

8 Comprehensive Treatment Strategies for Spinal Meningiomas

Robert J. Rothrock, Ori Barzilai, Yoshiya (Josh) Yamada, and Mark H. Bilsky

Key Points

- 1. Spinal meningioma is classically a WHO grade I psammomatous meningioma presenting in an elderly female in the thoracic spine.
- 2. Observation is a valid management strategy in an asymptomatic patient with an incidentally discovered, non-compressive tumor.
- 3. The goal of surgery is gross total resection along with the dural attachment (Simpson grade I) or gross total resection with cauterization of the dural attachment (Simpson grade II).
- 4. Stereotactic radiosurgery provides durable local control as both adjuvant and salvage therapies.
- 5. Rates of recurrence are directly related to both WHO grade and the initial extent of resection (EOR).

Introduction

Spinal meningiomas are the second most common intradural extramedullary tumor and are commonly encountered in neurosurgical practice. In the era of widespread access to magnetic resonance imaging (MRI), these tumors can be encountered incidentally, leaving patients and referring providers anxious for treatment recommendations. This chapter will systematically address considerations for diagnosis and management of spinal meningioma.

© Springer Nature Switzerland AG 2020 109

J. Moliterno, A. Omuro (eds.), *Meningiomas*, [https://doi.org/10.1007/978-3-030-59558-6_8](https://doi.org/10.1007/978-3-030-59558-6_8#DOI)

R. J. Rothrock \cdot O. Barzilai \cdot M. H. Bilsky (\boxtimes)

Department of Neurosurgery, Memorial Sloan-Kettering Cancer Center, New York, NY, USA e-mail: bilskym@mskcc.org

Y. (J.) Yamada

Department of Radiation Oncology, Memorial Sloan-Kettering Cancer Center, New York, NY, USA

Part I:Epidemiology, Pathology, and Diagnosis of Spinal Meningioma

Epidemiology

Although seen routinely in neurosurgical practice, spinal meningioma is a rare clinical entity. The incidence of intradural extramedullary spinal tumors is estimated to be 0.74 in 100,000 person-years [\[1](#page-11-0)]. Spinal meningiomas are thought to represent approximately 12% of overall meningiomas and account for 40–45% of all benign intradural extramedullary tumors [[1,](#page-11-0) [2\]](#page-11-1). Similar to intracranial meningiomas, the female to male ratio is 3:1 [[3\]](#page-11-2) with a peak incidence in the sixth and seventh decades [\[2](#page-11-1)]. Although more common in women, male sex has been associated with a higher rate of recurrence of spinal meningioma following surgery [\[4](#page-11-3)].

Risk factors for spinal meningioma include prior radiation exposure as well as syndromic predisposition, such as neurofibromatosis type 2 (NF2) [\[5](#page-11-4), [6](#page-11-5)]. Patients with these risk factors tend to present with spinal meningiomas at a younger age [\[4](#page-11-3)]. Spinal meningiomas have a predilection for the thoracic spine, but have been described in all areas of the spine, including cervical, lumbar, and sacral [\[7](#page-11-6), [8](#page-11-7)].

Histology and Pathophysiology

Meningiomas are thought to arise from arachnoid cap cells, which form the outer layer of the arachnoid, and facilitate the drainage of cerebrospinal fuid (CSF) into dural sinuses and veins [[9\]](#page-11-8). This hypothesis arose from the observation that normal arachnoidal cap cell clusters in older patients can form whorls and psammoma bodies identical to those found in meningiomas. However, the cell of origin for meningiomas has not been clearly defned. Others have proposed the possibility of fbroblastic origin, having to do with the mesodermal features observed in these tumors, and that they have sometimes arisen independently from dural attachment [\[10](#page-11-9)]. Meningiomas display both mesenchymal and epithelial-like features, which help establish diagnosis [[9\]](#page-11-8).

Several stereotypic genetic alterations have been identifed within spinal meningioma, but few have altered clinical practice. The most common is loss of heterozygosity of the 22q12.2 chromosomal region of the neurofbromatosis type 2 (NF2) gene, which can be found in 40 to 70% of sporadic meningioma as well as most NF2-associated meningiomas [[9,](#page-11-8) [11\]](#page-12-0). Other chromosomally related genes that have been implicated include the beta-adaptin BAM22 gene and the tissue inhibitor of metalloproteinase 3 (TIMP3) gene [\[12](#page-12-1), [13](#page-12-2)]. Additional genes have been associated with progression from low- to higher-grade meningioma, such as the 4.1 family member *DAL1* and the tumor suppressor in Lung Cancer-1 (*TSLC1*) gene [[14,](#page-12-3) [15\]](#page-12-4).

The World Health Organization defnes three grades of meningioma and a far greater number of histologically distinct morphologies [\[16](#page-12-5)]. Classically, the psammomatous subtype is the most common spinal meningioma, usually thoracic in location and predominantly observed in elderly female patients. Psammomatous

meningiomas express bone-related proteins, such as osteopontin, which are thought to contribute to their relatively high degree of calcifcation [\[17](#page-12-6)]. Conversely, clear cell meningiomas are found in younger patients and also predominantly thoracic in location [\[18](#page-12-7)]. Although they are histologically low grade, they are clinically more aggressive with higher rates of recurrence.

For the surgeon, the most important histopathological distinction remains grade I versus grade II/III tumors, the latter of which exhibit cellular atypia, pial invasion, and more aggressive clinical behavior (i.e., recurrence and local invasion) [[19\]](#page-12-8). Like their intracranial counterparts, spinal meningiomas are most commonly WHO grade I, but can also be atypical (WHO grade II/III). WHO grade III meningiomas, i.e., anaplastic meningioma, are differentiated by 20 or more mitoses per 10 highpower felds or the presence of frank anaplasia – defned as carcinoma-, melanoma-, or sarcoma-like histology [[9\]](#page-11-8).

Within grade I spinal tumors, the presence of a clear dural tail on preoperative imaging has been associated with high-grade transformation and invasive, recurrent behavior [[20\]](#page-12-9). WHO grade on initial pathology correlates with recurrence rates, with WHO grade I spinal meningioma undergoing a Simpson grade I or II resection demonstrating 0% recurrence at 10 years in one series [\[21](#page-12-10)]. In addition, given the indolent course of WHO grade I tumors, even with recurrence, symptoms typically develop slowly [[22\]](#page-12-11).

Imaging

Magnetic resonance imaging (MRI) is the preferred diagnostic imaging modality for spinal meningioma [[23](#page-12-12)]. Diagnostic consideration is raised with the presence of an intradural extramedullary lesion. Spinal meningiomas are 90% intradural extramedullary lesions, with only 10% manifesting either extradural or dumbbell (intradural/extradural) in location [[24](#page-12-13)]. Partial or complete calcifcation is highly suggestive of meningioma, but is only seen in approximately 5% of cases [[25](#page-12-14), [26\]](#page-12-15). MRI characteristically reveals avid enhancement with administration of gadolinium contrast. In addition, meningiomas are more specifcally hypo- or isointense on T2-weighted imaging compared to schwannomas, which are typically hyperintense [[27\]](#page-12-16).

Although the presence of a dural origin or tail to a lesion is specifc to meningioma, this is observed less frequently than in intracranial meningioma [\[23](#page-12-12)]. Common to all meningiomas, the blood supply is usually localized to the dural origin of the lesion. Large meningiomas may demonstrate signifcant intratumoral vasculature. Although rare, meningiomas may be fully extradural or even intramedullary [\[28](#page-12-17), [29\]](#page-12-18). In one case in our institutional series, a primary meningioma was resected in the extrapleural space over 6 cm from the closest dural margin.

Computed tomography (CT) can be a useful adjunct to delineate the degree of tumoral calcifcation or remodeling of local bony structures, both of which can be suggestive of a diagnosis of meningioma [[30\]](#page-13-0). CT can also be used to assess bone quality in patients who will require spinal instrumentation as an element of surgery.

Spinal angiography can be utilized in cases of hypervascular tumors as suggested by MRI, both to visualize the main vascular supply and to perform preoperative embolization [[31\]](#page-13-1). For lower thoracic tumors, spinal angiography may be important for determining the origin of the artery of Adamkiewicz, which is the dominant radiculomedullary feeding artery. This vessel commonly originates on the left side at T9 or T10, in 75% and 50% of cases, respectively; however, the location is variable [\[32](#page-13-2)]. Sacrifce of the artery of Adamkiewicz can be neurologically devastating to a patient, but alterations can be made to a surgical approach if the origin is determined preoperatively.

Terminology for Spinal Meningioma

The Simpson grade of resection describes the amount of residual tumor left after surgery [\[33](#page-13-3)]. Simpson grade I describes complete resection of the meningioma as well as its dural attachment. Grade II describes gross total resection of the meningioma with coagulation of the dural attachment. Grade III refers to gross total resection of tumor without coagulation of the dural attachment or any extradural component. Grade IV refers to subtotal tumor resection, and grade V is a biopsy or simple decompression surgery. The clinical importance of the Simpson grade for the purpose of intracranial neurosurgery is the direct correlation with rates of tumor recurrence if residual tumor is left behind [\[34](#page-13-4)]. Unlike the case of intracranial convexity meningiomas, it is often not feasible to obtain a Simpson grade I resection of a spinal meningioma given the high morbidity resulting from spinal CSF leak, which is more complicated with dural resection. Thus, Simpson grade II resection is often the primary surgical goal [[21\]](#page-12-10).

The McCormick grade describes the level of functional impairment from intradural tumor-associated myelopathy [\[35](#page-13-5)]. Originally developed in reference to spinal intramedullary ependymoma, this scale is now utilized for many intradural tumor studies. McCormick grade I refers to a neurologically "normal" patient with unimpaired and independent gait allowing for mild spasticity or refexes on exam. Grade II describes functionally independent gait in the presence of focal neurologic defcit or mild diffculty with balance (also severe pain). Grade III requires a cane or assistive device for ambulation with more severe neurologic deficit and/or bilateral upper extremity impairment. Grade IV describes a severely disabled patient who requires a wheelchair for mobility and with a severe/dense neurologic defcit.

Natural History of Spinal Meningioma

Defning the natural history of spinal meningioma has become more feasible with increasing use of MRI imaging and incidental discovery of clinically silent intradural extramedullary tumors [\[36](#page-13-6)]. While it can be diffcult to determine the histopathology of a tumor simply from imaging, the presence of a clear dural tail on MRI reliably denotes a spinal meningioma [[27,](#page-12-16) [37\]](#page-13-7). Interestingly, some studies have shown a higher growth rate for meningioma versus schwannoma over 5-year serial imaging follow-up [\[38](#page-13-8)]. Even with tumoral growth, however, there is no assurance that a relatively slow-growing spinal meningioma will become symptomatic. For those lesions that do become symptomatic in the presence of serial imaging and history, surgery is offered more promptly, especially with clinical deterioration.

Differential Diagnosis

The main differential diagnosis for an intradural extramedullary lesion on seen on MRI includes schwannoma, meningioma, neurofbroma, solitary fbrous tumor/ hemangiopericytoma, malignant peripheral nerve sheath tumor, leptomeningeal metastasis (solid or hematopoietic), as well as paraganglioma and myxopapillary ependymoma depending on location [[39\]](#page-13-9). In the absence of a history of cancer or a multisystemic syndrome, the main considerations are schwannoma, meningioma, and neurofbroma, with the frst two being the more common. As described earlier, the presence of a dural tail or calcifcation is considered more specifc for meningioma. Since both schwannoma and meningioma tend to avidly enhance with gadolinium, T2-weighted imaging is considered to be a differentiating factor, with hypointensity favoring meningioma. Using this method, however, only yields 80% sensitivity and 75% specificity [\[27](#page-12-16)]. In cases where both schwannoma and meningioma are considerations, operative planning should account for both scenarios. Intraoperative fndings often help differentiate the two entities, but histopathology is the gold standard for fnal diagnosis.

In patients presenting with suspected spinal meningioma at a young age, a diagnosis of NF2 should be considered, especially if multiple lesions are seen on MRI and without the stigmata of NF1 (which may instead favor multiple schwannoma/ neurofbroma). Multiple intradural extramedullary lesions should also raise consideration for schwannomatosis, which is more common in persons of Japanese and Ashkenazi Jewish descent [[40\]](#page-13-10). Discovery of multiple lesions should also prompt imaging of the entire neuraxis.

Part II: Management of Spinal Meningioma

Management of a suspected spinal meningioma is often dependent on the manner of discovery. Incidental lesions are usually managed expectantly with serial imaging, whereas lesions presenting with a neurologic defcit or severe biologic pain are typically operated at presentation.

Observation

With the increased use of CT and MRI, many intradural extramedullary lesions are incidentally discovered. Unlike intracranial meningiomas, spinal tumors do not usually penetrate the pia and do not result in spinal cord edema by direct invasion. Intramedullary spinal cord edema results from the compressive effects of the tumor and may not be manifest radiographically until the tumor is resected [\[41](#page-13-11)]. Whereas relatively small intracranial meningiomas can cause signifcant vasogenic edema (and become symptomatic), spinal meningiomas become symptomatic once they exhibit mass effect directly on the neural elements. For incidentally discovered suspected meningiomas, clinical and radiographic observation with serial visits and MRI is often the best management strategy. Even if growing on serial imaging, consideration must be given to the patient's age, comorbidities, and the relative size of the tumor if he or she is asymptomatic.

Surgical Indications

Surgical resection is indicated in patients with a progressive neurologic deficit, unremitting biologic pain or radiculopathy directly attributable to the tumor, or progressive growth on serial imaging with radiographic spinal cord compression. Biologic pain describes infammatory pain directly caused by the tumor, as opposed to mechanical pain which is caused by spinal instability. Biologic pain from a spinal tumor usually occurs at night when the body's endogenous steroid production is at its lowest. Similarly, biologic pain improves with the administration of exogenous steroids [[42\]](#page-13-12).

Surgery is not performed for diagnosis alone, given that the main differential considerations are also benign. One notable exception is in the case of a conus medullaris or flum terminale tumor in which myxopapillary ependymoma is a consideration. In this case there is a risk of tumor leptomeningeal seeding with delayed intervention [[43\]](#page-13-13). Progressive neurologic defcit is usually in the form of clinical myelopathy for cervical or thoracic tumors and multi-nerve distribution radiculopathy in the case of lumbar tumors. Because meningiomas are slow-growing tumors, they are extremely unlikely to present with an acute compression syndrome and require emergent surgery.

Surgical Goals

The goal of surgery is gross total resection if feasible (Simpson grade I), but at least Simpson grade II resection, given the durable low rates of recurrence with these results. In cases of ventral tumor where gross total resection is not safely achievable, small amounts of tumor should be left rather than cause a new or worsened neurologic defcit. Residual tumor can be observed and treated with stereotactic radiation if interval growth is seen.

Surgical Approach and Considerations

The surgical approach is determined by the location of the tumor. A posterior laminectomy or posterolateral approach can be employed for virtually all spinal meningiomas. Ventrally located cervical tumors are a category where some authors have advocated for resection using an anterior approach via a vertebrectomy [\[44](#page-13-14)]. Even though direct visualization of the tumor may be easier via an anterior approach, primary dural closure and reconstruction of dural defects is extremely diffcult. Despite potential benefts, this approach is rarely indicated. Recently, some authors have advocated for the use of minimal access surgery, including tubular retractor systems, especially in patients presenting at an advanced age [\[45](#page-13-15)]. The potential advantages include decreased muscle dissection and a better, watertight fascial closure. At present, this approach has not been widely adopted for meningiomas, but remains an area of active investigation in select centers.

Resection Techniques

Localization of the tumor is critically important. If available, intraoperative CT is a reliable localization technique. In the absence of this modality, the lumbar and cervical spine levels can typically be imaged with cross-table lateral plain radiographs or fuoroscopy. Thoracic tumors can be reliably localized using an anterior-posterior (AP) fuoroscopic image with a marker that lines up with the rib and pedicle at the level of interest. The correct level is identifed counting from either the T1 or T12 ribs.

Following exposure and radiographic confrmation of the level, laminectomy is performed with a high-speed drill using a 3 mm matchstick or diamond burr and 2 mm Kerrison Rongeur. Rarely, a posterolateral resection (i.e., laminectomy with unilateral facetectomy and pedicle resection) is required for exposure. Intraoperative ultrasound is used to ensure that the laminectomy has encompassed the superior and inferior extent of the tumor. All bone edges are waxed, and a hemostatic agent is placed in the lateral gutters.

At this point, the operating microscope or alternatively the exoscope is employed for dural opening and tumor resection. The durotomy is created with a 15 blade over a Woodson dental tool used to protect the intradural neural elements and tacked back with dural sutures. The dural opening is typically straightforward with the exception of calcifed meningiomas. The calcifed component can be dissected from the dura using micro-instruments, such as Rhoton 2 and 6 dissectors. Bisecting the dural calcifcation with a 1 mm side cutting or diamond drill bit allows for the safe removal of the tumor while preserving the dura and preventing spinal cord injury [\[46](#page-13-16)]. The soft tissue component of the meningioma is often deep to the calcifed mass, providing a safe margin of resection.

Following dural opening, the arachnoid is opened sharply, and the tumor is explored to defne the interface with the spinal cord. WHO grade I spinal meningiomas rarely violate the pia providing a plane of dissection. Defning the spinal cord margin is considerably more difficult in grade II/III meningioma, which may be invasive into the spinal cord. For ventrally located cervical and thoracic meningiomas with limited exposure, the dentate ligament may be sectioned to allow for slight rotation of the spinal cord during resection. This can be accomplished with 7-0 Prolene stitches placed through the dentate ligament and secured to the contralateral dura with a small vascular clip.

Safe resection is facilitated by intratumoral debulking using an ultrasonic aspirator and sharp resection. The tumor is dissected from the ventral spinal cord using Rhoton 2 and 6 instruments. Following decompression of the spinal cord, the meningioma is resected off the dural attachment using sharp dissection. This margin may be hypervascular, and bipolar cautery is used for hemostasis. Typically, the inner layer of the dura is resected with an 11 blade, and the dura is bipolar-cauterized to achieve a Simpson grade II resection. In some centers, the dura is resected to achieve a Simpson grade I resection.

Dural closure is accomplished with a 4-0 Surgilon or Prolene running suture. For cases in which dural resection is undertaken, reconstruction may best be accomplished with Dura-Guard (Baxter International, Minnesota) using a running 4-0 Surgilon stitch. Dura-Guard is a bovine pericardial patch graft that sews water tight and, in our experience, has little infection risk even when abutting contaminated spaces. Following dural closure, thrombin glue and gel foam are placed over the dura. A subfascial Jackson-Pratt drain is left in place to bulb suction for 24 h. The wound is closed in multiple layers to obliterate the dead space. The paraspinal muscles are closed with 2-0 Vicryls followed by a running 0 PDS in the fascia. The subcutaneous tissue is closed with 0 Vicryls and a 3-0 Monocryl running subcuticular stitch and a 3-0 Nylon running stitch for the skin (Fig. [8.1](#page-8-0)).

Immediate Postoperative Management

Several postoperative management issues require special attention. For 24 h, the head of bed (HOB) is raised no higher than 30 degrees to provide time for reconstitution of CSF. After 24 h, the HOB is slowly elevated, and patients are evaluated for headaches associated with intracranial hypotension. If headaches occur, the HOB is returned fat, and a more graduated head elevation is pursued. For persistent headaches in the absence of a known CSF leak, patients are placed on Fioricet and encouraged to ambulate. The subfascial Jackson-Pratt drain is kept on bulb suction for 24 h while the head of bed is low and then taken off suction and left in place for an additional 72 h.

Typically, patients are started on Decadron 3 days prior to surgery to reduce infammation associated with the tumor and to provide neural protection. Postoperatively, patients are placed on 6 mg Decadron 4 times a day. A rapid taper is performed every other day until 2 mg Decadron daily is achieved. At this time, 2 mg daily is continued for 5 days to prevent the sequelae of chemical meningitis.

Fig. 8.1 A 65-year-old female presenting with worsening ataxic gait and leg buckling. (**a**) T1-weighted MRI demonstrates a large isointense intradural extramedullary mass at T11. (**b**) STIR MRI demonstrates heterogeneous signal intradural extramedullary mass at T11. (**c**) Axial T2 demonstrates 90% canal compromise with high-grade cord compression. Green arrow highlights crescentic-appearing compressed spinal cord. Preoperative differential diagnosis included meningioma and schwannoma (tumor appeared to extend toward left T11–T12 foramen). Patient underwent bilateral T10 to T11 midline laminectomy and intradural tumor resection with Simpson grade I resection. Final pathology was WHO grade I meningioma. (**d**) Postoperative axial T1 postcontrast MRI demonstrates gross total resection. (**e**, **f**) Postoperative sagittal T1 post-contrast and T2-weighted MRI demonstrate gross total resection of tumor and re-expansion of the spinal cord. The patient's gait improved postoperatively, and she returned to work 3 months following surgery

Instrumentation

The use of spinal instrumentation is rarely required, but is increasingly being used for specifc indications. Instrumentation is not commonly required for lumbar and mid-thoracic tumor resection. There is evidence for the use of instrumentation for multilevel cervical laminectomies, laminectomies performed at the cervicothoracic or thoracolumbar junctions, or laminectomies performed across the thoracic apex to decrease risk of delayed spinal deformity [\[47](#page-13-17), [48\]](#page-13-18). In cases of laterally located tumors, resection of the facet joint or a portion of the pedicle may be required to adequately visualize the tumor resection, and in these cases spinal instrumentation is recommended [[49\]](#page-13-19). In the case of multilevel cervical exposure without violation of the facet joints, laminoplasty is an option [\[50](#page-14-0)]. Laminoplasty is likely effective in preventing development of delayed spinal deformity, but importantly is likely not effective in treating an already present spinal deformity in the context of intradural tumor resection [\[51](#page-14-1)].

Surgical Adjuvants

Intraoperative neurophysiologic monitoring (IOM) is often used as an adjuvant in the resection of intradural extramedullary tumors, including somatosensory evoked potentials (SSEP), electromyography (EMG), and motor evoked potentials (MEPs).

Epidural D-wave monitoring is not routinely employed in our center for these tumor types. MEPs are useful in avoiding iatrogenic injury if rotation of the spinal cord via the dentate ligaments is necessary for tumor resection. Intraoperative corrections to blood pressure and hypothermia can be made to address changes in MEP's that may not be specifcally related to manipulation of the spinal cord, but may improve neurologic outcomes. Changes in IOM, most importantly loss of MEPs, might infuence a surgeon to leave behind residual tumor in favor of avoiding potential iatrogenic injury during resection [[52,](#page-14-2) [53\]](#page-14-3).

Role of Radiation/Radiosurgery

Stereotactic body radiation therapy (SBRT) also known as stereotactic radiosurgery (SRS) has been described as initial therapy, adjuvant therapy, and salvage therapy for spinal meningioma [\[54](#page-14-4)]. In the case of meningioma, upfront radiosurgery is usually not the preferred treatment, given that small, non-compressive lesions are typically observed and larger symptomatic lesions should be resected. SRS can be considered as upfront treatment in patients who have major medical contraindications to open surgery. In addition, ideally, a 2–3 mm margin between tumor and spinal cord is required for an effective tumoricidal dose of radiation to the tumor [[55\]](#page-14-5). Thus, SBRT is well suited to patients who have already undergone surgical resection and who are more likely to have a safe margin. Defnitive SBRT dose is considered 21 Gy delivered in 3 fractions [[56\]](#page-14-6). Local 5-year control rates with SBRT for intradural tumors, including meningiomas, are reported from 70% to 100% [[56\]](#page-14-6).

Adjuvant SBRT or radiation therapy is considered when a signifcant amount of tumor is knowingly left at the time of surgery (i.e., Simpson grade III or higher). Adjuvant radiation is also considered in cases of WHO grade II or III meningioma, given the more aggressive clinical behavior and rates of recurrence [\[57](#page-14-7)]. As mentioned previously, SBRT is also used for salvage treatment in the case of recurrent tumor on serial imaging (Fig. [8.2](#page-10-0)).

Image-modulated radiation therapy (IMRT) and conventional fractionated radiation therapy have also been used in the treatment of spinal meningioma, but SBRT is the current preferred modality [\[58](#page-14-8)].

Chemotherapy

Chemotherapy is not commonly employed for treatment of spinal meningioma. In cases of invasive, atypical meningioma (WHO III), multiple agents have been employed including hydroxyurea, interferon α -2B, long-acting Sandostatin, and even multidrug sarcoma protocols [\[59](#page-14-9)]. Chemotherapy is used for salvage therapy in cases of highly aggressive tumors.

Fig. 8.2 A 58-year-old male presenting with neck pain, worst at night, without clinical myelopathy (biologic pain). (**a**) T2-weighted MRI demonstrating a large isointense intradural extramedullary mass at C6–C7. (**b**) Axial T2 demonstrates 90% canal compromise with high-grade cord compression. Patient underwent bilateral C6 to T1 midline laminectomy and intradural tumor resection with Simpson grade II resection (ventral dural base coagulated). Final pathology was WHO grade I meningioma. (c, d) Postoperative T1 post-contrast MRI demonstrates small known residual ventral dural attachment tumor. (**e**) Follow-up T1 post-contrast MRI at 1 year demonstrates recurrent/progressive tumor. Patient was treated with conventional fractionated radiation (54 Gy in 30 fractions). (**f**) Follow-up T1 post-contrast MRI 3 years after radiation shows durable local control with decrease in previously seen tumor volume. The patient remained neurologically intact at 5-year follow up

Part III: Outcomes for Spinal Meningioma

Surgical Outcomes

Surgical outcomes in patients undergoing resection of spinal meningioma are favorable, especially with Simpson grade I or II resection. Nakamura et al. reported a consecutive series of 43 patients who underwent Simpson grade I resection of spinal meningioma with 12.1-year follow-up, at which time 0 patients had tumor recurrence [[20\]](#page-12-9). Of 19 patients who had grade II resection, 6 patients (30%) had recurrence at 12-year follow-up. All patients with grade II resection had a ventral tumor location at initial surgery. Overall tumor recurrence at 12 years combining both Simpson grade I and II resection patients was 9.7% In a review of the literature combining 581 cases, Gottfried et al. reported postoperative clinical improvement of McCormick grade in 53% to 95% of patients following surgery for spinal meningioma [\[2](#page-11-1)]. Perioperative mortality was low $(0-3\%)$ as were rates of CSF leakage $(0-4\%)$.

Recurrence

As discussed earlier, rates of recurrence are low with Simpson grade I resection, but are more signifcant with Simpson grade II resection and higher (i.e., residual tumor at time of surgery). A relatively high percentage (45%) of recurrent spinal meningioma are not fully resectable, due either to tumor invasion or to scar tissue at the site of previous resection, favoring treatment with radiation therapy [\[7](#page-11-6)]. In patients without direct compressive symptoms from recurrence, we advocate for SBRT as the salvage treatment of choice in the setting of recurrent spinal meningioma. SBRT has durable high rates of local control in the setting of spinal meningioma [[54\]](#page-14-4).

Conclusion

Spinal meningiomas frequently present in elderly patients, in whom surgical intervention can have broader implications. Observation should be utilized when appropriate. Surgical treatment should be defnitive, with a goal of gross total resection. In cases where full resection cannot be safely achieved, stereotactic radiosurgery can offer high rates of local control.

References

- 1. Schellinger KA, Propp JM, Villano JL, McCarthy BJ. Descriptive epidemiology of primary spinal cord tumors. J Neuro-Oncol. 2008;87(2):173–9.
- 2. Gottfried ON, Gluf W, Quinones-Hinojosa A, Kan P, Schmidt MH. Spinal meningiomas: surgical management and outcome. Neurosurg Focus. 2003;14(6):e2.
- 3. Preston-Martin S. Descriptive epidemiology of primary tumors of the spinal cord and spinal meninges in Los Angeles County, 1972–1985. Neuroepidemiology. 1990;9:106–11.
- 4. Maiti TK, Bir SC, Patra DP, Kalakoti P, Guthikonda B, Nanda A. Spinal meningiomas: clinicoradiological factors predicting recurrence and functional outcome. Neurosurg Focus. 2016;41(2):E6. [https://doi.org/10.3171/2016.5.FOCUS16163.](https://doi.org/10.3171/2016.5.FOCUS16163)
- 5. Kok JL, Teepen JC, van Leeuwen FE, Tissing WJE, Neggers SJ, van der Pal HJ; DCOG-LATER Study Group, et al. Risk of benign meningioma after childhood cancer in the DCOG-LATER cohort: contributions of radiation dose, exposed cranial volume, and age. Neuro Oncol. 2018. <https://doi.org/10.1093/neuonc/noy124>*.*
- 6. Campian J, Gutmann DH. CNS tumors in Neurofbromatosis. J Clin Oncol. 2017;35(21):2378–85. [https://doi.org/10.1200/JCO.2016.71.7199.](https://doi.org/10.1200/JCO.2016.71.7199)
- 7. Solero CL, Fornari M, Giombini S, Lasio G, Oliveri G, Cimino C, Pluchino F. Spinal meningiomas: review of 174 operated cases. Neurosurgery. 1989;25(2):153–60.
- 8. Roux FX, Nataf F, Pinaudeau M, Borne G, Devaux B, Meder JF. Intraspinal meningiomas: review of 54 cases with discussion of poor prognosis factors and modern therapeutic management. Surg Neurol. 1996;46(5):458–63; discussion 463–4.
- 9. Mawrin C, Perry A. Pathological classifcation and molecular genetics of meningiomas. J Neuro-Oncol. 2010;99:379–91.
- 10. Ng THK, Chan KH, Mann KS, Fung CF. Spinal meningioma arising from a lumbar nerve root. J Neurosurg. 1989;70:646–8.
- 11. Ruttledge MH, Sarrazin J, Rangaratnam S, Phelan CM, Twist E, Merel P, et al. Evidence for the complete inactivation of the NF2 gene in the majority of sporadic meningiomas. Nat Genet. 1994;6(2):180–4.
- 12. Barski D, Wolter M, Reifenberger G, Riemenschneider MJ. Hypermethylation and transcriptional downregulation of the TIMP3 gene is associated with allelic loss on 22q12.3 and malignancy in meningiomas. Brain Pathol. 2010;20:623–31.
- 13. Peyrard M, Fransson I, Xie Y-G, Han F-Y, Ruttledge MH, Swahn S, et al. Characterization of a new member of the human /−adaptin gene family from chromosome 22q12, a candidate meningioma gene. Human Mol Genet. 1994;3:1393–9.
- 14. Nunes F, Shen Y, Niida Y, Beauchamp R, Stemmer-Rachamimov AO, Ramesh V, et al. Inactivation patterns of NF2 and DAL-1/4.1B (EPB41L3) in sporadic meningioma. Cancer Genet Cytogenet. 2005;162(2):135–9.
- 15. Surace EI, Lusis E, Murakami Y, Scheithauer BW, Perry A, Gutmann DH. Loss of tumor suppressor in lung cancer-1 (TSLC1) expression in meningioma correlates with increased malignancy grade and reduced patient survival. J Neuropathol Exp Neurol. 2004;63(10):1015–27.
- 16. Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, et al. The 2016 World Health Organization classifcation of tumors of the central nervous system: a summary. Acta Neuropathol. 2016;131(6):803–20. [https://doi.org/10.1007/](https://doi.org/10.1007/s00401-016-1545-1) [s00401-016-1545-1.](https://doi.org/10.1007/s00401-016-1545-1)
- 17. Hirota S, Nakajima Y, Yoshimine T, Kohri K, Nomura S, Taneda M, et al. Expression of bonerelated protein messenger RNA in human meningiomas: possible involvement of osteopontin in development of psammoma bodies. J Neuropathol Exp Neurol. 1995;54(5):698–703.
- 18. Colen CB, Rayes M, McClendon J Jr, Rabah R, Ham SD. Pediatric spinal clear cell meningioma. Case report. J Neurosurg Pediatr. 2009;3:57–60.
- 19. Harter PN, Braun Y, Plate KH. Classifcation of meningiomas—advances and controversies. Chin Clin Oncol. 2017;6(Suppl 1):S2. [https://doi.org/10.21037/cco.2017.05.02.](https://doi.org/10.21037/cco.2017.05.02)
- 20. Nakamura M, Tsuji O, Fujiyoshi K, Hosogane N, Watanabe K, Tsuji T, et al. Long-term surgical outcomes of spinal meningiomas. Spine (Phila Pa 1976). 2012;37(10):E617–23. [https://](https://doi.org/10.1097/BRS.0b013e31824167f1) doi.org/10.1097/BRS.0b013e31824167f1.
- 21. Setzer M, Vatter H, Marquardt G, Seifert V, Vrionis FD. Management of spinal meningiomas: surgical results and a review of the literature. Neurosurg Focus. 2007;23(4):E14.
- 22. Tsuda K, Akutsu H, Yamamoto T, Nakai K, Ishikawa E, Matsumura A. Is Simpson grade I removal necessary in all cases of spinal meningioma? Assessment of postoperative recurrence during long-term follow-up. Neurol Med Chir (Tokyo). 2014;54(11):907–13.
- 23. Abul-Kasim K, Thurnher MM, McKeever P, Sundgren PC. Intradural spinal tumors: current classifcation and MRI features. Neuroradiology. 2008;50:301–14.
- 24. Sevick RJ. Diagnostic neuroradiology. 1994. First Edition. By Anne G. Osborn. Published by Mosby-Year Book, Inc. 936 pages. Can J Neurol Sci. 1995;22:78.
- 25. Doita M, Harada T, Nishida K, Marui T, Kurosaka M, Yoshiya S. Recurrent calcifed spinal meningioma detected by plain radiograph. Spine (Phila Pa 1976). 2001;26(11):E249–52.
- 26. Zhu Q, Qian M, Xiao J, Wu Z, Wang Y, Zhang J. Myelopathy due to calcifed meningiomas of the thoracic spine: minimum 3-year follow-up after surgical treatment. J Neurosurg Spine. 2013;18(5):436–42. [https://doi.org/10.3171/2013.2.SPINE12609.](https://doi.org/10.3171/2013.2.SPINE12609)
- 27. Takashima H, Takebayashi T, Yoshimoto M, Onodera M, Terashima Y, Iesato N, Tanimoto K, et al. Differentiating spinal intradural-extramedullary schwannoma from meningioma using MRI T weighted images. Br J Radiol. 2018;91(1092):20180262. [https://doi.org/10.1259/](https://doi.org/10.1259/bjr.20180262) [bjr.20180262.](https://doi.org/10.1259/bjr.20180262)
- 28. Bettaswamy G, Ambesh P, Das KK, Sahu R, Srivastava A, Mehrotra A, et al. Extradural spinal meningioma: revisiting a rare entity. J Craniovertebr Junction Spine. 2016;7(1):65–8. [https://](https://doi.org/10.4103/0974-8237.176630) doi.org/10.4103/0974-8237.176630.
- 29. Sahni D, Harrop JS, Kalfas IH, Vaccaro AR, Weingarten D. Exophytic intramedullary meningioma of the cervical spinal cord. J Clin Neurosci. 2008;15:1176–9.
- 30. Lee JW, Lee IS, Choi KU, Lee YH, Yi JH, Song JW, et al. CT and MRI fndings of calcifed spinal meningiomas: correlation with pathological fndings. Skelet Radiol. 2010;39(4):345–52. [https://doi.org/10.1007/s00256-009-0771-1.](https://doi.org/10.1007/s00256-009-0771-1)
- 31. Shi HB, Suh DC, Lee HK, Lim SM, Kim DH, Choi CG, et al. Preoperative transarterial embolization of spinal tumor: embolization techniques and results. AJNR Am J Neuroradiol. 1999;20(10):2009–15.
- 32. Charles YP, Barbe B, Beaujeux R, Boujan F, Steib J-P. Relevance of the anatomical location of the Adamkiewicz artery in spine surgery. Surg Radiol Anat. 2011;33:3–9.
- 33. Simpson D. The recurrence of intracranial meningiomas after surgical treatment. J Neurol Neurosurg Psychiatry. 1957;20:22–39.
- 34. Nanda A, Bir SC, Maiti TK, Konar SK, Missios S, Guthikonda B. Relevance of Simpson grading system and recurrence-free survival after surgery for World Health Organization Grade I meningioma. J Neurosurg. 2017;126(1):201–11. <https://doi.org/10.3171/2016.1.JNS151842>.
- 35. McCormick PC, Torres R, Post KD, Stein BM. Intramedullary ependymoma of the spinal cord. J Neurosurg. 1990;72:523–32.
- 36. Nakamura M, Roser F, Michel J, Jacobs C, Samii M. The natural history of incidental meningiomas. Neurosurgery. 2003;53:62–71.
- 37. Liu WC, Choi G, Lee SH, Han H, Lee JY, Jeon YH, et al. Radiological fndings of spinal schwannomas and meningiomas: focus on discrimination of two disease entities. Eur Radiol. 2009;19(11):2707–15. <https://doi.org/10.1007/s00330-009-1466-7>.
- 38. Ozawa H, Onoda Y, Aizawa T, Nakamura T, Koakutsu T, Itoi E. Natural history of intraduralextramedullary spinal cord tumors. Acta Neurol Belg. 2012;112(3):265–70. [https://doi.](https://doi.org/10.1007/s13760-012-0048-7) [org/10.1007/s13760-012-0048-7](https://doi.org/10.1007/s13760-012-0048-7).
- 39. Koeller KK, Shih RY. Intradural extramedullary spinal neoplasms: radiologic-pathologic correlation. Radiographics. 2019;39:468–90.
- 40. Landi A, Dugoni DE, Marotta N, Mancarella C, Delfni R. Spinal schwannomatosis in the absence of neurofbromatosis: a very rare condition. Int J Surg Case Rep. 2011;2:36–9.
- 41. Salpietro FM, Alafaci C, Lucerna S, Lacopino DG, Tomasello F. Do spinal meningiomas penetrate the pial layer? Correlation between magnetic resonance imaging and microsurgical fndings and intracranial tumor interfaces. Neurosurgery. 1997;41:254–8.
- 42. Chwistek M. Recent advances in understanding and managing cancer pain. F1000Res. 2017;6:945.
- 43. Kraetzig T, McLaughlin L, Bilsky MH, Laufer I. Metastases of spinal myxopapillary ependymoma: unique characteristics and clinical management. J Neurosurg Spine. 2018;28(2):201–8. <https://doi.org/10.3171/2017.5.SPINE161164>.
- 44. Payer M. The anterior approach to anterior cervical meningiomas: review illustrated by a case. Acta Neurochir. 2005;147:555–60; discussion 560.
- 45. Iacoangeli M, Gladi M, Di Rienzo A, Dobran M, Alvaro L, Nocchi N, et al. Minimally invasive surgery for benign intradural extramedullary spinal meningiomas: experience of a single institution in a cohort of elderly patients and review of the literature. Clin Interv Aging. 2012;7:557–64.<https://doi.org/10.2147/CIA.S38923>.
- 46. Ruggeri AG, Fazzolari B, Colistra D, Cappelletti M, Marotta N, Delfni R. Calcifed spinal meningiomas. World Neurosurg. 2017;102:406–12. [https://doi.org/10.1016/j.](https://doi.org/10.1016/j.wneu.2017.03.045) [wneu.2017.03.045](https://doi.org/10.1016/j.wneu.2017.03.045).
- 47. Ahmed R, Menezes AH, Awe OO, Mahaney KB, Torner JC, Weinstein SL. Long-term incidence and risk factors for development of spinal deformity following resection of pediatric intramedullary spinal cord tumors. J Neurosurg Pediatr. 2014;13(6):613–21. [https://doi.org/1](https://doi.org/10.3171/2014.1.PEDS13317) [0.3171/2014.1.PEDS13317.](https://doi.org/10.3171/2014.1.PEDS13317)
- 48. Albert TJ, Vacarro A. Postlaminectomy kyphosis. Spine (Phila Pa 1976). 1998;23(24):2738–45.
- 49. Yasuoka S, Peterson HA, Laws ER, MacCarty CS. Pathogenesis and prophylaxis of postlaminectomy deformity of the spine after multiple level laminectomy. Neurosurgery. 1981;9:145–52.<https://doi.org/10.1227/00006123-198108000-00006>.
- 50. Dekker SE, Ostergard TA, Glenn CA, Cox E, Bambakidis NC. Posterior cervical laminoplasty for resection Intradural extramedullary spinal meningioma: 2-dimensional operative video. Oper Neurosurg (Hagerstown). 2019;16(3):392.<https://doi.org/10.1093/ons/opy204>.
- 51. Shi W, Wang S, Zhang H, Wang G, Guo Y, Sun Z, et al. Risk factor analysis of progressive spinal deformity after resection of intramedullary spinal cord tumors in patients who underwent laminoplasty: a report of 105 consecutive cases. J Neurosurg Spine. 2019;8:1–9. [https://doi.](https://doi.org/10.3171/2018.10.SPINE18110) [org/10.3171/2018.10.SPINE18110.](https://doi.org/10.3171/2018.10.SPINE18110)
- 52. Ghadirpour R, Nasi D, Iaccarino C, Giraldi D, Sabadini R, Motti L, et al. Intraoperative neurophysiological monitoring for intradural extramedullary tumors: why not? Clin Neurol Neurosurg. 2015;130:140–9. [https://doi.org/10.1016/j.clineuro.2015.01.007.](https://doi.org/10.1016/j.clineuro.2015.01.007)
- 53. Ghadirpour R, Nasi D, Iaccarino C, Romano A, Motti L, Sabadini R, et al. Intraoperative neurophysiological monitoring for intradural extramedullary spinal tumors: predictive value and relevance of D-wave amplitude on surgical outcome during a 10-year experience. J Neurosurg Spine. 2018;30(2):259–67. <https://doi.org/10.3171/2018.7.SPINE18278>.
- 54. Kufeld M, Wowra B, Muacevic A, Zausinger S, Tonn JC. Radiosurgery of spinal meningiomas and schwannomas. Technol Cancer Res Treat. 2012;11(1):27–34.
- 55. Tseng CL, Eppinga W, Charest-Morin R, Soliman H, Myrehaug S, Maralani PJ, et al. Spine stereotactic body radiotherapy: indications, outcomes, and points of caution. Global Spine J. 2017;7(2):179–97. [https://doi.org/10.1177/2192568217694016.](https://doi.org/10.1177/2192568217694016)
- 56. Kalash R, Glaser SM, Flickinger JC, Burton S, Heron DE, Gerszten PC, et al. Stereotactic body radiation therapy for benign spine tumors: is dose de-escalation appropriate? J Neurosurg Spine. 2018;29(2):220–5. <https://doi.org/10.3171/2017.12.SPINE17920>.
- 57. Noh SH, Kim KH, Shin DA, Park JY, Yi S, Kuh SU, et al. Treatment outcomes of 17 patients with atypical spinal meningioma, including 4 with metastases: a retrospective observational study. Spine J. 2019;19(2):276–84. <https://doi.org/10.1016/j.spinee.2018.06.006>.
- 58. Gerszten PC, Chen S, Quader M, Xu Y, Novotny J Jr, Flickinger JC. Radiosurgery for benign tumors of the spine using the Synergy S with cone-beam computed tomography image guidance. J Neurosurg. 2012;117(Suppl):197–202. [https://doi.org/10.3171/2012.8.GKS12981.](https://doi.org/10.3171/2012.8.GKS12981)
- 59. Moazzam AA, Wagle N, Zada G. Recent developments in chemotherapy for meningiomas: a review. Neurosurg Focus. 2013;35(6):E18. [https://doi.org/10.3171/2013.10.FOCUS13341.](https://doi.org/10.3171/2013.10.FOCUS13341)