SPINAL SOLITARY FIBROUS TUMORS: A SERIES OF FOUR PATIENTS: CASE REPORT

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OBJECTIVE AND IMPORTANCE: Intraspinal solitary fibrous tumors, which are rare mesenchymal tumors, have previously been reported as case reports. We review our experience and, to our knowledge, the first small institutional series with respect to clinical presentation, diagnosis, surgical management, pathohistological analysis, progression-free survival, and long-term outcome.

CLINICAL PRESENTATION: In this retrospective review, four patients (three male and one female) ranging in age from 17 to 59 years (mean, 38.5 yr) had spinal solitary fibromas located throughout the spinal canal. Three tumors were located in the thoracic region and one in the cervical spine. All patients presented with pain and paresthesia. Two patients had an associated spinal deformity. All had gross total resection as confirmed by postoperative imaging studies.

INTERVENTION: All four patients underwent surgical treatment for spinal solitary fibromas. None of these patients underwent irradiation or chemotherapy after surgery at our center; therefore, outcome was attributed to surgery alone. There were no surgical deaths, and the 5-year actuarial survival rate was 100%. At the most recent follow-up examination, neurological function was stable or improved in 90% of patients.

CONCLUSION: Patients with solitary fibrous tumors have a long survival. These tumors have an indolent course, and radiotherapy or chemotherapy seems to be unnecessary.

KEY WORDS: Intramedullary neoplasm, Outcome, Solitary fibroma, Spinal cord tumor, Surgery

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Intraspinal solitary fibrous tumors are extremely rare and benign mesenchymal neoplasms. Klemperer and Rabin (11) first identified this tumor as a localized form of pleural mesothelioma that seemed to be histologically suggestive of fibroma and responded well to surgery alone. These tumors have subsequently been reported arising in soft tissues elsewhere in the body, such as the peritoneum, and within the central nervous system. There have been several case reports of intracranial and intraspinal meningeal involvement. There also have been several case reports of intramedullary localization. However, the origin of this tumor still remains controversial, with arguments for a mesothelial or mesenchymal origin. Aside from these case reports, there has been no dedicated analysis of the prognosis and optimal management of these intraspinal tumors. The current study examines these two questions and, in particular, the effects of radical surgery on the progression-free survival and outcome.

PATIENTS AND METHODS

Between the years 1990 and 2003, four patients were diagnosed with spinal cord solitary fibrous tumors from our series of spinal cord tumors. A detailed neurological history and examination were completed on each patient. Patient outcome was assessed by a review of the hospital and outpatient charts. If a patient had not been evaluated within the previous 9 months, a telephone interview was conducted with a family member or the primary physician.

Of the four patients, three were male and one was female. The mean age at diagnosis was 38.5 years (range, 17–59 yr). The youngest

patient had two intraspinal lesions. This second tumor was small (9 mm in its widest dimension) and was located one vertebral level from the larger tumor.

The symptoms at presentation varied by the level of the tumor in the spinal canal. There were three tumors located in the thoracic region and one in the cervical region. The most common preoperative symptoms were pain (80%) and paresthesias (50%). Only two patients presented with a motor deficit or myelopathy. Interestingly, two patients experienced a spinal deformity or scoliosis before surgery. The medical histories, from the onset of symptoms to diagnosis, were generally rather long, with a mean duration of 16.5 months (range, 6–24 mo). There was no correlation between the character or duration of symptoms and the age of the patients (*Table 1*).

Diagnosis

All patients underwent spinal and cranial magnetic resonance imaging (MRI) studies before surgery. Those with a clinically apparent spinal deformity received x-rays of the spine to document the curvature for future comparison.

Treatment

All four patients underwent surgery using intraoperative neurophysiological monitoring with motor and sensory evoked potentials. Gross total resection was achieved in all patients. The extent of tumor removal, defined at the time of surgery with ultrasound or visually, was confirmed by MRI.

CASE REPORTS

Patient 1

A 41-year-old man sought care for gradually progressive weakness occurring for several months. The neurological examination revealed mild weakness in the right upper extremity. He had normal sensation; however, hyperreflexia in the lower extremities was present. MRI scans revealed an intradural mass in the cervical spine at C6–C7. It was slightly hyper- to isointense on T1-weighted sequences. T2-weighted sequences demonstrated a hypointense lesion with edema or gliosis rostral and caudal to the tumor. The mass demonstrated significant enhancement after intravenous injection of gadolinium (*Fig. 1*).

Surgery was performed via a standard laminectomy approach. The tumor was circumscribed and firm. The origin was intramedullary in location, but was eccentric to one side of the spinal cord. The plane between the tumor and the spinal cord was not well demarcated. The tumor was firm and not vascular in nature. It was removed in a gross total fashion, and the patient made an uneventful recovery. He was discharged 4 days after the operation. Postoperative imaging confirmed the total removal of the tumor. Neuraxis imaging did not demonstrate any further lesions.

Patient 2

A 17-year-old boy presented to the clinic with several months of back pain and a 1-week history of lower extremity weakness. MRI scans of the spine demonstrated two intraspinal lesions. The larger lesion was hypointense on T1- and T2-weighted sequences. There was significant spinal cord edema associated with the tumor. In addition, there was a tiny second tumor one segment rostral to the larger tumor on the other side of the cord. Preoperative x-rays demonstrated scoliosis. Surgery was performed for both tumors. The tumors were intramedullary in location, and both were firm and circumscribed, but were adherent to the lateral surface of the spinal cord. The consistency was similar to that of other spindle-cell-like tumors arising in the extramedullary space.

TABLE 1. Patient characteristics for solitary fibrous tumors[a]

Age (yr)/sex	Location	Duration of symptoms (mo)	Symptoms	MRI signal characteristics		Contrast enhancement	Follow-up (yr)
				T1-weighted	T2-weighted		
59/M	T5	24	Lower extremity numbness, paraparesis, paresthesias, and scoliosis	Iso	Hypo	Homogeneous	4.8
37/F	T2–T3	12	Dysesthesias and paresthesias	Iso	Hypo	Homogeneous	5.0
41/M	C6–C7	24	Dyesthesias and upper extremity weakness	Iso	Hypo	Homogeneous	3.5
17/M[b]	T5, T6	6	Scoliosis and spastic paraparesis	Iso	Hypo	Homogeneous	1.6

[a] Iso, isointense; Hypo, hypointense.
[b] Two separate intraspinal lesions.

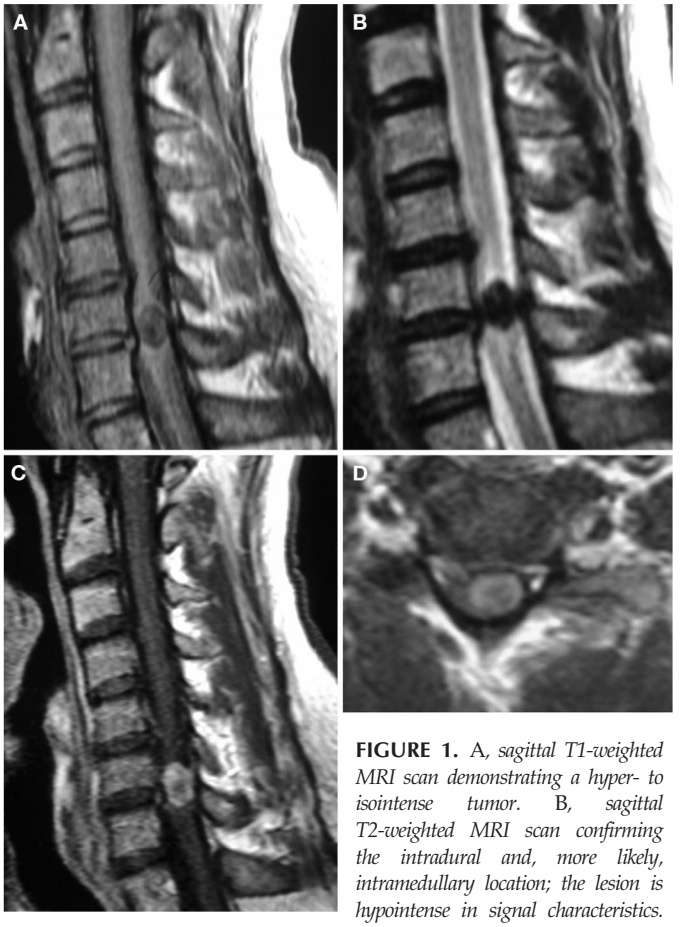

FIGURE 1. A, *sagittal T1-weighted MRI scan demonstrating a hyper- to isointense tumor.* B, *sagittal T2-weighted MRI scan confirming the intradural and, more likely, intramedullary location; the lesion is hypointense in signal characteristics.* C, *postgadolinium sagittal MRI scan revealing homogeneous enhancement.* D, *postgadolinium axial MRI scan demonstrating no distinct plane from the spinal cord.*

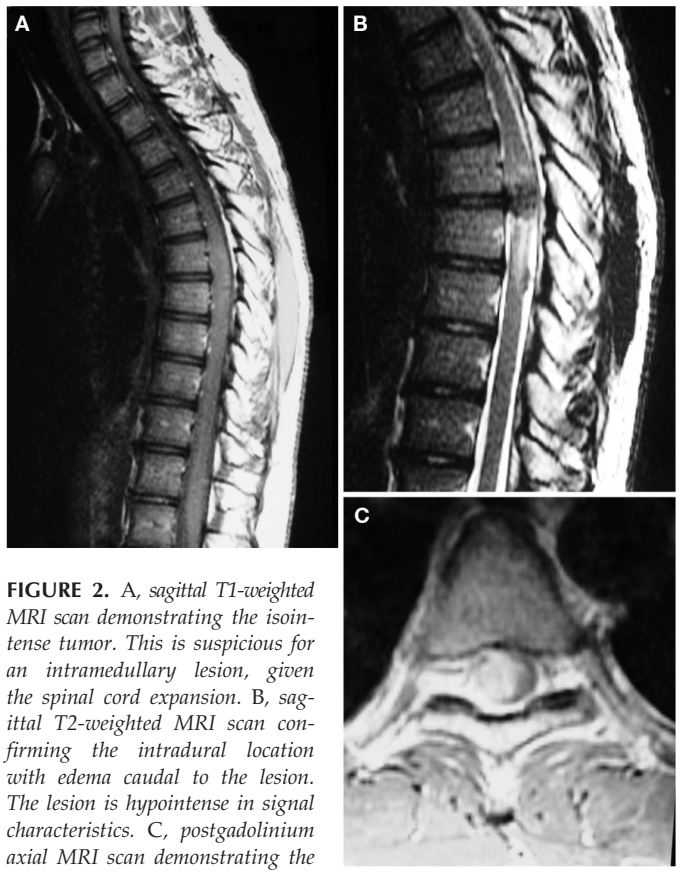

FIGURE 2. A, *sagittal T1-weighted MRI scan demonstrating the isointense tumor. This is suspicious for an intramedullary lesion, given the spinal cord expansion.* B, *sagittal T2-weighted MRI scan confirming the intradural location with edema caudal to the lesion. The lesion is hypointense in signal characteristics.* C, *postgadolinium axial MRI scan demonstrating the intramedullary tumor without a distinct plane from the spinal cord.*

The larger tumor was adherent, and no cleavage plane existed from within the spinal cord. The patient demonstrated motor improvement immediately after surgery. Postoperative imaging confirmed the total excision of the lesions. At the last follow-up examination, the patient was neurologically intact. His scoliosis is stable without progression (*Fig. 2*).

RESULTS

The preoperative diagnosis in all patients was an intradural tumor. The imaging characteristics were not characteristic for an intramedullary or intradural-extramedullary tumor. No tumor demonstrated any dural attachment on preoperative MRI scans or during surgery. On MRI scans, the tumors were hypointense on T1-weighted sequences and iso- to hypointense on T2-weighted sequences. All tumors demonstrated homogeneous enhancement after gadolinium administration; however, the margins were not sharp (as observed for meningiomas or schwannomas). At the time of surgery, the tumors seemed to arise from the pia-arachnoid. The tumor bulk was usually extramedullary or lateral in location; however, in two cases, it was intramedullary-exophytic in configuration. In any event, there was no clearly defined tissue interface between the tumor and the spinal cord.

Surgical resection differed significantly from resection of the more common intradural-extramedullary tumors, such as nerve sheath tumors and meningiomas. As a result of the firm attachment to or even exophytic growth from the cord, direct manipulation seemed to be dangerous because of potential injury to the cord. It turned out to be difficult to grab the mass with microinstruments and impossible to cut it with microscissors or to debulk it with the ultrasonic aspirator. The microsurgical neodymium:yttrium-aluminum-garnet contact laser (PhotoMedex, Montgomeryville, PA) was extremely helpful in cutting the firm tissue mass and removing it as well as in detaching it from the spinal cord.

There was no operative mortality (death within 1 mo of surgery) after radical resection. Gross total resection was accomplished for all patients. All the patients were alive at the most recent follow-up evaluation. The mean follow-up period was 3.7 years (range, 1.6–5.0 yr). There has been no tumor recurrence during the follow-up period.

Histological analysis was performed for all the lesions. All the tumors were stained with hematoxylin and eosin and with immunochemistry, including S-100, CD34, glial fibrillary

acidic protein, epithelial membrane antigen (EMA), neuron-specific enolase, vimentin, and proliferation-related labeling index (MIB-1). All the tumors were composed of spindle-shaped cells dispersed in a loose matrix, packed by bundles of stout reticular fibers. The amount of collagen and vascularity varied from one tumor to another. None of the tumors demonstrated increased cellularity or cellular pleomorphism. No pseudopalisading arrangement, whorl formation, or psammoma bodies were visualized. No giant cells or mitotic figures were observed. Immunohistochemistry stains demonstrated focal immunoreactivity to CD34 and vimentin. There was no staining to S-100, glial fibrillary acidic protein, EMA, neuron-specific enolase, or desmin. The MIB-1 proliferation index was low (*Fig. 3*).

DISCUSSION

This review provides new information regarding characteristics, prognostic factors, and treatment results for patients with intraspinal solitary fibrous tumors. In our small series, which represents a referral center, such tumors had a 3:1 male sex predilection; occurred predominantly in the thoracic and cervical spinal cord; usually involved one to two vertebral segments; and most often produced signs and symptoms consisting of pain, radiculopathy, and sensory or motor deficits. In our literature review, we found 13 previous reports of intraspinal solitary fibrous tumors. These tumors have been reported to occur within the intramedullary, intradural-extramedullary, and extradural compartments. All previous reports discuss one or two single-patient histories and have a relatively short follow-up period. These tumors were located predominantly in the thoracic region (*Table 2*).

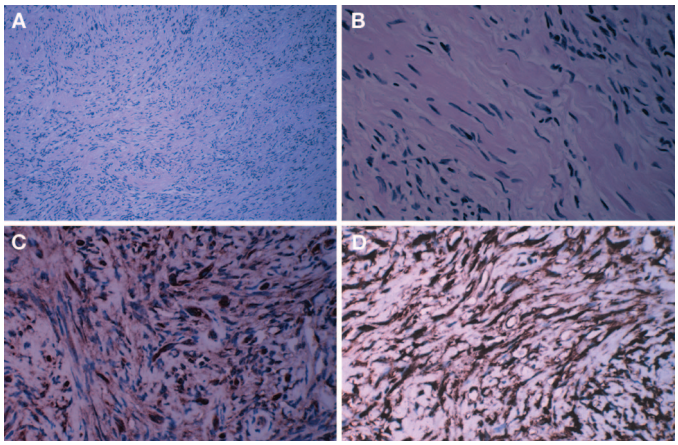

FIGURE 3. *Histological appearance of solitary fibrous tumors.* A, *tumor demonstrating biphasic architecture on immunohistochemical staining, with areas of collagen deposition (hematoxylin and eosin; original magnification, ×20).* B, *palisading pattern on immunohistochemical staining revealing the moderately cellular tumor with scattered vessels and a spindle-shaped appearance (hematoxylin and eosin; original magnification, ×100).* C, *diffuse staining with CD34 antibody in tumor cells (original magnification, ×100).* D, *vimentin staining showing focal staining in tumor cells (original magnification, ×100).*

The pathogenesis of this rare lesion is still unknown. The origin may be from perivascular connective tissue or from the pia-arachnoid as demonstrated in all our patients. All our cases were intradural nondural-based lesions. All seemed to be subpial in origin, similar to intramedullary exophytic tumors. During surgery, the pial surface of the spinal cord seemed to be involved by the tumor, suggesting this layer as the true origin for these tumors.

Solitary fibrous tumors must be histologically differentiated from meningioma, hemangiopericytoma, and other spindle-cell tumors. The distinct microscopic features for this tumor are the dense collagen bands and lack of specific architectural features such as psammoma bodies or whorls. Immunohistochemical stains are positive for CD34 antigen, whereas meningiomas are reactive for EMA and schwannomas for S-100. The staining of the tumor by CD34 and negative staining for S-100 and EMA distinguish solitary fibrous tumors from the other spindle-cell tumors. The exception is hemangiopericytomas, which may be positive for CD34 antigen; however, these tumors are typically hypercellular with necrosis and mitosis.

MRI scanning of these tumors revealed an isointense signal on T1-weighted sequences and a hypointense signal on T2-weighted sequences. Edema was only visualized in two patients. All the tumors enhanced homogeneously with gadolinium administration. The differential diagnosis of intraspinal solitary fibrous tumors includes intradural extramedullary tumors such as meningioma, schwannoma, and neurofibroma as well as intramedullary neoplasms. The characteristic T2 hypointense signal was not consistent with extradural or other intramedullary tumors. Solitary fibrous tumors are uniquely hypointense on this imaging sequence; thus, this sequence may generally indicate this tumor type.

Long-term Outcome

There have been only anecdotal reports suggesting that patients with spinal cord solitary fibrous tumors experience an indolent course (1–4, 6, 9, 10, 12, 13, 16–18). Because of the rarity of this tumor type, there is no series that investigates spinal cord solitary fibrous tumors. Our study supports the indolent course. These results confirm that patients with solitary fibrous tumors tend to survive for long periods after radical surgery alone. The survival of these patients is better than that reported for patients with other intramedullary neoplasms (5, 8, 14) and similar to that reported for patients with intradural extramedullary tumors such as meningiomas and schwannomas.

There are even fewer reports on the frequency of recurrence for spinal solitary fibrous tumors. There has been no recurrence in our series of patients, and there has been one report of recurrence of a conus tumor (4). The remaining case reports have relatively short-term or no follow-up. The overall rate of recurrence was 7.1% (*Table 3*). All our patients have been followed with routine semiannual or annual MRI scans. We think these tumors are indolent and can be followed with

TABLE 2. Summary of spinal solitary fibrous tumors[a]

Series (ref. no.)	Age (yr)/sex	Signs and symptoms at presentation	Location	Site	Follow-up
Carneiro et al., 1996 (4)	54/F	Progressive paraplegia	L1–L3	IM, EM	7 yr
	50/M	Bilateral leg pain, weakness, and paresthesias	?	ED	Death after recurrence
Alston et al., 1997 (1)	47/M	Brown-Séquard syndrome	T4–T5	IM	2 mo
Malek et al., 1997 (13)	33/M	Back and hip pain, and progressive myelopathy	T7–T8	EM	N/A
Brunori et al., 1999 (3)	38/F	Left lower extremity sciatica	T12–L1	EM	12 mo
Kanahara et al., 1999 (9)	62/M	Bilateral lower extremity paresthesias and decreased sensation	C6–C7	IM, EM	N/A
Donnellan et al., 2000 (6)	39/F	Back pain, and leg numbness	L1	ED	N/A
Mordani et al., 2000 (16)	33/M	Hand and feet paresthesia and paraparesis	C5	IM	18 mo
Vorster et al., 2000 (18)	51/M	Dysesthesia, paresthesias, and progressive paraparesis	T2–T3	EM	7 mo
Kurtkaya et al., 2001 (12)	70/F	Right leg weakness	T3	EM	12 mo
Obara et al., 2003 (17)	49/F	Quadriparesis	C2–C5	ED	12 mo
Bohinski et al., 2004 (2)	48/F	Neck pain and arm paresthesias	C4	IM, EM	10 mo
Kawamura et al., 2004 (10)	64/M	Right lower extremity weakness, Brown-Séquard syndrome, and hyperreflexia	T2–T3	IM	6 mo
Current study	59/M	Paresthesias, numbness, paraparesis, and scoliosis	T5	IM	4.8 yr
	37/F	Painful left foot, paresthesias, and ankle fracture	T2–T3	IM	5.0 yr
	41/M	Progressive dyesthesias in hands, and upper extremity weakness	C6–C7	IM	3.5 yr
	17/M	Spastic paraparesis, and scoliosis	T5, T6	IM	1.6 yr

[a] IM, intramedullary; EM, intradural-extramedullary; ED, extradural; N/A, not applicable.

annual imaging studies for several years and then every 5 years thereafter.

These tumors are typically isolated when found in the central nervous system. Although our youngest patient had multifocal disease with two intraspinal tumors, the literature does not support this presentation. However, there are reports of multifocal solitary fibrous tumors outside the central nervous system (15).

A comparison of the pre- and postoperative functional status at recent follow-up evaluation reveals that three patients were unchanged or improved. One patient developed worsening dysesthetic pain indicative of the intramedullary origin. The functional outcome of these patients does differ from that of patients with other intramedullary tumors as reported in previous studies (5, 7).

There is little information about the management of spinal cord solitary fibrous tumors. The extent of tumor resection did not influence the functional prognosis, because total resection was accomplished for all tumors. We advocate gross total resection for tumors that are well circumscribed and minimally involve the intramedullary spinal cord. Therefore, the overall survival rate, low recurrence rate, and good functional outcome observed are best attributed to the surgical extirpation of these tumors.

CONCLUSION

Our experience demonstrates that spinal cord solitary fibrous tumors are biologically indolent neoplasms. The tumor may be misdiagnosed unless an immunostaining panel is

TABLE 3. Summary of reported cases of spinal solitary fibrous tumors

No. of cases	17
Mean age	46.5 yr
Male/female ratio	1.4:1
Location	
Cervical	31.2%
Thoracic	56.3%
Lumbosacral	12.5%
Compartment	
Extradural	18%
Intradural-extramedullary	24%
Intramedullary	58%
Recurrence rate	7.1%

performed. Radical extirpation is associated with minimal morbidity and an excellent long-term prognosis. After radical resection is achieved, we recommend that patients be followed with semiannual to annual MRI scans thereafter. We do not advocate adjuvant radiotherapy because surgery alone seems to be curative.

REFERENCES

1. Alston SR, Francel PC, Jane JA Jr: Solitary fibrous tumor of the spinal cord. **Am J Surg Pathol** 21:477–483, 1997.
2. Bohinski RJ, Mendel E, Aldape KD, Rhines LD: Intramedullary and extramedullary solitary fibrous tumor of the cervical spine: Case report and review of the literature. **J Neurosurg (Spine)** 100:358–363, 2004.
3. Brunori A, Cerasoli S, Donati R, Giangaspero F, Chiappetta F: Solitary fibrous tumor of the meninges: Two new cases and review of the literature. **Surg Neurol** 51:636–640, 1999.
4. Carneiro SS, Scheithauer BW, Nascimento AG, Hirose T, Davis DH: Solitary fibrous tumor of the meninges: A lesion distinct from fibrous meningioma—A clinicopathologic and immunohistochemical study. **Am J Clin Pathol** 106:217–224, 1996.
5. Cooper PR, Epstein F: Radical resection of intramedullary spinal cord tumors in adults: Recent experience in 29 patients. **J Neurosurg** 63:492–499, 1985.
6. Donnellan RB, Govender D, Chite SH, Landers AT: An unusual presentation of solitary fibrous tumor. **Spine** 25:749–751, 2000.
7. Epstein FJ, Farmer J-P, Freed D: Adult intramedullary astrocytomas of the spinal cord. **J Neurosurg** 77:355–359, 1992.
8. Guidetti B, Mercuri S, Vagnozzi R: Long-term results of the surgical treatment of 129 intramedullary spinal gliomas. **J Neurosurg** 54:323–330, 1981.
9. Kanahara T, Hirokawa M, Shimizu M, Terayama K, Nakamura E, Hino Y, Mikawa Y, Manabe T, Abe S: Solitary fibrous tumor of the spinal cord: Report of a case with scrape cytology. **Acta Cytol** 43:425–428, 1999.
10. Kawamura M, Izawa K, Hosono N, Hirano H: Solitary fibrous tumor of the spinal cord: Case report and review of the literature. **Neurosurgery** 55:433, 2004.
11. Klemperer P, Rabin CB: Primary neoplasms of the pleura. **Arch Pathol** 11:385–412, 1931.
12. Kurtkaya O, Elmaci I, Sav A, Pamir MN: Spinal solitary fibrous tumor: Seventh reported case and review of the literature. **Spinal Cord** 39:57–60, 2001.
13. Malek AM, Weller SJ, Price DL Jr, Madsen JR: Solitary fibrous tumor presenting as a symptomatic intraspinal mass: Case report. **Neurosurgery** 40:844–847, 1997.
14. Minehan KJ, Shaw EG, Scheithauer BW, Davis DL, Onofrio BM: Spinal cord astrocytoma: Pathological and treatment considerations. **J Neurosurg** 83: 590–595, 1995.
15. Mohamed H, Mandal AK: Natural history of multifocal solitary fibrous tumors of the pleura: A 25-year follow-up report. **J Natl Med Assoc** 96:659–662, 2004.
16. Mordani JP, Haq IU, Singh J: Solitary fibrous tumour of the spinal cord. **Neuroradiology** 42:679–681, 2000.
17. Obara Y, Matsumoto M, Chiba K, Yabe H, Toyama Y, Mukai M: Solitary cervical fibrous tumor: Case illustration. **J Neurosurg** 98:111, 2003.
18. Vorster SJ, Prayson RA, Lee JH: Solitary fibrous tumor of the thoracic spine: Case report and review of the literature. **J Neurosurg** 92:217–220, 2000.

COMMENTS

The authors describe a series of four patients with intramedullary spinal cord fibrous tumors. This is an exceedingly unusual entity whose origin is unclear, as there was no apparent dural or arachnoid attachment. They are clearly distinct from spinal cord tissue and may be dissected from the adjacent cord tissue with maintenance or improvement of motor function, as the authors have demonstrated in this small group of patients. The presentation with sensory symptoms is a reflection of the central spinal location of these tumors adjacent to the spinothalamic tracts and their crossing fibers. Postoperative dysesthesias is similar to that observed with centrally located ependymomas and is again owing to perturbation of adjacent sensory pathways during operative resection. The authors state that the survival of patients with these tumors is better than that reported for other intramedullary tumors. However, survival for intramedullary tumors is very dependent on the histology. Hemangioblastomas and ependymomas can almost always be totally resected and patients with these lesions will have a normal survival, whereas the survival and subsequent deterioration of neurological function of patients with infiltrating glial tumors will be a function of tumor histology.

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Jallo et al. report a series of four patients suffering from spinal solitary fibrous tumors. It is the only series in the literature with such a long follow-up period in a very rare pathology. The experience of the authors on intraspinal cord tumors emphasizes the value of cautious surgical resection of those lesions without any adjunctive therapy. The data provided are unique in terms of magnetic resonance imaging, surgical management, histolology, pathogenesis, and follow-up. The authors should be commended for their excellent paper.

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The authors present a small series of four spinal solitary fibrous tumors, adding to the existing literature. Because this series will bring the total reported number of cases to 16, their experience with this neoplasm is instructive. Furthermore, the authors provide us with pearls on how to remove these tenacious lesions.

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