Intramedullary Spinal Cord Tumors: A Review and Discussion of Surgical Rationale

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ABSTRACT

Intramedullary spinal cord tumors (IMSCTs) are rare neoplasms of the central nervous system associated with severe neurological deterioration, morbidity, poor quality of life, and even death. The most common IMSCTs that we encounter are ependymomas, astrocytomas, and hemangioblastomas. Surgery remains the initial treatment of choice for IMSCTs. New and interesting therapies are currently under investigation and the extent to which they augment our current treatment paradigms remains to be seen but with the advent of novel therapies (nanomedicine, localizable therapies, and cell targeting) the future appears promising for lesions not amenable to gross total resection and a surgical cure. While complete resection is not always achievable, we must remain tempered by the paramount importance of our patient's neurological function, for it is their well-being and quality of life outcome that is the root purpose of all of the aforementioned endeavors.

KEY WORDS: Intramedullary tumor, surgical strategies, spinal cord

INTRODUCTION

Anton von Eiselsberg in Vienna, 1907, performed the first successful resection of an intramedullary spinal cord tumor (126).This was followed in 1911 by the beginning developments of what will be our current surgical concepts for intramedullary tumors by Elsberg and Beer. They envisioned a two-stage procedure for the treatment of these lesions. The first stage involved a surgical myelotomy with an unclosed durotomy, followed a week later by the second stage in which, due to the unclosed durotomy, the tumor would, by this point, be partially extruded from the spinal cord, thus facilitating resection (26). However, after their initial conceptions of surgical treatment of intramedullary tumors, there was a change in philosophy among the neurosurgical community, where a more conservative approach including decompression with laminectomy and/or duroplasty, was championed. This shift in methodology was in large part due to the morbidity of the surgical procedures and the advent of radiation therapy (131).

During the 1950s, as if to anticipate future directions, Greenwood began to develop instruments and concepts for what would later be termed "microsurgical" resection of these tumors, albeit largely abandoned during the conservative era (44-46). As microsurgery became mainstream, a resurgence of intramedullary tumor resection spread throughout the neurosurgery community in the 1980s, accompanied by both a lower morbidity associated with the surgical technique and the ability to achieve a more complete resection (27,36,48,87,118). As we move into the current era, with

new imaging technology, intraoperative monitoring, and perioperative care, surgery has again become the first step in the treatment of this disease.

In the following review, we discuss the most common intramedullary spinal cord tumors, providing diagnostic and therapeutic options for each type, followed by general surgical considerations

Intramedullary Spinal Cord Tumors

Intramedullary spinal cord tumors (IMSCTs) (Table 1) are rare neoplasms of the central nervous system (18) associated with severe neurological deterioration, morbidity, poor quality of life, and even death. These tumors represent only 2-4% of all primary tumors of the central nervous system with an incidence of 850 to 1700 cases per year in the United States. The most common IMSCTs that we encounter are ependymomas, astrocytomas, and hemangioblastomas (55).

Due to the eloquent nature of the surrounding nervous tissue, there is a significant risk of severe neurological injury to patients. Despite significant advances in the diagnosis and surgical management of IMSCTs, these tumors continue to present a major challenge for neurosurgeons both technically and conceptually (57,61,64,90,135). To this end, functional outcome should be of the utmost importance, and care must be taken to not only maximize surgical resection but to ensure maximum postoperative functional benefit.

The most common presenting symptom in patients with IMSCTs is pain. The pain may be localized to the back, either radicular or generalized, and the quality may change, often worsening in the evening. Additionally, they may be associated with sensory and motor disturbances, including bowel and bladder dysfunction (106). In young patients, IMSCTs may remain asymptomatic for a prolonged period of time before diagnosis, often manifesting as non-specific complaints such as clumsiness or even scoliosis (62). The non-specific symptoms require clinical suspicion to remain high and prompt proper imaging to make the diagnosis.

Current treatment paradigms for IMSCTs (Table 2) involve initial attempts at resection, followed by adjuvant therapies that are reserved for high grade histology, tumors not amenable to resection, tumor recurrence, and when surgery is otherwise contraindicated (124). There are three reasons surgery is indicated for these tumors: to prevent long term neurological demise, obtain a histological diagnosis, and provide specific oncological treatment for the tumor type harbored by the patient (75). With the advancement of surgical technique and equipment, more aggressive surgery is possible. However, resection remains dictated by the presence of a clear plane of dissection, as seen in ependymomas, or the lack there of, as seen in infiltrative tumors like astrocytomas (30), which limits gross total resectability, leading to higher rates of long term neurological disability with no added benefit for more aggressive approaches (10).

The most important characteristic in determining long-term neurological/functional outcome after surgery is the patient's preoperative neurological status (50,69). This is often evaluated using a standardized and validated rating system. The McCormick scale, which focuses on the neurological function and ambulatory status of the patient, is currently the most widely used evaluation system (19,20,90).

The second most important characteristic is tumor histology and grade, with the former also predictive of extent of resection, functional outcome and recurrence (69). Resection itself is generally considered a predictor of good outcome, albeit directly correlated with histology, one begetting the other. Conversely, poor outcome is often associated with the presence of syringomyelia or tumors harboring a cystic component (91).

Radiotherapy is often utilized in the treatment of high-grade tumors that prove not amenable to GTR and for lesions in which resection is contraindicated in adults and older children. This treatment, however, is not without risks. Radiation myopathy, spinal deformities, impaired growth, vasculopathy, radiation necrosis and secondary tumor formation have all been reported (11,18). Additionally, reports of efficacy have been conflicting (112).

Chemotherapy has been historically reserved for cases in which resection and radiotherapy were unsuccessful or contraindicated. In the pediatric population, however, chemotherapy plays a more prominent role (18) due to the sensitivity of this population to the effects of radiation and IMSCTs. To circumvent some of the difficulties associated with this treatment (systemic effects of the medication, dynamic process of CSF flow and pulsation, and traversing the blood-spinal cord barrier), new therapeutic strategies are being evaluated to improve drug localization and mitigate the aforementioned limitations (124). Unlike the pediatric population, there is no standardized pre- or

Table 1: Intramedullary Spinal Cord Tumors

Tumor	Incidence	Location	Prognosis
Ependymoma	Most common (50 to 60% of IMSCTs)	Cervical > thoracic > lumbar	Good
Myxopapillary ependymoma	Rare	Filum terminale & conus medullaris	Excellent
Astrocytoma	2nd most common overall, most common in children	Cervical > thoracic > lumbar	Poor
Hemangioblastoma	3rd most common but still rare; increased incidence in VHL	Cervical > thoracic > lumbar	Excellent
Ganglioglioma	Rare	Cervical > thoracic > lumbar	Good
Germ cell tumor	Very rare	Cervical > thoracic > lumbar	Good
CNS lymphoma	Rare	Cervical > thoracic > lumbar	Poor
Melanoma	Very rare (CNS only, no cutaneous melanoma)	Cervical > thoracic > lumbar	Poor (but better than cutaneous melanoma involving CNS)

Abbreviations: VHL – Von Hippel-Lindau disease.

Table 2: Current Evidence-Based Recommendations for Treatment of IMSCTs

Tumor	Treatment	Evidence-Based Classification*
Ependymoma	Primary: Surgical resection Secondary: RT Secondary: CT	Class I, Level of Evidence: C Class IIa, Level of Evidence: C Class IIb, Level of Evidence: C
Myxopapillary ependymoma	Surgical resection	Class I, Level of Evidence: C
Astrocytoma	Primary: Surgical resection Secondary: RT Secondary: CT	Class IIb, Level of Evidence: C Class IIa, Level of Evidence: C Class IIb, Level of Evidence: C
Hemangioblastoma	Surgical resection	Class I, Level of Evidence: C
Ganglioglioma	Surgical resection	Class I, Level of Evidence: C
Germ cell tumor	Primary: Surgical resection Secondary: RT (germinomas) Secondary: CT (non-germinomatous GCT)	Class I, Level of Evidence: C Class IIa, Level of Evidence: C Class IIa, Level of Evidence: C
CNS lymphoma	Intrathecal CT	Class IIb, Level of Evidence: C
Melanoma	Primary: Surgical resection Secondary: RT	Class I, Level of Evidence: C Class IIb, Level of Evidence: C

Abbreviations: CT – chemotherapy; **RT –** radiotherapy.

*The American Heart Association Evidence-Based Scoring System.

postoperative treatment regimen that has shown to improve survival. Agents used for intracranial high-grade glial tumor, such as bevacizumab (AVASTIN ®, Genentech, Inc.), have been applied to the spine but these are protocols that vary by institution. To date, there are no specific molecular targets for pirmary spinal tumors. Despite these new, novel therapies on the horizon, surgery remains the initial treatment of choice for IMSCTs with the exception of CNS lymphoma (Table 3).

Ependymoma

The ependymoma is the most common form of IMSCT. About 80-90% of IMSCTs are gliomas, of which approximately 70% are ependymomas and 30% are astrocytomas (102). 44% of ependymomas arise in the cervical cord and 23% in the upper thoracic cord, classically presenting as a male with chronic back pain (2). Ependymomas are typically slow growing and histologically exhibit a benign pathology (WHO Grade 1) for up to 50% of cases, but also have anaplastic variants that grow rapidly and exhibit aggressive pathology (WHO Grade III) (108,124).

Ependymomas classically show an easily identified plane of dissection in the majority of cases, which makes GTR the primary treatment option (124). Grossly, the ependymoma presents as a modestly vascular soft, encapsulated, reddish gray or yellow mass (110). Surgically, one method described to remove an ependymoma with a clear margin of demarcation from the spinal cord tissue involves using gentle blunt dissection, followed by tumor traction and spinal cord counter traction to remove the tumor, while incorporating cautery to manage fibrous adhesions and feeding vessels (88). Ependymomas tend to be hyperechogenic compared to normal tissue which allows them to be readily differentiated from spinal cord when using intraoperative ultrasound (62).

The well-demarcated ependymoma, when totally resected, has been shown to improve symptoms in over 90% of patients following surgery (48). Local control rates of 90-100% have been demonstrated following a gross total resection, but often complete resection is not possible due to considerations for patient side-effects that may result from a total resection surgery (88,90). There have been conflicting reviews on the benefits of GTR versus subtotal resection in regards to myxopapillary ependymomas (WHO Grade I), with some studies citing an improved survival rate with GTR and others displaying no benefit in outcomes; however the prognosis for adults has been shown to be excellent with high rates of survival (5,78). When generally regarding

Table 3: Authors' Proposed Management Strategy for IMSCTs

Tumor	Treatment
Ependymoma	- Attempt GTR based on plane of dissection and intraoperative neuromonitoring - If STR performed, add adjuvant radiation therapy
Myxopapillary ependymoma	- Attempt GTR based on plane of dissection and intraoperative neuromonitoring
Astrocytoma	- Assess plane of dissection, perform STR (biopsy) - If there is significant residual tumor or recurrence, add radiation therapy
Hemangioblastoma	- Preoperative spinal cord angiography for possible embolization of vascular feeders - GTR with a good plane of dissection
Ganglioglioma	- GTR - If there is recurrence, consider radiation therapy
Germ cell tumor	- Assess CSF for germ cell markers (hCG, AFP, PLAP) - Radiation therapy
CNS lymphoma	- High dose methotrexate and temozolomide - Diagnosis made with biopsy
Melanoma	- GTR - Postoperative radiation therapy

Abbreviations: AFP – alpha-fetoprotein; **CSF** – cerebrospinal fluid; **GTR** – gross-total resection; **hCG** – human chorionic gonadotropin; **PLAP** – placental alkaline phosphatase; **STR** – subtotal resection.

recurrence rates, studies have shown that GTR has been associated with a lower overall rate of tumor recurrence, prompting that GTR should continue to be the mainstay of treatment whenever possible and appropriate (34,117). When approaching myxopapillary ependymomas in children, many recent papers suggest similar overall clinical outcomes and long-term (>10 year) survival following surgical resection (12,13,85). A recent paper noted that the most common reasons for a subtotal resection were nerve root involvement and an absence of a clear surgical plane between the tumor and the spinal cord (12).

Tumors are classically evaluated and resected following patient complaints of objective and non-objective (typically pain) neurologic deficits caused by tumor growth. One study of 82 ependymoma resections for patients who did not have objective neurologic complaints has shown that preoperative complaints resolved in 30% of cases, stabilized in 60%, and worsened in 10% of cases, prompting the conclusion that surgical resection should be considered in patients with ependymomas who have not yet started displaying neurologic deficits (3).

The functional status of neurologic deficits is often graded according to a McCormick scale (20,90) (Table 4). In a follow-up study following radical excision of 164 intramedullary spinal cord tumors, clinical status and quality of life post-operatively were evaluated in 116 living patients with a mean follow-up time of 157.3 months. In the study, >60% of the patients were functioning at a Grade I or II level while 23% functioned at a Grade IV or V level (20). In a study of 68 ependymoma removals, 55 (81%) successfully underwent GTR; at long-term follow-up of the procedure, 25% (16 patients) showed improved presenting signs/symptoms, 66% (42 patients) remained unchanged, and 9% (6 patients) worsened [59]. One recent retrospective analysis of 24 intramedullary ependymoma GTRs found a mean McCormick score of 1.8 +/- 0.7 pre-surgery followed up with a mean McCormick score of 1.7 +/- 0.8 with no cases of recurrence 6 months following surgery (122). As this study does not show outcomes to be significantly improved, it is important to balance the neurological side-effects and risks of surgical intervention with the neurological side-effects and risks of not pursuing surgical intervention and allowing continued growth of the IMSCT. Surgical intervention with a goal of GTR to treat the intramedullary ependymoma has been correlated with improved patient outcomes as well as a decreased rate of relapse. One study finds that time-to-progression of a relapse ependymoma following surgery significantly correlates with extent of resection; overall this study noted overall 46% of patients to be progression-free 15 years post-surgery (of which 89% of pts were STR/Biopsy-only, and only 11% GTR) (42).

As with any spinal surgery, surgical outcomes following resection should be given consideration. One study of GTR of ependymomas found an overall complication rate post-GTR of 34% with the primary complications being wound infections or cerebrospinal fluid leaks (78). A study on resection of ependymomas in the pediatric population found primary complications following resection to be cerebrospinal fluid leaks, neurologic deficits, and risk of spinal column deformity (59).

Astrocytoma

Astrocytomas comprise 30-40% of intramedullary glioma (18). In children, however, the most common IMSCT is consistently found to be the astrocytoma (>50%), as ependymomas are relatively uncommon (18,102).

Table 4: McCormick Scale

Grade	Definition
1	Neurologically normal, mild focal deficits not limb function, mild spasticity/reflex abnormality, ambulates normally, normal gait
2	Mild sensorimotor deficit present affecting function of limb, functions and ambulates independently, mild gait difficulty
3	Moderate sensorimotor deficit affecting function of limb, ambulates independently w/aid, moderate gait difficulty
4	More severe sensorimotor deficit, may not function independently, requires assistance for ambulation
5	Severe deficit, paraplegia/quadriplegia

Astrocytomas are further classified as WHO Grade 1 pilocytic astrocytoma or WHO Grades 2,3,4 infiltrative astrocytomas (84). Although limited by sample size, one study of IMSCTs from December 1972 to June 2003 found astrocytoma lesions to be 31% WHO Grade 1, 48% WHO Grade 2, and 21% WHO Grade 3&4 (106). However, regardless of WHO Grade, astrocytomas have been noted to be infiltrative with poorly characterized boundaries (18). Similar to ependymomas, the primary treatment method for intramedullary astrocytomas is surgical, with a goal of GTR or STR as appropriate. Guidelines and benefits for use of chemotherapy and radiation therapy have not yet been clearly-defined, but these are often used as adjuncts to surgical therapy. Patient outcomes of GTR/STR correlate with tumor histology, as low-grade pilocytic astrocytomas are much more likely to be associated with defined tumor margins than are higher-grade astrocytomas (4).

Similar to ependymomas, the earliest symptom of the astrocytoma is often pain (124). Typical symptoms in children involve the insidious onset of diffuse/radicular pain that often wakes children from their sleep, motor deficits (clumsiness, falls, weakness), and a scoliosis curve of the spine; higher grade neoplasms typically exhibit a shorter prodrome (median: 4.5 months (62)) The symptoms of IMSCTs are often nonspecific and requires a thorough diagnostic workup prior to diagnosis.

As with all intramedullary tumors, the astrocytoma removal is approached with a laminectomy or osteoplastic laminotomy (4,62). The use of intraoperative ultrasound aids resection by allowing the surgeon to visualize the spinal cord in two dimensions, which is helpful for surgeries in which anatomical landmarks are limited and there is a small margin for error (31). When using intraoperative ultrasound, intramedullary astrocytomas and gangliogliomas have a similar echogenic pattern to the spinal cord, unlike the ependymoma which is readily hyperechogenic (62). A true plane between tumor and normal spinal cord tissue does not exist, and surgical manipulation of spinal cord tissue to search for a plane should not be undertaken as this can potentially be hazardous to patient outcomes (62,124). Astrocytomas are grossly described to have a gray-yellow or red, gray, glossy appearance with a poorly defined plane (62,110). In current practice, attempting a GTR for astrocytomas is not recommended, rather a STR should be attempted to prevent excessive damage of normal spinal cord parenchyma during the procedure (124). One review states that for malignant lesions, conservatively debulking the tumor in an inside-out fashion with an ultrasonic aspirator is recommended to preserve maximum motor function, rather than attempting a full resection (62). As with ependymomas, monitoring of sensory and motor evoked potentials during surgery is recommended.

Following a surgical approach, one study finds patients with pilocytic astrocytomas survived significantly longer than those with infiltrative astrocytomas (overall survival of 39.9 vs. 1.85 years); the study concluded that histologic type is the most important variable affecting prognosis regardless of surgical management type (92). More aggressive resection in this study was conducted on astrocytomas with more aggressive infiltrative histology, which may have played a role in subsequent patient outcomes. Another study of 46 astrocytomas noted that 30.7% of patients worsened compared to their pre-operative baseline symptoms, noting a statistically significant increased occurrence of poor outcomes correlated with a higher tumor grade (10). Another study in the pediatric age group, evaluated 55 pediatric intramedullary spinal cord tumors (29 astrocytoma cases), and found that extent of resection and tumor histology and grade were directly correlated with an improved 5 year "progression free survival" in patients with a GTR (4).

Regarding recurrence rate, a study by Karikari et al. noted that 47.6% of patients with a primary spinal cord astrocytoma experienced a recurrence versus only 7.3% of ependymomas; at long-term follow up of the astrocytoma cases, 4.8% improved, 47.6% remained the same, and 47.6% had worsened. It is important to note that a GTR was achieved in 90.9% of ependymoma cases but only 14.3% of astrocytoma cases; the paper concluded that pre-operative neurological status, tumor histology, and extent of resection were predictive of patient neurological outcomes (69). In one long-term study, GTR was achieved in 81% of Grade I astrocytomas, 12% of Grade II astrocytomas, and 0% of Grade III-IV astrocytomas; of these, in long-term follow-up, the presenting signs/symptoms improved in 26% of the Grade I patients and 10% of Grade II patients (106).

As a general trend, it is shown that astrocytoma resection is most beneficial for the patient with the most benign histologic tumor grade. In higher grade tumors, debulking can help to improve patient symptoms. The classic neurologic deficits exhibited by patients prior to surgery are sensory and motor deficits, which was shown to be improved equally by either GTR or STR in 55% of patients (40). Following

astrocytoma resection in children, frequent post-laminectomy spinal deformities such as scoliosis represent a serious post-operative complication, emphasizing the importance to follow these patients closely to watch for spinal deformities with continued growth (27,107). Neurologic decline has been widely noted as a side-effect following IMSCT resection. Of the sensory and motor deficits seen post-operatively in one study, 63% of motor-related symptoms resolved in a median time of 3 months and 50% of sensory deficits resolved in a median time of 7 months (4).

Hemangioblastoma

Following intramedullary ependymomas and astrocytomas, hemangioblastomas have been noted the third most frequent IMSCT, accounting for 2-8% of IMSCTs (60,91,94,110). Von-hippel Lindau disease (VHL) is strongly associated with hemangioblastoma; 20-30% of spinal cord hemangioblastoma patients also have VHL (18,97). Intramedullary hemangioblastomas are commonly located in the cervical spine (110). Often, hemangioblastomas are found to originate in the dorsal root entry zone near the dorsal roots or the spinal cord (82). Thus, initial symptoms are typically sensory in nature. One study found that the initial symptom seen for patient with hemangioblastomas was a sensory disturbance in 78% of patients, pain in 55% of patients and motor disturbances in 45% of patients (94). Like ependymomas, hemangioblastomas typically have well-demarcated margins that allow for an excellent prognosis with resection (1, 58).

Grossly, hemangioblastomas are richly vascular small tumors with a "sunset orange appearance" that rarely extend beyond 1-2 segments (88,110). Intramedullary hemangioblastomas can be approached with a laminotomy or hemilaminectomy (127). A risk of hemorrhage is present in the untreated hemangioblastoma; one study finds the etiologic odds to be 73% subarachnoid hemorrhage or 27% intramedullary hemorrhage (115). Classically, hemangioblastomas have been considered to pose severe surgical bleeding risks due to their origin (87). There have been many different methods described in the literature to circumvent this possible complication. Spinal angiography (for example digital subtraction angiography) is recommended by many authors to delineate the vessels feeding and draining the tumor (15,87). One paper notes their effective method of removing the tumor (midline dural incision to dorsal cord) in steps: first, coagulate the dominant arterial feeder vessels at the entrance of the tumor by bipolar coagulation with low power, followed by coagulation and shrinkage of the tumor, and lastly to coagulate the venous drainage; after this carefully excise the tumor (15). This method allows shrinking of the tumor and prevention of excessive bleeding during surgery.

Another study states that hemangioblastomas should be resected in a circumferential fashion, not debulked from within like the astrocytoma; the tumor surface may be initially coagulated to allow for manipulation (21,62). Circumferential detachment of normal spinal cord pia from the tumor capsule pia is conducted with gentle blunt traction-counter traction using surgical instruments, while fibrous extensions and feeding vessels are systematically isolated and cauterized (89). Some writers prefer a different method in which they note the demonstrable efficacy of embolization of hemangioblastoma feeding vessels using polyvinyl alcohol particles (with surgical resection 48-72 hours following embolization), which was shown to potentially decrease the morbidity and mortality of surgical resection of craniospinal hemangioblastoma (32).

About 50% of spinal hemangioblastomas have been noted to be associated with a syrinx (15) These hemangioblastomas favor resectability by nature, as the tumor's space-occupying edema and syrinx cause neurological symptoms by displacement rather than infiltration (111). One author concludes that additional opening of the syrinx or surgical removal is not necessary due to the fact that on removal of hemangioblastomas with a syrinx, the syrinx spontaneously stops growing and regresses in size following removal of the hemangioblastoma (94,111,125).

Generally speaking, surgical intervention is the primary course of treatment of an intramedullary hemangioblastoma due to the tumor's well-defined surgical margins. One study of 23 hemangioblastoma resection surgeries found that at the 6 month post-operative follow-up, 18 patients remained neurologically stable (17 in McCormick grade I and 1 in McCormick grade II) and 5 patients recovered to a better status (3 from grade III to II, 2 from grade II to I) (15). One study of 5 intramedullary hemangioblastoma with follow-up periods of 12-86 months found no additional neurological symptoms after surgery, as compared to preoperative status (127). One paper assessing 17 intramedullary vascular tumors (9 cavernomas, 8 hemangioblastomas) found these resections to have the most favorable results as 94.1% (16 of 17 patients) showed a good functional outcome with no or only slight neurological deficits and all were able to walk

independently post-operatively (112). A recent study of intramedullary hemangioblastoma resection concluded that safe surgical treatment should be considered the primary treatment modality; this study achieved GTR in 20 of their 21 patients with 57% of patients experiencing some form of long-term dysfunction after surgery (post-operative outcomes may be an product of the pre-operative state) (81).

Common complications include tumor recurrence (association to VHL disease) and a CSF fistula noted in one study (15). Minimal complications regarding postoperative functional outcomes were noted by one study of 17 vascular tumor removals, as all patients either improved or remained unchanged following the surgical resection (112).

Ganglioglioma

Intramedullary gangliogliomas are rare tumors that are typically benign and slow-growing (WHO Grade I or II), but have been shown to experience malignant transformation (7). One study of 58 gangliogliomas finds 40 classified as histological grade I, 16 grade II, and two grade III (79). They are typically found in the pediatric population and located in the cervical spine in 67% of cases (67,124).

The most common presenting symptoms of the ganglioglioma are paraparesis (limb weakness) and radicular pain, with an average duration of symptoms in adult patients of 7.8 years (63,67,83). One study found that 39% of ganglioglioma patients present with clear scoliosis (134). Similar to other intramedullary spinal cord tumors, the primary treatment of choice is surgical resection. GTR is commonly achieved, with one study noting a rate of 83.3% for spinal cord gangliogliomas (25). Another study notes that early surgical intervention has two main advantages: first, that you can undergo surgery with a better functional status and second, that resection is easier due to the smaller size (83).

Similar to astrocytomas, gangliogliomas have both a gross gray-yellow appearance and are resected in an inside-out fashion using ultrasonic aspiration (62,123). This similarity to astrocytomas is common in gangliogliomas, as one study notes that histologically, 41% of infratentorial gangliogliomas have a high histologic similarity to pilocytic astrocytomas as gangliogliomas contain astrocytes and neural cells (49). Surgical technique used in resection of gangliogliomas is similar to resection of astrocytomas. Using intraoperative ultrasound, intramedullary gangliogliomas have a similar echogenic pattern to the spinal cord (like astrocytomas) (62).

Gross total resection for gangliogliomas is associated with good patient outcomes and survival. One study notes a rate of operative morbidity of 37%, 5-year survival rates of 84%, and event-free survival rate at 5 years to be 36% for spinal cord gangliogliomas, concluding that radical surgery leads to improved long-term survival of patients with spinal gangliogliomas (79). Another study of 56 intramedullary ganglioglioma resections notes a 5-year actuarial survival rate of 88%, a 5-year progression-free survival rate of 67%, and neurological function in surviving to be stable or improved in 72% of all patients (63). Subtotal resection may be an effective treatment method to minimize neurologic dysfunction in cases where attempting GTR has a high risk of neurologic dysfunction due to tumor size and location (134).

Following surgery, special attention should be given to evaluation of spinal instability and subsequent deformity, as this is a cause of increased morbidity for patients with longitudinally extensive tumors (123). At resection, gangliogliomas are typically larger in size than other IMSCTs, a finding that may show a relationship with this finding pre- and post-operatively (124). A case report of 5 gangliogliomas removed by GTR found no evidence of recurrence or regrowth of the tumor in the mean follow-up period of 4.1 years (101).

Germ Cell Tumors

Tumors that arise from cells similar to the germinal cells found in the gonads, termed germ cell tumors (GCTs), are quite uncommon in the CNS, and the rate of primary intramedullary GCT is predictably very small. GCTs are classified pathologically into two subgroups: nongerminomatous GCT and germinomas.

Germinomas are more common, and are usually found in the pineal gland, suprasellar region, or basal ganglia (22,103). When they occur in the spinal cord, it is usually a spinal metastasis from a intracranial primary germinoma (14). Primary intramedullary spinal cord germinomas (PISG) are exceedingly rare, but they appear to affect patients at a relatively young age, as the average age in a recent study with 11 PISG patients was only 27.1 years (range: 14 – 48 years) (133).

On MRI imaging, T1-weighted images typically reveal a contrast enhancing, expanding mass, which can be associated with spinal cord atrophy (consistent with the atrophy associated with intracranial germ cell tumors) (86).

As these tumors often present in the thoracic spine, patients generally present with combined sensory and motor deficits, often in the lower extremities, which can progress to gait dysfunction and urological dysfunction. Given the lack of specificity associated with the imaging signs and clinical symptoms, these tumors are often initially misdiagnosed as ependymomas or astrocytomas. Clinicians can assess the levels of tumor markers such as AFP and Beta–hCG in both the serum and CSF during their patient work-up to try to focus their differential diagnosis. Additionally, the decline of these markers can be followed throughout treatment, although these markers are not very sensitive for tumor response (128,133). Following laminectomy, myelotomy, and surgical resection, PISG are identical to intracranial germ cell tumors, and an immunohistochemical analysis performed on tumor specimens typically demonstrates positivity for placental alkaline phosphatase (PLAP), c-KIT, and OCT4, but not AFP or b–hCG (53,96,133).

During surgical resection, these masses are often noted to have poor tumor-spinal cord interfaces, significant vascularity, and may have strong adhesions to the neural tissue, making exposure and complete resection difficult (133). Fortunately, multiple case studies have shown that these tumors respond favorably to chemotherapy and radiotherapy (8,86). For example, one retrospective study showed that post-operative chemotherapy with carboplatin and etoposide combined with low dose radiotherapy to the local spine resulted in symptom improvement in the vast majority of patients and no disease recurrence or dissemination was observed on imaging surveillance over a 6-year follow-up period (133). This same study also demonstrated that these tumors are so exquisitely radiosensitive that they respond to radiotherapy alone, albeit at higher doses, and similarly favorable clinical outcomes can be achieved. These long-term positive responses to combinatorial therapy advocate against aggressive surgical resection, as total resection is unnecessary for a complete clinical response and only increases the likelihood of a severe operative complication.

Unfortunately, nongerminomatous GTCs are not as radiosensitive as germinomas and thus generally require combinatorial chemoradiotherapy following surgical resection and are associated with a poorer 5-year survival rate (11).

Melanoma

Primary spinal cord melanomas are exceptionally rare, constituting only 1% of all CNS melanomas (33,77). In order to be classified as a primary spinal cord melanoma, there must be no evidence of melanoma outside of the CNS, there can be no tumor foci in other parts of the CNS, and a histological examination must confirm the diagnosis of melanoma (54). These tumors are thought to originate either from melanoblasts, which are derived from neural crest and are found in the primitive CNS during embryogenesis and remain present in the normal leptomeninges that penetrate the spinal cord with vascular bundles, or from neuroectodermal congenital rest cells (99,100). Primary intramedullary melanomas progress slower and have a better prognosis than cutaneous melanomas with CNS involvement (80).

When primary intramedullary melanomas are found, they are most frequently located in the thoracic portion of the cord. Patients with these tumors usually present similarly to those with more common IMSCTs, with symptoms like pain, weakness, and sensory and motor deficits. The paramagnetic melanin pigment in these tumors classically make them hyperintense on T1-weighted MRI imaging with mild homogenous enhancement with contrast, and hypointense on T2 scans (33).

These malignancies require a combinatorial treatment approach utilizing gross resection, radiotherapy, and possibly chemotherapy. Given the rarity of primary intramedullary melanoma, no major clinical trial or case series has evaluated possible treatment paradigms and thus no standardized treatment exists (39). While a radical surgical resection with intraoperative monitoring of somatosensory and motor evoked potentials is recommended, complete resection can be impossible to perform, and these tumors have been observed to recur in the same place they were first found. Multiple case studies have described dramatic quality of life improvements for patients with primary spinal cord melanoma following complete macroscopic tumor resection, highlighting the potential benefit surgery plays in the management of these tumors (38,72,73). However, melanoma recurrence was seen at the site of the original resection in some of these patients.

Intramedullary Spinal Metastases

Intramedullary metastases represent 1-3% of all intramedullary spinal tumors, and arise most frequently from lung, breast, or lymphoma primary malignancies (11). While the majority of tumors are discovered incidentally

at autopsy, if patients with these extremely rare tumors are symptomatic, typically their neurological deficits have an acute presentation that requires rapid examination and intervention. This acute onset of neurological dysfunction helps distinguish intramedullary spinal metastases from primary intramedullary tumors, which typically have a much slower clinical course (47). Imaging findings such as hyperintensity on T2-weighted images, which represents the tumor and surrounding edema, and gadolinium-enhancement of the tumor, are nonspecific and cannot delineate metastases from primary tumors (113,119). Occasionally, patients have been reported to present with symptoms of a Brown-Séquard lesion (9,66).

Currently, steroids and radiation are considered the gold standard therapy for intramedullary spinal metastases, although studies are lacking to describe the long-term survival and functional outcomes for these patients. Many patients undergo surgical resection/decompression, at which time a tissue diagnosis is made, and some will also receive chemotherapy (52); nevertheless, surgical treatment, radiation therapy, and chemotherapy are considered palliative in nature. As expected, the prognosis for patients with intramedullary metastases is grim, with one recent study finding a median survival time of 4 months and no patients achieving complete remission (51).

In the literature, there have been reports of well-circumscribed intramedullary spinal cord metastases that are amenable to GTR (35,71,98), however, GTR is often difficult due to lack of a clear dissection plane, infiltrative or indistinct margins, and heavy bleeding (105,120,129). The phenotypic features of the primary tumor likely contribute to the ability of the metastasis to be resected. Given the advanced state of the systemic malignancy and the risks involved with spinal cord operations, many surgeons are reluctant to attempt aggressive resections. One study demonstrated that neurological deficit, pain, and quality of life were significantly improved in a majority of patients following surgery, independent of the extent of resection (132). However, median survival is relatively unchanged and about 10% of the surgically treated patients deteriorated (119).

CNS Lymphoma

Intramedullary CNS lymphoma is a rare form of lymphoma with a prevalence of 2-3% of all non-Hodgkins lymphomas (NHL) and an even smaller proportion manifesting as spinal lymphoma with less than 40 cases reported resulting in myelopathy (56). It is usually an aggressive extranodal non-Hodgkin lymphoma (NHL) that resembles diffuse large B-cell lymphocytes. It can originate in the spinal cord, accompany tumors in other locations throughout the central nervous system, or occur as a part of systemic lymphoma (34,68). T1-weighted MRI shows homogenous contrast enhancement in an enlarged area of spinal cord while diffusion-weighted MRI demonstrate hyperintensity with an isointense to hypointense signal on apparent diffusion coefficient map (17,56). MR Spectroscopy identifies masses with diminished concentrations of N-acetylasapartate and elevated ratios (>3:1) of choline to creatine, similar to that of gliomas.

Due to its lack of localization, the primary treatment of intramedullary CNS lymphoma and other lymphomas of the neuraxis is the chemotherapeutic agent methotrexate; however, recurrence is common and often occurs within 2 months (37). High-dose methotrexate-based therapy has been shown to be effective in elderly patients suffering from primary CNS lymphomas, and even more so when combined with alkylating agents such as temozolomide (70). Thus, the development of chemotherapeutic agents in the treatment of primary CNS lymphoma is especially appealing, given that its diffuse nature makes surgical resection and radiotherapy more challenging. Surgical management is discouraged for primary CNS lymphoma of the spine given the response to chemotherapy and radiotherapy.

DISCUSSION

IMSCTs represent a difficult but rare pathology in a neurosurgeon's practice. Even though they often have significant neurological and functional consequences due to their eloquent location, rarity of incidence and technical difficulty of the surgical procedure, GTR remains the treatment of choice.

The initial imaging modality of choice is MRI with and without gadoliunium contrast agent with multi-planar views (43). This enables the surgeon to assess their location, configuration, presence or absence of peritumoral or intratumoral cysts, and texture (16,121). The T1-weighted imaging reveals the solid tumor component with pre- and post-contrast images are analyzed. The T2-weighted images reveal associated cysts, edema, and the CSF fluid spaces around the spinal cord. While it is not possible to make diagnostic decisions on imaging alone, several tumor types

have characteristic MR findings as mentioned in the previous sections, which can help the surgeon anticipate the type of resection expected. Astrocytomas and gangliogliomas are usually seen as eccentric, expansile lesions of the cord leading to asymmetry on axial imaging and express heterogeneous enhancement if present, though more often are non-enhancing. The tumor nodule of hemangioblastomas has marked enhancement with a frequently associated cystic component and surrounding edema with flow voids appreciated on T2-weighted imaging. Ependymomas express homogenous, intense enhancement. They are associated with cysts at the rostral and caudal ends, and hypointense areas on T2-weighted imaging at their poles, that represent hemosiderin deposits, often called "hemosiderin caps".

Preoperative evaluation of the patient is based on neurological and functional assessment. It is paramount to obtain preoperative and postoperative assesments in patients harboring intramedullary tumors. The McCormick scale (19,20,90), previously mentioned, is the standard evaluation technique that focuses on the neurological function and ambulatory status of the patient. As mentioned, the most common presenting symptoms is pain, diffuse more often than radicular, and is more prominent at night, thought to be due to the horizontal position assumed while sleeping, leading to mild venous congestion, increased pressure in the cord, with resultant meningeal irritation and activation of pain pathways (75). Younger children may present with decreased motor function or regression of motor abilities (19,20), while adolescents present with weakness, clumsiness and frequent falls. One third of patients present with spinal deformity (6,28), often associated with thoracic lesions with cystic components. Common exam findings for the majority of intramedullary tumor patients include spasticity, hyperreflexia, pathological reflexes, clonus, and increased muscle tone.

After the proper pre-operative imaging, neurological and functional evaluation are completed, surgical planning and preparation is performed. Classically, the patient is positioned prone in such a way as to decrease intraabdominal pressure (e.g. bolsters) in order to decrease intraoperative venous bleeding. For lesions cranial to T5 level, a Mayfield holder is often utilized to provide rigid fixation so to keep the head and spine in a neutral position throughout the operation. Recentley, Takami et. al. (122) have advocated a lateral oblique position for resection, with the advantage that any venous bleeding encountered during the procedure is allowed to follow the path of least resistance, and with the help of gravity, drains out of the operative field, thus negating the need for the non-dominant hand to continually harbor a suction device.

General anesthetic is achieved with opioid and propofol administration, adjusting dosages to keep the patient's blood pressure close to their awake value. Halogenated volatile anesthetics are avoided in these patient's due to their effect on intraoperative neurophysiological monitoring. Short acting muscle relaxants are given for intubation and allowed to wear off in order to allow for baseline neurophysiological levels to be obtained.

The Role of Intra-Operative Monitoring

Motor evoked potentials are especially important in IMSCT surgery. During dissection of the tumor, there is a distinct risk to injuring the motor tracts. Damage to this eloquent area is not always reflected in SEPs (41,65), and furthermore, changes in SEPs are common during the initial myelotomy (74), which have been shown to have no correlation to postoperative motor function (76). Additionally, contrary to SEPS, which provide delayed readings due to the averaging effect of the recordings and resultant delayed response to injury, MEPs provide near real-time feedback with high clinical correlation and allow corrective maneuvers to be deployed (93).

Motor evoked potentials (MEP) assess the function of the motor cortex and the descending motor pathways through the corticospinal tracts. In this technique, muscle activity is recorded after stimulation of the motor pathway through transcranial electrical motor cortex stimulation. In general, evoked potentials are described in terms of the post-stimulation latency in milliseconds and peak-to-peak amplitude in millivolts. These correspond to the time between the application of a stimulus and the occurrence of a peak in the waveform. These motor evoked potentials can be recorded from the spinal cord, either in the form of D-waves or from the peripheral muscles. It is the combination of these recordings that provide useful feedback to the surgeon. D-waves are recorded by an electrode placed in the epidural space caudal to the area of resection. These recordings represent a pool of high conduction velocity fibers within the corticospinal tracts that support locomotion. Muscle MEP responses are optimally recorded from muscles known to have strong pyramidal innervation, namely the thenar, hypothenar, tibialis anterior, and flexor hallucis brevis muscles.

Interpretation of both of these responses may help identify a window of reversibility should a change in recordings be found. According to some investigators, as long as the D-wave is preserved with an amplitude of at least 50% of the baseline value, a loss of muscle MEP during surgery correlates with a transient paraparesis or paraplegia (109). A decrease of more than 50% of the baseline D-wave amplitude is associated with a long-term postoperative motor deficit (93). The exact mechanism for this is unclear, however it is postulated that since the D-wave is generated exclusively by fast neurons of the corticospinal tracts and muscle MEP are generated by corticospinal as well as other descending pathways, an injury to the other pathways can be compensated by the corticospinal tracts but not vice versa.

Sala et al. reviewed their experience of intamedullary spinal cord tumor surgery with and without the use of monitoring. In this series, 50 surgeries performed before the introduction of MEP monitoring were compared to 50 consecutive operations performed with monitoring. Preoperative neurological status, histological findings, tumor location, and extent of removal were found to be independent of outcome. At the time of surgery, the disappearance of muscle MEP led to modification of surgical technique and surgery was not discontinued as long as D-wave amplitude remained greater than 50% of baseline. A decrement of more than 50% in D-wave amplitude without recovery was the major indication to stop surgery. At 1 year, patients in the monitoring group had significantly better outcomes, though the rates of early motor dysfunction at discharge were similar between groups.

Kothbauer et al. also performed a retrospective analysis of the pre- and postoperative motor status of 100 consecutive patients who underwent surgery for intramedullary tumors using intraoperative monitoring with D-wave and MEP. The authors report that the sensitivity of muscle MEP to detect postoperative motor deficits was 100% and its specificity was 91%. In this series, no motor deficits were detected postoperatively in patients with stable MEP. Patients with preserved D-wave amplitude up to 50% of the baseline amplitude with a complete loss of muscle MEP resulted in only transient paraplegia. Despite the transient weakness, all recovered within a few hours to a few weeks.

Intraoperative Strategies for Recovery of Potentials

In addition to surgical manipulation of the spinal cord, various physiologic factors can affect evoked responses. These include hypotension, hypoxemia, hypothermia, acidosis, low circulating blood volume, and electrolyte imbalance (116). When a change in responses occur, it is important to systematically assess for each of these.

Likewise, several surgical strategies may help recover lost or diminished potentials. Signals may recover spontaneously if surgery is stopped immediately after muscle MEP have disappeared or after D-wave amplitude has decreased significantly. Sala et al. describe a "stop and go" strategy during which surgery is stopped for half an hour or more to allow muscle MEP and/or D-wave to recover on their own before proceeding with surgery.

Irrigation of the surgical field with warm saline irrigation has also been shown to recover both SSEP and MEP (23). Whether this is due to the effect of temperature, irrigation, or a combination of the two is not clear. It is theorized that irrigation dilutes potassium that accumulates in the extracellular space and blocks conduction. This is based on experimental spinal cord injury models in which traumatic injury to the spinal cord induces a disruption of cell membranes with subsequent accumulation of potassium in the extracellular space.

Another mechanism of spinal cord injury during surgery is related to ischemia secondary to decreased perfusion pressure or vasospasm. The extent to which the spinal cord can tolerate a decrease in perfusion pressure is unknown. Applying papaverine locally to the spinal cord and increasing the mean arterial pressure may improve perfusion to the spinal cord and counteract any ongoing ischemia due to vasospasm or hypoperfusion.

After baseline motor and sensory feedback is obtain the patient is prepped and draped. A midline skin incision is centered at the midpoint of the lesion and extended rostrally and caudally. The raphe is split to allow retraction of the muscles. A laminectomy or osteoplastic laminectomy/laminotomy is performed. This is achieved with a high speed drill fit with a cutting bur, match stick bit, or craniotome attachment. Care is taken not to remove the lateral masses in the cervical spine or disrupt the pars articularis in lower segments to decrease the risk of post operative kyphosis. The amount of bone removal should correspond to the size of the solid component of the tumor, usually one level above and below the solid component, but does not need to include rostral or caudal cysts, unless their walls enhance. These are considered intratumoral cysts, as apposed to peritumoral

cysts found a the poles of the tumors and are usually composed of non-neopastic glial tissue (75). In pediatric patients, some authors prefer a laminotomy, with unilateral incision and unilateral laminotomy utilized for infants (16).

Once the bone is removed and the dura exposed, intraoperative ultrasound localizes and verifies the tumor location to plan the dural opening, allowing for further bone removal if necessary for adequate exposure (24). Prior to opening the dura, meticulous hemostasis is performed. Elongated strips of cottonoid or gel foam are placed at the lateral aspects of dural exposure in the bone opening to control venous bleeding.

Using loupe magnification or a microscope, the durotomy is made in the midline with the intent of keeping the arachnoid membrane intact. Dural retraction sutures with 4-0 nylon suture are placed for improved visualization. The arachnoid is then opened with micro-scissors and separated from the postero-lateral aspects of the spinal cord. The spinal cord is often rotated, expanded and distorted, making identification of the midline raphe difficult. Close visualization may reveal subtle color changes in the subpial spinal cord due to the presence of a tumor. Once either the lesion is identified by direct observation or referencing the preoperative imaging studies, the decision is made to use a midline or more lateral approach. The senior author prefers a midline approach unless the tumor is located in the posterior column or seen on the pial surface. Ependymomas, often centrally located, should be approached by a midline incision, compared to astrocytomas, which are often eccentric with occasional exophytic components that are better approached by a more direct, laterally oriented incision. If in doubt, however, a midline myelotomy is safer and should be the surgeon's first option. In cases of hemangioblastoma and cavernomas, they are commonly found at the pial surface, negating the need to perform a midline approach.

The first key step in dissection is to identify the dorsal median sulcus of the spinal cord. Observing the posterior spinal vein, a tortuous vessel that courses over the midline raphe, may identify this anatomical landmark. There is usually a convergence of vessels directed toward the midline, which may aid in identification of the surgical plane.

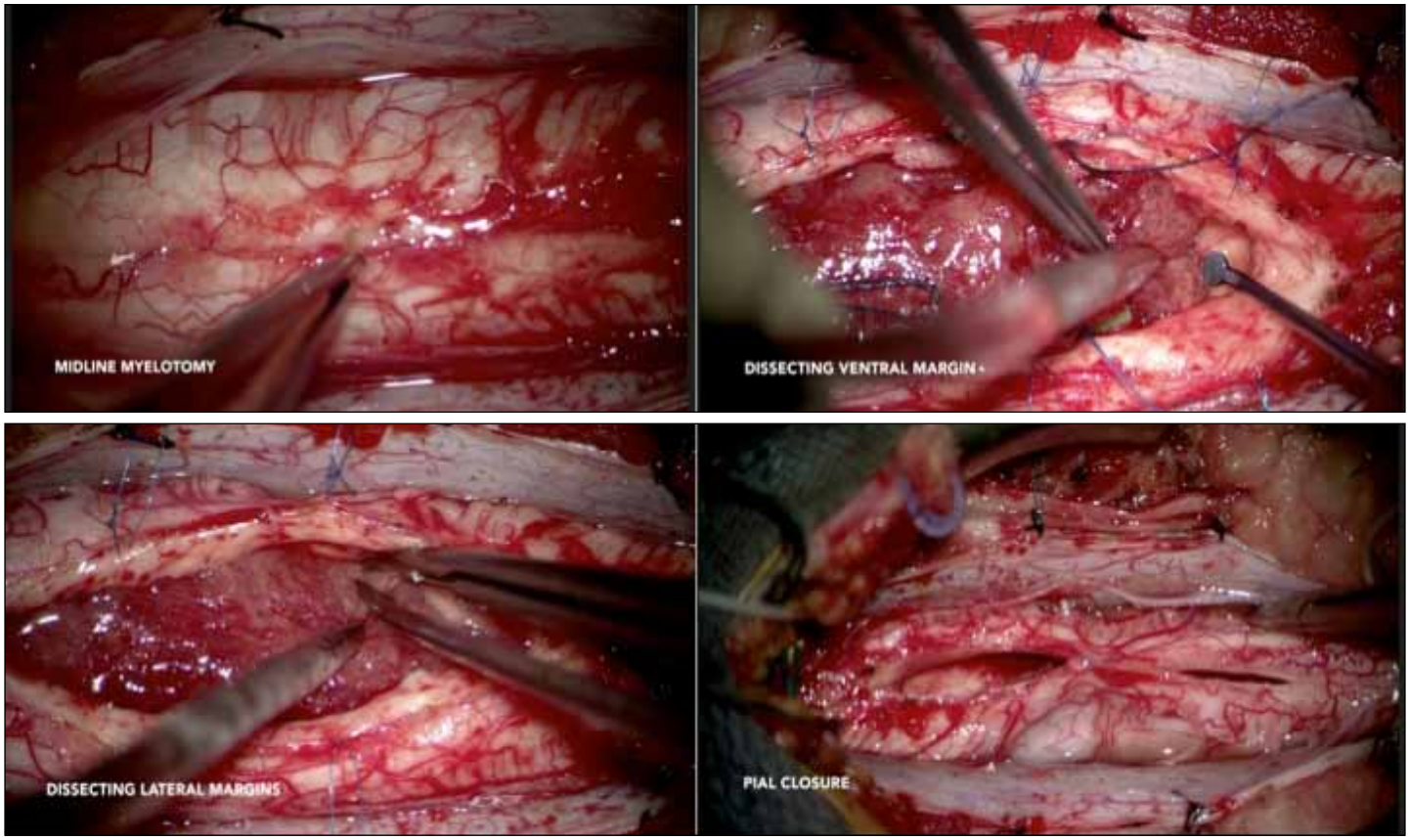

Figure 1: *Surgical technique is demonstrated. Midline myelotomy is first completed followed by the dissection of the lateral and then ventral margins. Ultimately after the tumor is resected the pia and dura mater and closed.*

As the dissection proceeds, care must be taken to preserve the longitudinally running vessels along the posterior column. They may be mobilized and gently retracted laterally. The dorsal columns are retracted and opened with microinstruments, extending to just beyond the rostral-caudal extent of the solid component of the tumor. SEPs should be continuously monitored through the dissection in attempt to preserve as much posterior column function as possible. Dorsal column mapping is now feasible and can help identify this dissection plane, especially for tumors that expand the cord and make identification of this raphe difficult with even high-powered magnification (95).

Once enough tumor has been exposed, a biopsy should be taken to confirm the histological diagnosis. Pathological suggestion of infiltrative or malignant tumor should be weighed heavily when deciding to continue depending also on the aggressiveness of the planned resection and the goal of functional outcome once the resection is completed.

If the surgeon decides to continue, the resection is continued either in an inside to out debulking following the identification of the cleavage plane, or following the tumor to either the pole in order to identify the tumor-spinal cord plane with subsequent dissection and separation the tumor from the spinal cord proper in a craniocaudal or caudalcranial manner. It is important to note however, that while most ependymomas have such a plane, only 30-40% of astrocytomas have a viable cleavage plan to allow further, more aggressive resection (114). Both astrocytomas and gangliogliomas have a heterogenous composition due to the lack of a true plan between the tumor and the spinal cord, GTR is surgically impossible without devastating neurological injury (29). In the instance of not being able to identify a proper cleavage plane, a biopsy only for histological diagnosis is pursued, with adjuvant therapy in the postoperative period. The risk of devastating neurological injury is too great in this instance and unnecessary removal should be avoided (58).

Once the cleavage plane is identified, the goal of the surgeon at this point should be GTR. The use of dual micro-forceps by the primary surgeon to dissect and advance the cleavage plane is often thought to be safer than the utilization of micro-dissectors in minimizing the local recurrent trauma to the surrounding spinal cord. If during the dissection, the plane becomes unrecognizable, or there are significant changes in the MEP or SEPs, the surgeon would be wise to pause the dissection in that area and move to a different location of the tumor and begin working again, with the plan to return to the difficult area once further dissection has been achieved.

The vascular pedicles of the tumor, which are branches of the anterior spinal artery, are often small, penetrating vessels passing through the anterior median raphe and adherent to the tumor, especially in the case of ependymomas. When removing the tumor and coagulating the pedicles' feeding arteries, care must be taken to not injure the anterior spinal artery (45). Often after resection of the tumor, bleeding will stop spontaneously, however, if hemostasis with bipolar cautery is necessitated, caution should be used to avoid retrograde thrombosis of the anterior spinal artery or local thermal injury to the cord.

The spinal cord is re-approximated with 6-0 nylon sutures and reconstitution of the arachnoid membrane is attempted. The dura is closed with running 4-0 nylon suture and if possible, the bone is re-approximated and secured with plating or sutures in younger patients. Superficial layers are closed in the usual fashion and the patient is placed on flat bed rest to decrease the risk of CSF leak.

There are other, specific considerations the surgeon must take into account depending on the particular type of lesion encountered. Ependymoma and astroytomas were discussed in the preceding paragraphs, but other lesions, such as hemangioblastomas and cavernomas have their own, specific surgical maneuvers that should be performed.

Hemangioblastomas are often visualized on the posterior or postero-lateral pial surface upon opening the dura. These lesions may be approached directly. If the lesion is located on the anterior aspect of the cord, one or several dentate ligaments may be resected, 6-0 nylon suture thrown through the ligamentous attachment and gentle rotational traction applied until the lesion becomes visible and access is adequate. As with vascular lesions elsewhere in the CNS, care must be taken not to coagulate the main draining vein at the onset of resection. If there is a large cystic component to the lesion, it might prove beneficial to deflate the cyst with needle aspiration prior to resection. The starting point of the dissection is identified as the area where the boundary of the tumor is located with the smallest number of associated vessels. Detachment of the tumor from the spinal cord is facilitated by cauterization of the tumor surface and applying gentle traction on the tumor to expose the tumor-spinal cord interface. The subsequently encountered vascular

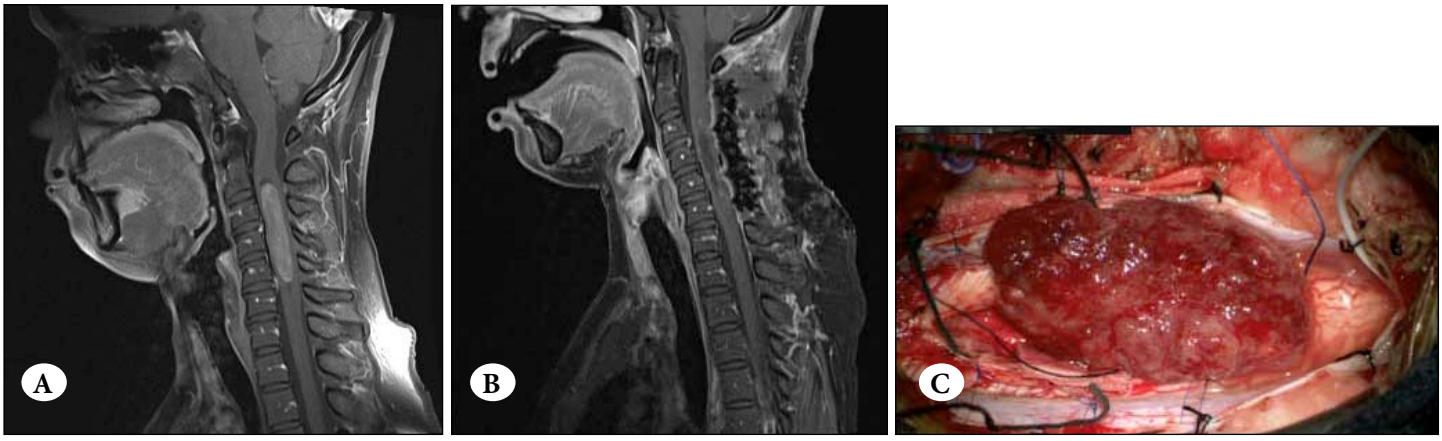

Figure 2: A) *An MRI of a sagittal T1 post-contrast image demonstrates an enhancing cervical ependymoma preoperatively.* ***B)*** *The post contrast T1 sagittal MRI demonstrates a gross total resection.* ***C)*** *The intraoperative image demonstrates the vascular ependymoma being microsurgically resected.*

connections are coagulated in a systematic, stepwise approach. This process is completed until the tumor can be removed en bloc. The same approach is also utilized for cavernomas. If the hemangioblastoma is associated with a large cystic cavity, opening the cavity, removing the fluid, and resecting the nodule is sufficient for this particular component of the lesion.

Post-operative care is provided in a dedicated neuro-intensive care unit if available. MRI imaging is obtained within 48 hours of the completion of surgery to evaluate the extent of resection. Adequate analgesics and anti-inflammatory medications are provided and early mobilization and deep vein thrombosis prophylaxis is implemented.

Intramedullary spinal cord tumor surgery carries a not insignificant risk of complications. The most feared complication is paralysis. The risk of occurrence of post-operative neurological motor dysfunction is directly related to the patient's preoperative neurological function. Patients with significant motor dysfunction preoperatively are more likely to have postoperative deterioration (93). One third of patients will have short-term motor dysfunction, resolving in hours to days (76). More protracted, long term dysfunction is directly related to preoperative neurologic function (93). This is the justification for the theory of operating on intramedullary tumors as early as possible to ensure attempted resection occurs prior to the development of severe, disabling neurological deficits.

Post operative spinal deformity may occur after surgery in the form of scoliosis or kyphosis (107,130,136), which is especially important in children. It is believed that osteoplastic laminectomies reduce the incidence of spinal deformity in children (1,104). Nonetheless, it is important to follow children with standing plain films. If there is a development of progressive deformity, in addition to surgical causes, a repeat MRI must be obtained to rule our recurrent tumor.

CONCLUSION

Surgery remains the initial treatment of choice for intramedullary spinal cord tumors (IMSCTs). New and interesting therapies are currently under investigation and the extent to which they augment our current treatment paradigms remains to be seen but with the advent of novel therapies (nanomedicine, localizable therapies, and cell targeting) (124) the future appears promising for lesions not amenable to GTR and a surgical cure. While complete resection is not always achievable, we must remain tempered by the paramount importance of our patient's neurological function, for it is their well-being and quality of life outcome that is the root purpose of all of the aforementioned endeavors.

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Manuscript submitted November 05, 2015.

Accepted January 17, 2016.

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