Childs Nerv Syst (2008) 24:735–739
DOI 10.1007/s00381-007-0513-4

ORIGINAL PAPER

Spinal deformity after resection of cervical intramedullary spinal cord tumors in children

Matthew J. McGirt · Kaisorn L. Chaichana · Frank Attenello · Timothy Witham · Ali Bydon · Kevin C. Yao · George I. Jallo

Received: 23 August 2007 / Published online: 30 October 2007

Abstract
Objective Progressive spinal deformity after cervical intramedullary spinal cord tumor (IMSCT) resection requiring subsequent fusion occurs in many cases among pediatric patients. It remains unknown which subgroups of patients represent the greatest risk for progressive spinal deformity.
Materials and methods The data for 58 patients undergoing surgical resection of cervical IMSCT at a single institution were retrospectively collected and analyzed for development of progressive spinal deformity requiring fusion. The association of all clinical, radiographic, and operative variables to subsequent progressive spinal deformity as a function of time was assessed via Kaplan–Meier plots and Log-rank and Cox analyses.
Results Mean age at the time of surgery was 11 ± 6years. Eleven (19%) patients required subsequent fusion for progressive spinal deformity at a median [interquartile range (IQR)] of 4 (2–6) years after IMSCT resection. Five (36%) of 14 patients with preoperative scoliosis or loss of lordosis developed postoperative progressive spinal deformity compared to only 6 (13%) of 44 patients with normal preoperative sagittal and coronal balance, $p = 0.06$. Patients <13years of age were more than three times more likely to develop postoperative progressive deformity, $p = 0.05$. Decompression spanning both the axial cervical spine (C1–C2) and the cervico–thoracic junction (C7–T1) increased the risk for progressive spinal deformity fourfold, $p = 0.04$. Number of spinal levels decompressed, revision surgery, radiotherapy, involvement of C1–C2 or C7–T1 alone in the decompression, or any other recorded variables were not associated with progressive postoperative spinal deformity.
Conclusion Patients possessing one or more of these characteristics should be monitored closely for progressive spinal deformity after surgery.

Keywords Intramedullary · Spinal cord tumor · Deformity · Scoliosis · Kyphosis

Introduction

Intramedullary spinal cord tumors (IMSCTs) account for approximately 35% of all intraspinal tumors in children [2]. Approximately 50% of these tumors are located in the cervical spine [4, 5]. In pediatric patients, cervical IMSCTs, more so than other spinal locations [7], are more commonly associated with postoperative spinal deformity, with rates between 24 and 100% in several series [6, 7, 10, 11]. However, it remains relatively unknown which subgroups of patients with cervical IMSCTs represent the greatest risk for progressive spinal deformity and who may benefit from very close observation and early spinal fusion. We report the single largest study using statistical analyses to evaluate risk factors for progressive spinal deformity after cervical IMSCT resection in pediatric patients.

M. J. McGirt (✉) · K. L. Chaichana · F. Attenello · T. Witham · A. Bydon · G. I. Jallo
Department of Neurosurgery, Johns Hopkins School of Medicine, 3553 Newland Rd., Baltimore, MD 21218, USA
e-mail: mmcgirt1@jhmi.edu

K. C. Yao
Department of Neurosurgery, Tufts-New England Medical Center, Boston, MA, USA

Materials and methods

Fifty-eight consecutive patients with cervical IMSCTs surgically treated at a single institution were reviewed. Their data were retrospectively collected and long-term follow-up was obtained with a focus on the development of progressive spinal deformity. All clinical and radiological variables at presentation were recorded. Patients underwent pre- and postoperative imaging studies in all cases. Each patient's neurological exam was documented at hospital discharge and at a 3-month follow-up visit. For long-term follow-up evaluation, patients and their families were contacted by phone by a physician or trained nurse practitioner and completed a questionnaire assessing symptoms, functional status, and subsequent medical occurrences. The functional status was graded according to a modified McCormick scale (MMS) [12] During the follow-up period, patients demonstrating symptomatic 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 technique

Children undergoing primary or revision surgery underwent laminectomy spanning the length of the tumor in all cases. Only medial facet joint exposure was need in the vast majority of cases, and an effort was made to preserve the facet joint capsule in all cases. Facetectomy was not performed in any cases. Ultrasonic aspiration was invariably used to excavate the tumor from the inside outward until its interface with the white matter was reached. Sensory-evoked and motor-evoked potentials were used. Resection was deemed complete once the interface with the white matter was reached or halted if a sustained decrease in motor evoked potentials >50% of baseline occurred. Gross total removal of the tumor was attempted in most patients. Cases with no residual contrast enhancement on postoperative magnetic resonance imaging (MRI) were classified as gross total resection. The resection was defined as subtotal if residual contrast enhancement was noted on immediate postoperative MR image.

Statistical analysis

For inter-group comparison, the Student's t test was used for parametric data, and the Mann–Whitney U test was used for non-parametric data. Percentages were compared via chi-squared tests and the Fisher exact test when indicated. The association of all recorded variables to progressive postoperative spinal deformity requiring fusion was assessed via Kaplan–Meier plots and log-rank analysis for categorical variables and Cox survival analysis for continuous variables (Statview, SAS, 1998).

Results

Patient population

Fifty-eight patients underwent surgery for a cervical IMSCT during the reviewed period and were followed for a median [interquartile range (IQR)] of 6 (2–9) years. Presenting demographics, operative, and radiographic variables are summa-

Table 1 Summary of demographics, presenting symptoms and signs, radiographic, and operative variables in 58 consecutive pediatric patients undergoing resection of an intramedullary spinal cord tumor

Demographics	Number (%)
Male	38 (66%)
Age (mean±SD)	11±6.2
Age <13	30 (52%)
Symptoms	
Symptoms <1 month	9 (16%)
Symptoms >12 months	12 (21%)
Unilateral symptoms	17 (29%)
Upper extremity symptoms only	14 (24%)
Urinary symptoms	11 (19%)
Sensory symptoms	39 (67%)
PreOp MMS, median (interquartile range)	2 (2.3)
PreOp Dependent Function	14 (24%)
PostOp MMS, median (interquartile range)	2 (2.3)
Loss of cervical lordosis or scoliosis (Cobb>10°)	14 (24%)
Syrinx	44 (76%)
Operative	
Laminectomy Levels	7±3
Laminectomy extends to C2 or C1	26 (45%)
Laminectomy stops at C7–T1 junction	9 (16%)
Laminectomy crosses C7–T1 junction	31 (53%)
Laminectomy extends to C2/C1 and crosses C7–T1 junction	6 (10%)
Prior resection	11 (19%)
Gross total resection	42 (72%)
Subtotal resection	14 (24%)
Biopsy	2 (3%)
PostOp radiotherapy	26 (45%)
PostOp chemotherapy	12 (21%)
Pathology	
Astrocytoma	29 (45%)
Ganglioglioma	15 (26%)
Ependymoma	8 (14%)
Ganglioneurocytoma	6 (10%)
Follow-up	
Tumor recurrence	16 (28%)
Time to tumor recurrence (mean±SD, months)	48±24
MMS score at 3 months PostOp	3 (2,4)

MMS Modified McCormick Score

rized in Table 1. Mean age at the time of tumor resection surgery was 11 ± 6years. Eleven (19%) patients had undergone prior surgery for their IMSCT. The median (IQR) preoperative MMS was 2 (2–3). Fourteen (24%) patients presented with loss of cervical lordosis or scoliosis (Cobb angle >10°).

The mean ± SD number of laminectomy levels was 7 ± 3. The axial cervical spine (C1 or C2) was included in the laminectomy in 26 (45%) cases. Laminectomy crossed the cervico–thoracic junction (C7–T1) in 31 (53%) cases. The cervico–thoracic junction and axial cervical spine were both surgically decompressed in 6 (10%) cases. Gross total resection was achieved in 42 (72%) patients.

Incidence of progressive spinal deformity

Eleven (19%) patients required subsequent fusion for progressive postoperative spinal deformity at a median (IQR) of 4 (2–6) years after IMSCT resection, Fig. 1. Five (36%) of 14 patients with preoperative scoliosis (Cobb angle >10°) or loss of lordosis developed progressive postoperative spinal deformity compared to only 6 (13%) of 44 patients with normal preoperative sagittal and coronal balance, $p = 0.06$, Fig. 2a. In patients with scoliosis or loss of lordosis, the deformity resolved postoperatively in two (14%) cases and remained stable in seven (50%) cases. Patients <13 years of age at the time of IMSCT resection were over three times more likely to develop postoperative progressive deformity [relative risk(95% CI); 3.86(1.0–5.4), p=0.05)], Fig. 2b. Surgical decompression of both the axial cervical spine (C1 or C2) and the cervico–thoracic junction was associated with a fourfold increased likelihood of subsequent progressive deformity [relative risk(95% CI); 4.27(4.3–21.2), p=0.04)], Fig. 2c. Number of spinal levels decompressed, revision surgery, radiotherapy, or involvement of C1–C2 or C7–T1 alone in the decompression was not associated with postoperative deformity. No other variables were associated with postoperative progressive spinal deformity.

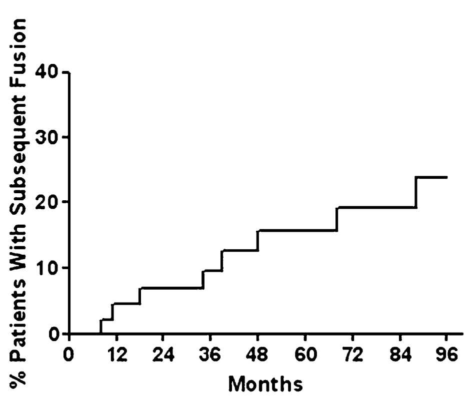

Fig. 1 Overall incidence of progressive spinal deformity requiring fusion as a function of time after surgical resection of cervical intramedullary spinal cord tumor

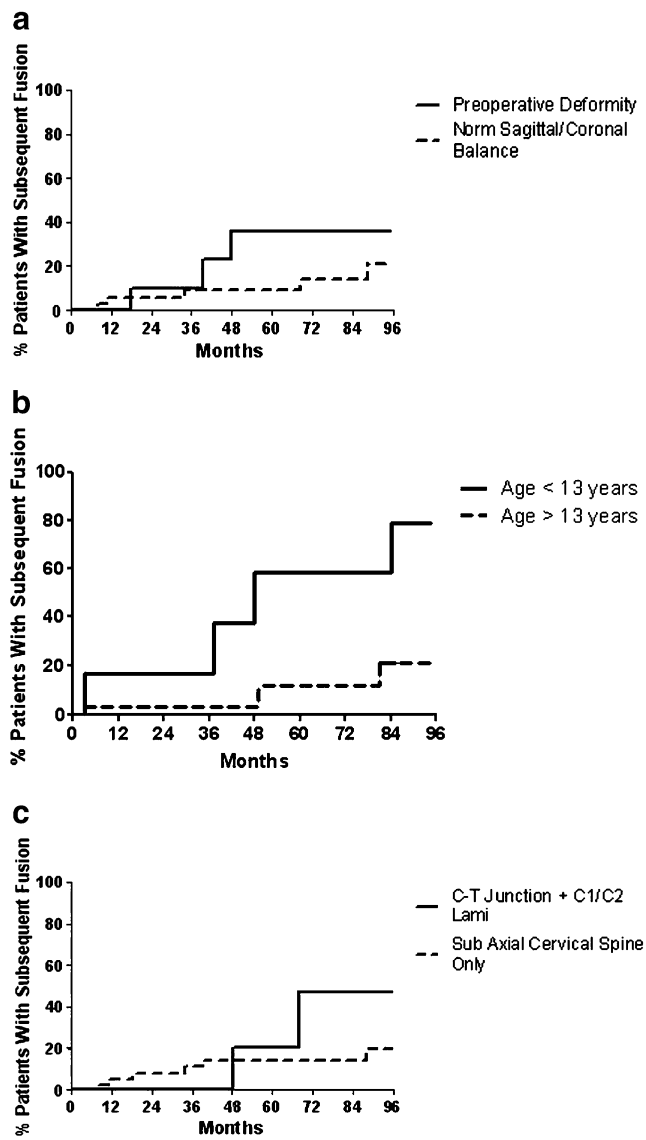

Fig. 2 Estimated Kaplan–Meier plots of the incidence of progressive spinal deformity requiring fusion after resection of intramedullary spinal cord tumors. **a** Patients with either preoperative loss of lordosis or scoliosis (Cobb angle >10°) had a higher incidence of progressive spinal deformity. **b** Patients <13 years of age were more likely to experience progressive postoperative deformity. **c** Surgical laminectomy involving both the axial cervical spine (C1 or C2) and the cervico–thoracic junction were more likely to experience progressive postoperative deformity

Discussion

In this study, we identified variables predictive of progressive spinal deformity requiring fusion in pediatric patients after cervical IMSCT resection. Age less than 13 years at the time of surgery, presence of preoperative scoliosis or loss of lordosis, and decompression spanning both the axial cervical spine (C1–C2) and the cervico–thoracic junction (C7–T1) increased the risk for progressive spinal deformity requiring fusion. The number of spinal levels decompressed, revision surgery, radiotherapy, involvement of C1–C2 or

C7–T1 alone in the decompression, or any other recorded variables were not associated with progressive postoperative spinal deformity.

Spinal deformity after laminectomy procedures is relatively common in the pediatric population. In children, postlaminectomy deformities can occur at any spinal level but are more common in the cervical spine [6, 16, 17]. Bell et al. [3] reported that 46 of 89 (52%) children with Chiari malformations developed spinal deformity after cervical laminectomy. De Jonge et al. [6] documented that 21 of 21 patients (100%) undergoing cervical laminectomy for either intradural or extradural tumors developed postoperative deformity. In addition, Yeh et al. [18] reported that 3 of 12 patients (25%) with cervical IMSCTs developed postoperative deformity after a mean follow-up period of 45 months, which is similar to our study. Despite these studies, little is known about the risk factors associated with spinal deformity after cervical IMSCT resection in children. Small cohort sizes and short follow-up times have prevented more accurate statistical analyses. It, therefore, remains relatively unknown which subgroups of patients with cervical IMSCTs represent the greatest risk for progressive spinal deformity and who may benefit from close observation and early spinal fusion.

Age appears to be a contributing factor to the development of postoperative spinal deformity. The reason for this is likely multifactorial [6, 7, 10, 11]. The facet complexes in the cervical spine have a more horizontal orientation in children as compared to a more vertical orientation in adults [7]. Children also have more lax ligamentous structures and a greater proportion of cartilage in their spine [7]. These features, in combination with the growing spine, accentuate the altered spinal biomechanics caused by IMSCT resection [7, 14] and likely underlie the increased incidence of spinal deformity observed in children <13 years of age in our study.

The presence of preoperative scoliosis or loss of lordosis was also associated with the development of postoperative cervical deformity. This has been reported by other studies as well [9, 10]. Kaptain et al. [9] reported that the risk of postoperative deformity approximately doubled when the spine was deformed preoperatively in adult patients undergoing laminectomy for cervical myelopathy. It is likely that preoperative deformity leads to asymmetric loading on the spine, which results in documented alteration in the composition of the vertebral bodies, vertebral discs, and paravertebral musculature [8, 13, 15]. These irreversible changes consequently contribute to the progressive deformity after surgical resection [8, 13, 15].

The location of the surgery also appears to be associated with postoperative deformity. Decompression spanning both the axial cervical spine (C1–C2) and the cervico–thoracic junction (C7–T1) increased the risk for progressive spinal deformity requiring fusion. Previous studies have documented that patients who undergo laminectomy at the craniocervical junction for both Chiari malformations [1] and spinal (intradural or extradural) tumors [10] were at increased risk of developing spinal deformity. Surgery at these high stress areas may predispose to postoperative deformity by impairing the spine's ability to withstand the stress associated with cervical motion [7].

It is noteworthy that only 11 of 58 patients (19%) required fusion. This is consistent with prior studies [4, 5, 18]. As the majority of cases did not require fusion and fusion of the growing spine is more prone to failure and limits the use of MRI imaging, fusion at the time of tumor resection is often not warranted [7]. According to this study, the most dramatic risk factors for postoperative deformity are age <13 years, presence of preoperative deformity, and surgery involving the craniocervical and cervicothoracic junction. Patients that possess all of these factors may warrant fusion but at the very least require close monitoring for the development of progressive deformity.

Conclusions

The incidence of cervical deformity in the pediatric population after IMSCT resection is relatively common. The high incidence and associated morbidity necessitate understanding which factors are associated with progressive spinal deformity. This study is the largest of its kind to statistically analyze the factors associated with spinal deformity after cervical IMSCT resection in children. The factors that significantly contributed to postoperative deformity were patient age <13 years, presence of preoperative scoliosis or loss of lordosis, and/or surgery involving the axial cervical spine (C1–C2) and the cervico–thoracic junction (C7-T1). Patients possessing one or more of these characteristics should be monitored closely for progressive spinal deformity after surgery.

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