anteriorly

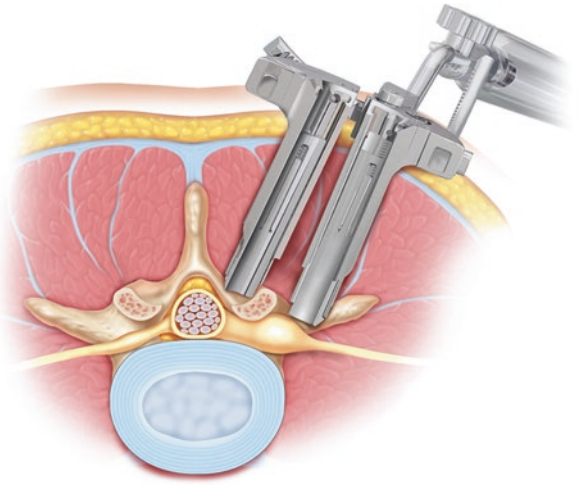

Fig. 11.4 Illustration of a minimally invasive spine surgery approach with an expandable tubular retractor. (Originally published in Lu et al. J Neurosurg Spine 2009 [50]; reprinted with permission)

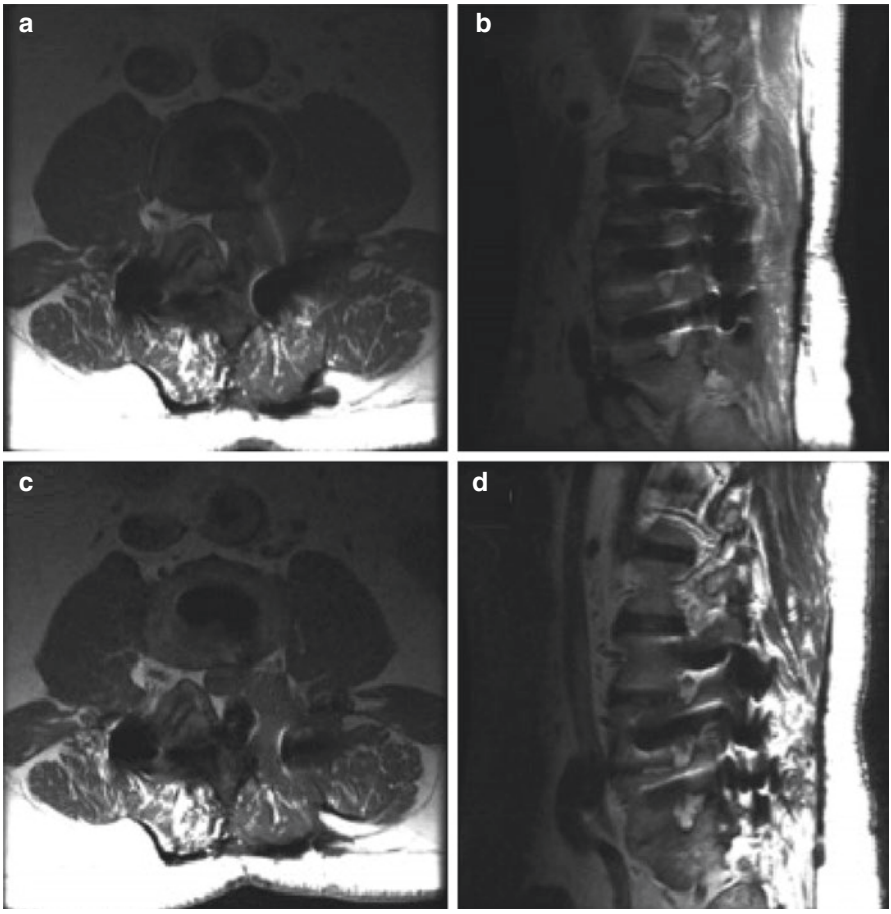

Fig. 11.5 Images showing the results of a minimally invasive approach with an expandable tubular retractor to resect a left L3–L4 schwannoma found 2 years after a spinal fusion. By using a tubular retractor placed via a Wiltse approach, we did not have to remove any of the hardware. Preoperative axial (**a**) and sagittal (**b**) MRI imaging. Postoperative axial (**c**) and sagittal (**d**) images following gross total resection of tumor. (Originally published in Lu et al. J Neurosurg Spine 2009 [50]; reprinted with permission)

11.5 Other Considerations

11.5.1 Dumbbell Tumors

Schwannomas can take the form of a "dumbbell" shape, which is a term to describe a tumor that is split into two different spaces (eg, a tumor that is both intradural and extradural). Dumbbell schwannomas are typically found in the cervical spine [1, 10]. A study of 118 spinal dumbbell tumors found that 18% of schwannomas were

at the C2 nerve root, which was the highest incidence of any nerve root [49]. Dumbbell schwannomas can be destructive to the vertebra and incorporate multiple nerve roots; therefore, some studies recommend removal as soon as identified regardless of whether or not the patient is symptomatic [52].

Although spinal dumbbell tumors are not particularly rare [49], they present surgical challenges because of the unique exposure required. In the cervical spine, a posterior approach can be used, especially if the foramen is involved. A study of 41 cases of dumbbell schwannomas in the subaxial cervical spine found that gross total resection with minimal postoperative neurological deficit was possible, if the extraforaminal component was smaller than 5.4 mm in its longest diameter [53]. Gross total resection is possible by using a modified posterior midline exposure, followed by a single-sided laminectomy and facetectomy, which gives access to the intraspinal, foraminal, and extraforaminal spaces for optimal tumor removal [54]. Others advocate use of a posterior approach to resect as much tumor possible while leaving the foraminal portion unresected with the goal of preservation of the facet joint [55]. The rationale is to maintain low recurrence rates while minimizing potential complications from facet disruption. Dumbbell tumors have been removed through a hemilaminectomy with or without facetectomy and possible fusion, depending on assessment of stability [49]. The use of microsurgical or endoscopic techniques for transforaminal resection may help preserve stability and joint integrity, precluding the need for a fusion [56].

When debulking a dumbbell tumor, it is beneficial to start with resection of the extradural component. The invagination of the dural ring may resemble intradural extension during the operation, but there may not actually be any intradural component [57]. By debulking the extradural portion initially, the dural ring can be carefully examined for intradural extension before the dura is opened. Schwannomas are encapsulated tumors, which allows for potential removal without sacrificing the nerve root or undertaking an unnecessary durotomy.

11.5.2 Sacral Schwannoma

Tumors in the sacrum are rare and often asymptomatic but can present with a variety of neurological deficits like bowel or bladder dysfunction (Fig. 11.6) [58]. Only approximately 50 sacral schwannomas have been described in the literature [59]. Indeed, these tumors can become massive with expansion into the spinal canal and pelvis before causing severe-enough symptoms for patients to seek evaluation. Their size can make them difficult to manage [58]. MRI is often required to make a diagnosis because radiograph can be inconclusive [60]. The rarity of these tumors has made surgical management somewhat controversial, as aggressive resections may lead to a low chance of recurrence, but a high probability of neurological deficits [60, 61]. Therefore, conservative management may be more appropriate in some circumstances and surgical judgment should be utilized.

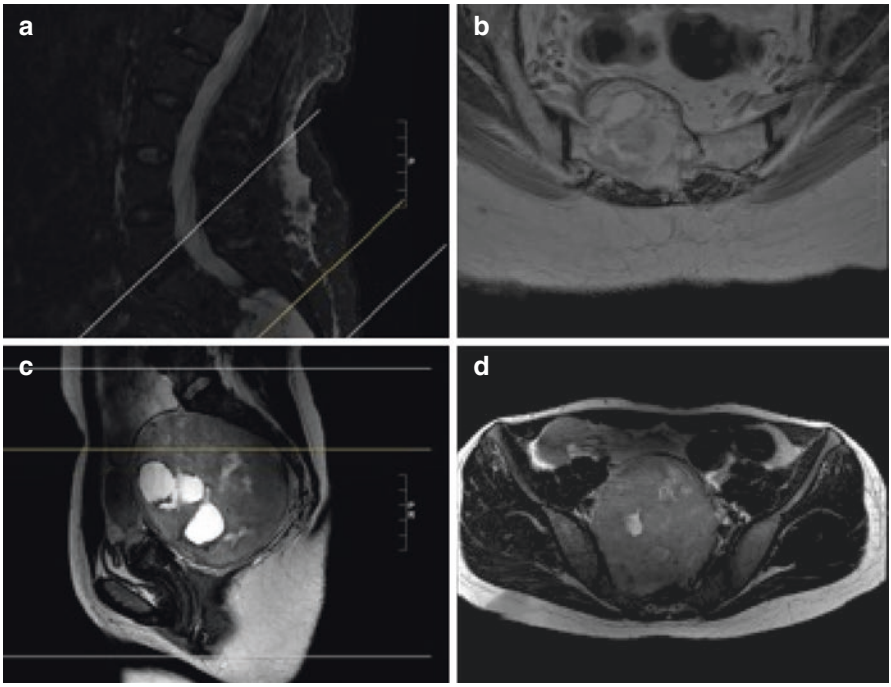

Fig. 11.6 MRI sequences of 2 patients with large sacral tumors. Sagittal (**a**) and axial (**b**) images of a large right-sided sacral tumor from the first patient. Sagittal (**c**) and axial (**d**) images of a giant sacral schwannoma

11.5.3 Intraoperative Neural Monitoring

The use of intraoperative neural monitoring is important for appropriate surgical management as it has become standard of care for resecting these tumors. Recent guidelines have been outlined for surgery on the spinal column and cord [62]. Specifically, multimodality intraoperative monitoring (MIOM) (eg, somatosensory and motor evoked potentials) is recommended to assess spinal cord integrity with motor evoked potentials being superior to somatosensory evoked potentials for assessment of spinal cord integrity. In our experience, intraoperative neural monitoring can also help with diagnosing whether the nerve root is involved or if the spinal cord is damaged prior to resection. Monitoring also helps avoid functional motor fibers during the procedure.

11.5.4 Recurrence

As with other benign tumors, gross total resection of spinal schwannomas is curative while subtotal resection permits possible recurrence. The rate of recurrence in spinal schwannomas is roughly 5% at 2 years postoperatively [23, 63]. Schwannomas

have a wide variability in growth rate and can increase in size by 5% annually [64]. A retrospective analysis of 169 patients with spinal schwannomas found that the risk factors for recurrence were higher number of spanned levels, increasing tumor size in the cranial-caudal direction, and tumor location in the cervical or sacral regions [63]. However, residual tumor size may not correlate with the rate of recurrence. A study of 27 patients found that postoperative residual mass did not correlate with significant tumor regrowth at 2-year follow-up. Alternatively, in this cohort, tumors that had a high Ki-67 labeling index value—a cellular marker for proliferation—were more likely to recur than those with low values [65]. In another study of 32 patients with giant spinal schwannomas (defined as tumors extending at least 2 vertebral levels intraspinally or 2.5 cm extraspinally), those who underwent gross total resection were less likely to have tumor recurrence compared with patients who underwent subtotal resections [66].

11.5.5 Complications

Resection of spinal nerve sheath tumors is safe with respect to major morbidity and mortality, but the complication rate is relatively high. A study of 199 patients who underwent resection for spinal nerve sheath tumors, including 163 schwannomas, described the complication rate as 32% with new or worsening sensory deficits as the most frequent complication [67]. Another study of 187 cases of spinal schwannoma described the late complication rate as roughly 21% [11]. Severe pain, spinal deformity, and spinal arachnoiditis (ie, pain disorder derived from the arachnoid) were the most common complications. Furthermore, cerebrospinal fluid leak is a possible complication, especially in cases with dumbbell schwannomas.

11.6 Conclusion

Spinal schwannomas are relatively common primary encapsulated nerve sheath tumors. The main presenting symptom is pain, although other complaints are possible, such as neurological deficits. We described the typical surgical approach for intradural and extradural tumors. Often, a posterior open midline incision followed by laminectomy or hemilaminotomy/hemilaminectomy with or without facetectomy and fusion is necessary for gross total resection—the mainstay of curative treatment. Subtotal resections can lead to recurrence, although the rate of recurrence is relatively low. Common complications include worsening sensory deficits or severe pain.

11.7 Surgical Pearls

- Intraoperative monitoring can help distinguish whether the associated nerve root can be sacrificed without postoperative motor deficits. However, it is important to note that potential sensory or pain deficits cannot be detected this way.

- Intramedullary schwannomas are exceedingly rare and may not require myelotomy, if the origin of the tumor is the dorsal root entry zone.
- Extradural tumors and intradural extramedullary tumors in certain instances can be reached via a minimally invasive approach with tubular retractors, although nerve monitoring is very limited with this approach.
- Dumbbell schwannomas should be resected extradurally first and then intradurally; resection in this order will spare the need for durotomy in case there is not actually an intradural portion of the tumor.
- Intraoperative neural monitoring is important for diagnosing nerve root involvement or spinal cord damage, as well as helping to avoid damaging functional motor fibers.

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