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ORIGINAL PAPER

Surgical management of long intramedullary spinal cord tumors

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Abstract
Object Spinal cord tumors represent approximately 10–20% of primary central nervous system tumors. Only 20–30% of primary intradural tumors are intramedullary. The incidence of longitudinally extensive tumors involving the cervical, thoracic, and lumbar spine is very low (<1% of intramedullary lesions); hence, little literature exists on the management of this entity.
Materials and methods We retrospectively reviewed all patients undergoing surgical resection of longitudinally extensive intramedullary spinal cord tumors involving the majority of the spinal cord between 1990 and 2002. Clinical, radiographic, operative, and outcome variables were retrospectively recorded and reported.
Results Thirteen patients (eight male, five female) were included in the study. Mean age was 15 years (range, 3–45) at the time of the initial resection. Gross total resection was achieved in eight cases and subtotal resection in five cases. Pathology revealed astrocytoma in six cases (two pilocytic, four grade II), gangliogliomas in four cases, oligodendroglioma in two cases (one anaplastic), and lipoma in one case. One (8%) patient died from progression of anaplastic oligodendroglioma, and two (15%) underwent reoperation for recurrent tumor (ganglioglioma, grade II astrocytoma). With a mean of 3.4 years (range, 1–12) after surgery, the modified McCormick score (MMS) had worsened in only two (15%) patients, improved in three (23%) patients, and remained stable in seven (54%) patients compared to preoperative MMS. Five (38%) patients required fusion for progressive spinal deformity.
Conclusion Gross total resection of holocord and longitudinally extensive intramedullary spinal cord tumors can be achieved with preservation of long-term neurological function in many cases. Serial imaging is recommended to guide subsequent resection for tumor recurrence and stabilization of progressive spinal deformity.

Keywords Intramedullary spinal cord tumor · Holocord · Pediatric

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Introduction

Tumors of the spinal cord occur with one tenth the incidence of intracranial neoplasms—approximately three to ten per 100,000 population [1]. Although they arise more frequently in the pediatric population and are seldom reported in individuals more than 60 years of age, these

are lesions that affect individuals of all ages. The subset of holocord and longitudinally extensive spinal tumors (lesions involving the majority of the spinal cord) represent a rarer and more challenging condition for which there has been little recommendations and treatment protocols reported in the literature [2–4, 7, 8, 11–15]. Prior studies have almost entirely been case reports limiting the ability to assess surgical outcome [2–4, 7, 8, 11–15]. Advancement in magnetic resonance imaging (MRI) technology, neurophysiologic monitoring, and surgical equipment have greatly enhanced our ability to treat this rare tumor type safely and effectively [2–4, 13]. It is our premise that aggressive surgical resection is an applicable approach to the surgical management of these lesions [4]. In this report, we present our experience with the treatment of 13 patients with holocord and longitudinally extensive tumors and their outcomes.

Materials and methods

All cases of holocord and longitudinally extensive intramedullary spinal cord tumors (lesions involving the majority of the spinal cord) surgically treated at the New York University Hospital between 1990 and 2002 were retrospectively reviewed. The medical, radiological, surgical, pathological, and follow-up clinical records from these patients were reviewed. The medical information analyzed for each patient included age at presentation, symptoms at presentation, duration of symptoms, previous treatment, adjuvant therapy, and immediate and late outcome. The Modified McCormick Score (MMS) [9, 10] was recorded preoperatively, postoperatively, and at last follow-up. During the follow-up period, patients demonstrating continued progression of structural kyphotic or scoliotic curves on radiographs despite external bracing underwent attempted curve correction and instrumented fusion. Patients with stable spinal deformity underwent curve correction and fusion only if accompanied by radicular symptoms, significant back pain, or functional limitation thought to arise from their spinal deformity.

Surgical approach

Patients operated on for the first time underwent an osteoplastic laminotomy with subsequent bone replacement while those with prior laminectomy underwent laminectomy. The surgery was performed in either a single setting or in a two-staged technique, starting in 1999. The goal of staging the procedure was to limit surgeon fatigue and ensure a safe and radical resection of the tumor. Ultrasound aspiration was used to excavate the tumor from the inside outward until its interface with the white matter was reached. Resection was deemed complete once the interface with the white matter was reached or if a sustained decrease in motor evoked potentials greater than 50% of baseline occurred. Ultrasonography was routinely employed before and after resection for localization and to assure the extent of the resection. Gross total removal of the tumor was attempted in all patients. Extent of resection was based solely on comparison of pre- and immediate postoperative MRI studies. Gross total removal was defined as removal of at least 95% of the tumor. Subtotal removal was defined as 80 to 95% resection. Sensory- and motor-evoked potentials were used routinely for all the operations.

Results

Presentation

Thirteen patients (eight male, five female) were included in the study. Mean age was 15 years (range, 3–45) at the time of the initial resection. At the time of diagnosis, all lesions were located in the cervical and thoracic spine. Four (31%) lesions extended down to the first lumbar segment. Tumor spanned a mean±SD 16±2 spinal levels. The characteristics for this patient population are summarized in Table 1. The most common indicators for radiographic imaging were pain, sensory, motor deficits, or gait abnormalities. The most common presenting symptoms were pain and sensory deficits with significant weakness usually evolving much later. Pain was predominately along the spinal axis. The mean preoperative MMS [9, 10] for the study group was 2.4. Before surgery, two patients were classified as MMS grade I, five as grade II, two as grade III, and four as grade IV. Five (38%) patients underwent prior biopsy to guide radiotherapy before being referred to our institution. These patients were referred for further treatment because of clinical or radiographic progression (Fig. 1).

Surgical results

Nine (69%) patients had an osteoplastic laminoplasty performed, and four (31%) had traditional laminectomy as the surgical approach. Gross total resection was achieved in eight cases and subtotal resection in five cases. Pathology revealed astrocytoma in six cases (two pilocytic, four grade II), gangliogliomas in four cases, oligodendroglioma in two cases (one anaplastic), and lipoma in one case. Two (15%) patients had very large tumors, an 8-year-old patient (C3–L1) and a 24-year-old (C1–T12) patient. For these two cases, the surgery was staged, and the cervical resection was done first, followed approximately 2 weeks later by the thoracic resection.

Table 1 Characteristics and outcomes of 13 patients undergoing resection of holocord intramedullary spinal cord tumors

Age, sex	Level	Surgery	Preoperative MMS grade	Postoperative MMS grade	Histology	Follow-up (months)	Outcome	Comments
3, F	C2–T11	Laminoplasty	1	1	LGA	30		
14, M	C4–T9	Laminoplasty	1	1	LGA	150		
4, F	C4–T10	Laminoplasty	3	3	LGA	88	Recurrent disease (C7-T7)	Spinal fusion for deformity
8, M	C3–T12	Laminoplasty Staged	2	3	Oligodendroglioma	9		Preoperative Bx
13, M	C4–L1	Laminoplasty	2	2	JPA	31		Spinal fusion for deformity
11, F	C6–L1	Laminectomy	2	3	JPA	20		Preoperative Bx, spinal fusion for deformity
15, M	C1–L1	Laminoplasty	4	4	Ganglioglioma	26	Recurrent disease (C5-T2)	Spinal fusion for deformity
36, F	C6–L1	Laminectomy	2	1	Lipoma	30		Spinal fusion for deformity
3, M	C3–T10	Laminoplasty	4	4	LGA	13		
45, F	C1–T9	Laminectomy	4	4	Ganglioglioma	32		Preoperative Bx, VP shunt
24, M	C1–T12	Laminoplasty Staged	4	4	Anaplastic Oligodendroglioma	25	Death	Disseminated tumor, death
7, M	C1–T10	Laminectomy	2	1	Ganglioglioma	40		Preoperative Bx, VP shunt
12, M	C1–T8	Laminoplasty	3	2	Ganglioglioma	29		

MMS Modified McCormick Scale

Adjuvant therapy

A total of six patients received radiotherapy as adjuvant treatment for their intramedullary spinal cord tumor. Five (38%) patients underwent prior biopsy and received radiotherapy before presenting to our institution. Only one patient received postoperative radiotherapy after surgery at our institution. This patient was treated for the anaplastic oligodendroglioma. The remaining patients were treated with surgery alone.

Outcome analysis

There was no perioperative mortality. A cerebrospinal fluid leak requiring wound revision surgery occurred in one case. All patients were available for follow-up review with a mean of 3.4 years (range, 1–12) after surgery. Spinal deformity requiring surgical correction occurred in five patients. The incidence of fusion for spinal deformity was similar between the laminectomy and laminoplasty groups (two [50%] vs five [55%]). Recurrence or progression of the tumor occurred in four (31%) patients. Two underwent reoperation (ganglioglioma, grade II astrocytoma), and one received chemotherapy for leptomeningeal spread (anaplastic oligodedroglioma) who eventually died from tumor progression 25 months after surgery. Two patients developed hydrocephalus postoperatively and had ventriculoperitoneal shunts placed. By last follow-up, disease progression was similar in patients receiving gross total vs subtotal resection. The eight patients (100%) who received a gross total resection showed no evidence of tumor progression on MRI. Four patients (80%) undergoing subtotal resection had evidence of residual but stable disease.

With a mean of 3.4 years (range, 1–12) after surgery, MMS [9, 10] had worsened in only two (15%) patients, improved in three (23%) patients, and remained stable in

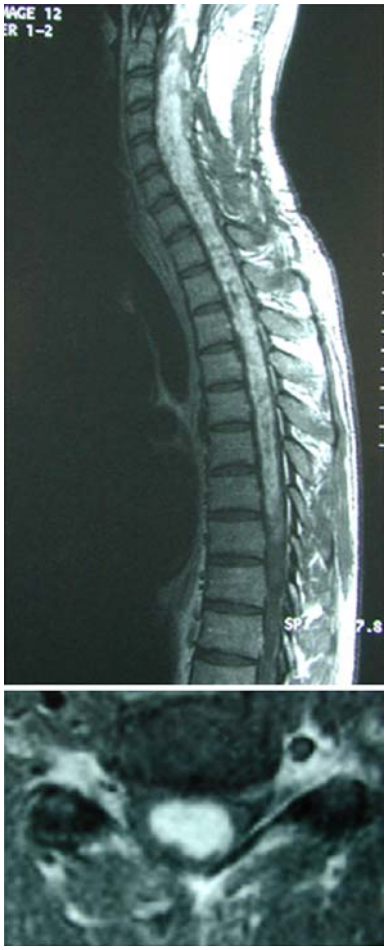

Fig. 1 Sagittal and axial (C5) postcontrast T1 MR images of an intramedullary spinal cord tumor involving 15 spinal levels

seven (54%) patients compared to preoperative MMS [9, 10]. Of the three patients that improved, all had their surgery within 1 year of developing signs and symptoms that led to the diagnosis of their holocord lesion.

Discussion

The management for holocord intramedullary tumors remains debated. Although the majority of these tumors are benign, the optimal treatment for these rare lesions is not yet defined in the literature. The same guidelines established for treating intramedullary tumors remain applicable for holocord tumors [5, 6, 9]. Early treatment remains necessary in preventing further progression of symptoms and possibly improving neurological function for these large and extensive lesions [2–4, 7, 8, 11–15]. However, there are additional challenges that face the surgeon performing the extensive resection required in removing a holocord tumor.

Although only 2 of our 13 patients had staged operations, it is felt that it may be beneficial to stage holocord surgery in the future [2, 4]. Two variables that might encourage the surgeon to opt for staging the resection are a large lesion and/or a large patient. The two cases that were staged in our series were an 8-year-old patient (C3–L1) and a 24-year-old (C1–T12). In general, if more than 15 spinal levels are involved and the patient is not very small in size, staging allows the surgeon to focus on a smaller lesion, which will cut down on anesthesia time, and this may result in a more complete resection. The two operations typically will entail a cervical operation and a thoracic operation.

Follow-up for these patients requires special attention to spinal deformity and instability. This represents an increased source of morbidity for patients with longitudinally extensive tumors, which can be corrected if careful follow-up is maintained postoperatively. In this cohort, five patients ultimately had undergone fusion for spinal deformity by last follow-up, demonstrating the common occurrence of progressive spinal deformity in these patients.

The clinical and functional status of the patients was graded using the modified McCormick classification scheme [9, 10]. Seven patients experienced no change in their grade, three patients improved by one grade, and two patients deteriorated by one grade. Of the three patients that improved, all had their surgery within 1 year of developing signs and symptoms that led to the diagnosis of their holocord lesion. The two patients that deteriorated both underwent surgery more than 1 year after displaying signs and symptoms of an intramedullary lesion and experienced their decline perioperatively.

All patients in the series harbored low-grade tumors with one exception. The patient with the high-grade tumor was diagnosed with an anaplastic oligodendroglioma. This patient ultimately died because of disseminated tumor spread 2 years after surgery. The good long-term tumor control and achievement of gross total resection in the majority of these cases were a result of benign tumor pathology. Complete resection and long-term disease control are far less attainable in high-grade lesions, representing a subset of patients less applicable to our cohort.

Aggressive surgical resection with the goal of complete resection can be achieved for holospinal cord tumors and longitudinally extensive tumors of low-grade pathology. Long-term neurological function can be preserved in the majority of these cases. A two-staged resection in tumors of the holocord is an affective option to limit surgeon fatigue and maximize safe radical resection based on surgeon preference. By 3 years after surgery, stable disease control can be achieved in both cases of gross total and subtotal resection of low-grade tumors. Serial imaging is recommended to guide subsequent resection for tumor recurrence and stabilization of progressive spinal deformity.

References

1. Abbott R, Feldstein N, Wisoff JH, Epstein FJ (1992) Osteoplastic laminotomy in children. Pediatr Neurosurg 18:153–156
2. Benzel EC, Mirfarkhraee M, Hadden T, Fowler M (1987) Holocord astrocytoma: a two-staged operative approach. Spine 12:746–749
3. Chacko AG, Chandy MJ (2000) Favorable outcome after radical excision of a 'Holocord' astrocytoma. Clin Neurol Neurosurg 102:240–242
4. Epstein F, Epstein N (1981) Surgical management of holocord intramedullary spinal cord astrocytomas in children. J Neurosurg 54:829–832
5. Epstein FJ, Farmer JP, Freed D (1992) Adult intramedullary astrocytomas of the spinal cord. J Neurosurg 77:355–359
6. Hoshimaru M, Koyama T, Hashimoto N, Kikuchi H (1999) Results of microsurgical treatment for intramedullary spinal cord ependymomas: analysis of 36 cases. Neurosurgery 44:264–269
7. Irikura T, Johki T, Tanaka H, Nakajima M, Yasue M, Sakai H, Nakamura N (1990) Holocord astrocytoma—case report. Neurol Med Chir (Tokyo) 30:966–971
8. Lau BH, Lin MI, Sung TC, Wei CP, Peng HL, Lee CC (1998) Holocord intramedullary spinal cord astrocytoma: report of one case. Zhonghua Min Guo Xiao Er Ke Yi Xue Hui Za Zhi 39:48–50
9. McCormick PC, Stein BM (1990) Intramedullary tumors in adults. Neurosurg Clin N Am 1:609–630
10. McCormick PC, Torres R, Post KD, Stein BM (1990) Intramedullary ependymoma of the spinal cord. J Neurosurg 72:523–532
11. Pagni CA, Canavero S, Gaidolfi E (1991) Intramedullary "holocord" oligodendroglioma: case report. Acta Neurochir (Wien) 113:96–99
12. Pluchino F, Lodrini S, Lasio G, Allegranza A (1984) Complete removal of holocord subependymoma. Case report. Acta Neurochir (Wien) 73:243–250
13. Razack N, Jimenez OF, Aldana P, Ragheb J (1998) Intramedullary holocord lipoma in an athlete: case report. Neurosurgery 42:394–396 discussion 396–397
14. Seol HJ, Wang KC, Kim SK, Kim H, Choe G, Chi JG, Cho BK (2001) Intramedullary immature teratoma in a young infant involving a long segment of the spinal cord. Child's Nerv Syst 17:758–761
15. Vles JS, Grubben CP, van Ooy A, Weil EH (1990) Holocord astrocytomas in childhood. Clin Neurol Neurosurg 92:361–364