Neurosurgical Review (2019) 42:371–388
https://doi.org/10.1007/s10143-018-0957-x

REVIEW

# Intradural spinal tumors in adults—update on management and outcome

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Received: 24 October 2017 / Revised: 16 January 2018 / Accepted: 6 February 2018 / Published online: 17 February 2018


## Abstract

Among spinal tumors that occur intradurally, meningiomas, nerve sheath tumors, ependymomas, and astrocytomas are the most common. While a spinal MRI is the state of the art to diagnose intradural spinal tumors, in some cases CT scans, angiography, CSF analyses, and neurophysiological examination can be valuable. The management of these lesions depends not only on the histopathological diagnosis but also on the clinical presentation and the anatomical location, allowing either radical resection as with most extramedullary lesions or less invasive strategies as with intramedullary lesions. Although intramedullary lesions are rare and sometimes difficult to manage, well-planned treatment can achieve excellent outcome without treatment-related deficits. Technical advances in imaging, neuromonitoring, minimally invasive approaches, and radiotherapy have improved the outcome of intradural spinal tumors. However, the outcome in malignant intramedullary tumors remains poor. While surgery is the mainstay treatment for many of these lesions, radiation and chemotherapy are of growing importance in recurrent and multilocular disease. We reviewed the literature on this topic to provide an overview of spinal cord tumors, treatment strategies, and outcomes. Typical cases of extra- and intramedullary tumors are presented to illustrate management options and outcomes.

**Keywords** Intradural · Spinal · Intramedullary · Meningioma · Ependymoma · Astrocytoma · Neurofibroma · Schwannoma · Dumbbell tumor

## Introduction

Among central nervous system tumors, only about 15% of tumors occur intraspinally. These are mostly benign tumors with 60% occuring extradurally (ED), about 30% occurring intradural extramedullarly (ID-EM) and only 10% occurring as true intramedullary spinal cord tumors (IMSCT). Apart from the location in relation to the dura and spinal cord, the level at which the tumor occurs plays an important role in clinical presentation and treatment strategy (Table 1) [1].

## Clinical symptoms and scores

The first symptom in adults is often pain, often pronounced in a recumbent position or at night. In ID-EM tumors, neurological symptoms develop over time once the spinal cord is compressed and depend on the location of the tumor. In these mostly antero- or posterolaterally growing tumors, Brown-Sequard Syndrome is common. IMSCTs tend to present first with pain and dysesthesia. As with ID-EM tumors, the level of the tumor dictates the neurological symptoms [2]. Due to the late onset and sometimes non-specific nature of neurological symptoms, the diagnosis of these tumors is often delayed. Early magnetic resonance imaging (MRI) is important to guide the timely initiation of treatment [3].

Clinical scores based on neurological function have proven effective to describe and compare the patient's functional

**Electronic supplementary material** The online version of this article (https://doi.org/10.1007/s10143-018-0957-x) contains supplementary material, which is available to authorized users.

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**Table 1** Overview of the most frequent intradural spinal tumors

| Location | Entity | | Region | Female/male ratio | Age | Distribution |
|---|---|---|---|---|---|---|
| ID-EM tumors (30%)* | Meningiomas | | 80% thoracic | 1/5 | 5th–7th decade | 50%** |
| | Nerve sheath tumors | Schwannomas<br>Neurofibromas | Cervical = lumbar | 1/1 | 4th–5th decade | 30%** |
| | Filum terminale ependymomas | | 100% lumbar | 1/2 | 3rd–5th decade | 15%** |
| | Miscellaneous: dermoids, epidermoids, lipomas, teratomas, neuroenteric cysts | | | | | 5%** |
| IMSCT (10%)* | Ependymomas | | 45% cervical | 1/2 | 4th decade | 55%*** |
| | Astrocytomas | | 70% thoracic | 2/3 | 3rd decade | 40%*** |
| | Miscellaneous: gangliogliomas, oligodendrogliomas, subependymomas, hemangioblastomas, neurocytomas, metastases | | | | | 5%*** |

*Of all spinal tumors, **Among all ID-EM Tumors, ***Among all IMSCT

status pre- and post-treatment as an important outcome parameter. The routine use of these scores in clinical practice and documentation is advised (McCormick score [4], Klekamp and Samii Score [5], Frankel score [6]). We refer to the McCormick score throughout this paper unless indicated otherwise.

## Diagnostics

Since the introduction of the MRI, the diagnosis and therefore incidence of intradural spinal tumors have risen. High-resolution sagittal and axial images after gadolinium injection are state of the art. Table 2 shows the MRI characteristics of the most common intradural spinal tumors. T2-weighted imaging (T2WI) helps detect intramedullary lesions while the suppression of high CSF signal with FLAIR images is useful for detecting subtle intramedullary lesions. Diffusion-weighted imaging (DWI) is recommended for detecting cytotoxic edema [1] and gradient echo sequences help to visualize blood products and calcifications. Complete imaging of the neuroaxis is recommended after detection of a lesion, especially with ependymomas.

CT scans are relevant for imaging the osseous spine. This modality is useful when primary intradural tumors infiltrate the vertebrae or when larger lesions require extensive approaches through bony structures and require stabilization. CT also shows intralesional calcifications, which can occur in meningiomas, gangliogliomas, and sometimes ependymomas and may alter the surgical planning. Angiography may be useful in hemangioblastomas and paragangliomas as well as in some meningiomas to visualize the blood supply if embolization is desired, or to assess relevant vascular compression [7]. CT-myelography is used if MRI is contraindicated as in patients with a pacemaker. Plain radiographs are useful in select cases. CSF analyses by lumbar puncture may be important to rule out differential diagnoses (e.g., myelitis and other neuroinflammatory diseases).

## Intradural extramedullary tumors

### Meningiomas

The most common primary extramedullary tumors are meningiomas, which account for 25% of all spinal neoplasms [8]. They are most likely to occur in the thoracic spine in a posterolateral position and are more frequent in older and female patients. Meningiomas can be histologically classified as psammomatous, fibroblastic, or meningothelial. These generally well-circumscribed, slow growing lesions can be treated successfully with surgical resection. Nevertheless younger patients tend to develop more complex meningiomas with a mortality rate up to 10% even when treated [9].

GTR is the treatment of choice for spinal meningiomas. Yoon et al. reported the Simpson Grade of resection as a good predictor for recurrence with no recurrent disease seen in their series after Simpson Grade I to III resections [10]. Invasion of the arachnoid or pia is an independent negative prognostic factor [11]. Advanced age does not seem to be contradictive to a good outcome [12]. Minimally invasive approaches should be used where possible because they are associated with a better postoperative course. However, severe intratumoral calcification might lead to a more extensive approach in order to remove a hard tumor safely [13]. As in cranial cases, 5-ALA can be used to identify remnant tumor [14]. Radiosurgery is reserved for cases where surgical treatment is not possible or for cases with recurrent, residual, or multiple lesions, as with other benign intraspinal tumors [15].

Surgical treatment of spinal meningiomas has a favorable outcome in the majority of cases. When resected completely, functional outcome is excellent and recurrence rates are low [16]. An analysis of 80 patients with spinal meningioma treated surgically with intraoperative monitoring (IOM) by Setzer et al. showed a good outcome with improved

**Table 2** MRI characteristics of intradural spinal tumors

| | T1 | T2 | Contrast enhancement | Special features |
|---|---|---|---|---|
| Intradural lesions | | | | |
| Meningiomas | Hypo- or isointense | Hyperintense | ++<br>Homogeneous | Calcifications, dural tail |
| Nerve sheath tumors | Hypo- or isointense | Hyperintense | ++<br>Homogeneous | 15% extradural growth |
| Dermoids | Hypo- or hyperintense | Hyperintense | –<br>Thin enhancement around the periphery might be seen | Less likely to show diffusion restriction than epidermoids |
| Epidermoids | Isointense | Isointense or slightly hyperintense | –<br>Thin enhancement around the periphery might be seen | Califications are rare<br>White epidermoids occur due to hemmorhage or high protein content |
| Neuroenteric cysts | Variable depending on protein content | Variable depending on protein content | None | |
| Teratoma | Mixed signal from different components | Mixed signal from different components | Solid soft tissue components enhance | |
| Paraganglioma | Isointense | Hyperintense | ++<br>Homogeneous | Hemorrhage is common (hemosiderin cap sign)<br>Flow voids are common |
| Lipomas | Hyperintense | Hyperintense | –<br>None | Hypointense in fat-supressed sequences |
| Leptomeningeal metastasis | Isointense | Hyperintense | ++<br>Homogeneous | Sugar coating of spinal cord and nerve roots |
| Intramedullary lesions | | | | |
| Ependymomas | Hypo- or isointense | Hyperintense | ++<br>Homogeneous | Cystic in 50%, syrinx |
| Astrocytomas | Hypo- or isointense | Hyperintense | ++<br>Heterogeneous | Cystic in 30%, leptomeningeal spread |
| Ganglioglioma | Hypo- and hyperintense (mixed) | Hyperintense | +<br>Heterogeneous | Calcification is common |
| Subependymoma | Hypo- or isointense | Hyperintense | None<br>mild in some cases | Origin is the central canal<br>Calcifications are possible |
| Hemangioblastoma | Isointense (hypo- and hyperintensity possible) | Iso- or hyperintense | ++<br>Homogeneous | Tumor cyst or syrinx is common |
| Lymphoma | Isointense | Hyperintense | ++<br>Homogeneous | |
| Metastasis | Hypointense | Hyperintense | ++<br>Homogeneous | Well-defined lesions, cysts are rare |

or unchanged neurological status in 93.5% of patients. Other studies also showed positive outcomes in over 90% of patients [11, 17–22].

Schaller reported a 79% restored neurological function in a group of 33 patients. In this group, the psammomatous subtype correlated with a less favorable outcome while age under 60 years, a posterior or lateral tumor position and short duration of preoperative symptoms correlated with good outcome [23]. Riad et al. reported a neurological improvement in 87% and a recurrence rate of 8% in their series [24] but long-term follow-up is necessary for late recurrences. Even in cases with severe preoperative deficits, good outcomes can be expected [25].

## Nerve sheath tumors (schwannomas, neurofibromas)

The second most common extramedullary tumors are nerve sheath tumors, spinal schwannomas, or neurofibromas. Schwannomas are slow-growing lesions that arise from the sensory dorsal rootlets in most cases. The majority of lesions are intradural but they can also grow extradurally (10%) or combined intra-extradurally (10–15%). Schwannomas tend to develop an hourglass shape due to bony impression at the neural foramen during their growth and are then called dumbbell tumors (Fig. 1). They present first with radicular pain and later result in motor deficits. The recurrence rate after surgical resection is low. Malignant dumbbell tumors are rare (2.5%) and are more common in children under the age of 10 [26].

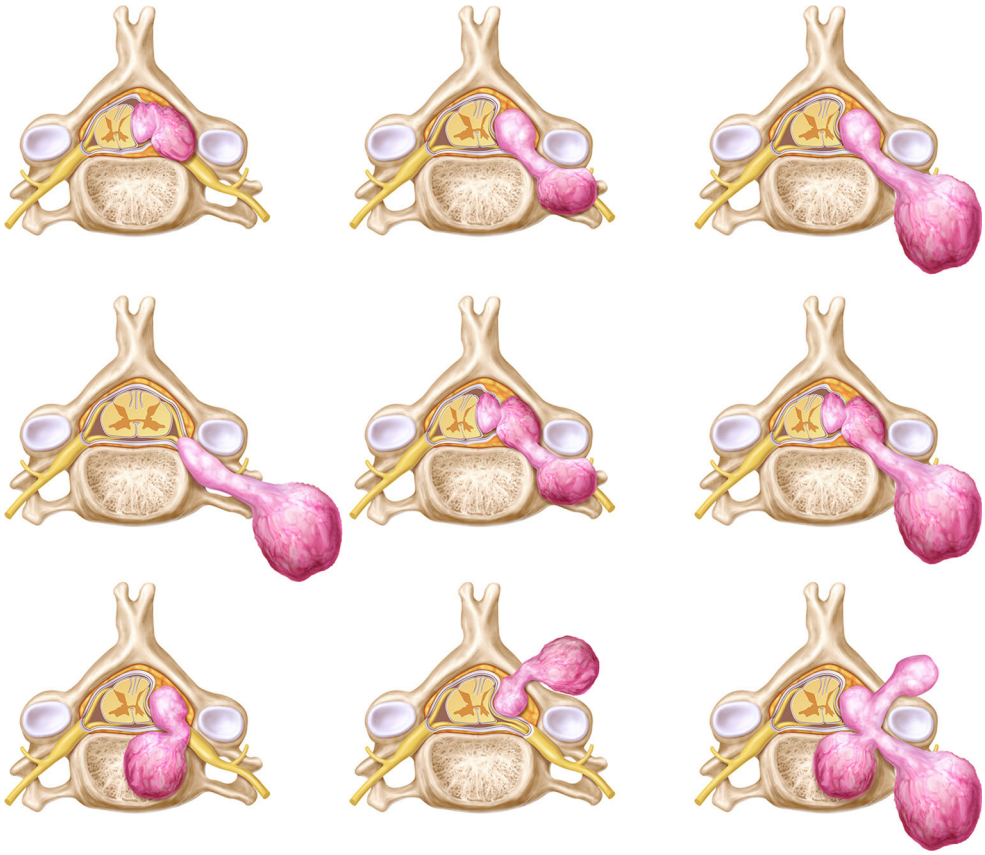

**Fig. 1** Classification of dumbbell spinal tumors, modified from Asazuma et al. (Surgical Strategy for Cervical Dumbbell Tumors Based on a Three-dimensional Classification, Spine, Volume 29, Number 1, pp. E10-E14. 2003) showing nine types of dumbbell tumors from left to right (Typ I, Typ IIa, Typ IIb, Typ IIc, Typ IIIa, Typ IIIb, Typ IV, Typ V, Typ VI). A posterior approach is feasible in types I, IIa, IIIa and sometimes in types IV and V while a combined anterior-posterior approach is required in types IIb, IIc, IIIb, and VI. Tumors invading at multiple levels, extradural intervertebral tumors (type IV), and multidirectionally eroding tumors (type VI) require spinal reconstruction

If associated with neurofibromatosis (NF), an autosomal dominant neurocutaneus disorder, nerve sheath tumors are likely to be neurofibromas. In this case, they incorporate other cells in addition to schwann cells, are unencapsulated, and tend to occur in multiple locations, engulfing the nerve rather than displacing it. The management of NF differs because the goal is to achieve control of symptoms and local disease. Therefore in most cases only symptomatic tumors are considered for resection. Intradural spinal tumors are more common in NF2 than in NF1. NF should be considered in pediatric patients with a nerve sheet tumor and in patients with additional spinal or cranial tumors (especially vestibular schwannomas and meningiomas), skin lesions (nodules, dermal neurofibromas, café au lait spots), or first degree relatives with NF. The workup includes ophthalmic examination, genetic studies, auditory evaluation, and MRI of the whole neuroaxis to rule out additional tumors. These patients should be managed by a multidisciplinary care team consisting of neurosurgeons, neurologists, geneticists, opthalmologists, dermatologists, plastic surgeons, and endocrinologists. Table 3 provides an overview of neurocutaneous tumor syndromes.

Nerve sheath tumors are also primarily treated surgically in most cases. Tumors with large extraforaminal extension may require more extensive approaches and sometimes even stabilization. Therefore these tumors are associated with complications more often [27]. Minimally invasive approaches require very detailed preoperative planning and have to be tailored to the origin (root), location, and size of the lesion [28]. As mentioned, management of NF with multiple lesions is more complex. Surgical intervention is considered only when neurological deficits are evident and disease control is needed. Stereotactic radiosurgery is becoming important to patients with NF for long-term disease control and its low complication rates [29]. In the rare case of intramedullary schwannomas, surgery is the treatment of choice and results in good outcomes if GTR is achieved [30].

Nerve sheath tumors can be removed safely and effectively through microsurgery. Fernandes et al. reported a GTR rate of 96% and a recurrence rate of 3.3% in a series of 30 patients [31]. A large series with 367 cases by Lenzi et al. with a median follow-up of 10 years (range 1–20) reported full or partial recovery in the great majority of patients with root pain resolving in all but nine patients. In this group, sphincter compromise led to the worst prognosis and persistent back pain was the most frequent complication [32].

## Filum terminale ependymomas

Around 40% of spinal ependymomas are primary extramedullary, occur within the filum terminale, and are mostly myxopapillary (WHO Grade I).

**Table 3** Neurocutaneous disorders/tumor syndromes (phakomatoses)

| Disorder | NF1 | NF2 | NF3 | von Hippel-Lindau disease | Tuberous sclerosis complex |
|---|---|---|---|---|---|
| aka | Von Recklinghausen's disease | MISME syndrome (multiple inherited schwannomas, meningiomas, and ependymomas | Schwannomatosis | Familial cerebello retinal angiomatosis | Bourneville–Pringle disease |
| Genetics | ▪ Autosomal dominant<br>▪ 50% de novo mutations<br>▪ Neurofibromin protein decreased<br>▪ *NF1* gene on long arm chromosome 17 | ▪ Autosomal dominant<br>▪ 50% de novo mutations<br>▪ Protein merlin<br>▪ Chromosome 22 | ▪ Autosomal dominant<br>▪ *SMARCB1* gene<br>▪ Chromosome 22 | ▪ Autosomal dominant<br>▪ 20% de novo mutations<br>▪ Protein pVHL<br>▪ Chromosome 3p25 | ▪ Autosomal dominant<br>▪ 80% de novo mutations<br>▪ Proteins hamartin and tuberin<br>▪ Chromosome 9q34/16p13 |
| Clinical criteria include | ▪ Six or more café au lait macules<br>▪ Two or more neurofibromas or one plexiform neurofibroma | See Manchester, Baser, NIH, and National Neurofibromatosis Criteria<br>Bilateral acoustic schwannomas | 2 or more pathologically ascertained schwannomas without symptoms of 8th nerve dysfunction at age > 30 years | Two or more characteristic lesions, two hemangioblastomas | Major criteria<br>▪ Facial angiofibroma, cortical tuber, subependymal giant cell tumor and<br>▪ Minor criteria Cerebral white matter radial migration lines |
| Associated tumors include | Neurofibromas, optic pathway gliomas, brainstem gliomas, cerebellar lesions | Schwannomas, neurofibromas, meningiomas | Schwannomas | Hemangioblastomas, endolymphatic sac tumors, renal cell carcinomas, pheocromocytomas, neuroendocrine tumors | Facial angiofibromas, renal angiolipoma, cortical tubers, subependymal nodules, subependymal giant cell astrocytoma (SEGA) |
| Intradural spinal lesions | Spinal neurofibromas and plexiform neurofibromas | Spinal meningiomas, neurofibromas and Schwannomas | Schwannomas | Spinal hemangioblastomas | Very rare, chordomas and subependymal tumors |

In filum terminale ependymomas, integrity of the tumor capsule along with en bloc resection of the tumor seem to be the factors that lead to an optimal surgical outcome and a reduced risk for local recurrence [33, 34]. Klekamp demonstrated a significantly higher proportion of local recurrence in patients with unencapsulated tumors compared to those with encapsulated filum ependymomas at long-term follow-up [34]. The authors of this review consider en bloc tumor removal more important than the utilization of limited approaches. In limited approaches, the risk or necessity of tumor debulking to facilitate tumor removal may lead to CSF seeding and result in a high frequency of subtotal resection (STR) in case of secondary surgery [34]. Therefore, we adopt the surgical attitude of Nakamura et al. that "myxopapillary ependymoma should be treated assuming that this tumor is malignant," and practice resection techniques that avoid violating the capsule and spare neurological function [35]. Prior to surgery, a bipolar tumor (secondary tumor manifestation along the filum [36]) should be ruled out by MR imaging. If such a tumor is present, it should be treated with an extended resection of the filum including all nodular tumors. CSF dissemination from a filum ependymoma might occur prior to surgery by capsule breach or exfoliation of ependymal cells, but the prognostic value of this prior dissemination is unclear [34, 37]. Suspicion of CSF seeding on MR imaging during the radiological diagnosis of a filum terminale tumor should lead to cranial and total-spine MR imaging and CSF sampling including cytospin to rule out an intrinsic brain tumor (e.g., a cerebral ependymoma with drop metastasis) [37].

Radiotherapy is recommended if GTR was not possible. In these cases, radiotherapy leads to acceptable disease control [38, 39]. Filum terminale ependymomas are associated with a higher complication rate than other benign entities in this region [40]. Although chemotherapy is administered in cases of recurrent disease, evidence for its benefit as a primary treatment is lacking.

## Miscellaneous

Other tumors such as dermoids, epidermoids, lipomas, teratomas, neuroenteric cysts, paragangliomas, and leptomeningeal metastases can occur intradural extramedullarly or

juxtamedullarly in the case of spinal cord lipomas. They comprise about 5% of ID-EM tumors. Due to their rare occurrence, the literature contains mostly analyses of small series.

Epidermoids can be associated with spinal cord dysraphism or caused iatrogenously by lumbar puncture or surgery if epidermal cells are displaced into the spinal canal [41]. They have to be resected en bloc, along with the capsule, to prevent recurrence. Adherence of the capsule to neural elements poses the risk of neurological deficits [42, 43]. Spontaneous or iatrogenous rupture of the cyst can cause chemical meningitis [44]. Van Aalst et al. concluded that intraspinal dermoid and epidermoid tumors do not differ in clinical practice and suggested identical management and new nomenclature as, "spinal cutaneus inclusion tumors" [45].

A recent publication by Pang details the management of spinal cord lipomas, recommending GTR for asymptomatic dorsal and transitional lipomas in children and for all symtomatic lipomas of all ages [46].

Paragangliomas are vascular, neuroendocrine tumors of the autonomic nervous system mostly found in the lumbar region. They are uncommon in children and rarely produce catecholamine. Preoperative embolization should be considered prior to microsurgical resection [7].

Intraspinal teratomas are composed of ectodermal, endodermal, and mesodermal elements and are more frequent in children and neonates than in adults. Radical resection is recommended [47].

Neuroenteric cysts result from incomplete resorption of the neurenteric canal and usually occur in a ventral location and in the thoracic region. First line treatment is surgical resection. In the case of STR and recurrence, cyst-peritoneal (CP) and ventricle-peritoneal (VP) shunts are considered as second line treatment. Risk factors for recurrence are STR and mucin-secreting cells [48].

Spinal metastases are either drop metastases from intracranial processes or from distant solid tumors and most commonly affect the lumbar spine. Management depends on the primary tumor and prognosis is poor in most cases [49].

Important differential diagnoses for ID-EM tumors include inflammatory processes, arachnoid cysts, and vascular malformations.

## Intramedullary spinal cord tumors

As in ID-EM tumors, "smart" GTR [50] is the primary goal of surgery in IMSCT [51]. In malignant cases where GTR is barely possible, adjuvant radio- and chemotherapy is used. While histopathology and extent of resection are significant predictors for PFS, the preoperative functional status is the most important predictor for long-term functional outcome [52]. Age and the region of spinal cord in which the tumor is present are also important factors for functional outcome [53].

### Ependymomas

Intramedullary ependymomas are mostly benign tumors (WHO Grade II), which are most likely found in the cervical spine. They are slightly more frequent in males and occur during the 3rd and 6th decade. Histopathologically, ependymomas can be classified as myxopapillary, papillary, cellular, epithelial, and mixed. WHO Grade III intramedullary ependymomas are very rare [54]. The primary treatment of choice is surgical resection [55]. While the role of chemotherapy has not yet been established, adjuvant radiotherapy has been shown to prolong PFS and should be considered in cases of STR [56].

In a cumulative analysis of data from studies reporting more than 50 surgical cases of intramedullary ependymomas [57–64], the rate of GTR is related to the caseload of ependymomas (trend; $p = 0.054$, ANOVA) or all glial IMSCT operated upon ($p = 0.011$, ANOVA; JMP12.1.0, SAS, USA). In a retrospective cohort of 100 patients with intramedullary ependymomas, Klekamp revealed a significant influence of surgical experience on the rate of GTR, but not on the avoidance of surgical complications [61]. While some authors favor en bloc resection, others favor internal decompression and piecemeal resection [61], depending on tumor size. Meticulous control of the anterior vascular supply of the tumor is of utmost importance to maintain spinal cord function [61]. To lower the risk of dissemination, the authors of this article recommend en bloc resection whenever safely possible. While adjuvant radiation in ependymomas is often used after STR, the role of radiation after GTR is debatable. Sgouros et al. [65] failed to detect a significant effect of radiotherapy on progression or recurrence, and Tarapore et al. [66] identified a negative effect of radiotherapy on survival after STR. Therefore, reoperation in case of the first recurrence should be considered. Radiotherapy may be indicated in case of a repeated STR, thus reducing the long-term risks of radiation myelopathy especially in children and young adults.

Histology and extent of resection are considered to be the most important prognostic factors for the post-surgical outcomes of ependymomas [63]. Safaee et al. observed a recurrence rate of 40% in ependymomas that had not been resected completely compared to a recurrence rate of 20% after GTR in children and young adults [67]. This resembles long-term survival data published by Tarapore et al. [66] and Lee et al. [63] with 90% 10-year survival after GTR and only 80% 10-year survival after STR. In this study, radiation therapy after STR decreased survival (70% at 10 years). A recent analysis by Li et al. found tumor length to be a relevant factor in postoperative outcome, with longer tumors (> 10 cm) showing reduced extent of resection and elevated risk for postoperative deficits

[68]. In a series of 67 intramedullary cases, Kucia et al. described a worse outcome in patients after STR with an overall complication rate of 34% where infection and CSF leak were the most common complications [62]. In a series of 39 intramedullary ependymomas, tumors in the thoracic region had a worse outcome than tumors of the cervical or lumbar region [58, 61]. Postoperative neuropathic pain occurs in approximately one third of patients and might depend mainly on the alteration of the cord by the tumor itself, because preoperative neuropathic pain is the main predictor of postoperative pain. Other predictors of postoperative pain include permanent morbidity and preoperative McCormick grade [61]. Long-term neurological function after microsurgery may be improved or stabilized in 26 and 66% of patients, respectively, with only 9% of patients experiencing worsened neurological function [50].

## Astrocytomas

Astrocytomas comprise about 60% of all primary intramedullary tumors. They develop in childhood or later in adulthood within the 3rd and 5th decade with a slight male predominance (1.5:1). Their dominant location is the thoracic spine. Spinal astrocytomas in children are mostly benign while those in adults can be more commonly WHO Grade III and IV lesions. Complete surgical resection at this stage is seldom possible and the recurrence rate is as high as 50% over 5 years. Some authors favor biopsy and radiation because surgery carries a risk of neurological deficits and has limited survival benefit even after aggressive resection, but advances in surgical techniques and intraoperative monitoring have improved outcomes [69, 70].

In spinal astrocytomas, histological type is the main prognostic factor for outcome. Tumor histology affects its biological behaviour (WHO grade) as well as the probability of finding a plane of dissection and thereby the potential for GTR (e.g., the behavior of pilocytic astrocytoma versus diffuse astrocytoma) [50, 60, 70, 71].

Low-grade tumors have low surgery-related complication rates and a good long-term outcome when operated early. The outcome of malignant spinal astrocytomas remains poor. An analysis of the SEER database by Adams et al. revealed a median survival of 13 months for patients with primary malignant astrocytomas of the spinal cord and a 5-year survival rate of 18.7%. Patients with anaplastic astrocytomas had a median survival of 17 months whereas those with glioblastomas had a median survival of 10 months [72], which resembles the prognosis of high-grade glioma of the brain [73, 74]. In most high-grade tumors, GTR is hardly possible and STR or biopsy followed by radiation and chemotherapy is the recommended standard treatment [58, 75]. Successful GTR in 44% of patients with spinal anaplastic astrocytoma was reported in a study by McGirt et al. based on postoperative MR imaging, but no cases of GTR were reported for patients with glioblastoma. Survival did not significantly improve with GTR, but a positive trend was reported for anaplastic astrocytomas [74]. Additionally, the risk of neurological deficit is associated with the extent of surgical intervention [69]. Therefore GTR remains reserved for selected cases [76].

Chemotherapy has been shown to have a significantly effect on PFS (but not OS) in infiltrative astrocytomas in a retrospective study by Fakhreddine et al. with multivariate analysis in a retrospective cohort study [70, 71].

In pilocytic astrocytomas, solid portions and cysts seem to displace rather than infiltrate the cord, allowing for potential GTR [77]. However, extensive cyst formation or repetitive hemorrhage with inflammation might obviate GTR. Since radiation therapy may not add to tumor control in this entity [70], treatment with GTR should be attempted, but guided by intraoperative neuromonitoring. Identification of a distinctive tumor plane during microdissection improves resectability in low-grade astrocytomas [4, 78]. Therefore the combination of a distinct resection plane during surgery and low-grade (pilocytic) histology on the intraoperative fast stain mandates the attempt of maximum safe resection under intraoperative neuromonitoring. Ardeshiri et al. reported better resectability and functional outcome for cervically located astrocytomas [79].

In case of a non-discernable resection plane, patients with infiltrative astrocytoma may benefit from tumor debulking for spinal cord decompression. However, there are controversial data and even reports of reduced survival after STR in infiltrative astrocytomas [70]. Due to the deficits related to surgery, some authors favor biopsy and radiation [69, 70].

Additional expansion duraplasty, along with myelotomy and biopsy, may prolong clinical progression and provide a beneficial window for initiating radiation therapy. With progression of neurological deficits after radiation, the effect of expansion duraplasty is at best limited. Therefore the elevated perioperative risk of local and systemic complications (e.g., CSF leak, wound dehiscence, pneumonia, thrombosis/pulmonary embolus) in patients usually already treated with cortisone and local radiation therapy must be taken into account prior to performing an expansion duraplasty.

Special consideration regarding treatment strategies should be given to the very rare holocord intramedullary tumors. Extended midline myelotomy, biopsy, and expansive laminoplasty may result in favorable mid-term control in low-grade astrocytoma [80], and successful GTR resection with improvement of neurological function [81], both outcomes which were reported in pediatric cases. For children and adolescents, Ebner et al. reviewed their experience and the literature and favored an attempt for GTR if feasible based on intraoperative neuromonitoring, since many cases were pilocytic astrocytomas with favorable prognosis. For these lesions, resection might be achieved via a single or staged

procedure including a holocord midline myelotomy and expansion duraplasty to facilitate tumor expansion posteriorly, thereby improving tumor resection during a second stage procedure [82]. Adjuvant therapy should be reserved for cases with tumor regrowth at follow-up [83, 84].

### Miscellaneous

Gangliogliomas, subependymomas, hemangioblastomas, neurocytomas, lymphomas, and metastases comprise only 5% of IMSCTs. Due to their rare occurrence, only a few cases are reported in the literature. Spinal hemangioblastomas are frequent in patients with von Hippel-Lindau disease and are treated surgically if symptomatic. Kim et al. presented an algorithm on the management of spinal cord hemangiomas suggesting resection in asymptomatic cases for tumors of a certain size (> 500 $mm^3$) or with interval growth [85].

Spinal gangliogliomas are slow-growing WHO grade I or II tumors predominantly occurring in young adults and typically involving long segments of the spinal cord. Gangliogliomas can form cysts and are sometimes associated with bony changes. GTR might be difficult in cases of extensive tumor growth with associated scoliosis further complicating the management [86].

Intramedullary lymphomas are secondary to leptomeningeal disease in most cases and are managed with radiation and or chemotherapy. Repeated CSF sampling is recommended to obtain histopathological diagnosis before commencement of treatment [87].

Spinal subependymomas mostly occur in the cervical spine and are often found in an eccentric location within the spinal cord. Surgical nuances in the treatment of these tumors are presented in a recent paper by Tan et al. [88].

Spinal neurocytomas are extremely rare and benign in most cases. They usually occur in the cervical spine. Resection is the treatment of choice. In cases of histological atypia, close follow-up is recommended and radiation is considered in cases of recurrence [89, 90].

In intramedullary spinal metastasis, resection is reserved for well-selected cases. Resection may preserve function without improving overall prognosis [91].

Important differential diagnoses for IMSCT include cavernomas, spinal cord infarction, multiple sclerosis, myelitis, and contusion.

## Therapeutic management

Once an intradural spinal tumor is diagnosed, treatment should start as soon as possible. After diagnostics and surgical planning, resection is performed. A watchful monitoring approach is used in NF with multiple lesions and no evidence of neurological deficits. In elderly and very old patients with suspected meningioma, a wait-and-scan strategy may be used, since a significantly high neurological complication rate is reported in those patients with severe intratumoral calcification [21]. When surgery is performed, the goal is a GTR while maintaining or restoring function. Several minimally invasive approaches have been published recently. Although minimally invasive approaches have advantages, small surgical windows or endoscopic removal carries the risk of STR and the dissemination of tumor cells. Therefore the approach has to be tailored to ensure GTR. To lower the risk of neurological deficits, IOM is recommended in all intramedullary tumors as well as in ID-EM tumors [92], especially for ventral lesions and tumors of the conus medullaris [3]. The main goal of IOM is to identify neurologic deficits at a reversible stage and to guide intraoperative decisions. Most tumors can be safely resected via standard microsurgical posterior approaches. For ventral tumors, ventral approaches should be considered [93].

## Intraoperative techniques

### Intraoperative monitoring

As with lesions occurring in eloquent regions of the brain [94], intraoperative neurophysiological monitoring (IOM) of spinal cord lesions has been proven safe and effective.

SSEP induced at the tibial or the median nerve provides constant monitoring of the dorsal columns with a time-delay from signal averaging that is dependent on the sampling rate (e.g., 5/s rate, 100 stimuli: 20-s delay). This monitoring technique harbors a high sensitivity for sensory function, but prediction of neurological deficit is limited (Table 4) because SSEP alone does not reliably monitor motor function.

MEP monitors the corticospinal tract in discontinuous but very short intervals (2–10 s). Transcranial stimulation elicits an electric potential build up in fast conducting motor fibers within the lateral and anterior corticospinal tract (recorded by an epidurally positioned intraspinal electrode; eMEP, aka D-wave) and a compound muscle action potential (mMEP) [92]. A decrease in D-wave amplitude of greater than 50% correlates with motor deficits. Combining mMEPs with D-wave monitoring offers the most comprehensive approach to assess the integrity of the spinal motor tracts [95–97]. Using this technique, permanent motor deficits and paraplegia occur in less than 5% and less than 1%, respectively [98]. In pediatric patients, intraoperative changes from prophasic to biphasic waveforms in MEP monitoring were associated with a postoperative decrease in motor function [99].

When single nerve roots need to be monitored, EMG is the best choice. EMG offers very high sensitivity, but limited prediction of functional changes [3].

**Table 4** Overview of neuromonitoring for intradural spinal tumors

| Method | Monitors | Sensitivity (%) | Specificity (%) | PPV (%) | NPV (%) |
|---|---|---|---|---|---|
| SSEP | Dorsal columns | 52 | 100 | 100 | 97 |
| mMEP<br>eMEP/D-wave | Corticospinal tract, anterior horn motor neurons<br>Fast conducting fibers in the corticospinal tract | 100 | 96 | 96 | 100 |
| EMG | Nerve root function | 46 | 73 | 3 | 97 |

While SSEP monitoring is user-friendly, it lacks the specificity to detect deficits in motor function. This limitation can be overcome by combining SSEP with m/eMEP and/or EMG monitoring. A prospective analysis of SSEP, EMG, and MEP monitoring for predicting postoperative deficits by Kelleher et al. showed a 52% sensitivity, 100% specificity, a PPV of 100%, and a NPV of 97% for SSEP monitoring. MEP sensitivity was 100%, specificity was 96%, PPV was 96%, and NPV was 100% while EMG sensitivity was 46%, specificity was 73%, PPV was 3%, and NPV was 97% [100]. As none of these methods provide a global assessment of the spinal cord, multi-modality monitoring is necessary to enhance precision (Table 4).

Although IOM is been used routinely in thoracolumbar surgery, it has not been universally accepted. Given that neurological deficits can occur throughout the duration of the operation, Forster et al. highlighted the importance of IOM beyond tumor removal in a retrospective study of 203 patients. Neuromonitoring should start prior to laminotomy and continue after tumor resection because critical changes in monitoring can occur during laminotomy, dural opening, dural closure, and laminoplasty in 9% of patients [101]. Surgical effects on neuromonitoring and strategies for management of significant alterations of monitored function are discussed below with a focus on IMSCTs.

## Surgical technique

### ID-EM tumors

For intradural extramedullary tumors, the surgical approach should aim to control the origin of the respective tumor. Direct control after dural opening is feasible with a posterior or laterally located tumor matrix (e.g., highly vascularized tumors or tumors that are difficult to debulk such as partly calcified meningiomas) with no mobilization of the spinal cord (see Case 1).

Tumors with an anterolateral or anterior matrix might be accessible posterolaterally in cases with a sufficiently large lateral tumor extension, which in turn might allow debulking and reaching its anterior matrix without moving the cord medially or posteriorly. Sectioning of the dentate ligaments might further release the cord and enable some cord rotation (but not traction) to improve anterior control.

Without medial dislocation of the spinal cord by the tumor itself, a posterior or posterolateral approach might not suffice to control the tumor matrix (and its vascular supply, which might disturb a clear microsurgical field by constant oozing) and an anterolateral or anterior approach should be considered (see Case 2). Anatomical regions with obstacles for an anterior approach (i.e., craniocervical junction, upper thoracic spine, sacral region) might be addressed via a lateralized posterolateral approach, including facet joint resection, which warrants reconstruction by instrumentation at the end of the procedure in most cases.

Schwannomas nearly exclusively originate from the posterior sensory root. Unless the tumor has extended into the dorsal root gangion, the anterior motor root might be decompressed and saved anatomically (Fig. 2). A T-shaped dural opening towards the root sleeve improves visualization and root-sparing resection. Lateral dural defects at the radicular dural sleeve might be controlled by a pull-through soft tissue plug (i.e., autologous muscle or subcutaneous fat) from the dural inside to outside, which is attached to the epidural tissue/dura with a stitch. This technique supports a facet-sparing combined intra-extraspinal approach in dumbbell-shaped tumors.

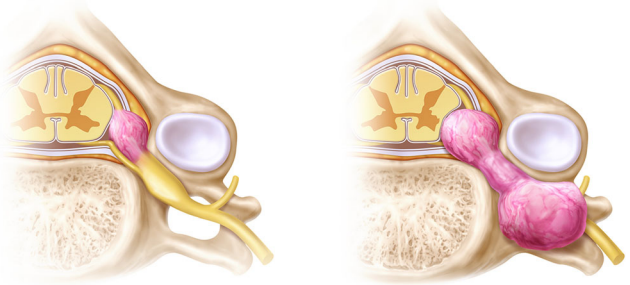

**Fig. 2** Microsurgical considerations for intradural spinal schwannomas modified from McCormick (Intradural extramedullary tumors, In: Schmidek and Sweet: Operative Neurosurgical Techniques—Indications, Methods, and Results. Ed. Quinones-Hinojosa A. 6th Ed, Vol. 2, Chapter 187, pp. 2127–2133, Elsevier, Philadelphia, 2012). *Left side*: schwannoma arising from the posterior root displacing the anterior root and ending at the level of the dorsal root ganglion. In this case the anterior root can be spared. *Right side*: schwannoma extending beyond the dorsal root ganglion involving the dorsal and anterior root. If microsurgically resected, permanent loss of motor function is inevitable in these cases because the anterior root cannot be spared

In case of resection of the dura, meticulous dural repair is absolutely mandatory. Synthetic duraplasty sutured into the dural defect by a non-absorbable monofilament suture (e.g., Prolene) might reduce the risk of secondary tethering of the spinal cord, but clearcut evidence in favor of one specific material is still missing. In spinal meningiomas, resection of dura is only recommended if it can be done without altering the risk for CSF leak or other complications [102, 103] and is feasible in the posterior or posterolateral attachment. Given that about three out of four spinal meningiomas are attached at or anterior to the level of the lateral dentate ligaments [21], spinal duraplasty is technically more demanding. The increased perioperative risk does not outweigh potential long-term benefits in PFS [103]. Reconstruction of a normal sized or even widened subarachnoid space is very important in order to reduce adhesions, tethering, and the risk of secondary syringomyelia as a rare complication [104].

## IMSCT

For IMSCTs, evidence-based guidelines recommend the combination of intraoperative monitoring of SSEP, mMEP, and eMEP (D-wave) as predictors for neurological outcome [105]. Intraoperative neuromonitoring significantly improves clinical outcome with surgical resection of intramedullary ependymomas, less so in astrocytomas [106]. Rapid neurological deterioration indicates an urgent surgical decompression and resection, which can be performed without IOM in order to salvage function [107]. In patients with major neurological impairment, monitoring might not be possible due to signal loss preoperatively. Therefore, preoperative SSEP and mMEP might help to plan for the intraoperative monitoring set-up. Additionally, these modalities are useful in monitoring the resection of lower thoracic IMSCTs or conus tumors where D-wave monitoring is not possible.

After dural opening, mapping the posterior columns might help to identify the physiologic midline sulcus for myelotomy in asymmetrically localized IMSCTs, but this is rarely needed [108]. In IOM, relevant changes (reduction of amplitude, increase of latency) of SSEP or MEP must be discussed with the technical and anesthesiology team with a written document to rule out or correct for changes in medication (anesthetic agents and levels, neuro-muscular blocking agents), drop of mean arterial pressure (i.e., spinal cord perfusion pressure), body temperature, or oxygenation (hemoglobin levels, ventilation settings). Next, technical reasons should be excluded in less than 5 min, and in critical stages of surgery (e.g., cord-tumor-interface preparation), a warning should be given to the surgeon and anesthesiologist to pause resection and re-check baselines.

Reduction of SSEP amplitude by 50% or an increase in latency by 10% of baseline after myelotomy indicates the risk of functional compromise and should prompt the release of any cord tension (pial sutures) or local cord compression (preparation technique), changing the side of preparation and watching for recovery of the SSEP signal. SSEP is highly sensitive, but limited in specificity. SSEP amplitude reduction above 50% or a transient signal loss may go along with full recovery, while a permanent intraoperative signal loss indicates new neurological deficit in 50 to 75% of cases [109].

Dissection close to the corticospinal motor tracts may lead to signal changes in mMEP and D-wave. D-wave monitoring is robust and predictive for motor function outcome. Reduction of the D-wave amplitude by more than 50% compared to that during skin incision predicts a permanent motor deficit and must be avoided at any cost. Thus, reduction of the D-wave amplitude should not occur or must prompt a comprehensive team reaction: pausing of dissection, warm saline irrigation of the surgical site, elevation of mean blood pressure by 10 to 20 mmHg, and waiting for signal recovery [110]. Reduction or loss of mMEP predicts a temporary deficit and may be accepted with D-wave amplitude at baseline if further resection seems necessary in cases with presumed low-grade histology and a plane for dissection. Gentle surgical manipulation must be very closely guided by D-wave monitoring in order to avoid a permanent deficit. Alternatively, a staged secondary intervention may be considered. When D-wave recovery does not occur, surgery should be finished without further tumor removal and an expansion duraplasty might be considered in cases with larger tumor remnants or cord swelling. Secondary surgery might be planned with a definite histological diagnosis, maybe after rehabilitation (depending on the achieved surgical decompression of the cord) and after developing a sound hypothesis for the drop of D-wave amplitude in order to prevent or mitigate the cause during the next surgical attempt.

For IMSCT, safe entry zones into the cord have been described in detail [111–113]. The posterior midline myelotomy is feasible in most intrinsic spinal cord tumors with a central or slight paramedian extension (see Case 3). For lateralized IMSCTs posterior to the level of the attachment of the dentate ligament, the posterolateral sulcus (dorsal root entry zone) is feasible [111]. Anterior or anterolateral intramedullary tumors, which are very rare, may need an anterior approach when feasible with a paramedian anterior myelotomy (Fig. 3) [93].

Access to the spinal canal should be large enough in length (through laminotomy or laminectomy) and cover the extension of the solid tumor part in order to control protrusion of the cord and tumor into the dural opening by the immediate release of CSF and/or opening of a tumor cyst. Intraoperative ultrasonography helps to reconfirm sufficient exploration of the dura in relation to tumor extension and to identify an ideal starting point for myelotomy (at the center of the tumor). Myelotomy should respect the vasculature in general, but a draining vein over the posterior midline sulcus might be obliterated. Especially at the posterior midline, the sulcal

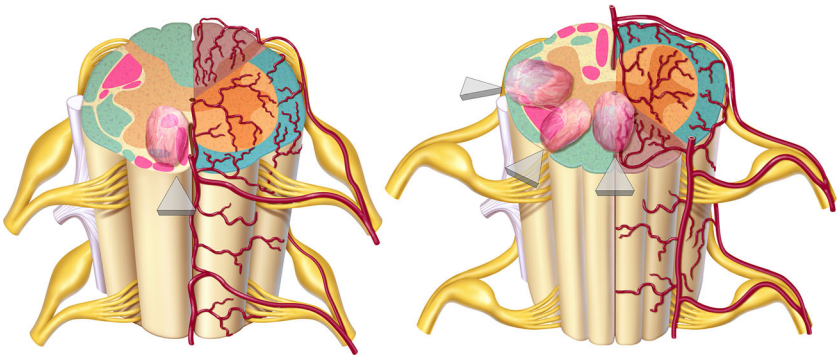

**Fig. 3** Cross-section of the spinal cord showing ascending (pink) and descending (green) tracts, the ligamentum denticulatum (on the left) and the vascular supply (on the right). *Left*: anterior view with blood supply from the anterior spinal artery (predominantly supplying the motor area). The paramedian anterior myelotomy is indicated to resect tumors situated anteriorly or anteriolaterally. *Right*: posterior view showing one of two posterior spinal arteries and the surgical approaches to the spinal cord from right to left; posterior midline myelotomy, posterolateral myelotomy at the dorsal root entry zone (posterolateral sulcus), and lateral approach with entry point at the level of the dentate ligament

microvasculature guides the microsurgical separation of the posterior columns down to the tumor. In cases of attempted maximum tumor resection, the myelotomy should be extended in length to the poles of the tumors for full visualization and preparation (see Case 3) [61]. Myelotomy is performed by sectioning the arachnoid and bluntly dissecting within the plane while avoiding focal pressure on the adjacent tissue (by using, e.g., plated forceps or broad blunt bipolar but always preparing a shallow and long, but not deep and small approach).

Preparation of the tumor-spinal cord interface depends on differences in color, texture, and vascularity and might be performed with microdissectors or ultrasonic aspiration under high magnification. Pial retention sutures at the posterior cord attached to the dura or supported by weights (i.e., mini-bulldog clamps) might minimize repetitive alteration of the dissected surface of the cord, but stretching of the cord tissue by the pial sutures must be avoided.

Surgery might start with debulking of the tumor by ultrasonic aspiration. A fast-stain histology evaluation is recommended. Identification of the tumor-spinal cord interface starts on the lateral tumor borders. Gaining early control of the anterior extension of the tumor at the tumor poles eases intraoperative orientation especially in cases of cystic tumor poles. The decision to aim for GTR might be based on two factors: malignancy verified by fast-stain histology and the continued identification of a plane of dissection. In cases of a non-malignant or inconclusive grade of malignancy on fast stain, "smart" resection (relying totally on a distinctive tumor–cord interface with cessation of resection with loss of this guiding plane [50]) and IOM-guided resection are mandatory. Bipolar coagulation should be avoided to reduce the risk of spinal cord alteration. Identification and preservation of the anterior spinal cord artery or its tributaries to the anterior cord is absolutely mandatory and damage by traction, evulsion of tributaries, or direct coagulation must be avoided. Reconstruction of the cord by pial sutures is advised to minimize the risk of posterior tethering of the dorsal columns and formation of a secondary syringomyelia. Duraplasty should be used to expand the CSF space in cases with tumor remnants or residual cord swelling to improve CSF flow around the cord [61].

## Radiosurgery

Surgery remains the first choice treatment for spinal intradural tumors. Radiotherapy has been used for years especially after STR in intramedullary ependymomas or astrocytomas with conventional fractionation and doses of 45 to 50 Gy, limited by the risks of radiation myelopathy and gastrointestinal or fertility compromise and with mixed effects on tumor control [114]. In recent years, stereotactic radiosurgery for spinal lesions, first described by Hamilton et al. in 1995 [115], has become an effective alternative for patients with recurrent or residual disease or in cases where surgery is contraindicated. Several authors reported excellent results [116–118]. Gerszten et al. observed no subacute or long-term spinal cord toxicity in 41 patients treated for benign intradural spinal tumors [119]. Sachdev et al. reported on 103 benign intradural tumors treated with radiosurgery with only one tumor progression during the mean follow-up period of 33 months [29]. Shin et al. showed promising results for intramedullary metastases without detecting any radiation toxicity during follow-up [120]. A recent systematic literature review on stereotactic radiosurgery for intramedullary spinal cord tumors concluded that the technique is safe and effective in selected cases [121].

Due to technical advances like the cone beam CT image guidance used in a case series of ID-EM tumors published by Monserrate et al. [122], radiosurgery has become more effective and accurate which is reflected in satisfactory results and low complication rates.

However, long-term data are still lacking and because the therapeutic effect of radiosurgery sets in slower and is less pronounced than surgery, radiosurgery remains a first choice treatment alternative only in a select group of patients [123]. While radiosurgery on spinal tumors is an established and well-described procedure [124, 125], the associated treatment-related toxicity on adjacent organs which are at risk in about 25% of cases has to be taken into consideration [126]. The same is true for radiosurgery on intradural spinal lesions, especially on benign lesions. The risk of spinal cord toxicity and radiation myelopathy has to be discussed with the patient. Nevertheless, radiosurgery will most likely be developed to play a more important role in the treatment of spinal intradural lesions as it has with cranial tumors.

While stereotactic radiosurgery seems to pose more importance in future treatment regimens, conventional radiotherapy has been used to treat spinal tumors, especially extensive intramedullary tumors, with satisfying results [127].

## Outcome (quality of life)

The treatment strategies discussed above lead to satisfactory outcomes for intradural lesions. Functional outcome factors and survival have been discussed with the clinical entities as they pertain to the timing of surgery, grade of resection, age, location, perioperative management, and complications.

In ID-EM tumors, the anatomical location of the tumor is a prognostic factor. A study by Mehta et al. found that tumors of the upper anterior thoracic spine have the highest rate of surgical complications [128]. While ventral positioning of the tumor has been described as a negative prognostic factor by some [129], others have not described this relationship [130].

In IMSCT, other factors such as the longitudinal extension of the intramedullary tumor (extension beyond three vertebral levels [131]) have been identified as prognostic factors in addition to tumor location and histopathological diagnosis.

Quality of life with IMSCT, as a patient's perspective on treatment and outcome, will gain more attention. The postsurgical neurological status and quality of life in patients with IMSCT, measured using EQ-5D and PDQ and its improvement above the minimal clinically important difference (MCID), also depends on the preoperative neurological status (modified McCormick Score; for EQ-5D), preoperative EQ-5D or PDQ, histology (ependymoma), tumor location (cervical location as a negative factor), and length of stay (EQ-5D). Additionally, the incidence of surgical complications (CSF leak, syrinx formation) was a significant factor for QOL (EQ-5D), thus highlighting the importance of proper treatment planning and surgical technique for the patient [132].

## Illustrative cases

### Case 1: intradural spinal meningioma

A 58-year-old man presented to our Neurosurgery department with headaches and neck pain not associated with any neurologic deficits. The MRI with contrast of the cervical spine showed a huge left-sided intradural extramedullary tumor suspicious for a meningioma at the level of C1/C2 with severe cord compression (Fig. 4). The surgery was planned with SSEP and mMEP monitoring. We performed a C1 laminectomy, C2 left hemilaminectomy, and bony widening of the foramen magnum on the left. The dura was opened in a C-shaped fashion to the left side. The tumor was partly calcified and therefore debulking was not possible. The dural entrance of the left vertebral artery was controlled and the tumor matrix at the spinal dura was sharply resected. The tumor was mobilized and removed en bloc (Simpson grade II) with preservation of spinal veins by stripping the arachnoid off the tumor capsule. The patient was discharged home on postoperative day 6 with mild neck discomfort and normal neurological status. MRI revealed complete tumor removal.

### Case 2: intradural spinal schwannoma

A 19-year-old boy developed weight loss and slowly progressing tetraparesis. Neurological examination revealed tetraparesis grade 4 MRC with spastic gait disturbance and brisk tendon reflexes. Severe cord compression from a right-sided dumbell-shaped tumor at C3 was seen on MRI. After an unsuccessful attempt to address the anterolaterally located intraspinal tumor via a posterior approach at an outside institution, the patient was referred to our department (Fig. 5). Using a right-sided anterolateral approach, the tumor was removed via the widened neural foramen without facet drilling while maintaining direct control of the vertebral artery (Video 1). A transient chemical meningitis was successfully treated with cortisone after negative CSF cultures at the rehabilitation department. The patient regained full motor and sensory function and started a voluntary social year at a medical institution. He had no tumor recurrence at long-term (8 years) follow-up.

### Case 3: intramedullary spinal ependymoma

A 62-year-old presented with mild gait disturbance to the outpatient department. Neurological examination revealed mild sensory deficits in the lower limbs, very mild spinal ataxia, and no motor deficits. MRI showed an intramedullary mass at the level of C2 to C7 with central location, cystic widening of the central canal at the lower pole, a fairly distinct lesion border, and an inhomogenous contrast enhancement, suggesting ependymoma as the likely diagnosis (Fig. 6). The tumor was removed en bloc via a multi-level laminotomy by

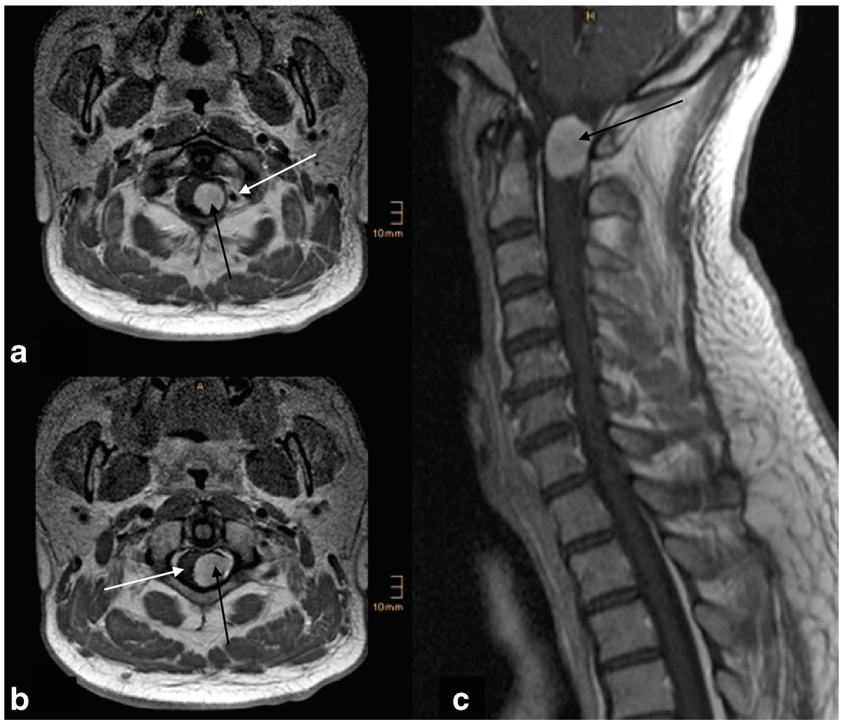


**Fig. 4** Spinal intradural meningioma (black arrow) originating from the perivascular dura at the entrance of the left vertebral artery at C1 (VA, white arrow at Fig. 1a). The spinal cord is shifted to the right side (white arrow on Fig. 1b). The tumor was removed en bloc via a posterior approach with sharp dissection from the dura (Simpson Grade II)

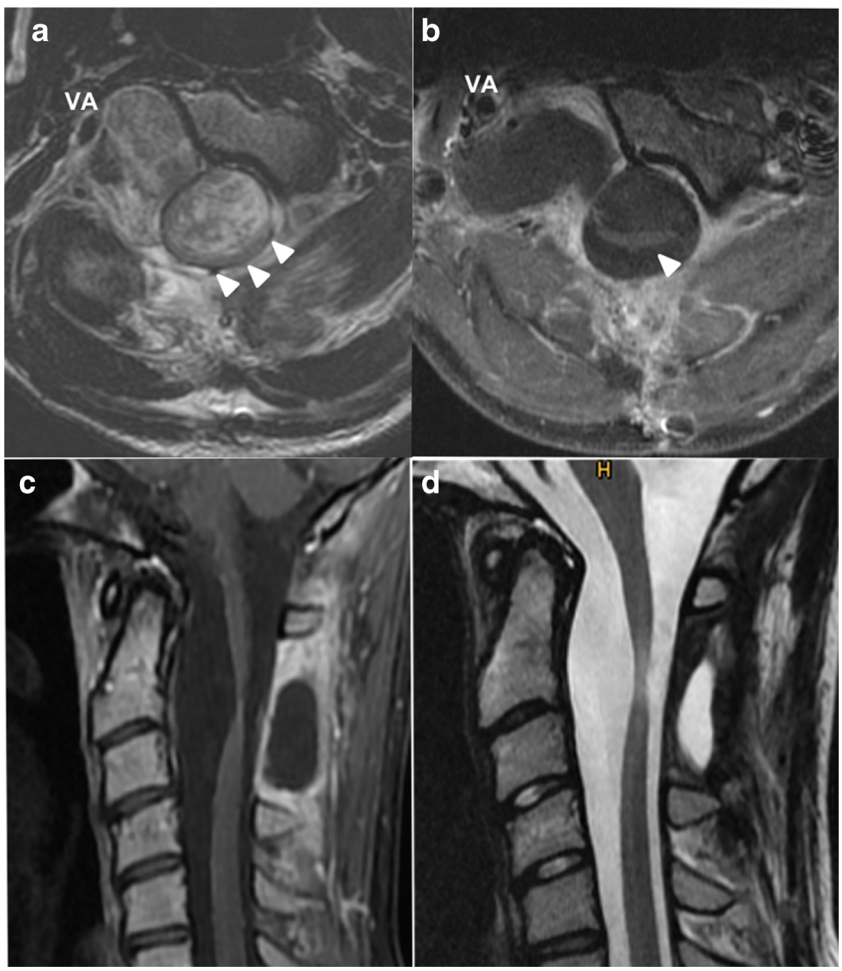


**Fig. 5** Right-sided dumbell-shaped schwannoma at C2/3 with severe spinal cord compression. The spinal cord is stretched out at the posterior border of the spinal canal in a semilunar shape (**a**, white arrows). The extradural part reaches anterior and medially to the vertebral artery (**a**, VA). The tumor was removed completely via a right-sided anterolateral transforaminal approach with transient translocation of the vertebral artery (**b**, VA). The spinal cord was decompressed (**b**, white arrow; see Video 1). Fluid collection within the dorsal extradural midline remained from an unsuccessful posterior approach performed at another institution (**c**)

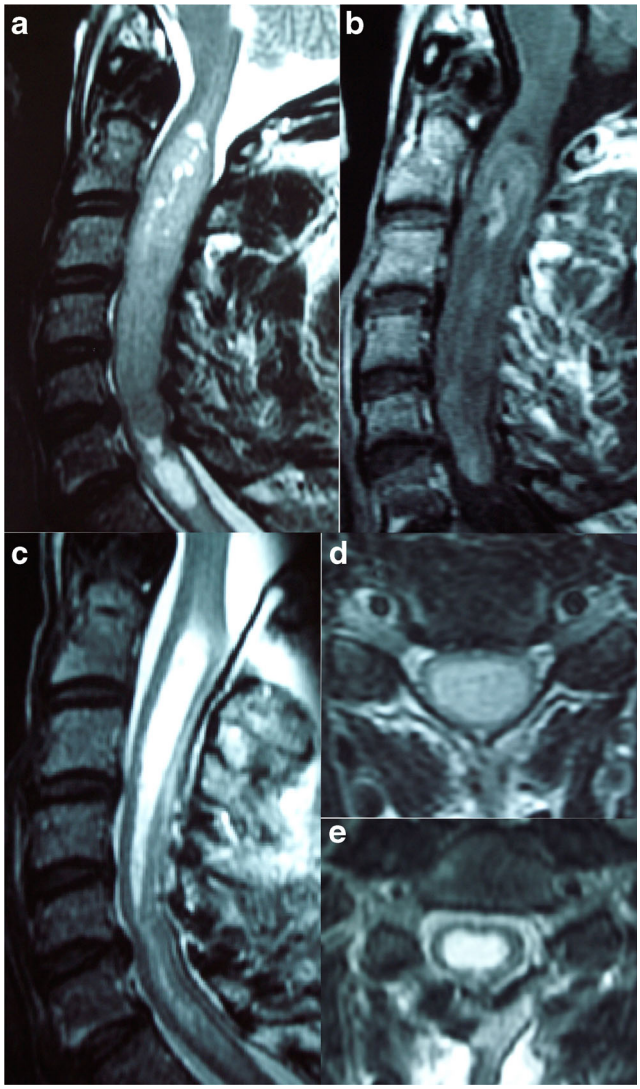


**Fig. 6** Intramedullary ependymoma WHO II, C2 to C6. The tumor was removed en bloc via a midline laminotomy approach and midline myelotomy supported by neuromonitoring (see Video 2). The tumor is bordered by a small syrinx cavity at the top and the bottom (**a**, T2w MRI), enhances irregularly with gadolinium (**b**, T1w MRI with gadolinium) and is located centrally within the spinal cord (**d**, T2w MRi). After resection, the spinal cord is decompressed and the resection cavity filled with CSF (**c** and **e**, Tw2 MRI)

**Table 5** Key considerations in the management of intradural spinal tumors

| Important considerations |
|---|
| - Early MRI |
| - Early resection |
| - Neuromonitoring in all intramedullary tumors |
| - Neuromonitoring in all ventrally located or large intradural tumors |
| - Function-preserving surgery |
| - GTR whenever possible |
| - Adjuvant chemo- and radiation therapy in malignant or recurrent cases |

microsurgical dissection with IOM (Video 2) and was diagnosed as a spinal cord ependymoma, WHO II. Postoperative MRI showed complete resection and restored cervical lordosis with laminotomy (Fig. 6). Gross and fine motor and sensory skills including hand-writing and buttoning of the shirt were intact 1 week after tumor removal (Video 2). The patient was monitored without further intervention and remained free of recurrence at long-term follow-up.

## Conclusion

Spinal lesions can occur intra- or extradurally. Intradural lesions can be further divided into intramedullary and extramedullary. Meningiomas and nerve sheath tumors comprise the most common extramedullary tumors while ependymomas and astrocytomas are the most common intramedullary lesions. While GTR can be achieved in most benign lesions, a more function-preserving approach is chosen in malignant intramedullary lesions. Advances in microsurgery, neuromonitoring, and adjuvant therapies have led to excellent outcomes in benign tumors while outcomes are less favorable in malignant disease. When treating intradural lesions, we recommend early resection with IOM, and carefully tailored function-preserving approaches in combination with radio- and chemotherapy in cases of malignant transformation (Table 5).

**Acknowledgements** The authors thank Alexander Bock, MD, Head of Department of Neuroradiology, Vivantes Klinikum Neukölln, Berlin, Germany, for close clinical cooperation and for providing MR images. We also thank Matthew Holt, Medical Illustrator, for the outstanding medical illustrations.

**Funding** None.

## Compliance with ethical standards

**Conflict of interest** On behalf of all authors, the corresponding author states that there is no conflict of interest.

**Ethical approval** All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. For this type of study, formal consent is not required.

**Informed consent** Informed consent was obtained from all individual participants for whom identifying information is included in this article.

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