Solitary fibrous tumors of the spine: a pediatric case report with a comprehensive review of the literature

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Solitary fibrous tumors of the spine are rare lesions. Their description in the literature is limited to case reports and small case series. While generally benign lesions, they can recur and occasionally occur as malignancies. Here the authors present the case of a 10-year-old boy, the youngest patient and first preadolescent reported thus far, with this condition. In addition, they perform a comprehensive review of all previously published cases of spinal solitary fibrous tumors.

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KEY WORDS solitary fibrous tumor; spine; central nervous system; pediatric; oncology

SOLITARY fibrous tumors (SFTs) were initially described by Klemperer and Rabin in 1931.³⁶ These lesions are most commonly found in the pleura and abdomen/pelvis. One large single-center review suggests a nearly even split between benign and malignant pathologies.¹⁹ A recent review of the Surveillance, Epidemiology, and End Results (SEER) database found 804 cases reported over nearly 40 years. Tumors in these patients were thoracic/abdominal/pelvic in 35.3%, central nervous system (CNS) in 24.5%, head/neck in 19.9%, and extremity in 16.2%. Most reported cases did not include pathological classification, and a plurality of patients (49.4%) underwent surgery without adjuvant therapy.⁷⁹

Carneiro and colleagues described the first 2 cases of spinal SFT, both of which were treated with surgery.¹² Few comprehensive reviews of these lesions exist. Bisceglia et al. found 220 cases of CNS SFTs, 50 of which occurred in the spine.⁵ Fargen et al. reviewed 189 cases of CNS SFTs, including 46 spinal SFTs, and added 1 other spinal SFT case from their own experience.²²

Spinal SFT in pediatric patients is quite rare. The youngest patient with spinal SFT reported thus far was a 17-year-old male,³⁰ and there have been only 2 other previously reported cases of spinal SFT in patients younger than 21 years of age.^{11,71} Here we describe the case of an

intradural extramedullary spinal SFT in a 10-year-old boy. In addition, we identified 81 unique patients with spinal SFT in the literature and comprehensively review these reports.

Case Report

History and Examination

A 10-year-old, previously healthy male was admitted to the neurology service for evaluation of right upper extremity weakness. On examination, he was unable to adduct his right shoulder and had Medical Research Council Grade 4/5 strength in the other muscle groups of his right upper extremity. Otherwise, his motor and sensory examinations were normal. He was hyperreflexic in the right upper and lower extremities with a positive Babinski sign on the right. Magnetic resonance imaging studies revealed an intradural extramedullary tumor on the right side at C1–3 that was hypointense on T1- and T2-weighted sequences with homogeneous enhancement (Fig. 1A–C). Preoperative diagnosis was meningioma or, less likely, nerve sheath tumor.

Operation

The patient was taken to surgery where a C1–3 lami-

ABBREVIATIONS CNS = central nervous system; GTR = gross-total resection; SFT = solitary fibrous tumor; STR = subtotal resection.

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FIG. 1. Preoperative axial T2-weighted MR image (A) demonstrating hypointensity of the lesion and cord compression. Preoperative sagittal (B) and coronal (C) T1-weighted MR images with contrast demonstrating homogeneous enhancement of the tumor. Postoperative coronal T1-weighted MR image (D) with contrast demonstrating GTR and reexpansion of the spinal cord.

nectomy was performed for excision of the tumor. After performing two-thirds laminectomies at each level, a paramedian durotomy was made. The firm, rubbery pale tumor was intradural extramedullary without dural attachment. It was located between the dorsal and ventral nerve rootlets. It was adherent to the lateral aspect of the spinal cord but could be dissected free of pia mater without injury to the cord and did not have an intramedullary component. The tumor had a pial attachment at its caudal end. The tumor also appeared to be attached to the right dentate ligament at C-1. Gross-total resection (GTR) was achieved in a piecemeal fashion (Fig. 2). There were no intraoperative complications.

Postoperative Course

Postoperative MRI confirmed GTR (Fig. 1D). The patient was discharged home on postoperative Day 4 with outpatient physical therapy. At the time of discharge, he had trace residual weakness in his right hand and otherwise had full strength with no new neurological deficits. At 4.5 months after surgery, he had regained full strength, MRI showed no evidence of recurrent disease, and dynamic cervical spine radiographs showed no evidence of instability (Fig. 3). One year after surgery, the patient had a normal neurological examination, no evidence of recurrence on MRI, and maintained stability on flexion and extension radiographs of the cervical spine.

Light microscopy examination of the tissue showed a

FIG. 2. Intraoperative photographs of tumor resection. A: Initial view of tumor with dorsal nerve roots overlying the lesion. B: Pial attachment at caudal end of tumor. C: Attachment of tumor to pia of spinal cord. D: Tumor has been completely resected. Figure is available in color online only.

spindle cell neoplasm with alternating cellular and collagenous areas. A few blood vessels were well demarcated with thin walls and showed branching, that is, staghorn vessels (Fig. 4A–C). A few nuclei had nucleoli, but the cell population was overall monomorphous without significant cytological atypia. Mitotic count was variable between 1 and 5/10 hpf on multiple counts in different areas. No necrosis, hemorrhage, or hypercellularity was seen. The neoplasm was diffusely and strongly positive for Bcl-2 and CD34 (Fig. 4D) with a diffuse nuclear staining for STAT6 (Fig. 4E). The Ki-67 proliferation index was variable between 10% and 15% in various areas (Fig. 4F). Epithelial membrane antigen and S100 protein were negative. This appearance was consistent with SFT with atypical features.

Review of the Literature

A PubMed search was performed for SFTs of the spine. In addition, the reference section of each paper was reviewed, allowing for the identification of additional cases. We identified 65 previously published papers with 83 cases.^{1–4,7–14,16,17,20–22,24–35,37–40,42,43,46–59,62,64–74,76,78,80,81} Two of these papers clearly duplicated patients (4 reports of 2 unique patients).^{14,76} A third reported patient was a suspected duplication.^{12,57} Eliminating these duplications and adding our own report yielded 82 reported cases of spinal SFT (Table 1).

Seventy-eight cases reported age at presentation. The majority of these cases occurred in adults (94.8% in patients over the age of 21 years). Patients ranged in age from 10 years (our patient) to 83 years (median 51.5 years). Seventy-eight cases contained information on patient sex. There is a slight male predominance in the reported cases of spinal SFT (56.4% male vs 43.6% female; Table 2).

Seventy-six reported cases indicated spinal level. The plurality of lesions occurred in the thoracic region (42.1%), followed by the cervical region (27.6%) and lumbar region (13.2%). The remaining tumors occurred in the sacrum,⁵⁰ across junctional levels,^{1,11,13,22,28,46} or at multiple levels.^{35,47,67,68} Sixty-eight reported cases contained information on the spinal compartment involved. The plurality

FIG. 3. Postoperative flexion (A) and extension (B) cervical spine radiographs demonstrating no evidence of dynamic instability.

(35.3%) was intradural extramedullary, followed by extradural (26.5%). Of these extradural tumors, 6 were primarily extraspinal or bone as opposed to primarily intraconal (Table 2).^{27,31,40,51–53}

Sixty-one reported cases contained information on MRI appearance. Tumors in all of these cases exhibited some degree of enhancement with intravenous contrast, most of them homogeneously enhancing. Most of the lesions were either hypointense or isointense on T1- and T2-weighted imaging, although rare tumors were hyperintense on one or the other of these sequences. A few reports indicated preoperative diagnoses, most often meningioma or nerve sheath tumor.^{2,10,43,64,65,72,73} One hemorrhagic lesion in the conus was thought to be either a myxopapillary ependymoma or cavernous malformation.⁷⁴

Seventy-five of the reported cases contained information regarding treatment. Patients in all of these cases underwent surgery. Gross-total resection was achieved in 82.7% of patients, and subtotal resection (STR) was performed in 17.3%. Four patients (3 GTR and 1 STR) underwent preoperative embolization of the tumor.^{9,40,53,67} One GTR patient also underwent intraoperative embolization.⁷¹ Two of the GTR outcomes were achieved in 2 stages of surgery.^{10,51} Four patients (3 GTR and 1 STR) required spinal fusion at the initial presentation.^{26,31,51,71} A single patient with a malignant SFT was treated with chemotherapy and radiation after STR.⁶⁷ Otherwise, chemotherapy and radiation were reserved for recurrences and/or metastases.^{9,26,46,50,51}

Forty-seven patients had reports of intraoperative assessments of tumor origin. In most cases, the origin was either the spinal cord or the dura mater, with fewer reports of tumors arising from nerve roots. Four patients, including our own, had attachment to the pia mater.^{3,9,29} Four patients had tumors arising from bone or periosteum,^{26,52,67,71} 2 patients had tumors involving the arachnoid,^{10,22} 1 patient's tumor seemed to arise from the venous plexus.²⁷

Seventy-eight cases indicated tumor histopathology. Sixty-nine patients (92%) had SFT without mention of malignant or other unusual features on pathology. Five

FIG. 4. Microscopic findings. **A:** Low-power view with peculiar, well-defined, branching blood vessels (asterisk) and alternating dark and light areas reflecting variable cellularity and collagen. **B:** Spindle cell morphology of the neoplastic cells and mitotic activity (arrow). **C:** Thick, keloid-like collagen bundles and decreased cellularity in some areas. **D:** Diffuse and strong CD34 positivity. **E:** Nuclear STAT6 expression. **F:** Higher-than-expected Ki-67 proliferation index. H & E (A–C), original magnifications $\times 40$ (A), $\times 200$ (B–C), $\times 100$ (D–F). Figure is available in color online only.

patients (6.7%) had malignant SFT.^{9,24,35,51,67} Three of the previously reported cases included unusual pathological findings such as retiform or papillary features,⁷⁰ focal pleomorphism,⁷ and lipomatous features.¹ Our patient's tumor demonstrated atypical features without reaching the criteria for malignant SFT.

Outcome depended largely on histology and extent of resection. Four of the patients with malignant SFT had outcome data reported. All 4 died of their disease, one at 2 months,²⁴ one at 6 months,⁶⁷ one at 9 months,⁵¹ and one at 4 years.⁹ Fifty-six patients with SFT had outcome data reported. Follow-up times ranged from 2 months to 21 years. Twenty-two patients had follow-up of 1 year or less, and 45 patients had follow-up of 5 years or less. Outcome of SFT without malignant features seemed to depend largely on extent of resection. Nine SFT patients with STR had information on outcome. Three of these patients had only 6 months of follow-up and no recurrence or progression.^{28,34,62} Of the remaining 6 patients, 5 exhibited local disease progression between 6 months and 21 years after the first presentation.^{8,12,26,37,46} The final patient developed a new spinal lesion 14 years after the original surgery.⁵⁶ Forty-six patients treated with GTR had data on outcome and recurrence. Of these patients, 41 (89.1%) had no recurrence at a median of 1.2 years of follow-up (range 2 months–7 years). One of these patients had residual dis-

TABLE 1. Literature summary of spinal SFT cases

Authors & Year	Age (yrs)/ Sex	Level	Compartment	Treatment	Histology	Last FU (yrs)	Outcome	Notes
Carneiro et al., 1996	50/F	NR	IDEM & IDIM	STR	SFT	6.0	Recurrence	Died 1 wk after surgery for recurrence
	54/F	Lumbar	NR	GTR	SFT	7.0	Disease free	Died of unrelated causes
Alston et al., 1997	47/M	T4–5	IDIM	GTR	SFT	0.2	Disease free	
Kuchelmeister et al., 1997	64/M	C-5	IDEM & IDIM	GTR	SFT	NR	NR	
Malek et al., 1997	33/M	T7–8	IDEM	GTR	SFT	NR	NR	
Brunori et al., 1999	18/M	O–C3	ED & IDEM	GTR	SFT	1.0	Disease free	
	46/F	T12–L1	IDEM	GTR	SFT	0.3	Disease free	
Hasegawa et al., 1999	39/M	NR	NR	GTR	SFT	9.0	Local recurrence at 5 yrs & 9 yrs	
Kanahara et al., 1999	62/M	C6–7	IDEM & IDIM	Surgery	SFT	NR	NR	
	46/F	C4–5	ED & IDEM	GTR	SFT	NR	NR	
Donnellan et al., 2000	39/M	L-1	ED	GTR	SFT	NR	NR	
Mordani et al., 2000	33/M	C-5	IDIM	GTR	SFT	1.5	Disease free	
Vorster et al., 2000	51/M	T2–3	IDEM	GTR	SFT	0.6	Disease free	
Kurtkaya et al., 2001	70/F	T-3	IDEM	GTR	SFT	1.0	Disease free	
Endo et al., 2003	63/F	C2–4	ED	GTR	SFT	NR	NR	Dumbbell tumor
Obara et al., 2003	49/F	C2–4	ED	GTR + preop embo	SFT	1.0	Disease free	Primarily extraspinal
Tihan et al., 2003	NR	NR	IDIM	NR	NR	NR	NR	
	NR	NR	IDIM	NR	NR	NR	NR	
	NR	NR	IDEM	NR	NR	NR	NR	
	NR	NR	IDEM	NR	NR	NR	NR	
Bohinski et al., 2004	49/F	C-4	IDEM & IDIM	GTR	SFT w/ focal pleomorphism	0.8	Disease free	
Caroli et al., 2004	54/M	C7–T1	IDEM	GTR	SFT	1.3	Disease free	
Hirakawa et al., 2004	52/M	C1–2	ED	GTR	SFT	2.5	Disease free	Primarily extraspinal
Kawamura et al., 2004	64/M	T2–3	IDEM & IDIM	STR	SFT	0.5	No progression	
Kim et al., 2004	55/M	C-6, T-3, T4–5, L-5, S-2	ED	STR	Malignant SFT	NR	NR	Prior posterior fossa tumor
Piana et al., 2004	67/M	L1–2	ED	GTR	SFT	NR	NR	
Pizzolitto et al., 2004	36/M	T7–8	IDEM	GTR	SFT	1.5	Disease free	
	47/M	C-4	IDIM	GTR	SFT	1.0	Disease free	
Miyashita et al., 2004	63/F	C-7, T3–5	IDEM	GTR (1 lesion)	SFT	NR	Stable residue	T-5 lesion resected, had tentorial lesion 15 yrs prior
Jallo et al., 2005	41/M	C6–7	IDIM	GTR	SFT	3.5	Disease free	
	17/M	T-5, T-6	IDIM	GTR	SFT	1.6	Disease free	2 separate lesions
	59/M	T-5	NR	GTR	SFT	4.8	Disease free	
	37/F	T2–3	NR	GTR	SFT	5.0	Disease free	
Ogawa et al., 2005	63/F	T-11	IDEM	GTR	SFT	NR	NR	
Ogungbo et al., 2005	53/M	C3–4	IDEM & IDIM	GTR	SFT	NR	NR	
Pakasa et al., 2005	27/M	T5–7	IDEM & IDIM	STR	SFT	14.0	New sacral lesion at 14 yrs treated w/ GTR	

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TABLE 1. Literature summary of spinal SFT cases

Authors & Year	Age (yrs)/ Sex	Level	Compartment	Treatment	Histology	Last FU (yrs)	Outcome	Notes
Metellus et al., 2007	56/M	C7–T1	NR	STR	SFT	3.5	Local progression at 2 yrs treated w/ GTR	
	57/M	L-2	NR	GTR	SFT	8.5	Recurred at 8.5 yrs, treated w/ GTR & chemo	
	57/M	T-9	NR	GTR	SFT	1.8	Disease free	
	44/F	T-12	NR	GTR	SFT	4.2	Disease free	
	72/F	T-12	NR	GTR	SFT	0.8	Disease free	
	44/F	T-7	NR	GTR	SFT	1.7	Disease free	
Nakamura et al., 2007	28/F	Cervical	ED	GTR	SFT	5.0	Disease free	Arising dorsally from lamina & facets
Hashimoto et al., 2008	71/F	C4–5	ED	STR + anterior fusion	SFT	4.0	Progressed at 6 mos & 2.5 yrs	Required additional resection & fusion surgeries
Muñoz et al., 2008	35/M	Sacral	NR	GTR	SFT	8.0	Local recurrence w/ pulmonary mets at 5.5 yrs	Malignant pulmonary mets at 3.5 yrs; pulmonary/hepatic mets at 6.5 yrs
Shin et al., 2008	40/F	C6–7	ED & IDEM	GTR	SFT	0.7	Disease free	Dumbbell tumor
Arantes et al., 2009	22/M	T1–2	IDEM	GTR	SFT	1.5	Disease free	
Ciappetta et al., 2010	75/F	T6–7	IDEM & IDIM	GTR	SFT	2.0	Disease free	
Ishii et al., 2009	63/F	C-5	IDIM	GTR	SFT	1.2	Disease free	
Kakimaru et al., 2009	75/F	T8–10	ED	GTR + dorsal fusion	SFT	3.0	Disease free	Dumbbell tumor w/ large extraspinal component
Aftab et al., 2010	38/M	L4–S2	ED	STR	SFT w/ lipomatous features	0.4	Stable residue	
Cincu et al., 2010	58/M	T3–5	IDEM	GTR	SFT	NR	NR	
Vassal et al., 2011	52/F	T8–9	ED & IDEM	GTR	SFT	5.2	Disease free	
Fargen et al., 2011	62/M	O–C2	IDEM	GTR	SFT	0.5	Disease free	
	28/F	C2–3	IDEM & IDIM	GTR	SFT	2.0	Small radiographic recurrence	Lost to FU
Guo et al., 2012	60/M	Thoracic	Intracanal	Surgery	Malignant SFT	0.2	Deceased from metastatic disease	Metastatic from renal primary
Takenouchi et al., 2011	45/F	C-3, C7–T1	IDEM	GTR (1 lesion)	SFT	0.4	Stable residue	Prior posterior fossa tumor
Bouyer et al., 2012	56/F	T10–11	ED	STR	SFT	5.0	Progressed at 6 mos, recurred at 4 yrs	
Mariniello et al., 2012	67/M	C4–7	IDEM	GTR	SFT	1.0	Disease free	
	75/F	T6–7	IDEM	GTR	SFT	1.0	Disease free	
Wu et al., 2012	50/F	L-2	IDEM	GTR	SFT	NR	NR	Arising from filum terminale
Chen et al., 2012	43/M	T11–12	NR	GTR	SFT	14.7	Recurred at 14.7 yrs	
	63/M	L4–S2	NR	GTR	SFT	2.8	Disease free	
Brigui et al., 2013	56/M	T6–7	IDEM	GTR	SFT	2.4	Disease free	

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TABLE 1. Literature summary of spinal SFT cases

Authors & Year	Age (yrs)/ Sex	Level	Compartment	Treatment	Histology	Last FU (yrs)	Outcome	Notes
Brigui et al., 2013	69/M	T7–8	ED	GTR + preop embo	Malignant SFT	4.0	Deceased from metastatic disease	Primary pulmonary lesions; developed kidney/extremity/visceral mets
Montano et al., 2013	56/F	L1–2	IDEM	GTR	SFT	1.0	Disease free	
Son et al., 2013	54/F	T-8, S1–2	ED	STR + preop embo + postop radiation/chemo	Malignant SFT	0.5	Deceased from metastatic disease	Presented w/ multifocal disease (skull, spine, pelvis)
Tomek et al., 2013	66/M	T11–12	ED	STR	SFT w/ retiform & papillary features	4.1	Progressed at 18 mos	
Hwang et al., 2014	48/M	C7–T1	IDEM	STR	SFT	0.5	No progression	
Kobayashi et al., 2014	40/M	C4–5	IDEM & IDIM	STR	SFT	21.0	Progression at 21 yrs	
Nagano et al., 2014	57/F	L4–5	ED	GTR + fusion (2 stages)	Malignant SFT	0.8	Deceased from metastatic disease	Local & metastatic recurrence at 3 mos
Robert et al., 2014	49/F	T9–10	IDEM & IDIM	STR	SFT	0.5	No progression	
Sebaaly et al., 2014	32/F	L3–4	IDEM	GTR	SFT	1.3	Disease free	
Tsutsumi et al., 2014	19/M	C-1	ED	GTR + intraop embo + fusion	SFT	1.0	Disease free	Primarily bone, had parapharyngeal lesion 10 yrs prior
Yuan et al., 2014	48/M	T-9	ED & IDEM	GTR	SFT	NR	NR	Dumbbell tumor
Basaran et al., 2015	67/M	L-3	IDEM	GTR	SFT	1.0	Disease free	
Bruder et al., 2015	83/F	T8–9	IDIM	GTR (2 stages)	SFT	0.7	Disease free	
Sade et al., 2015	43/M	Thoracic	IDEM	Surgery	SFT	NR	NR	
Walker et al., 2015	47/F	L-1	IDIM	GTR + preop embo	SFT	0.5	Disease free	Hemorrhagic lesion in conus medullaris
Lavrador et al., 2015	23/F	T11–12	ED	GTR + preop embo	SFT	0.5	Disease free	Primarily extraspinal, dumbbell
Zaldivar-Jolissaint et al., 2016	33/M	C1–4	ED	GTR	SFT	7.0	Disease free	
Present case	10/M	C1–3	IDEM	GTR	SFT w/ atypical features	1.0	Disease free	

Chemo = chemotherapy; ED = extradural; embo = embolization; FU = follow-up; IDEM = intradural extramedullary; IDIM = intradural intramedullary; mets = metastasis; NR = not reported; O = occiput.

ease elsewhere but no recurrence of the resected lesion.⁶⁸ Three patients had single local recurrences at 2 years,²² 8.5 years,⁴⁶ and 14.5 years.¹⁴ Another patient developed local recurrences twice, once at 5 years and again at 9 years.²⁵ The final patient developed malignant transformation of his tumor with pulmonary metastases at 3.5 years and local spinal recurrence and recurrent pulmonary and hepatic metastases at 5.5 years.⁵⁰

Discussion

Solitary fibrous tumors are lesions of mesenchymal origin that differentiate primarily along fibroblastic lines. Characteristically, they demonstrate spindle cells in a col-

lagen background. They are now thought to exist along a spectrum with hemangiopericytomas, representing a more cellular and vascular subtype of the same tumor category. The majority of SFTs exhibit benign histology and behavior; however, malignancy does occur and is evidenced by a high mitotic count, increased cellularity, hemorrhage and/or necrosis, pleomorphic nuclei, and foci of dedifferentiation.¹⁸

A borderline increase in the mitotic rate and a high proliferation index indicate atypical features of the SFT in our patient. Malignancy in SFT is not well described, and the criteria are mainly developed for the other end of the spectrum, that is, hemangiopericytoma.^{23,44} Nonethe-

less, a large series on soft tissue and pleural SFTs showed that patients 55 years of age or older with tumors 15 cm or larger and with a mitotic count of 4 or more per 10 hpf had a high risk of metastasis and death,¹⁸ while a proliferation index of 10% or more was found to be associated with malignancy in another study.⁶ The importance of the proliferation index has been emphasized in predicting prognosis in a series of CNS SFTs,¹⁴ as well as in a meta-analysis of CNS SFTs.⁵ Authors of the latter study concluded that a proliferation index of 5% or more may indicate a higher likelihood of recurrence.⁵

Solitary fibrous tumor was first described as a tumor arising from the pleura in the thoracic cavity. In this location, the majority of these tumors are benign. However, 10%–20% will exhibit locally aggressive behavior or malignancy. Surgery is the preferred method of treatment and carries with it a 5-year survival of close to 100%. Adjuvant therapy is reserved for metastatic or recurrent disease. Chemotherapy and radiation are only moderately effective. In the thorax, most tumors that recur do so within 2 years, and imaging is recommended every 6 months for 2 years followed by annual imaging. Malignant SFT carries a poor prognosis, with 63% of tumors recurring despite GTR and most patients dying within 2 years of diagnosis.¹⁵

A recent single-center review of 82 patients with SFT demonstrated a slight female predominance (57%).¹⁹ Among this study cohort there was a nearly even split between benign and malignant histologies. However, patients were identified through a search of a sarcoma database at a tertiary care cancer center that likely skewed the series toward a malignant profile. The median survival of patients with benign tumors was 15.5 years with a 5-year survival of just over 75%. The median survival of patients with malignant tumors was 7.8 years with a 5-year overall survival of 55%. While some patients with a benign histology did receive chemotherapy and radiation, these modalities were mostly reserved for patients with malignant SFT.

The literature on spinal SFT is limited to case reports and small case series. Therefore, we cannot comment on its incidence, although it does appear to be a very rare diagnosis. Likewise, since existing reports include very small numbers of patients, it is difficult to determine epidemiology, ideal treatment, follow-up, and overall prognosis. However, based on our review of 82 patients, we can attempt to draw some conclusions.

Not surprisingly, most of the tumors occur in the thoracic spine, as is seen with other spine neoplasms. The intradural extramedullary compartment of the spine is the most common space to find these tumors, although they are also frequently reported in the extradural space and intramedullary space with an exophytic component. Imaging characteristics, particularly for extradural and intradural extramedullary tumors, tend to mimic meningiomas and/or nerve sheath tumors. The tumors can arise from any tissue, and their origin is not always evident on imaging or during surgery.

With regard to management, the overwhelming majority of patients are treated with surgery. Gross-total resection should be the goal and can be curative. Subtotally resected tumors tend to recur, although the number of patients who underwent STR in our series is small. There

TABLE 2. Demographics and tumor characteristics

Variable	Value
Age (yrs)	
Mean (SD)	49.8 (15.4)
Median (range)	51.5 (10–83)
Sex	
Male	44 (56.4%)
Female	34 (43.6%)
Level	
Occipitocervical	2 (2.6%)
Cervical	21 (27.6%)
Cervicothoracic	3 (3.9%)
Thoracic	32 (42.1%)
Thoracolumbar	1 (1.3%)
Lumbar	11 (14.5%)
Lumbosacral	1 (1.3%)
Sacral	1 (1.3%)
Multiple	4 (5.3%)
Compartment	
Extradural	18 (26.5%)
Extradural & intradural extramedullary	5 (7.4%)
Intradural extramedullary	24 (35.3%)
Intradural extramedullary & intramedullary	11 (16.2%)
Intramedullary	10 (14.7%)

does not appear to be a role for adjuvant therapy for the benign SFT patient who has had a successful GTR. Adjuvant therapy for residual benign SFT is probably also of little benefit given the overall good prognosis of these tumors and the literature on SFTs in other locations suggesting that adjuvant therapy is of little value for benign SFT. Malignant SFT, on the other hand, appears to have a very poor prognosis regardless of intervention. There were very few malignant SFT cases in our series, but the patients in these cases all fared poorly. In these patients, strong consideration should be given to resection followed by adjuvant therapy, although it is not clear how much impact this will have on life expectancy.

Despite its benign histology, SFT can recur. This has been seen with SFTs in other nonspinal locations. Most patients in our series had very short follow-ups. Interestingly, the tumors that did recur seemed to do so late (2 years, 8.5 years, 5 and 9 years, and 14.5 years), suggesting that there may be value in the long-term follow-up of patients. In addition, 3 of the patients with spinal SFT had prior intracranial lesions and presented with spinal lesions consistent with drop metastases. All of these lesions had benign histologies. The lag time between intracranial disease and the diagnosis of spinal disease was 10 years,⁶⁸ 14 years,⁵⁶ and 15 years.⁴⁷

Solitary fibrous tumors in any location are much more common in adults than in children. In the pleura, it seems that less than 2% of SFTs occur in children. In addition, children have a lower incidence of malignant pleural SFT than adults do.⁴⁵ Solitary fibrous tumors have been report-

ed in other locations in children including the mesentery,⁷⁵ scalp,⁶¹ kidney,⁷⁷ orbit,⁴¹ shoulder,⁶⁰ and brain.⁶³ Prior to this report, the youngest reported patient with spinal SFT was 17 years of age³⁰ and only 2 other reported patients were younger than 21 years of age.^{11,71} Our patient is the first documented preadolescent child with spinal SFT.

The major limitation of this paper is its reliance on previously published case reports and case series. These previous reports have all been selected for publication based on unusual features of the cases. There is inevitably publication bias that introduces selection bias to this report. In addition, previous authors have been selective about the information they have included in their reports. Unfortunately, given the rarity of SFTs of the spine, larger studies are unlikely to be performed.

Conclusions

Solitary fibrous tumors of the spine are rare neoplasms. No large series exist. However, given this review of reported cases, it appears that the majority of these lesions are benign and should be treated with GTR alone. Even under these circumstances, however, the tumors can recur and thus long-term follow-up should be considered.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Albert. Acquisition of data: both authors. Analysis and interpretation of data: Albert. Drafting the article: Albert. Critically revising the article: both authors. Reviewed submitted version of manuscript: Albert. Approved the final version of the manuscript on behalf of both authors: Albert. Statistical analysis: Albert. Study supervision: Albert.

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