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ORIGINAL PAPER

Neurological outcome after resection of intramedullary spinal cord tumors in children

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Abstract
Objective With modern surgical advances, radical resection of pediatric intramedullary spinal cord tumors (IMSCT) can be achieved with preservation of long-term neurological function. Clinical and radiographic risk factors predictive of postoperative neurological outcome may serve as a guide for surgical risk stratification.
Materials and methods We prospectively reviewed the outcomes of 16 consecutive cases of pediatric IMSCT resection at a single institution. Clinical, radiographic, and operative variables were analyzed as predictors of postoperative neurological function defined by the modified McCormick score (MMS).
Results Sixteen children 10±5 years old presented with median (interquartile range) MMS score of 2 (1–2) with IMSCTs (eight cervical, eight thoracic) involving 4±2 levels. Pathology revealed astrocytoma in 12 cases (three pilocytic, four grade II, three gradeIII, two GBM), gangliogliomas in two, ependymoma in one, and gliosis in one case. At median follow-up of 7 months, six (38%) patients experienced improved neurological function, eight (50%) remained stable, one (6%) experienced a delayed decrease in neurological function (GBM progression), and one (6%) died (GBM progression). Five (31%) patients developed persistent dysesthetic symptoms. Four (80%) patients with cystic tumors experienced neurological improvement compared to only two (18%) patients with noncystic tumors, $p<0.05$. Preoperative steroid use (odds ratio, OR [95% confidence interval, CI]=18.0 [1.24–260.1], $p=0.03$) and cystic tumor (OR [95%CI]=18.0 [1.24–260.1], $p=0.03$) predicted neurological improvement after surgery.
Conclusion Radical resection of pediatric IMSCTs can be achieved with low incidence of neurological injury. Sensory syndromes frequently occur after pediatric IMSCT resection and frequently affect patient's quality of life. Tumors with compressive cysts may identify patients more likely to experience improved neurological function after surgical resection.

Keywords Intramedullary spinal cord tumor · Outcome · Pediatric · Predictors

Introduction

Intramedullary spinal cord tumors (IMSCTs) are relatively rare tumors that account for 4–10% of central nervous system tumors but account for approximately 35% of pediatric intraspinal tumors [1, 2, 26]. Conventionally, conservative management consisted of biopsy, dural decompression, and radiation therapy. This approach avoided sequelae associated with iatrogenic injury to adjacent neurological tissue but yielded only moderate improvement in disease progression and survival. Recent advances in microsurgical techniques, operative equipment, neuroimaging, and intraoperative neurophysiology have made more aggressive resection of IMSCT feasible [3, 4, 9, 10, 18]. In

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fact, this approach has been associated with increased long-term survival and improved quality of life [3, 4, 15, 17, 25].

While these developments now allow radical resection in the majority of cases, overall morbidity rates remain high [5, 23]. Many studies have examined prognostic factors associated with functional outcome after IMSCT resection in adults [5, 11, 15, 23, 25]. However, studies in the pediatric population are sparse. We set out to identify preoperative variables that may identify patients most likely to experience improvement in neurological deficits with surgical resection of IMSCTs.

Materials and methods

Sixteen consecutive pediatric patients undergoing surgery for IMSCTs at a single academic institution were prospectively reviewed. Demographics, clinical presentation, comorbidities, preoperative magnetic resonance imaging (MRI) characteristics, intraoperative records, pathological data, and neurological function at discharge and at clinic follow-up were recorded. Pre- and postoperative neurological function was defined by the modified McCormick score [21]. Preoperative steroid use was defined as oral dexamethasone given several days before surgery. Radical resection was defined as removal of at least 95% of the tumor based on comparison of the pre- and immediate postoperative MRI. The resection was defined as subtotal removal if 80 to 95% of the tumor was resected. Improvement in neurological function was defined as both (1) an improvement in deficit on physical exam and (2) patients and their families described a subjective improvement in symptoms. Sensory discomfort arising from burning, tingling, or hyperesthesia was defined as dysesthetic symptoms and documented if they affected quality of life according to patients and their families. A tumor-associated cyst was defined as an eccentrically located cavitary lesion within the tumor mass (Fig. 1). A tumor-associated syrinx was defined as a fluid-filled cavity extending centrally within the spinal cord rostral and/or caudal to the tumor (Fig. 1).

Surgical technique

All patients underwent osteoplastic laminotomy for access to the IMSCT. Only medial facet joint exposure was performed, and an effort was made to preserve the facet joint capsules in all cases. The interspinous ligaments at the rostral-most and caudal-most levels of the laminoplasty segment were removed with a Leksell rongeur. Bilateral laminotomies were then made at a width equal to the spinal canal in a caudal-to-rostral direction spanning the entire

Fig. 1 Mid-sagittal T1 postcontrast MRI demonstrating an intramedullary spinal cord tumor with both tumor-associated cysts and syringomyelia. Syringomyelia can be seen immediately caudal to the enhancing tumor. The paracentric tumor associated cysts can be seen both within and immediately rostral to the enhancing tumor

laminoplasty segment utilizing the osteotome drill with a footplate. The tumor was approached and removed via a midline dural incision spanning the entire length of the tumor in all cases. Sensory-evoked and motor-evoked potentials were used. Resection was deemed complete once interface with the white matter was reached or if a sustained decrease in motor-evoked potentials greater than 50% of the baseline occurred. Tumor-associated syrinx were not exposed or surgically manipulated in any case because of frequent resolution after tumor resection alone. After tumor resection and primary dural closure, the laminae were reapproximated and attached with titanium microplates. Instrumented or in situ fusion was not utilized in any case.

Statistical analysis

For intergroup comparison, the Student's *t* test was used for parametric data, and the Mann–Whitney *U* test was used for nonparametric data. Percentages were compared via chi-squared tests or the Fisher exact test where appropriate. The univariate association of all recorded variables to postoperative neurological improvement was assessed via logistic regression analysis (Statview, [24]).

Results

Patient population

Sixteen patients underwent resection of an IMSCT. Mean± standard deviation (SD) age was 10±5 years, and nine

(56%) patients were male. The mean duration of symptoms was 8 months (range 1–36) before resection. The median (interquartile range, IQR) McCormick grade at presentation was 2 (1–2). Eight (50%) patients presented primarily with motor symptoms while eight (50%) presented primarily with sensory symptoms. One (6%) and two (12%) patients received preoperative chemotherapy and radiotherapy before surgery, respectively. Five (31%) patients were started on oral decadron at least 1 week before surgery. The spinal location of the IMSCT was primarily cervical in eight (50%) patients and primarily thoracic in eight (50%) patients, and the location crossed the cervicothoracic junction in four (25%) patients. Tumor spanned 4±2 (mean±SD) spinal levels and was associated with a syrinx in five (31%) cases. A tumor associated cyst was evident on preoperative MRI in five (31%) cases. Radical resection (>95%) was achieved in ten (72%) of non-glioblastoma multiforme (GBM) cases. Subtotal resection (80–95%) was achieved in four (28%) of the non-GBM cases and in both cases of GBM. Surgical pathology revealed astrocytoma in 12 cases (three pilocytic, four grade II, three grade III, two GBM), gangliogliomas in two cases, ependymoma in one case, and gliosis in one case.

Outcome

At median (IQR) follow-up of 7 months (1–24), the median (IQR) modified McCormick grade [21] was 1 (1–2). Six (38%) patients experienced improved neurological function, eight (50%) remained stable, one (6%) experienced a delayed decrease in neurological function (GBM progression), and one (6%) died (GBM progression). No patient experienced an acute decline in neurological function immediately after IMSCT resection. Five (31%) patients developed persistent dysesthetic symptoms that felt to affect quality of life by last follow-up.

Four (80%) patients with cystic tumors experienced neurological improvement compared to only two (18%) patients with noncystic tumors, $p<0.05$, Fig. 1. Preoperative steroid use (odds ratio, OR [95% confidence interval, CI]=18.0 [1.24–260.1], $p=0.03$) and cystic tumor (OR [95%CI]=18.0 [1.24–260.1], $p=0.03$) were associated with neurological improvement after surgery. All other clinical and radiographic variables (age, gender, duration symptoms, tumor size, location or pathology, presence of syrinx, degree of resection, radiotherapy) were not associated with neurological improvement at last follow-up.

Discussion

In this study, we identified clinical and radiographic variables predictive of neurological improvement in 16 consecutive pediatric patients undergoing IMSCT resection. These variables remain relatively uninvestigated and may serve as a guide for surgical risk stratification. For non-GBM cases, more than 40% of patients experienced improvement in their neurological deficits by a median of 7 months after surgery, while neurological function remained stable in all other patients. Preoperative steroid use and the presence of tumor-associated cysts on MRI each markedly increased the likelihood of neurological improvement with surgical resection.

In the adult IMSCT literature, several studies have attempted to determine predictors of neurological outcomes after IMSCT resection. Intramedullary ependymomas, which primarily occur in adults, are associated with better neurological outcomes compared to intramedullary astrocytomas [5, 13]. The use of intraoperative monitoring and postoperative radiation therapy have also been linked to improved morbidity, while the presence of arachnoid scarring [14], spinal atrophy [14], and solid tumor components [6] have been associated with increased morbidity. These studies, however, involve adult patients, and factors associated with neurological outcome in the pediatric population remain largely unstudied.

IMSCTs in pediatric patients are inherently different than in adults. Histologically, infiltrating astrocytomas are the most common type of tumor in children, as opposed to adults who more commonly have intramedullary ependymomas [3, 4, 15, 16, 25]. Astrocytomas are often eccentrically located and, as a result, cause an asymmetric enlargement of the spinal cord [16]. The pediatric spine is also more susceptible to postoperative deformity because of lax ligamentous structures, horizontal orientation of the facet complexes, and the dynamic growth of the immature spine [12, 22, 28]. Furthermore, children more often present with insidious, nonspecific symptoms of diffuse and non-localized pain, motor deficits, and, in young infants, motor regression [3, 4, 16]. Adults, however, typically present with sensory deficits, including dysesthesia, extremity numbness, and myelopathy [10, 15, 25]. These intrinsic differences between IMSCTs in adults and children highlight the importance of identifying outcome predictors solely for pediatric IMSCTs to draw conclusions most applicable to pediatric patients.

The overwhelming majority of patients in this study had improved or maintained neurological function after IMSCT resection at last follow-up. The two patients who worsened neurologically had GBM and deteriorated secondary to tumor progression. These results are similar to previous pediatric studies [3, 4]. Constantini et al. [4] reported that approximately 75% of the 164 children and young adults had improved or maintained neurological function at last follow-up. In a separate study, Epstein et al. documented that 25 of 27 patients under 3 years of age had improved or

maintained neurological function after IMSCT resection. Therefore, this study and others support the finding that IMSCT can be safely resected in pediatric patients and often lead to improved and/or maintained neurological function at last follow-up [3, 4].

Patients who were administered preoperative dexamethasone several days before surgery in this study were more likely to improve their neurological function at last follow-up. Steroids presumably function by decreasing both tumor-associated inflammation and spinal cord edema, hypothetically making tumors more amenable to resection and decreasing the chance that surrounding neurological tissue would be damaged [7, 8, 27]. Steroids have been shown to decrease surgical pain, length of stay, and improve functional outcome after routine spine surgery [19, 20]. Furthermore, steroids improve neurologic outcomes after surgery for metastatic spinal cord compression [7, 8, 27]. The findings in our study also support the use of steroids in the preoperative period.

Finally, patients who possess IMSCTs with associated macroscopic cysts were more likely to have improved neurological outcomes after surgical resection. Prior studies have noted the presence of cysts but do not address their predictive value or effect on neurological outcomes [3, 4]. Nevertheless, the detection of cysts on preoperative neuroimaging may indicate a better likelihood of improved outcomes. This may be due to the fact that cysts typically occur at the tumor–spinal cord interface allowing safer tumor margin identification and resection [18]. Cystic components of IMSCT may also contribute to compressive spinal cord injury that may be acutely relieved at the time of decompression and resection. Decompression of compressed adjacent spinal cord not yet infiltrated with the tumor may underlie the greater improvement observed in patients with intratumor cysts.

Conclusion

Prior studies on IMSCTs primarily involve adult patients. Studies that exclusively focus on pediatric patients are limited, although IMSCTs in pediatric patients are distinctly different than adults. This study supports the finding that IMSCTs can be safely and radically resected in the pediatric population, with the slight majority experiencing improvement in deficits. Tumors with compressive cysts may identify patients more likely to experience improved neurological function after surgical resection.

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