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CLINICAL STUDY - PATIENT STUDY

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.

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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, treatment, and tumor histology with event free and overall survival [10, 12, 13, 18, 20, 23–27]. To our knowledge, no series has specifically attempted to correlate duration of symptoms, neurological examination abnormalities, and specific neuroradiographic features with malignancy and overall survival. To address these issues, we have performed a retrospective analysis of all previously healthy pediatric patients seen at our institution from 1995 to present with newly diagnosed PSCT. The diverse presentations, duration of symptoms, radiographic findings, and outcomes presented in our series expands our current knowledge of this rare pediatric neoplasm.

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 schwanommas 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 × height × width × 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).

Table 1 Clinical features of primary spinal cord tumors (PSCT) of childhood

Pt	Age at diagnosis (years)	Sex	Symptom duration (months)	Chief complaint	Physical exam abnormalities	Spinal level	Pathology diagnosis	Treatment[a]			Progressive disease	Survival
								S	C	XRT		
1	13	M	5	Right extremity weakness	RUE/RLE weakness, atrophy, fasciculations, shoulder drop	C5–T2	Pilocytic astrocytoma	GT	No	Yes	No	Yes
2	11	M	7	Lower back pain, difficulty ambulating	Minimal hip flexion weakness bilaterally	L4–S2	Anaplastic ependymoma	GT	No	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
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
5	15	F	1	Back pain, left lower extremity weakness	LLE weakness, hyperreflexia, Babinski	T8–T10	Glioblastoma multiforme	ST	Yes	Yes	Yes	No
6	9	M	18	Back Pain	Minimal LLE weakness	T4–T10	Fibrillary astrocytoma	ST	Yes	Yes	Yes	No
7	5	F	0.25	Toe walking	LLE weakness, absent rectal tone	L3–L5	Lymphoblastic lymphoma	B	Yes	No	No	Yes
8	11	M	0.75	Difficulty ambulating	Bilateral LE weakness, hyperreflexia, clonus, Babinski	T3–T5	Langerhans cell histiocytosis	GT	Yes	No	No	Yes
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
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
11	11	M	4	Back pain	Bilateral LE weakness, areflexia	L2–L3	Ependymoma	GT	No	No	No	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
13	1.5	F	4	Neck stiffness	Increased tone neck flexors	C1–C7	Pilocytic astrocytoma	ST	Yes	No	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
15	5	F	2	Urinary incontinence, difficulty ambulating	Bilateral LE weakness, decreased rectal tone	S1–5	Ependymoma	B	Yes	Yes	Yes	No

Table 1 continued

Pt	Age at diagnosis (years)	Sex	Symptom duration (months)	Chief complaint	Physical exam abnormalities	Spinal level	Pathology diagnosis	Treatment[a]			Progressive disease	Survival
								S	C	XRT		
16	17	M	1	Back pain, lower extremity weakness, constipation	Bilateral LE weakness, hyperreflexia, Babinski, deceased rectal tone	Thoracic cord holosyrinx	Pilocytic Astrocytoma	ST	Yes	Yes	No	No
17	17	M	12	Back pain	Bilateral hip flexion weakness	Cauda equina	Myxopapillary ependymoma	GT	No	Yes	No	Yes
18	2.5	M	7	Nuchal tremor	Head tilt, decreased tone bilateral UE, depressed reflexes, decreased strength	T1–T6	Diffuse fibrillary astrocytoma	GT	No	Yes	No	Yes
19	1	F	0.25	Progressive LE weakness	LE plegia, areflexia, absent sensation, absent rectal tone	C1–S5	Embryonal tumor	ST	Yes	No	Yes	No
20	17	M	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
21	10	M	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
22	0.75	F	4	Early handedness, head tilt	Head tilt, RUE weakness, hyperreflexia	C2–T2	Glioblastoma multiforme	ST	Yes	Yes	Yes	Yes
23	9	M	12	Difficulty with ambulation	LE dorsiflexion/plantar flexion weakness, R patellar hyporeflexia, bilateral Babsinki	T9–L1	Fibrillary astrocytoma	ST	No	Yes	No	Yes
24	8	F	0.25	Back pain, LE weakness	LE weakness, hypotonia, areflexia, absent rectal tone	L3–L5	Ependymoma	GT	Yes	Yes	Yes	Yes
25	7	M	15	Neck pain	RUE hemiatrophy, minimal weakness, depressed reflexes	Medulla-T1	Pilocytic astrocytoma	ST	No	No	No	Yes

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

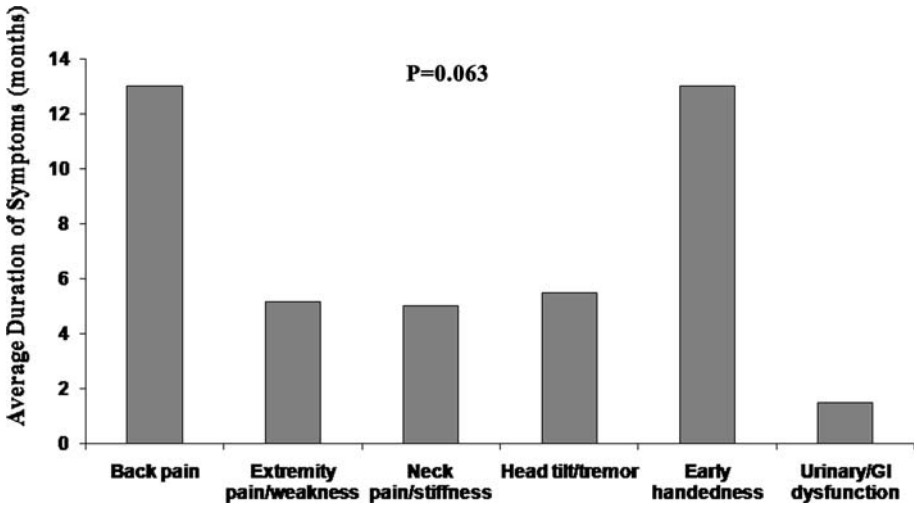

Fig. 1 Average duration of symptoms of primary spinal cord tumors (PSCT) of childhood. The average duration of neurological complaints of PSCT is shown. There was no significant difference between specific symptom type and duration by ANOVA analysis ($P = 0.063$)

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 Barré 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 ($P = 0.22$).

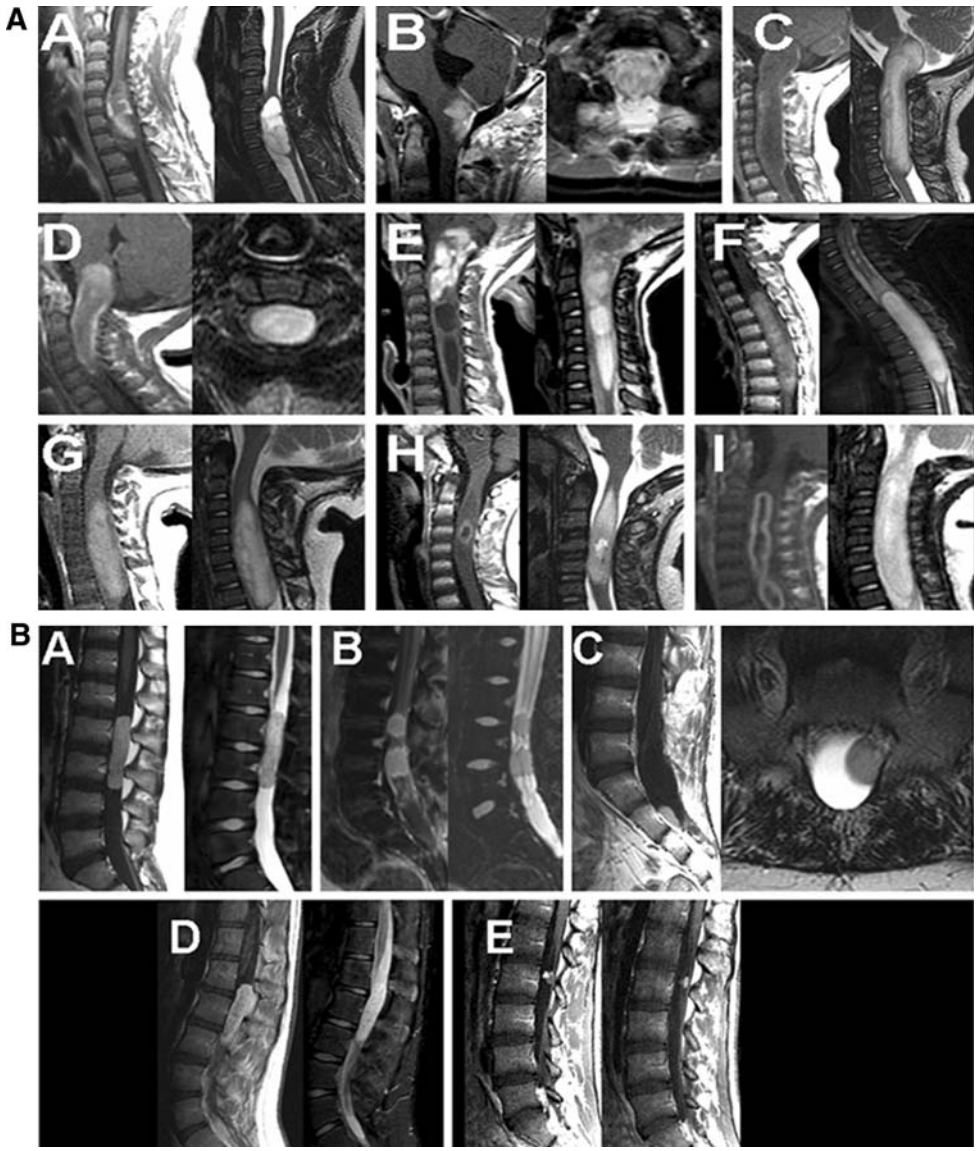

Fig. 2 MRI characteristics of glial and ependymal primary spinal cord tumors (PSCT). *Top panel* **a** Pediatric spinal cord astrocytomas. *A–E* Pilocytic astrocytoma. Note the wide variability of imaging features. All lesions *A–E* are well-defined on T2 (T1)-weighted images. Only *A* and *E* have appearance of nodular enhancement in conjunction with a cyst, no cysts are seen in lesions *B*, *C*, *D*. The enhancement patterns vary from irregular (*A*, *E*), homogeneous (*B*), no significant enhancement (*C*) to rim-enhancement (*D*), the latter usually more typical of a higher grade lesions. *F*+*G* Fibrillary astrocytoma. These lesions share a pattern of sausage-like expansion of the spinal cord, similar to pilocytic astrocytoma *C*, but with diffuse contrast enhancement. Edema of the adjacent spinal cord is only seen in lesion *F*. *H*+*I* Anaplastic (*H*) and high-grade (*I*) astrocytoma. Both high grade astrocytomas are rim-enhancing; edema of the adjacent spinal cord is more prominent in *I* when compared to *H*. *Bottom panel* **b**. Pediatric spinal cord ependymoma. Sagittal T1, T2, and axial T2 are shown in *A–C* compared to a patient with myxopapillary ependymoma in *D*. These lesions are only different in the T2 appearance (bright in patient *D*) and inhomogeneous contrast enhancement in patient *D*. Interestingly, 2/5 patients presented with multiple lesions (patients *B*+*E*)

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

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

	Pathology	Spinal level	Tumor location	Tumor volume (mm3)	MRI signal[a]		Gadolinium enhancement	Edema
					T1	T2		
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 histiocytosis	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

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

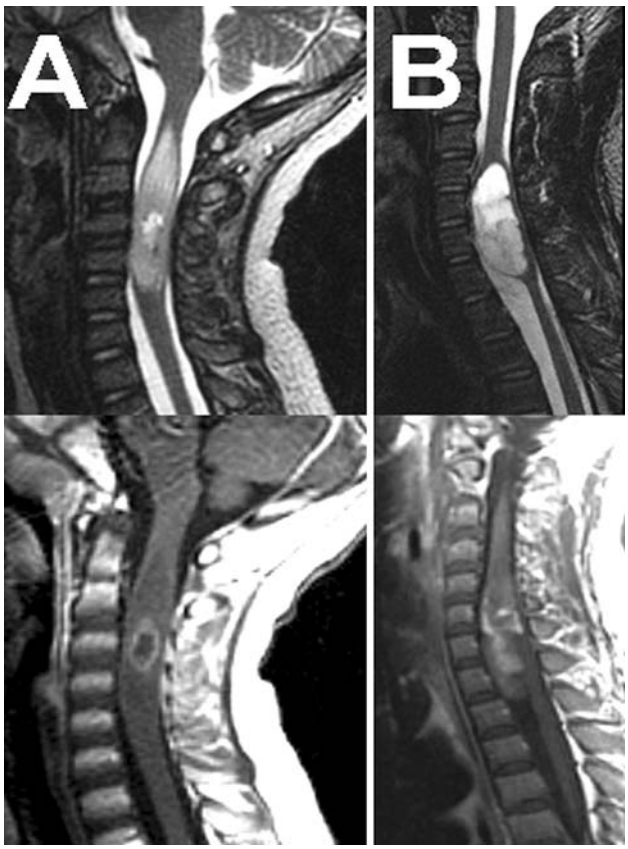

Fig. 3 Detection of spinal cord tumor-related edema on MRI. Examples of the presence or absence of edema in two cases of pilocytic astrocytoma are shown. **a** *Edema present* Note the small central rim-enhancing lesions surrounded by bright T2 (*top*) and dark T1 (*bottom*) signal, compatible with edema. **b** *Edema absent* Note there is no increased T2 (*top*) or dark T1 (*bottom*) signal beyond the well-defined border of this lesion

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

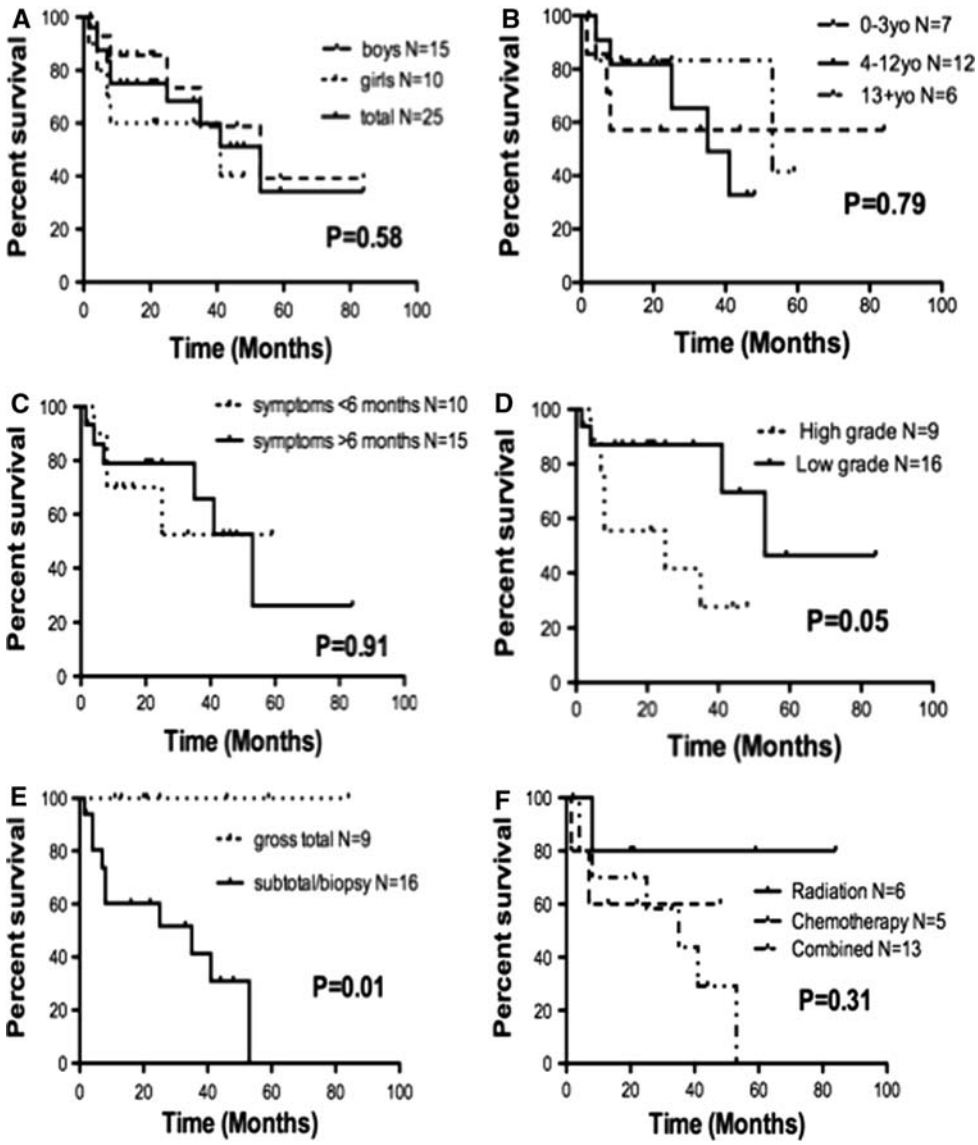

Fig. 4 The effects of symptomatology, tumor grade, and treatment on overall survival of pediatric primary spinal cord tumors. Kaplan–Meier survival analysis was stratified according to gender (**a**), age (**b**), duration of symptoms (**c**) tumor grade (**d**), extent of resection (**e**), and adjuvant therapies (**f**). There was no correlation between overall survival and gender ($P = 0.58$), age ($P = 0.79$), or duration of symptoms greater or less than 6 months ($P = 0.91$). High grade malignancy was associated with poorer survival ($P = 0.05$) as was gross total resection ($P = 0.01$). Adjuvant chemotherapy and radiation therapy, either alone or in combination, had no effect on overall survival in our series ($P = 0.31$)

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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