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REVIEW PAPER

Intramedullary spinal cord tumors in children

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Abstract *Introduction:* Pediatric intramedullary spinal cord tumors are rare and make up only a small percentage of all central nervous system neoplasms. *Discussion:* These neoplasms are predominantly benign histologically, regardless of size, with a large percentage being astrocytomas or gangliogliomas. Ependymomas, which are common in adults, are relatively uncommon in children. Contemporary management of these tumors has generated much controversy. These neoplasms occur primarily in the cervical region, with pain or a motor deficit as the chief complaint. Surgery on these neoplasms can safely be performed using modern surgical adjuncts such as the ultrasonic aspirator, contact laser and neurophysiological monitoring. A radical resection of these tumors results in a good long-term outcome, since the majority are histologically benign. Adjuvant radiation therapy should only be administered for high-grade or malignant tumors. *Conclusion:* The majority of spinal cord tumors are benign; however, malignant tumors have a dismal outcome and surgery in these children should be limited to a conservative debulking. An attempt should be made for radical surgery in hope of avoiding radiation and chemotherapy for the benign tumors. Spinal deformity is a concern in these patients since 35% of children may require a stabilization procedure.

Keywords Astrocytoma · Intramedullary · Spinal cord · Spine · Surgery

Introduction

Intramedullary spinal cord tumors are rare central nervous system neoplasms. In particular the intramedullary location is an uncommon site for tumors with only 35–40% of all intraspinal tumors in children found in this location. The first successful resection of an intradural tumor, a fibro-myxoma, was accomplished in 1887 by Victor Horsley [30], and the first successful resection of an intramedullary spinal cord tumor was performed in 1907 by Anton von Eiselsberg in Austria [18]. However, the first report of an intramedullary tumor appeared in 1911 by Charles Elsberg in New York [19]. Elsberg described a two-stage strategy for the removal of these intramedullary tumors. At the initial operation a myelotomy would be performed, the surgeon would then return 1 week later to remove the spinal cord tumor. This technique allowed the neurosurgeon to remove only the extruded portion of an intramedullary tumor.

Following these initial reports, pioneering neurosurgeons advocated a more aggressive and radical tumor surgery. However, the postoperative neurological morbidity was quite significant as it included surgery at the wrong levels, cerebrospinal fluid leaks, infection, paralysis, and in some cases death. Thereafter, many subsequent neurosurgeons again recommended a more conservative strategy with biopsy, dural grafting, and radiation therapy regardless of the histological diagnosis and age of the patient [67]. With the advent of modern neurosurgical instruments, operating microscope, imaging technology, and intraoperative neurophysiology, the treatment strategy for these intramedullary neoplasms has again become more

Table 1 Histological diagnoses of pediatric intramedullary neoplasms at a single institution between 1991 and 1998

Tumor type	Children
Juvenile pilocytic astrocytoma	4 (3%)
Fibrillary astrocytomas	45 (38%)
Low grade	32
Anaplastic	10
Glioblastoma	3
Ependymoma	14 (12%)
Oligodendroglioma	1
Mixed glioma	3
Ganglioglioma	31 (27%)
Miscellaneous	14 (12%)
Neuronal tumors	10
Hemangioblastoma	3
Others	1
Total	113

aggressive. This is particularly important as the majority of intramedullary tumors are histologically benign [11, 53] and the radical removal results in long-term survival with an acceptable morbidity [11, 14, 20, 21, 25, 31, 32].

A review of the spinal cord database at a single institution between 1991 and 1998 yielded 294 cases in adults and children [53]. The 294 tumors included 117 removed from children under the age of 21 years, and 177 from patients 21 years and older (Table 1). The majority of these pediatric tumors were operated upon by the senior author (F.E.). The most common single tumor type in this study population was the fibrillary astrocytoma, which accounted for 45 or 39% of all tumors. There were 31 gangliogliomas, 19 ependymomas, 5 of which were myxopapillary ependymomas located in the cauda equine. Significantly, in this large series of children there were only 13 high-grade tumors.

Clinical presentation

Intramedullary tumors may remain asymptomatic for a long time or cause nonspecific complaints, which make the diagnosis difficult in children. The onset of symptoms is often insidious and symptoms are typically present for many months. High-grade or malignant neoplasms typically have a shorter prodrome (median, 4.5 months). Some children may complain of symptoms following a trivial injury. The most common symptom is pain that may be diffuse or radicular in nature. There is no characteristic feature for the pain associated with an intramedullary tumor, although children will complain of nocturnal pain that awakens them from sleep. This complaint should be a heralding sign to all practitioners. Young infants may even present with abdominal pain, enduring extensive gastrointestinal investigations before receiving the diagnosis of intramedullary tumor.

Children may present with some motor deficits, which can initially be perceived as clumsiness, weakness, or frequent falls. In young infants, this may manifest as motor regression such as refusal to stand or crawl after having learned to walk. Complaints of sensory dysfunction are quite uncommon in children.

One third of patients may initially present with scoliosis [69]. The scoliosis curve is not specific to any direction. Children with scoliosis typically have paraspinal pain, which is otherwise unusual for intramedullary tumors.

Diagnostic studies

Magnetic resonance imaging (MRI) is the study of choice to identify an intramedullary spinal cord neo-

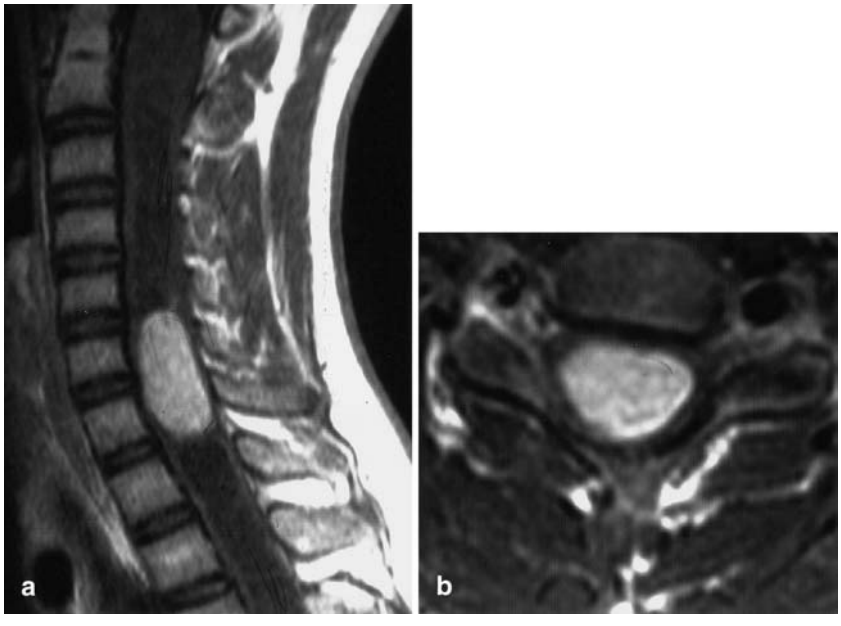

Fig. 1a, b MRI in a 13-year-old boy who presented with neck pain lasting for several months. Histological diagnosis was a ganglioglioma. **a** Sagittal T1-weighted image demonstrates the enhancing cervical tumor associated with a rostral and caudal cyst. **b** Axial T1-weighted image demonstrates the eccentric location of this large tumor

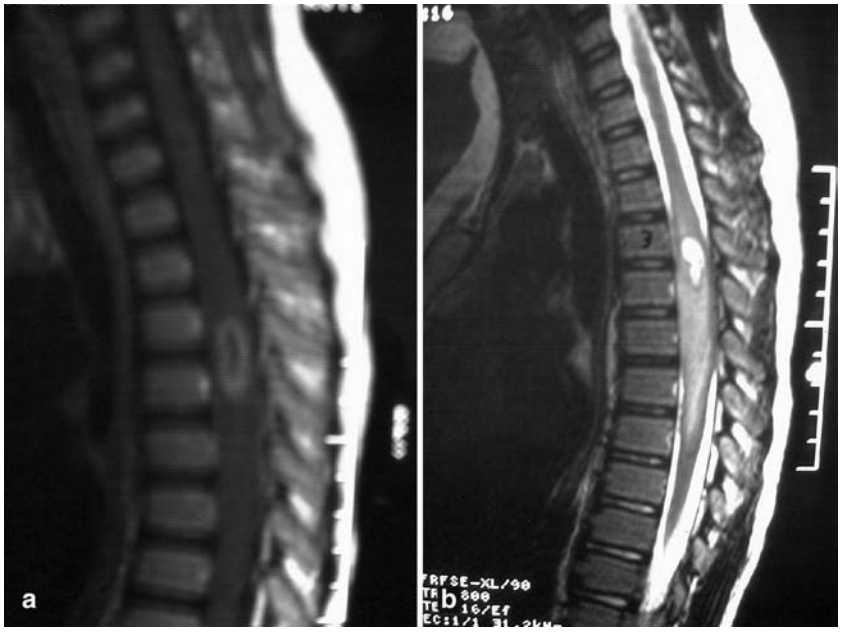

Fig. 2a, b MRI in an 11-year-old girl with rapid onset back pain and scoliosis. Histological diagnosis was a glioblastoma. **a** T1-weighted sagittal image with contrast demonstrates the enhancing tumor within the thoracic region. **b** T2-weighted sagittal image confirms the tumor with central necrosis and more extensive signal change

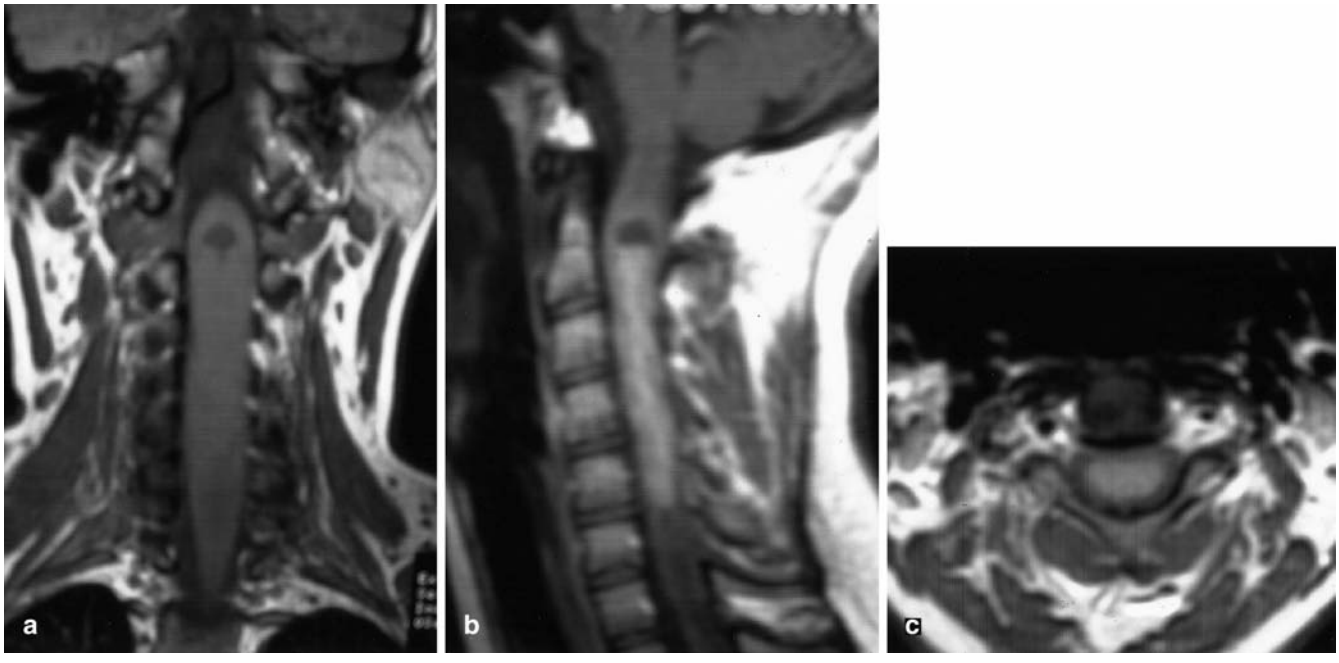

Fig. 3a–c MRI in a 19-year-old male who presented with numbness and finger clumsiness. Histological diagnosis was an ependymoma. **a** Coronal T1-weighted MRI demonstrates a rostral cyst and expansile cervical tumor. **b** Sagittal T1-weighted MRI demonstrates the enhancing tumor from C2–C5. **c** The axial T1-weighted images with contrast demonstrate the characteristic central location of this tumor type

plasm [6, 15]. MRI should be performed with intravenous contrast agents and in multiple planes. These images demonstrate the solid tumor component, associated cysts, edema, and on occasion the scoliosis. Although MRI does not provide the histological diagnosis, there are some typical patterns of appearance for intramedullary tumors. Astrocytomas and gangliogliomas have a heterogeneous enhancement pattern (Fig. 1). These tumors are often eccentrically located and produce an asymmetric enlargement of the spinal cord. Malignant tumors do not have characteristic imaging features, although extensive T2-weighted signal change is highly suggestive (Fig. 2). Ependymomas, on the other hand, tend to enhance brightly and homogenously with contrast (Fig. 3) and are centrally located within the spinal

cord. Often ependymomas are associated with rostral and caudal cysts.

Plain radiographs are mandatory only in children who present with scoliosis. These initial films serve as a baseline for the future management of spinal deformity. In addition, children who undergo an extensive laminotomy or laminectomy should be followed with serial radiographs and we therefore advocate immediate postoperative films.

Surgical management

Surgical instruments

The traditional method of removal of an intramedullary neoplasm is now supplemented by special microinstruments. These instruments have become essential for the microsurgical resection of spinal cord tumors.

The Cavitron ultrasonic aspirator (CUSA) uses high-frequency sound waves to fragment and then suction tumor from the tip of this device [10, 26]. This surgical adjunct now has microtips that allow for intraspinal use [37]. This allows for tumor removal with only minimal manipulation of the adjacent spinal cord tissue.

The laser is another excellent surgical instrument for intramedullary surgery. The Nd:YAG Contact Laser System (SLT, Montgomeryville, PA) has a hand piece and various contact probes. The contact probes are useful as a scalpel to perform the myelotomy, demarcate the glial-tumor interface and to remove any residual fragments. Unlike other laser systems, there is minimal associated char and smoke generation. This contact laser is effective in preventing small arteriole or capillary bleeding. This laser essentially works as a microinstrument [40].

Surgical technique

There have been many publications illustrating the technique for intramedullary spinal cord tumor removal [4, 39]. The surgical approach for all intramedullary tumors is a laminectomy or osteoplastic laminotomy. For resection of cervical or cervicothoracic tumors, the child's head is fixed in a Sugita head holder. The child is positioned on soft gel rolls to minimize any venous hypertension. We avoid the horseshoe headrest for fear of facial and eye abrasions.

A craniotome is used to cut the laminae bilaterally. In children who have been previously operated upon, the laminae may not be present for repositioning. The opening is made large enough to expose the solid component of the tumor. The rostral and caudal cysts do not need to be fully exposed. This opening is planned with X-ray control and verified with intraoperative ultrasound. The ultrasound allows the surgeon to visualize the spinal cord in two dimensions [23]. Intramedullary astrocytomas and gangliogliomas have a similar echogenic pattern to the spinal cord. However, the cord will appear expanded in the area of the tumor. In contrast, ependymomas tend to be hyperechogenic and can readily be differentiated from spinal cord.

The ultrasound is helpful in identifying the associated cyst(s). If the bone removal is not adequate to visualize the full extent of the tumor, the laminotomy is extended prior to opening the dura. The dura is then opened in the midline. The spinal cord is expanded and may occasionally be rotated. The asymmetric expansion and rotation of the spinal cord may make the identification of the midline difficult. In those cases of an asymmetric tumor and rotated spinal cord, a myelotomy may be performed through the dorsal root entry zone.

The neoplasm is typically located several millimeters underneath the dorsal surface. The contact laser is used to perform the myelotomy with minimal neural injury. Intramedullary tumors have differences in appearance, such as texture and color, which help the neurosurgeon differentiate the tumor type.

Astrocytomas or gangliogliomas have a gray-yellow appearance. A true plane between tumor and normal spinal cord does not exist. The surgeon should make no effort to define this "true" interface because it results in hazardous manipulation of normal spinal cord tissue. Ependymomas are typically red-gray in color and these tumors are well demarcated from the surrounding spinal tissue. This interface can be separated with a plated bayonet [22] or the scalpel probe on the contact laser [40].

Once the tumor is exposed a biopsy is taken for immediate histological examination. This information may, if a malignant glioma or inflammatory process is suspected, be crucial in deciding the extent of tumor resection. For malignant gliomas a more conservative approach, to limit any potential motor deficits, is undertaken. The goal is a debulking with preservation of motor function.

Tumor removal for low-grade astrocytomas and gangliogliomas begins after the initial myelotomy is performed. An internal debulking with the CUSA is done to reduce tumor volume. Resection of astrocytomas is initiated at the mid-portion rather than the tumor poles. The rostral and caudal poles are the least voluminous and manipulation at these locations may be the most dangerous to the normal cord tissue. Then, using the suction or contact laser, the tumor is gently removed from the surrounding spinal tissue. These tumors do not have a cleavage plane, although in some areas a plane may exist between tumor and normal spinal tissue. These tumors tend to displace the motor tracts anteriorly or laterally. The surgeon should be aware of these pathways during tumor resection.

The rostral-caudal length of the tumor does not influence the functional outcome after tumor resection. We

have found the removal of small tumors with a wide girth to be more difficult and hazardous than the holocord narrow tumors. This observation corresponds to previous reports that spinal cord atrophy is a poor prognostic factor in terms of neurologic dysfunction [34].

Miscellaneous tumors

Hemangioblastomas are quite uncommon in children unless associated with von Hippel-Lindau disease [62]. Hemangioblastomas in the spinal cord, regardless of size, are often associated with significant edema and syrinx formation [12] These lesions should be resected in a circumferential fashion. The tumor surface can be coagulated to allow for the manipulation of the lesion; however, this tumor should not and cannot be debulked from within. Cavernous malformations, similar to hemangioblastomas, are typically located on the dorsal surface of the spinal cord [14]. These lesions are typically located in the cervical cord. This vascular malformation is resected in an inside-out fashion, similar to the technique for astrocytomas. These lesions do not usually bleed during the resection, thus the CUSA or suction cautery can be safely used for their removal. These vascular malformations are quite uncommon in children. When these lesions present in the pediatric population, there is a high chance of multiple intracranial lesions [14].

Intramedullary lipomas require a different strategy from glial neoplasms [46]. Although this tumor may appear well demarcated from the adjacent spinal cord tissue, these lesions are densely adherent. Thus, total removal is fraught with neurological compromise. The contact laser is used to debulk these tumors. The laser vaporizes the fatty tissue without any surgical trauma to the spinal cord.

Following intramedullary tumor removal, the dura is closed primarily in a watertight fashion. If an osteoplastic laminotomy is performed the laminae are replaced and secured with titanium or absorbable miniplates. One tissue layer must be closed in a cerebrospinal fluid tight fashion. The muscle and fascial closure must not be under tension [71]. Children who have had previous surgery and radiation therapy are at increased risk of wound dehiscence and cerebrospinal fluid leak.

Intraoperative neurophysiological monitoring

Intraoperative monitoring with motor evoked potentials (MEPs) allows direct monitoring of the corticospinal tracts [3, 7, 13, 45, 55]. This monitoring is predictive of functional motor outcome for intrinsic spinal cord tumor surgery [45]. Motor potentials are evoked with transcranial stimulation of the motor cortex. A single electrical impulse results in the direct activation of fast-conducting axons. This potential, called D-wave [58], is recorded by an epidurally placed electrode just distal to the intramedullary tumor. The D-wave amplitude is a relative measure of the number of functioning fast-conducting corticospinal fibers. When this amplitude drops 50%, then 50% of these fibers have been injured [55]. The muscle MEPs are elicited with a short high-frequency train of 5–7 electrical stimuli [66]. The responses are recorded with needle electrodes from limb muscles with heavy pyramidal innervation, such as the thenar muscles, the tibialis anterior, and short toe flexors. These motor potentials follow an on-off pattern; their presence indicates intact motor control and their absence or loss is highly indicative of temporary loss of motor function [45].

Table 2 Interpretation of motor-evoked potentials (*MEP*) during intramedullary tumor surgery

D-wave	Muscle MEP	Motor status
Decreased <50%	Unchanged	Unchanged
Decreased <50%	Uni- or bilateral loss	Transient motor deficit
Decreased >50%	Bilateral loss	Prolonged motor deficit

The D-wave and muscle MEPs must be interpreted together [43, 44, 45]. Loss of muscle MEPs during a tumor resection indicates temporary disruption of motor function. The incremental change of the D-wave amplitude allows for further interpretation and resection of motor outcome in cases of intramedullary tumors. As long as the D-wave amplitude remains above 50%, the patient is likely to awaken with a motor deficit; however, the deficit will disappear within hours, days, or weeks. The intraoperative monitoring using these two electrophysiological parameters has significantly improved the safety of complete resections of intramedullary neoplasms (Table 2).

Somatosensory evoked potentials (SEPs) are used to assess the functional integrity of the sensory system. The correlation of SEPs with pre- and postoperative motor function is poor [43]. In many cases of intramedullary tumors, the SEPs disappear after the myelotomy is performed.

Outcome following surgery

Neurologic outcome

The most feared complication following intramedullary tumor surgery is paralysis. The incidence of this occurrence is related to the preoperative motor status. Patients who have no or minimal preoperative motor deficits have less than 1% incidence of this postoperative complication. Almost all patients undergoing gross total re-

section of intramedullary spinal cord tumors experience some postoperative deterioration of neurological function, and about one-third have a significant temporary motor deficit. This neurological deterioration typically disappears within a few weeks [5, 29, 43, 45].

In our group of over 200 children, there were no deaths due to surgery. The tumors were located throughout the spinal axis. Most children remained at their preoperative grade level or improved. Of the patients who deteriorated, 60% deteriorated by only one functional grade. The children whose grade deteriorated already had significant preoperative motor deficits. At last follow-up, mean 14.2 years, more than 65% of the children were functioning at a Grade I or II level on the modified McCormick Scale [51]. Thus, it is essential that children with known intramedullary tumors be operated upon before the development of severe neurological deficits.

It appears that neurologic improvement after surgery is more likely in patients undergoing total resection than in patients undergoing partial resection [68]. In our series, the extent of gross total resection (>95%) or subtotal resection (80–95%) did not significantly affect the long-term outcome. Only patients who underwent a partial resection (<80%) fared significantly worse than those with radically removed tumors. These conclusions regarding intramedullary astrocytomas have been supported by others [56].

Table 3 Survival of patients with low-grade astrocytomas treated with radiation reported in the literature

Reference	Patients	Survival rate (%)	
		5-year	10-year
[42]	9	89	89
[28]	14	60	50
[47]	12	91	91
[8]	16	60	40
[63]	6	50	50
[36]	8	86	57

Oncologic outcome

Despite gross total resections for intramedullary neoplasms, residual microscopic fragments are left in the resection bed. These residual fragments may remain dormant or involute over time. There is abundant evidence that radiation has deleterious effects on the nervous and osseous system [9, 17, 49]. The alterations in motor- and sensory-evoked potentials in patients who have received radiation therapy have been documented [55]. Some authors still recommend radiotherapy for all intramedullary neoplasms regardless of the histological diagnosis [54, 57]. In a review of the literature, several papers advocate radiation therapy for low-grade astrocytomas. Although there is a limited number of patients in each series, with different radiation protocols, the overall 5-year survival was 50 to 91% (Table 3) [8, 28, 36, 42, 47, 63]. Our overall 5-year survival with radical surgery alone for low-grade tumors (astrocytomas and gangliogliomas) was 88%. Thus, there is no evidence that radiation therapy improves the outcome of low-grade astrocytomas or ependymomas [24, 25, 35, 38, 64]. Therefore, we only recommend radiation therapy for malignant tumors, for children with documented postoperative rapid tumor regrowth, and in those cases where substantial tumor remains and further surgery is not safely feasible. Unfortunately, despite adjuvant radiation therapy these malignant neoplasms invariably progress.

Chemotherapy has rarely been used for intramedullary tumors. There are no large studies documenting the effectiveness of this modality for pediatric intramedullary tumors (Table 4) [2, 16, 27, 33, 48, 52]. Although adjuvant chemotherapy is considered standard therapy for low-grade intracranial tumors, there is little justification to use this modality alone for low-grade intramedullary tumors that can safely be removed.

Spinal deformity

Scoliosis and kyphosis may evolve following laminectomy [60, 69, 70]. Many children, approximately two-

Table 4 Reported cases of chemotherapy responses in children with intramedullary spinal cord tumors (*CDDP* cisplatin, *CPA* cyclophosphamide, *JPA* juvenile pilocytic astrocytoma, *LGA* low-grade astrocytoma, *PCBZ* procarbazine, *VCR* vincristine, *VP-16* etoposide)

Reference	Patients	Histology	Chemotherapy	Follow-up (months)
[48]	1	LGA	Carboplatin/VCR	14
[27]	5	JPA 3 LGA 2	Carboplatin/VCR	22 (mean)
[16]	6	JPA 2 LGA 1 Oligoastrocytoma 3	Carboplatin/PCBZ/VCR/CPA/VP-16/CDDP	40.6 (mean)
[52]	2	LGA 2	CDDP, VP-16 (1), Carboplatin (1)	1–226
[33]	3	JPA 2 Ganglioglioma 1	Carboplatin	27.3 (mean)

thirds, developed a spinal deformity following laminectomy for their intramedullary neoplasm. However, only one-third of these children required a stabilization procedure. The only significant factors associated with progressive deformity were cystic tumors, prior radiotherapy, and age less than 7 years. The mean time to an orthopedic stabilization procedure was 3.4 years in our series. Other examined parameters such as location, histology, extent of laminectomy, and imaging characteristics were not significantly associated with subsequent stabilization surgery. Spinal deformity has been reported as a complication of radiation therapy used to treat epidural tumors [41, 61]. There was a higher rate of spinal deformity in children who were irradiated at a younger age and at doses greater than 20 cGy for neuroblastomas [50]. In another study of 58 patients younger than 25 years who underwent laminectomies for intraspinal tumors, deformity occurred in 46% of patients younger than 15 years and in only 6% of patients older than 15. In addition, there was a higher incidence for a progressive deformity in the cervical region [65]. Several authors have recommended osteoplastic laminotomies for all children to reduce the incidence of spinal deformity [1, 59]; however, we could not evaluate this parameter in our series. It is essential that patients with intramedullary neoplasms be followed for possible progression of spinal deformity. All patients should undergo routine serial plain radiographs.

Conclusion

Surgical resection for intramedullary neoplasms has evolved since the initial reports. With the advent of microsurgical techniques, imaging technology, and intraoperative electrophysiology, the radical resection of intramedullary neoplasms is a safe and effective treatment. In particular, the electrophysiological monitoring of motor pathways is extremely helpful in achieving a radical resection for these intramedullary tumors. The functional outcome of surgery is best correlated with the preoperative status, thus surgery should be performed early prior to the onset of severe motor deficits. The majority of these neoplasms are typically benign in children. Adjuvant radiation and chemotherapy should only be administered to malignant or inoperable intramedullary tumors.

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