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Original Article

Clinical Characteristics and Treatment Outcomes of Long-Level Intramedullary Spinal Cord Tumors: A Consecutive Series of 43 Cases

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Objective: Long-level intramedullary spinal cord tumors (LIMSCTs) cause complex treatment issues. However, LIMSCTs have rarely been analyzed separately. The authors reported a large case series of LIMSCTs and analyzed the clinical characteristics and treatment outcomes.

Methods: The medical data of patients with LIMSCTs at our institution between January 2015 and December 2019 were retrospectively reviewed. Demographics, tumor size and location, pathology, extent of resection, and neurological functional status were collected.

Results: A total of 43 consecutive cases were included. Twenty-three cases (53.5%) of LIMSCTs were ependymal tumors. All patients with ependymal tumors achieved gross total resection (GTR). In ependymal tumor cases, 3 cases (13%) of ependymal tumors experienced postoperative neurological deterioration, and 66% of them showed an improvement at follow-up; 25.6% were low-grade astrocytic tumors. The rates of GTR, subtotal resection (STR) and partial resection (PR) were 63.6%, 27.3%, and 9.1%, respectively. Twenty-seven percent cases showed postoperative neurological worsening, and 33% of them had an improvement at follow-up; 20.9% were high-grade astrocytic tumors. The excision rates were 44.4% for GTR, 44.4% for STR, and 11% for PR, respectively. Fifty-five percent cases showed postoperative neurological worsening, and none of them had an improvement at follow-up.

Conclusion: In this series, all LIMSCTs were gliomas. Aggressive tumor resection did not increase the risk of long-term functional deterioration in ependymal tumors and low-grade astrocytic tumors, but in high-grade astrocytic tumors, patients had a higher risk of neurological deterioration and difficulty in recovery. In ependymal tumors and low-grade astrocytic tumors, patients can achieve long-time survival after performing aggressive tumor resection.

Keywords: Intramedullary tumors, Long level, Ependymoma, Astrocytoma, Outcome

INTRODUCTION

Long-level intramedullary spinal cord tumors (LIMSCTs) are defined as tumors that involve at least 5 spinal vertebral segments.[1] Generally, most intramedullary tumors present clinically due to nerve compression rather than tumor invasion.[2] In rarer cases, intramedullary tumors can be long-segmental localized. Long-level lesions gradually progress neurologically. Patients may suffer from weakness, sense disorders, and sphincter defects and eventually become significantly disabled, morbidly ill, and even die.

Given that patients with long-level intramedullary lesions suffer more neurological deterioration and postoperative complications, some surgeons may choose conservative manage-

ment.[3,4] These conservative protocols cannot prevent disease progression.[5,6] In addition, several studies have also noted that aggressive resection increases the overall survival of patients with intramedullary tumors, and the earlier the radical resection of intramedullary tumors is performed, the greater the chance of preserving the patient's neurological function.[7] To date, the optimal management for LIMSCTs remains unclear.

In this study, we reported our experience with LIMSCTs, which has rarely been discussed separately in previous literature.[8,9] Based on a series of 43 patients treated in our institution, we analyzed the tumor characteristics, extent of resection and clinical outcomes and attempted to provide answers on optimal treatment strategies for LIMSCTs.

MATERIALS AND METHODS

1. Study Patients

This study was a retrospective analysis of data from surgical cases who underwent microsurgical LIMSCT resection at Beijing Sanbo Brain Hospital between January 2015 and December 2019. The collective patient databases were analyzed to determine cases of intramedullary tumors in which the tumor involved at least 5 spinal vertebral levels or longer. Patients with terminal filum tumors or cauda equina tumors were excluded. Data were collected from our computerized database, follow-up appointments and telephone interviews.

A total of 43 cases were included in this analysis. Patient data, including age, sex, symptoms, tumor size and location, pathology, extent of resection, neurological functional status, and follow-up outcomes, were recorded. We used the modified McCormick Scale[10] (MMS) to evaluate the neurological function of each patient at the first presentation, postoperation, and follow-up visits. The location of each LIMSCT was categorized as cervical, cervicothoracic (tumor that involved C7 and T1 level), thoracic, thoracolumbar (tumor that involved T12 and L1 level), and holocord involvement (tumor involved cervical, thoracic, and lumbar levels).

This study protocol was approved by the University Institutional Review Board (SBNK-YJ-2020-006-03). Informed written consent was obtained from all participants.

2. Histological Evaluation and Pathological Findings

All histological specimens were reviewed by senior neuropathologists at our institution. Specimens were independently reviewed by one neuropathologist and then examined by another to confirm the diagnosis. We used the World Health Organization (WHO) 2016 classification of central neural system tumors for diagnosis and classification.[11] For patients who underwent operation before 2016, tumor specimens were assessed according to the same classification system to maintain unified histological grading criteria.

3. Surgical Technique

Our operative technique has been described previously in detail.[12] Laminoplasty that spanned the length of the intramedullary lesion was performed in all patients. A C1 laminectomy was performed for tumor sections at the C1 level. In each case, the medial facet joint was exposed, and an effort was made to maintain the facet joint capsules. Bilateral laminectomies were performed using an ultrasonic bone scalpel. The spinous processes, interspinous ligaments, and ligamentum flavum of the planned laminoplasty section were kept intact. Using intraoperative neuro-monitor mapping, a midline dural and spinal cord incision was performed spanning the whole length of the tumor. The tumor was approached gently and resected piecemeal. After removing the tumor, the dura was closed via 5-0 absorbable sutures. Mini titanium plates were used to fix the laminae. Finally, the paraspinal muscles were sutured, and the skin incision was closed. In addition, 3 patients who suffered from LIMSCT complicated with scoliosis were treated by a 1-stage operation of tumor resection and scoliosis correction. Gross total resection (GTR) was attempted in all patients. GTR is defined as excision of ≥95% of the tumor, as evidenced by a clean operative field at the end of the surgery or absence of residual enhancement signal on postoperative magnetic resonance imaging (MRI).[5,12,13] The procedure was considered a subtotal resection (STR, 80%–95% resection) if a small tumor piece was deliberately left in place due to intraoperative conditions or a retained fragment was detected on postoperative MRI. Partial resection (PR) was defined as less than 80% tumor removal, which was rare in this case series. Intraoperative neurophysiological monitoring, including motor evoked potentials (MEPs) and somatosensory evoked potentials (SEP), was performed in all patients. The warning criteria were set for a 50% amplitude reduction of SEP or a 50% amplitude reduction of MEP. The permanent 75% amplitude reduction of MEP was used to stop the operation.[14-16]

4. Statistical Analysis

Statistical analyses were performed using GraphPad Prism ver. 8.0 (GraphPad Software Inc., La Jolla, CA, USA). Percentages were calculated for categorical data, and medians and in-

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terquartile ranges (IQRs) were calculated for continuous data. The Mann-Whitney test was used to compare nonparametric data between the groups. For percentage analysis, the chi-square test was used. Kaplan-Meier analysis for overall survival was performed with log-rank tests. The differences were considered statistically significant if the p-value was <0.05.

RESULTS

1. Demographic and Clinical Characteristics

A total of 43 consecutive cases underwent LIMSCT resection in the study period (Table 1). Twelve of them (27.9%) were pediatric patients (age ≤18 years), whereas 31 patients (72.1%) were adults. Nineteen patients (44.2%) were female, and 24 patients (55.8%) were male.

The most common presenting symptoms in this series were combined symptoms (65.1%) followed by sensory dysfunction (20.9%) and motor deficits (14.0%). In 19 cases (44.2%), the prodrome time persisted for ≤1 year from the initial symptomatology to surgical treatment. In 13 patients (30.2%), the prodrome time lasted 1 to 3 years. In 11 patients (25.6%), the prodrome time lasted >3 years. The major sensory symptoms included pain, extremity numbness, and dysesthesia. Major motor deficits included extremity weakness, motor loss and muscle atrophy. The median preoperative MMS score in our series was 2 (IQR, 2–3).

2. Tumor Location, Resection, and Pathology

Based on preoperative imaging, patients most commonly presented with a lesion that involved the cervicothoracic spine. Thirteen tumors (30.2%) were located in the cervical spine, 21 tumors (44.8%) were located in the cervicothoracic spine, 4 tumors (9.3%) were located in the thoracic spine, 4 tumors (9.3%) were restricted to the thoracolumbar spine, and 1 tumor (2.3%) had a holocord tumor (Fig. 1A).

In total, 23 cases (53.5%) were ependymal tumors, and 20 cases (46.5%) were astrocytic tumors. The detailed distribution of histopathological diagnosis was subependymoma in 2 cases (4.6%), ependymoma in 21 cases (48.8%), pilocytic astrocytoma (PA) in 3 cases (7%), diffuse astrocytoma in 8 cases (18.6%), anaplastic astrocytoma in 2 cases (4.6%), glioblastoma in 3 cases (7%), and diffuse midline glioma in 4 cases (9.3%) (Fig. 1B, C). Overall, GTR was achieved in 34 cases (79.1%), and STR was achieved in 7 cases (16.3%). Only 2 patients (4.6%) underwent PR. All patients with ependymoma achieved GTR. The rates of GTR, STR, and PR of low-grade astrocytomas (WHO grade I and II) were 63.6%, 27.3%, and 9.1%, respectively. The excision rates for high-grade astrocytomas (WHO grade III and IV) were 44.4% for GTR, 44.4% for STR, and 11% for PR. High-grade tumors had a significantly lower possibility of achieving complete tumor resection than low-grade tumors ($p=0.004$).

Table 1. Summary of characteristic of patients with LIMSCTs

Characteristic	Value
Age	
Pediatric (≤18 yr)	12 (27.9)
Adult (>18 yr)	31 (72.1)
Sex	
Male	24 (55.8)
Female	19 (44.2)
Clinical symptom	
Sensory	9 (20.9)
Motor	6 (14.0)
Combined	28 (65.1)
Symptom duration in years	
≤1	19 (44.2)
1–3	13 (30.2)
>3	11 (25.6)
Classification of tumor (extent of resection)	
Ependymal tumor WHO I and II (GTR 100%)	23 (53.5)
Astrocytic tumor WHO I and II (GTR 63.6%, STR 27.3%, PR 9.1%)	11 (25.6)
Astrocytic tumor WHO III and IV (GTR 44.4%, STR 44.4%, PR 11.1%)	9 (20.9)
Extent of tumor involvement (spinal levels)	
5	16 (37.2)
6–8	23 (53.5)
11–13	3 (7.0)
Holocord	1 (2.3)
Preoperative MMS score	2 (2–3)
Length of stay in days	20 (17–25)
Postoperative adjuvant therapy	
Chemotherapy	3 (7.0)
Radiotherapy	4 (9.3)
Combined	4 (9.3)
Postoperative MMS score	3 (2–3)
Follow-up length in months	34 (25–60)

Values are presented as number (%) or median (interquartile range).
LIMSCT, long-level intramedullary spinal cord tumor; WHO, World Health Organization; GTR, gross total resection; STR, subtotal resection; PR, partial resection; MMS, modified McCormick Scale.

3. Surgical-Related Outcomes and Neurological Function

No patient died because of operative-related procedures. The median length of hospitalization was 20 days (IQR, 17–25 days). As reported in previous studies,[12] the modified MMS was used to best express the change in operation-induced neurological function. Overall, most patients (27 cases, 62.8%) had only a minor neurological deficit (MMS II) at presentation. In contrast, 8 cases (18.6%) had MMS III, and 5 cases (11.6%) had MMS IV. The median preoperative MMS score was 2 (IQR, 2–3), and the postoperative MMS score was 3 (IQR, 2–3). Overall, no significant difference was noted between the pre- and postoperative MMS scores (p = 0.1067). In ependymal tumor cases, 3 cases experienced postoperative neurological deterioration, and 2 of them showed an improvement at the last follow-up. Of the low-grade astrocytic tumor cases, 3 cases showed postoperative worsening in their neurological status, and one of them had an improved grade at the last follow-up. Of the high-grade astrocytic tumor cases, 5 cases experienced postoperative functional worsening. Unfortunately, functional improvement was not observed in those patients. No further

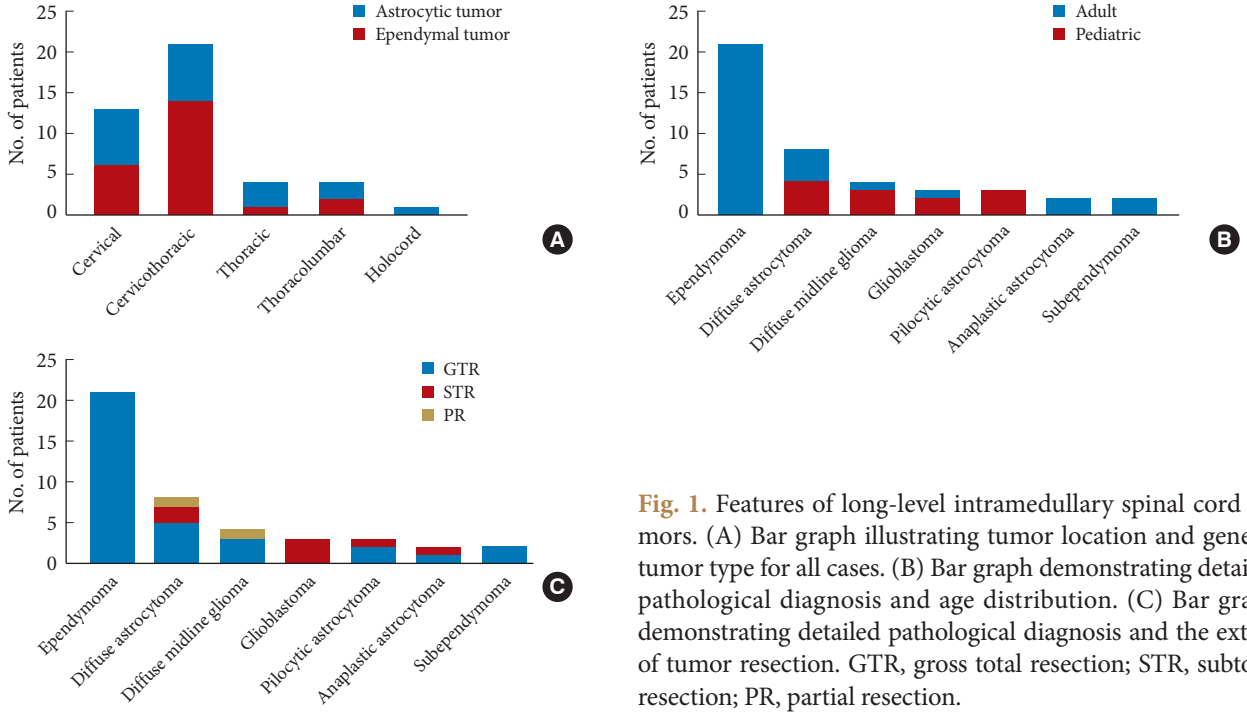

Fig. 1. Features of long-level intramedullary spinal cord tumors. (A) Bar graph illustrating tumor location and general tumor type for all cases. (B) Bar graph demonstrating detailed pathological diagnosis and age distribution. (C) Bar graph demonstrating detailed pathological diagnosis and the extent of tumor resection. GTR, gross total resection; STR, subtotal resection; PR, partial resection.

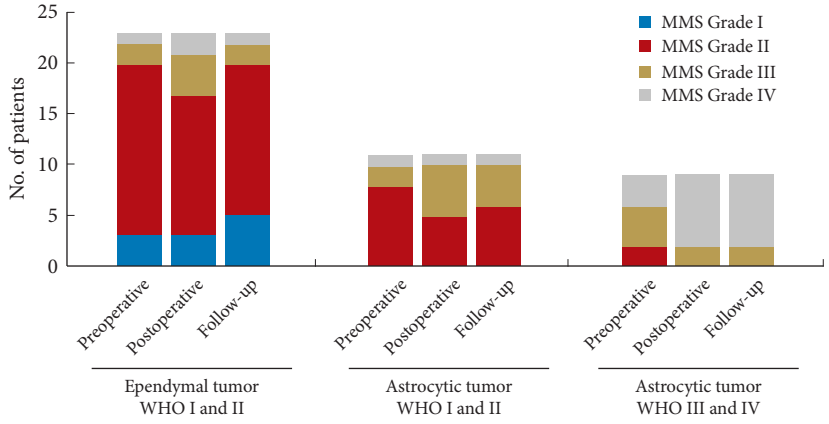

Fig. 2. The neurological status at the perioperative, postoperative, and last follow-up periods based on MMS is illustrated separately as ependymal tumor WHO grades I and II, astrocytic tumor WHO grades I and II, and astrocytic tumor WHO grades III and IV. MMS, McCormick Scale; WHO, World Health Organization; GTR, gross total resection.

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neurological functional decrease in MMS was found in the follow-up duration (Fig. 2). Three patients encountered postoperative pneumonia. These complications were successfully treated by subsequent management.

All 43 patients had clinical follow-up, and the median follow-up duration was 34 months (IQR, 25–60 months). In the whole group, 39 patients (90.7%) remained alive at the time of this study. Four patients died of disease progression and respiratory failure, including 2 WHO grade IV astrocytic tumors (a 6-year-old girl died 20 months after surgery, and a 13-year-old boy died 24 months after surgery), one WHO grade III astrocytic tumor (a 34-year-old woman died 6 months after surgery) and one WHO grade II astrocytic tumor (a 2-year-old girl died 30 months after surgery). One patient with diffuse midline glioma was alive despite recurrence after adjuvant therapy at the 21-month follow-up. The remaining patients were progression-free during the follow-up period. Survival was statistically correlated with tumor pathological grade ($p = 0.004$). The following variables did not affect mortality independently: age, tumor location, tumor span, and extent of tumor resection. Four patients (9.3%) with astrocytic tumors who underwent incomplete tumor resection received adjuvant radiation therapy (3 cases of WHO grade II and 1 case of WHO grade III). Three patients (7.0%) with WHO grade IV astrocytic tumors who underwent complete tumor resection underwent adjuvant chemotherapy. Four patients (9.3%) with WHO grade IV astrocytic tumors with incomplete resection received combined therapy. The major determinant of overall survival was the WHO grade of the tumor in LIMSCTs (Fig. 3). An illustrative case of LIMSCT is presented in Fig. 4.

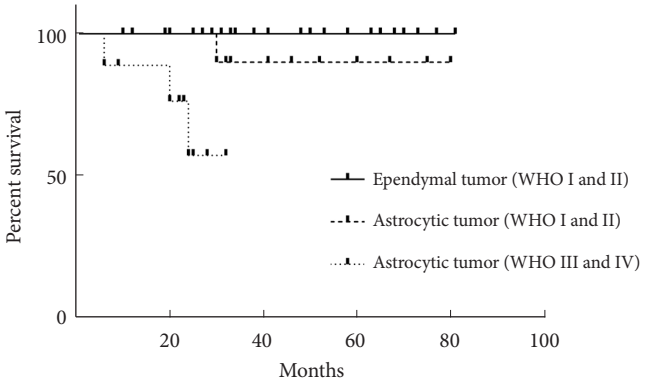

Fig. 3. Kaplan-Meier analysis for overall survival with regard to ependymal tumor WHO grades I and II, astrocytic tumor WHO grades I and II, and astrocytic tumor WHO grades III and IV. WHO, World Health Organization.

DISCUSSION

Generally, intramedullary tumors comprise approximately 20%–30% of all primary spinal cord tumors.[17,18] Some rare cases present with neurological defects due to a long-level extent of tumor involvement, which complicates the management strategies. However, the clinical characteristics and surgical outcomes for LIMSCTs have rarely been studied before. In this study, we presented a series of 43 consecutive patients who had LIMSCT and underwent tumor resection at our institution. To our knowledge, this represents the largest series of patients with LIMSCT involvement who underwent tumor resection.

The general characteristics of long-segment intramedullary neoplasms are somewhat different from those of general intramedullary neoplasms. According to other papers, intramedullary tumors are more common in adults and less common in

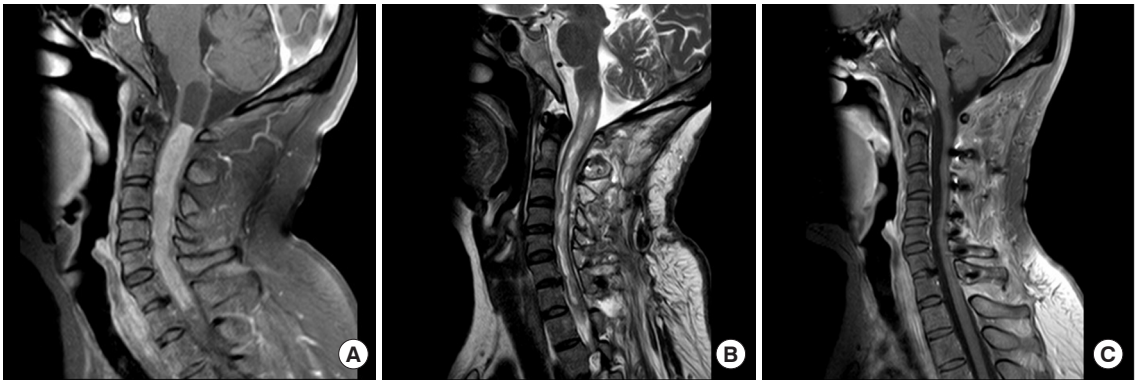

Fig. 4. (A) A 48-year-old female patient. Preoperative imaging indicated a long-level intramedullary tumor. (B) Gross total resection was achieved, and the pathology result was ependymoma WHO grade II. (C) Thirty-month follow-up imaging showed no tumor recurrence. WHO, World Health Organization

children. Only approximately 10%–13% of intramedullary tumors occur in children.[7,12] However, in this series, approximately 30% of patients with long-segment intramedullary tumors were children, which was significantly greater than that generally noted for intramedullary tumors. The possible explanation for this finding is that the pathological properties of intramedullary tumors in children are often poor, and the degree of malignancy is high. Malignant tumors are more likely to grow rapidly and infiltrate the spinal cord. In addition, children often have higher neurological adaptability, and early numbness and weakness do not receive sufficient attention. Therefore, the proportion of children with long-segment intramedullary tumors is higher than that noted for general intramedullary tumors.

In our series, most LIMSCTs involved the cervicothoracic and cervical levels. All the long-segment intramedullary tumors were gliomas, and no other types were observed. Ependymal tumors and astrocytic tumors occurred in 53.5% and 46.5% of all patients, respectively. All patients with ependymal tumors were adults, whereas pediatric patients exhibited a higher proportion of astrocytic tumors. This finding is compatible with previous studies.[19,20] Ependymomas are the most frequent tumors, representing half of the patients in this series. Many studies have demonstrated the characteristics of ependymomas.[21,22] Typically, intramedullary ependymomas are solitary tumors located centrally in the spinal cord. Ependymomas rarely show infiltrative growth. The relatively well-defined interface between the lesion and the surrounding spinal cord tissue facilitates the aggressive resection of ependymomas. In this study, we achieved GTR in all patients with long-level ependymomas. No recurrence was observed at the last follow-up of each patient. Interestingly, 2 patients were diagnosed with subependymoma in this study. Subependymomas are indolent, benign tumors that have been described as having a distinct pathological pattern.[23] Jain et al.[24] concluded that surgical treatment can provide long-term tumor control. In the current case of subependymoma, the patient received GTR and achieved long-term progression-free status at the 50-month follow-up duration. Compared with ependymal tumors, preserving the functional status in patients with astrocytic intramedullary tumors is comparatively difficult. Astrocytomas are infiltrating tumors. Therefore, aggressive resection cannot be achieved in most cases.[25-27] In this series, we were able to achieve GTR in 63.6% of patients with long-level low-grade astrocytic tumors and in 44.4% of patients with high-grade astrocytic tumors. Considering the long-level extent of tumor involvement, the clinical outcome is still encouraging. The medical prognosis is relatively poor in astrocytic tumors, especially those with high histological grade. Overall survival was mainly affected by the WHO grade of the tumor in LIMSCTs.

Despite their rarity, intramedullary tumors can lead to severe neurological function defects prior to medical treatment.[28,29] In addition, patients may have no specific complaints or even remain asymptomatic for a long period, which increases the difficulty of early diagnosis.[2,7] In our current series, most patients experienced combined symptoms of both sensory and motor defects. Neuropathic pain, numbness and muscle weakness were the most frequent complaints in our patients. There is a high index of suspicion for intramedullary tumors when patients experience intractable pain. Although there was a trend that patients with LIMSCTs had a longer prodrome duration compared to other studies, neurological function grade was not statistically associated with the length of prodrome time. Most patients showed a relatively mild neurological defect (MMS grade II) when they presented to our institution. However, 5 patients experienced severe dysfunction (MMS grade IV) due to rapid deterioration. Previous studies have indicated that early removal of an intramedullary tumor increases the possibility of preserving neurological function.[5,30] In our experience, all patients with ependymoma achieved GTR. The rates of GTR, STR, and PR of low-grade astrocytomas were 63.6%, 27.3%, and 9.1%, respectively. The above excision rates for high-grade astrocytomas were 44.4% for GTR, 44.4% for STR, and 11% for PR. In this case series, intraoperative neurophysiological monitoring, including MEPs and SEPs, was performed in all patients. The warning criteria were set for a 50% amplitude reduction of SEP or a 50% amplitude reduction of MEP. The permanent 75% amplitude reduction of MEP was used to stop the operation.[14-16] When there were significant changes in MEP, the operation was suspended temporarily. Warm saline solution was used to irrigate the surgical field, and the blood pressure was increased to at least 60 mmHg. Several literatures have studied and reviewed different monitoring techniques.[14] It is showed that intraoperative neuromonitoring facilitates more aggressive tumor resection given its high sensitivity and specificity and the D-wave signal also exhibits potentials for the prediction of the neurological status.[14,31,32] However, the optimal criteria for monitoring techniques remains controversial and a small tumor piece may be deliberately left in place due to intraoperative conditions. After aggressive tumor resection, disease recurrence is rare. In the ependymal tumor group (WHO grade I and II), 13% of patients experienced postoperative neurological deterioration, and 66.6% of them showed an improvement at the last

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follow-up. In the low-grade astrocytic tumor group (WHO grade I and II), 27.3% of patients showed postoperative worsening in their neurological status, and one-third of them had improved their grades at the last follow-up. In the high-grade astrocytic tumor group (WHO grade III and IV), 55.6% of patients experienced postoperative functional worsening. Sadly, functional improvement was not observed in those patients. Our results indicated that low-level ependymal tumors and low-grade astrocytic tumors achieve a relatively better functional grade after tumor resection, whereas long-level high-grade astrocytic tumors are associated with a poor postoperative neurological status. The functional outcome was not affected by the extent of tumor resection in low-grade ependymal tumors and astrocytic tumors. During the follow-up period, no further functional aggravation was witnessed. In addition, high WHO grade tumors were significantly related to high mortality, which should not be neglected. For the treatment of high-grade gliomas, some institutions prefer conservative treatment strategies, such as biopsy with adjuvant therapy.[33] For a sensitive patient population, adjuvant therapies and immunotherapy may represent alternative options.[34,35] However, the role of adjuvant therapies for high-grade intramedullary tumors still needs to be discussed.[36,37] Tumor resection remains the mainstay of spinal cord tumors.[7,38] In this study of LIMSCTs, we found that long-level tumor extent did not affect the benefits of surgical management. In contrast, the limitation of conservative treatment strategies is that the process cannot prevent tumor progression and finally cause intractable deterioration in patients with intramedullary tumors.[6,39-41] Therefore, we strongly advocate that GTR, or even STR, should be attempted in patients with LIMSCTs, especially in ependymal tumors and low-grade astrocytic tumors, and a beneficial outcome can be expected.

In our case series, no postoperative progressive spinal scoliosis or kyphosis was observed. Three cases of long-segment intramedullary low-grade astrocytomas complicated with severe preoperative scoliosis were treated via a 1-stage operation. Laminectomy was performed for the entire tumor area, and complete resection of the tumor was performed; transpedicular screws were used to correct scoliosis. Patients achieved long-term tumor-free survival, and neurological function was basically preserved at the last follow-up. No serious short- or long-term complications resulted from the primary procedure. The correction of scoliosis prevents the development of spinal deformities and facilitates a return to normal life. Specifically, we encountered a unique case of primary holocord PA. PA is a subtype of glioma with a well-circumscribed, slow-growing, and cystic nature.[42] Primary holocord PAs are extremely rare.[43] We performed spinal internal fixation and posterior spinal fusion as well as laminoplasty. Even after performing spinal fixation, laminoplasty is still recommended. The reinsertion of the lamina plays an important role in reconstructing posterior spinal alignment, avoiding epidural scarring, preventing cerebrospinal fluid leakage, and facilitating reoperation if tumor recurrence occurs.[44,45]

There were several limitations in this study. In addition to the nature of a retrospective study, all of the patients in this study underwent surgery in a single institution, which limited the generalizability. Additionally, the limited population of patients with LIMSCTs also leads to bias. Moreover, the true disease progression rate may be underestimated based on a mean follow-up duration of 37 months. To counter these limitations, further prospective randomized studies with large populations of patients with LIMSCTs are needed.

CONCLUSION

In the current case series, all LIMSCTs were gliomas, among which ependymoma was the main proportion type, and no other tumors were observed. LIMSCTs are most common in adults, whereas the proportion of children is increasing compared to general intramedullary tumors. All ependymal tumors achieved GTR, whereas astrocytic tumors exhibited a lower rate of complete tumor resection, especially in high-grade tumors. Aggressive tumor resection did not increase the risk of long-term neurological functional deterioration in low-grade ependymal tumors and astrocytic tumors. However, in high-grade astrocytic tumors, patients had a higher risk of neurological deterioration and difficulty in recovery. With attempting GTR, patients can expect a low disease progression rate in low-grade ependymal tumors and astrocytic tumors, and complete tumor removal remains the primary goal of treatment. We believe this protocol provides a good review of clinical characteristics and treatment outcomes for patients with LIMSCTs.

NOTES

Conflict of Interest: The authors have nothing to disclose.

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Author Contribution: Conceptualization: T Fan; Data curation: DZ, XZ, CL, YW, KW; Formal analysis: DZ, XZ, CL, YW, KW; Funding acquisition: TF; Methodology: DZ, WF; Project administration: TF; Visualization: DZ, WF; Writing - original draft: DZ; Writing - review & editing: TF, WF.

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