Primary spinal cord tumors of childhood: effects of clinical presentation, radiographic features, and pathology on survival

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Abstract To determine the relationship between clinical presentation, radiographic features, pathology, and treatment on overall survival of newly diagnosed pediatric primary spinal cord tumors (PSCT). Retrospective analysis of all previously healthy children with newly diagnosed PSCT at a single institution from 1995 to present was performed. Twenty-five pediatric patients (15 boys, average 7.9 years) were diagnosed with PSCT. Presenting symptoms ranged from 0.25 to 60 months (average 7.8 months). Symptom duration was significantly shorter for high grade tumors (average 1.65 months) than low grade tumors (average 11.2 months) ($P=0.05$). MRI revealed tumor (8 cervical, 17 thoracic, 7 lumbar, 7 sacral) volumes of 98–94,080 mm$^3$ (average 19,474 mm$^3$). Homogeneous gadolinium enhancement on MRI correlated with lower grade pathology ($P=0.003$). There was no correlation between tumor grade and volume ($P=0.63$) or edema ($P=0.36$) by MRI analysis. Median survival was 53 months and was dependent on tumor grade ($P=0.05$) and gross total resection ($P=0.01$) but not on gender ($P=0.49$), age of presentation ($P=0.82$), duration of presenting symptoms ($P=0.33$), or adjuvant therapies ($P=0.17$). Stratified Kaplan–Meier analysis confirmed the association between degree of resection and survival after controlling for tumor grade ($P=0.01$). MRI homogeneous gadolinium enhancement patterns may be helpful in distinguishing low grade from high grade spinal cord malignancies. While tumor grade and gross total resection rather than duration of symptoms correlated with survival in our series, greater than one-third of patients had reported symptoms greater than 6 months duration prior to diagnosis.
**Keywords**  Pediatric spinal cord tumor - Intraspinal tumor - Childhood spinal tumor

**Abbreviations**
CNS  Central nervous system  
PSCT  Primary spinal cord tumor

**Introduction**

Primary spinal cord tumors (PSCT) are rare central nervous system (CNS) neoplasms in childhood that occur at a frequency of 0.19 per 100,000 person-years according to the Central Brain Tumor Registry of the United States [1]. The incidence varies by age, and increases 1.6 times from 0–4 years old (0.17 per 100,000 person-years) to ages 15–19 (0.28 per 100,000 person-years) [1]. Pediatric PSCT account for <6% of all CNS tumors [2], and have a roughly similar male to female predominance [3–5]. The initial approach to diagnosis and management of PSCT has been extensively reviewed [2, 6, 8–21] and is dependent on anatomical location (intramedullary, extramedullary intradural, and extradural) and pathology. Much of our understanding of the clinical presentation, diagnosis, treatment, and survival features of PSCT comes from small series of patients due to the low incidence. A few larger series of combined multi-institutional PSCT patients have been reported according to specific tumor type [2, 17, 22]. Several smaller pediatric series of PSCT have been published correlating presentation, diagnosis, treatment, and survival features of PSCT comes from small series of patients due to the low incidence. A few larger series of combined multi-institutional PSCT patients have been reported according to specific tumor type [2, 17, 22]. Several smaller pediatric series of PSCT have been published correlating presentation, diagnosis, treatment, and survival features of PSCT comes from small series of patients due to the low incidence. A few larger series of combined multi-institutional PSCT patients have been reported according to specific tumor type [2, 17, 22]. Several smaller pediatric series of PSCT have been published correlating presentation, diagnosis, treatment, and survival features of PSCT comes from small series of patients due to the low incidence. A few larger series of combined multi-institutional PSCT patients have been reported according to specific tumor type [2, 17, 22].

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**Methods**

**Clinical information**

All spinal cord tissue specimens at Children’s National Medical Center in Washington, DC, from 1995 to present were available for retrospective analysis and approved by the Institutional Review Board. A total of 45 patients were identified with spinal cord lesions diagnosed between 1995 and present. Neurodevelopmental tumors (dermoids, epidermoids, and teratomas), lesions associated with tethered cord (lipomas, fibrous bands, hemartomatous tissue, and fibrolipomatosis), sacrococcygeal teratomas, epidermoid cysts, and tumors related to neurofibromatosis Type 1 or Type 2 were excluded from the study. Patients with non-PSCT (i.e. drop metastasis from brain neoplasms) were excluded from the analysis. No patients in our study had meningiomas or schwannomas that were not associated with Neurofibromatosis. Of the 45 total spinal cord samples, 25 patients were diagnosed with PSCT and were available for analysis. Information including age, sex, presenting symptoms, duration of symptoms, neurological examination, and treatment were collected and utilized in the overall clinical analysis.

**Neuroradiographic investigation**

Standard MRI sequences of pediatric spinal cord tumors using a 1.5-T magnet were reviewed by three non-blinded pediatric neuroradiologists (NK, AZ, and GV). Of the 25 patients with available clinical information, 20 patients had complete imaging studies available for analysis. The following neuroimaging features were used for quantitative analysis: tumor location, size, contrast enhancement, and presence of edema. Tumor volume was measured in depth, height, and width. Volume (mm$^3$) was calculated as: depth $\times$ height $\times$ width $\times$ 0.5 and grouped in subcategories of small ($\leq$1,000 mm$^3$), medium (1,001–9,999 mm$^3$), and large ($\geq$10,000 mm$^3$) for statistical analysis.

**Pathological investigation**

All pathology diagnosis were made by a pediatric neuropathologist. Select cases used for the clinical and radiographic analysis were re-reviewed by two pediatric neuropathologists (MS, EJR). Hematoxylin and eosin stained sections were re-reviewed as were other routine histochemical and immunohistochemical preparations. Neoplasms were classified and graded based on World Health Organization criteria.

**Statistical analysis**

Data were analyzed using Fisher’s exact test to compare proportions, and $t$-test for independent samples to compare means. Kaplan–Meier Survival and ANOVA analysis were performed using GraphPad 5.0 Software (San Diego, CA). Stratified Kaplan–Meier analysis was performed using SPSS software (Chicago IL).
| Pt | Age at diagnosis (years) | Sex | Symptom duration (months) | Chief complaint | Clinical exam abnormalities | Spinal level | Pathology diagnosis | Treatmenta | Progressive disease | Survival |
|----|-------------------------|-----|--------------------------|-----------------|---------------------------|-------------|-------------------|-------------|---------------------|----------|
| 1  | 13                      | M   | 5                        | Right extremity weakness | RUE/RLE weakness, atrophy, fasciculations, shoulder drop | C5–T2       | Pilocytic astrocytoma | GT No Yes No | Yes                 | Yes      |
| 2  | 11                      | M   | 7                        | Lower back pain, difficulty ambulating | Minimal hip flexion weakness bilaterally | L4–S2       | Anaplastic ependymoma | GT No Yes Yes | Yes                 | Yes      |
| 3  | 14                      | M   | 24                       | Low back pain, difficulty ambulating | RLE weakness, dermatomal sensory loss, decreased reflexes | T8–L2       | Pilocytic astrocytoma | GT No No No | Yes                 | Yes      |
| 4  | 5                       | M   | 0.5                      | Left extremity weakness, neck pain | LUE proximal > distal weakness, normal sensation/reflexes | C1–C5       | Anaplastic astrocytoma | ST Yes Yes Yes | No                  | No       |
| 5  | 15                      | F   | 1                        | Back pain, left lower extremity weakness | LLE weakness, hyperreflexia, Babinski | T8–T10      | Glioblastoma multiforme | ST Yes Yes Yes | No                  | No       |
| 6  | 9                       | M   | 18                       | Back Pain | Minimal LLE weakness | T4–T10      | Fibrillary astrocytoma | ST Yes Yes Yes | No                  | No       |
| 7  | 5                       | F   | 0.25                     | Toe walking | LLE weakness, absent rectal tone | L3–L5       | Lymphoblastic lymphoma | B Yes No No | Yes                 | Yes      |
| 8  | 11                      | M   | 0.75                     | Difficulty ambulating | Bilateral LE weakness, hyperreflexia, clonus, Babinski | T3–T5       | Langerhans cell histiocytosis | GT Yes No No | No                  | No       |
| 9  | 6                       | M   | 0.25                     | Leg pain, abdominal pain | Bilateral proximal LE weakness, areflexia, sensory level | T11         | Primitive neuroepithelial tumor | B Yes Yes Yes | No                  | No       |
| 10 | 0.75                    | F   | 0.25                     | Bilateral lower extremity weakness | Bilateral LE plegia, areflexia, sensory level, decreased rectal tone | T12–S5      | Primitive undifferentiated neoplasm | ST No Yes Yes | No                  | No       |
| 11 | 11                      | M   | 4                        | Back pain | Bilateral LE weakness, areflexia | L2–L3       | Ependymoma | GT No No No | Yes                 | Yes      |
| 12 | 1.5                     | F   | 0.5                      | Refusal to walk, neck stiffness | Head tilt, nuchal rigidity, minimal LUE weakness | C3–T1       | Fibrillary astrocytoma | ST Yes Yes Yes | Yes                 | Yes      |
| 13 | 1.5                     | F   | 4                        | Neck stiffness | Increased tone neck flexors | C1–C7       | Pilocytic astrocytoma | ST Yes No Yes | Yes                 | Yes      |
| 14 | 1                       | F   | 13                       | Early handedness, delayed motor milestones | Mild R hemiparesis, hyperreflexia, increased tone | Midbrain-C5 | Pilocytic astrocytoma | B Yes No Yes | No                  | No       |
| 15 | 5                       | F   | 2                        | Urinary incontinence, difficulty ambulating | Bilateral LE weakness, decreased rectal tone | S1–5        | Ependymoma | B Yes Yes Yes | Yes                 | No       |
| Pt | Age at diagnosis (years) | Sex | Symptom duration (months) | Chief complaint | Physical exam abnormalities | Spinal level | Pathology diagnosis | Treatment | Progressive disease | Survival |
|----|-------------------------|-----|--------------------------|----------------|---------------------------|-------------|-------------------|-----------|---------------------|---------|
|    | 16                      | 17  | 1                        | Back pain, lower extremity weakness, constipation | Bilateral LE weakness, hyperreflexia, Babinski, decreased rectal tone | Thoracic cord holosyrinx | Pilocytic Astrocytoma | ST Yes Yes No No | No       | No       |
|    | 17                      | 17  | 12                       | Back pain | Bilateral hip flexion weakness | Cauda equina | Myxopapillary ependymoma | GT No Yes No No | No       | Yes      |
|    | 18                      | 2.5 | 7                        | Nuchal tremor | Head tilt, decreased tone bilateral UE, depressed reflexes, decreased strength | T1–T6 | Diffuse fibrillary astrocytoma | GT No Yes No No | No       | Yes      |
|    | 19                      | 1   | 0.25                     | Progressive LE weakness | LE plegia, areflexia, absent sensation, absent rectal tone | C1–S5 | Embryonal tumor | ST Yes No Yes No | No       | No       |
|    | 20                      | 17  | 60                       | Intermittent low back pain, R thigh radicular pain | Hip flexion weakness, patellar hyperreflexia | L2–cauda equina | Ependymoma with myxopapillary features | GT No No No Yes | No       | Yes      |
|    | 21                      | 10  | 3                        | Lower back pain | LE weakness, hyperreflexia, Babinski, Sensory level up to T8, decreased rectal tone | T5 | Primitive Neuroepithelial tumor | ST Yes Yes Yes No | No       | No       |
|    | 22                      | 0.75| 4                        | Early handedness, head tilt | Head tilt, RUE weakness, hyperreflexia | C2–T2 | Glioblastoma multiforme | ST Yes Yes Yes Yes | Yes      | Yes      |
|    | 23                      | 9   | 12                       | Difficulty with ambulation | LE dorsiflexion/plantar flexion weakness, R patellar hyporeflexia, bilateral Babinski | T9–L1 | Fibrillary astrocytoma | ST No Yes No Yes | Yes      | Yes      |
|    | 24                      | 8   | 0.25                     | Back pain, LE weakness | LE weakness, hypotonia, areflexia, absent rectal tone | L3–L5 | Ependymoma | GT Yes Yes Yes Yes | Yes      | Yes      |
|    | 25                      | 7   | 15                       | Neck pain | RUE hemiatrophy, minimal weakness, depressed reflexes | Medulla-T1 | Pilocytic astrocytoma | ST No No No No | No       | Yes      |

*S* Surgery, *C* chemotherapy, *XRT* radiation therapy, *GT* gross total resection, *ST* subtotal resection, *B* biopsy
Results

Clinical features of primary spinal cord tumors of childhood

We retrospectively reviewed the records of 25 consecutive pediatric patients seen at a single institution from 1995 to present newly diagnosed with PSCT. As summarized in Table 1, the average age at presentation was 7.9 months (range 1–5 years; 15 boys). Thoracic cord was the most commonly involved location ($N=17$) followed by cervical ($N=9$), lumbar ($N=7$), and sacral/cauda equina ($N=7$). The most common presenting features were back pain (15/25) and weakness (13/25). In children less than 3 years old, head tilt, delayed motor milestones, and early handedness were the predominant presenting symptoms. There was no difference between age of presentation and symptoms of pain and weakness ($P=0.17$), however, specific neck complaints including pain, weakness, rigidity, or tremor were significantly observed in younger patients (average 2.5 years; range 1.5–5 years) ($P=0.05$). The average reported duration of symptoms was 7.8 months, ranging from 1 week (acute lower extremity pain/weakness) to 5 years (chronic low back pain). There was no significant difference between duration of symptoms and symptom type ($P=0.06$), but early handedness and back pain were present the longest prior to diagnosis (Fig. 1). There was no correlation between symptom duration and age of presentation ($P=0.95$). When stratified according to specific age groups (0–3 years, 4–12 years, and 13–18 years) duration of symptoms were not different ($P=0.11$). Boys had a longer reported duration of symptoms prior to diagnosis than girls (11.3 vs. 2.9 months) ($P=0.03$). While there was no correlation between length of presenting symptoms and anatomical location ($P=0.30$), there was a difference between length of symptoms and tumor grade. Patients with high grade tumors had a shorter duration of symptoms (average 1.65 months, range 0.25–7 months) than patients with low grade tumors (average 11.1 months, range 0.25–60 months) ($P=0.05$). There was no difference between tumor grade and age ($P=0.71$) or gender ($P=0.10$). The most common neurological abnormality was change in muscle tone or strength, followed by abnormal reflexes (7 hyper, 9 hypo/absent). Four patients had evidence of a sensory level on examination along with hypo or absent reflexes, mimicking transverse myelitis or Guillain Barre’ syndrome.

Neuroradiographic analysis of primary spinal cord tumors of childhood

We performed a detailed neuroradiographic analysis including tumor volume, T1/T2 signal characteristics, gadolinium enhancement patterns, and the presence of edema in 20 patients with newly diagnosed PSCT who had sufficient image sequences for interpretation. Typical and atypical neuroradiographic features of spinal cord astrocytomas and ependymomas, the most common tumors in our series, are illustrated in Fig. 2. As summarized in Table 2, the most common spinal tumor location was intramedullary ($N=11$) followed by extramedullary intradural ($N=8$) and epidural ($N=1$). Eighty percent (4/5) of ependymomas analyzed in our series had an extramedullary component; half of which had multiple lesions. Quantitative volumetric analysis revealed ranges from 98 to 94,080 mm$^3$ (average 19,474 mm$^3$). There was no difference between low grade tumor volume (average 19,868 mm$^3$) and high grade tumor volume (average 15,676 mm$^3$) ($P=0.63$) at the time of diagnosis. When stratifying for evidence of edema (illustrated in Fig. 3), there was no correlation with tumor grade ($P=0.22$).
Likewise, neither T2 hyperintensity \((P = 1.0)\) nor T1 hypointensity \((P = 0.11)\) were significantly associated with grade. Homogeneous gadolinium enhancement was found significantly more in low grade tumors \((P = 0.003)\). Rim gadolinium enhancement, on the other hand, did not correlate with tumor grade \((P = 0.098)\).

Effects of symptomatology and treatment on survival

The median overall survival of our series of PSCT was 53 months (range 1.5–53 months; 10 deaths) with a median follow up 21 months (Fig. 4). Despite the earlier presentation of girls in our series, there was no affect of
gender on survival (Median survival 53 months boys; 41 months girls) \((P = 0.58)\) (Fig. 4a). There was no correlation between age of diagnosis and survival \((P = 0.35)\), nor was there a difference when stratified according to specific age group \((P = 0.79)\) (Fig. 4b). Duration of symptoms did not affect overall survival; given the wide range of presenting neurological symptoms. Those patients with symptoms greater than 6 months had an average survival of 48 months compared to 35 months for symptoms greater than 6 months \((P = 0.91)\) (Fig. 4c). Of the 10 deaths in our series, the average time of presentation was 3.9 months compared to 10.4 months for those who survived \((P = 0.08)\). As expected, patients with high grade tumors (median survival 25 months) had significantly poorer survival than those with low grade tumors (median survival 53 months) \((P = 0.05)\) as shown in Fig. 4d. In addition to having no correlation with tumor grade, tumor volume did not correlate with overall survival in our series \((P = 0.13)\).

Compared to patients with biopsy or subtotal resection, patients with gross total resection had 100% survival (Fig. 4e) \((P = 0.01)\). Thirty-six percent of patients in our series had a gross total resection (9/25). Of these patients, three had residual post operative weakness. Of the 25 patients with surgical intervention (gross/subtotal resection, biopsy) 10 had some degree of post operative weakness, 8 of which resolved within months of surgery. The most severe complication was the development of Brown-Sequard syndrome in a patient with a lumbar sacral diffuse fibrillary astrocytoma.

Since non-surgical adjuvant treatments were not standardized, a generalized stratification of chemotherapy, radiation, or combined therapies were used for survival analysis. Three of 25 patients had adjuvant chemotherapy

### Table 2 Radiographic features of primary spinal cord tumors (PSCT) of childhood

| Pathology               | Spinal level | Tumor location     | Tumor volume (mm\(^3\)) | MRI signal\(^a\) | Gadolinium enhancement | Edema |
|-------------------------|--------------|--------------------|-------------------------|-----------------|------------------------|-------|
| 1 Pilocytic astrocytoma | C5–T1        | Intramedullary     | 21,660                  | ↓               | ←                      | Irregular | No    |
| 2 Pilocytic astrocytoma | C1–C3        | Intramedullary     | 12,000                  | ←               | ↓↑                    | Homogenous | No    |
| 3 Anaplastic ependymoma | S2           | Extramedullary Intradural | 2,211         | ←               | ←                      | Homogenous | Yes   |
| 4 Anaplastic astrocytoma| C2–C5        | Intramedullary     | 11,832                  | ↓               | ↑↑                    | Rim enhancing | Yes   |
| 5 Langerhans cell histiocytes | T4      | Epidural           | 11,160                  | ←               | ←                      | Not performed | No    |
| 6 Primitive neuroepithelial tumor | T8–T12 | Intramedullary     | 6,600                   | ↓↑              | ↓↑                    | Irregular | Yes   |
| 7 Primitive neuroectodermal tumor | S3     | Extramedullary Intradural | 1,056           | ←               | ↑↑                    | Not performed | No    |
| 8 Primitive undifferentiated tumor | T10–S1 | Extramedullary Intradural | 13,520          | ↓               | ↓↑                    | Irregular | Yes   |
| 9 Ependymoma            | L2–L3        | Extramedullary Intradural | 11,856          | ↓               | ↑                      | Homogenous | No    |
| 10 Fibrillary astrocytoma | C3–T1     | Intramedullary     | 21,504                  | ↑               | ↑                      | Irregular | No    |
| 11 Pilocytic astrocytoma | C1–C7      | Intramedullary     | 21,560                  | ↑               | ↑                      | Irregular | Yes   |
| 12 Pilocytic astrocytoma | C1–C5      | Intramedullary     | 94,080                  | ↑               | ↑                      | Irregular | No    |
| 13 Myxopapillary ependymoma | L1–S1 | Extramedullary Intradural | 21,630          | ↓               | ↑                      | Irregular | No    |
| 14 Pilocytic astrocytoma | T7–T8       | Intramedullary     | 98                      | ←               | ↓↑                    | Homogenous | No    |
| 15 Fibrillary astrocytoma | T1–T6      | Intramedullary     | 21,097                  | ↓               | ↑                      | Irregular | Yes   |
| 16 Embryonal tumor      | C5–S2       | Extramedullary Intradural | 70,200          | ←               | ←                      | Irregular | No    |
| 17 Ependymoma with myxopapillary features | L2     | Extramedullary Intradural | 2,736           | ↓               | ↑                      | Irregular | No    |
| 18 Glioblastoma multiforme | C2–T2    | Intramedullary     | 4,212                   | ↓               | ↑                      | Rim enhancing | Yes   |
| 19 Ependymoma            | L3–L5       | Intramedullary     | 4,920                   | ↓               | ↑                      | Homogenous | No    |
| 20 Pilocytic astrocytoma | C1–T1       | Intramedullary     | 35,552                  | ↓               | ↑                      | Irregular | Yes   |

\(C\) Cervical, \(T\) Thoracic, \(L\) Lumbar, \(S\) Sacral

\(^a\) ↑, Hyperintense; ↓, hypointense; ←, isointense
alone without evidence of relapse. Six of 25 had adjuvant radiation therapy alone (two fibrillary astrocytoma, one anaplastic astrocytoma, one pilocytic astrocytoma, one PNET, one myxopapillary ependymoma); of these two had progressive disease. Combined radiation and chemotherapy were used in 40% of patients (10/25), 90% of whom had either metastatic disease at diagnosis or eventually had progressive disease. As shown in Fig. 4f, adjuvant chemotherapy and radiation either alone or in combination had no significant effect on overall survival ($P = 0.31$). While the specific cause of death was not known for each of the 10 patients, 4 had complications secondary to pneumonia and sepsis.

Discussion

The average duration of presenting symptoms of 7.8 months in our series of PSCT is similar to previous reports ranging from 2 to 9 months [18, 21, 22]. Bouffet et al. reported 11% (8/73) of patients with primary spinal astrocytomas had greater than 3 years of symptoms prior to presentation. While pain and weakness were the predominant presenting features in many patients, more subtle findings such as early handedness can delay diagnosis particularly in younger patients. A common set of presenting complaints among younger patients in our series involved the neck and included pain and torticollis, as has been reported in two younger patients with PSCT [28]. In older patients, chronic back pain has been associated with delayed diagnosis of PSCT [22, 23], similar to our findings. The variability of reflexes (hypo/hyper/absent) on neurological examination was not particularly helpful in establishing tumor location or grade compared to more sensitive findings of tone and strength. Ultimately, duration of presenting symptoms did not correlate with outcome as has been reported [23]. However, shorter duration of symptoms is associated with higher grade tumors in our series and has been associated with poor survival in the series reported by Bouffet et al. [22].

One of the strengths of the current study is the detailed radiographic analysis performed on a subset of patients where neuroimaging studies were complete. It seems counterintuitive that there was no correlation between tumor volume and tumor type, grade, or survival. This suggests that tumor location itself as opposed to size may be an important factor in achieving gross total resection and hence improved survival. One set of factors that may associated with spinal cord tumor grade are specific patterns of gadolinium enhancement. Our observations of homogeneous gadolinium enhancement associated with low grade tumors has been reported [29]. However, in the context of predictors of survival, this may be an important finding. Due to our small number of patients studied, it is difficult to make generalizations. A multi-institutional series of collaborative neuroradiographic data on PSCT is ultimately necessary to validate our results.

One of the major factors associated with survival in our series of PSCT was degree of surgical resection. While 35% of PSCT are intramedullary (65% in our series), making total resection at times technically challenging, it is a feasible option [7, 14, 30–33]. However, as reported in our series, post operative complications, although temporary, can be associated with significant morbidity. Radical excision of intramedullary tumors has been reportedly associated with both an increase in survival and improved quality of life [6, 31–34], but are dependent on tumor type and grade. Long term control or cure can be achieved for some intramedullary ependymomas by total/subtotal resection alone [9, 11, 17, 21]. This is in contrast to infiltrating astrocytomas where the role of subtotal resection is less clear [4, 9, 17, 21, 31] but may be better than biopsy alone [35]. Only through collaborative studies involving

![Fig. 3](image-url) Detection of spinal cord tumor-related edema on MRI. Examples of the presence or absence of edema in two cases of pilocytic astrocytoma are shown. a Edema present Note the small central rim-enhancing lesions surrounded by bright T2 (top) and dark T1 (bottom) signal, compatible with edema. b Edema absent Note there is no increased T2 (top) or dark T1 (bottom) signal beyond the well-defined border of this lesion.
large number of patients will we be able to meaningfully assess the extent of surgical resection on survival.

One of the major criticisms of the current study in addition to the small sample size and retrospective study design, is the lack of uniformity of adjuvant therapies. While neither chemotherapy nor radiation alone or in combination affected overall survival in our series, there remains great debate regarding the role of adjuvant therapies in PSCT. There are some who avoid adjuvant therapy in cases of total resection [36, 37]. In the case of radiation therapy, favorable outcome results have been reported in patients with low grade spinal astrocytomas and ependymomas [38–44]. However, in patients with low grade astrocytomas with incomplete resection, the role of radiation therapy is unclear [22]. With regards to adjuvant chemotherapy, there is no proven efficacious regimen for any given pathological subtype or location.

A major hurdle in our understanding of PSCT, is a lack of fundamental knowledge of the biology of the tumor. It is naïve to assume the biological pathways that govern oncogenesis in the brain can be applied to the spinal cord. Furthermore, small amounts of tissue obtained during biopsy or resection can limit the number of non standard genetic/biochemical tests necessary to fully understand the biology of the tumors. While fortunately the incidence if PSCT is quite low, the mortality associated with PSCT calls for a more collaborative approach to our understanding and treatment of pediatric spinal cord tumors.
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