doi: 10.2176/jns-nmc.2024-0145

NMC Case Report Journal 11, 297-303, 2024

Recurrence of Solitary Fibrous Tumor in the Spinal Cord Following Gross Total and Subtotal Resection: A Case Report of Recurrence 19 Years of Post-total Resection and Systematic Literature Review

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Abstract

Spinal cord solitary fibrous tumors (SFTs), previously known as hemangiopericytoma (HPC), represent exceedingly rare neoplasms. Strategies for their management, such as appropriate follow-up duration, remain controversial due to their propensity for recurrence despite extended periods of quiescence.
We report a 51-year-old male presenting with new-onset back pain and gait disturbances, who had undergone gross total resection (GTR) of an SFT within the thoracic spinal cord 19 years ago. Magnetic resonance imaging of the thoracic spine revealed recurrent tumors at the T7 level within the spinal cord. Subsequent resection achieved GTR. A comprehensive literature review was undertaken to assess the benefits of different resection extents (gross total removal (GTR) vs. subtotal removal (STR)), adjuvant radiation therapy, and the optimal duration of postoperative follow-up.
Since 1960, 46 cases, including the present one, have reported recurrent spinal SFT/HPC following GTR and STR. Statistical analyses demonstrated that neither the type of resection nor adjuvant radiation therapy significantly impacted median recurrence-free survival in this cohort. Given their unpredictable behavior, meticulous lifelong follow-up following successful resection appears crucial for managing these tumors effectively.

Keywords: solitary fibrous tumor, hemangiopericytoma, total resection, adjuvant radiation therapy, long-term follow-up

Introduction

Solitary fibrous tumors (SFTs) primarily manifest intracranially, originate from mesenchymal tissue, and have a high recurrence rate. SFTs that occur in the spinal cord are considerably rare. Although complete surgical resection is considered the most advantageous approach for patients, debates on the efficacy of adjuvant radiation therapy and the optimal duration of postoperative follow-up persist. Within the literature, 45 instances of recurrent spinal SFTs following either gross total resection (GTR) or subtotal resection (STR) have been documented. Notably, most recurrences transpired within 5 years of post-initial resection, with two cases recurring after an interval exceeding 15 years subsequent to the initial subtotal resection procedure. We contribute an additional case of recurrent spinal SFT after initial GTR. In this instance, recurrent tumors manifested with spinal cord dysfunction 19 years after the initial surgery.

Case Report

A 31-year-old male presenting with gait disturbance sought evaluation from a neurologist. T1-weighted magnetic resonance (MR) images following Gd-DTPA administration revealed a homogenously enhanced mass in the thoracic spinal cord centered at T6. The mass lesion appeared to be located in the subpial region with an extra-

Received June 5, 2024; Accepted August 10, 2024

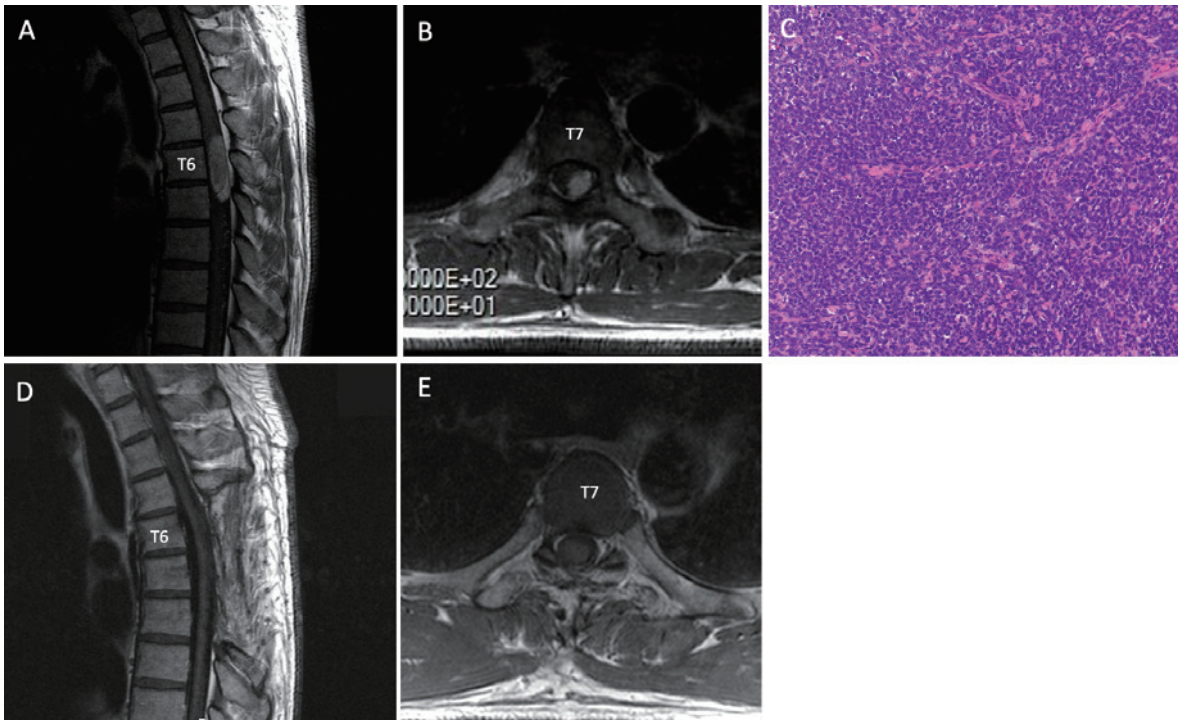

Fig. 1 Preoperative and Postoperative follow-up magnetic resonance imaging (MRI) of the first surgery.
A: Sagittal T1-weighted MR image after injection of Gd-DTPA before the first surgery revealed an enhanced tumor with an irregular margin by contrast.
B: Axial T1-weighted MR image after injection of Gd-DTPA before the first surgery revealed an enhanced mass in the spinal cord. The mass appeared to be located in the subpial region with extramedullary extension.
C: Photomicrographs of the specimen from the first surgery revealed atypical cells with short spindles or rounded nuclei, proliferating alongside staghorn-like vessels. These findings were consistent with hemangiopericytoma/solitary fibrous tumor (H&E staining).
D: Sagittal T1-weighted MR image at 4-year postoperative follow-up revealed no recurrent tumors.
E: Axial T1-weighted MR image at 4-year postoperative follow-up revealed no recurrent tumors.

medullary extension (Fig. 1A & B). The patient subsequently underwent thoracic laminectomy from T6 to T8 and tumor removal at a different medical facility. According to the operative note provided by the facility, intraoperative findings disclosed a subpial tumor with extramedullary extension. The surface of the mass was cauterized, and it was then circumferentially dissected from the pia mater and detached from the pial feeding arteries and draining veins. After the internal decompression by the cavitron ultrasonic surgical aspirator, the subpial spinal cord tumor and its extramedullary component were removed in a piece-by-piece fashion. In contrast to clear border between the rostral end of the tumor and the spinal cord, the caudal end of the tumor was strongly adherent to the pia and spinal cord. The mass was finally gross-totally resected without resulting neurological deficits. Microscopically, atypical cells with short spindles or rounded nuclei proliferated with staghorn-like vessels and mitotic activity was very low (Fig. 1C). Histopathological analysis confirmed a diagnosis of hemangiopericytoma. Following tumor removal, the patient experienced complete resolution of symptoms, and subsequent MR images after 4 years of the primary surgery revealed no recurrent mass (Fig. 1D & E). However, the patient was lost to follow-up.

The patient reported experiencing paresthesia in the right lower extremity and left ankle dorsiflexion impairment 19 years after the primary surgery. Progressive symptoms, along with new-onset middle back pain, prompted the patient to seek medical attention half a year after the new symptom started. Spinal MR images revealed the recurrent spinal cord tumor intramedullary at the T7 level, accompanied by a cystic component at the T5-6 level and marked edema surrounding the lesions (Fig. 2A-C). Subsequently, tumor resection through previous T6-8 laminectomy was performed at our hospital 19.5 years after the primary treatment. Once the cystic portion rostral to the mass was opened by sharp dissection of overlying pial layer, xanthochromic fluid was evacuated from the cyst. This procedure decreased the tension of the swollen spinal cord. Our attention was then directed to the tumor resection. The mass was adherent to the spinal cord. This finding suggested that the mass was a locally recurrent tumor

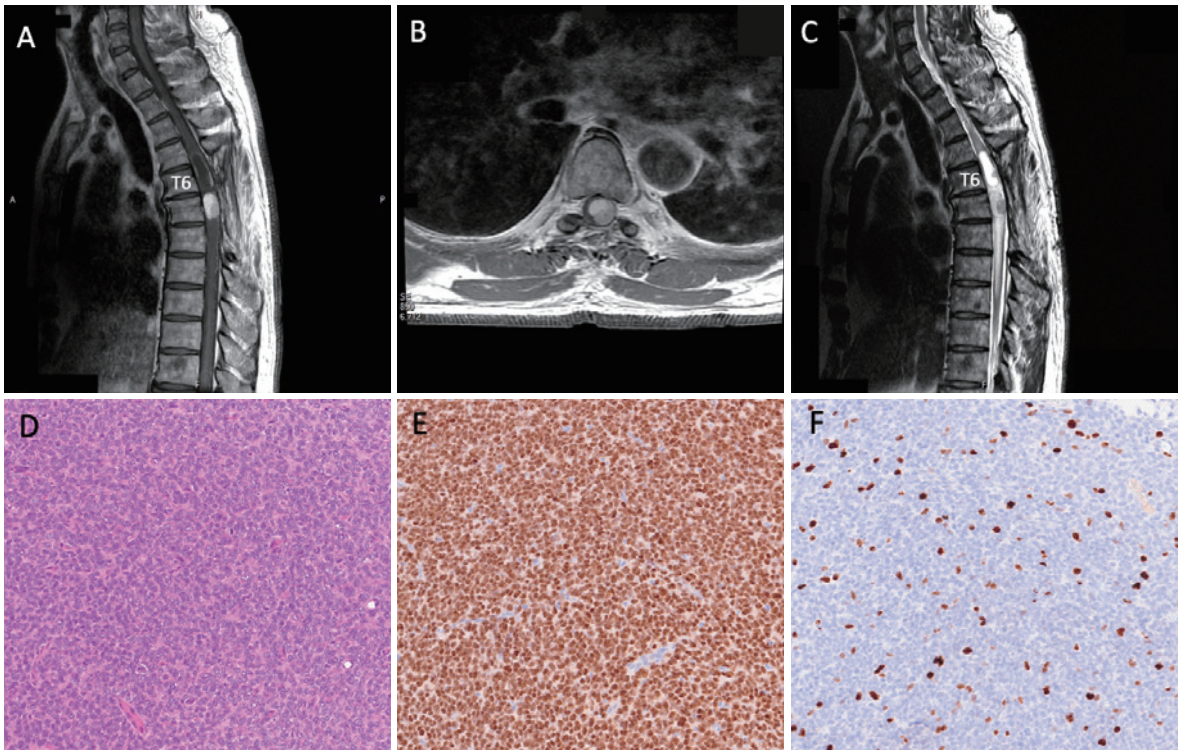

Fig. 2
A-C: Preoperative magnetic resonance imaging (MRI) before the second surgery.
A: Sagittal fat-suppressed T1-weighted MR image after an administration of Gd-DTPA demonstrated an enhanced mass lesion at the T7 level.
B: Postcontrast axial fat-suppressed T1-weighted MR image demonstrated a heterogeneously enhanced tumor with a mass effect on the spinal cord.
C: Preoperative sagittal T2-weighted MR image revealed hyperintense signals in the cystic lesion above the tumor at the T5 and T6 levels, accompanied by a tumor with iso-hypointensity at the T7 level.
D-F: Pathological specimen.
D: In the specimen from the second surgery, atypical cells with short spindle or round nuclei proliferated in alveolar or fascicular formations around small vessels with hyalinization.
E: Immunohistochemistry revealed STAT6 positive (nuclear stain).
F: Ki-67 labeling index was 12.4% in hot spot.
The findings above were consistent with solitary fibrous tumor, CNS WHO grade II.

rather than a newly developed one. As in the previous surgery, the mass was cauterized and circumferentially dissected from the surrounding pia and the spinal cord. The tumor was then removed in a piece-by-piece fashion. The surgery again resulted in GTR of the mass lesion and total collapse of the cyst. The patient experienced resolution of gait disturbance and other symptoms such as paresthesia postoperatively. Follow-up MR imaging revealed the cavity after tumor resection in the spinal cord without evidence of residual enhanced mass, and the patient successfully returned to work after surgery. We are currently considering adjuvant radiation therapy following the second surgery.

Histological analysis of the resected tumor revealed atypical cells with short spindle or round nuclei that proliferated in alveolar or fascicular formations around small vessels with hyalinization (Fig. 2D). Immunohistochemistry revealed STAT6 positive (nuclear stain) (Fig. 2E), and Ki-67 labeling index was 12.4% in a hot spot (Fig. 2F). We diagnosed this tumor as the SFT, CNS WHO grade II based on morphology and immunohistochemistry.

Literature Review

A systematic literature review was conducted in accordance with the PRISMA checklist[1]. The PubMed database was searched using the terms "spinal," "spine," "solitary fibrous tumor," "hemangiopericytoma," "recurrence," and "recurred" in combination in March 2024. The inclusion criteria were as follows: (1) the presence of at least one human case of recurrent SFT or HPC in the spine and (2) the availability of clinical data before and after the initial surgery. The exclusion criteria encompassed (1) cases reporting metastatic tumors from intracranial or other lesions, (2) lack of clarification on the extent of tumor removal

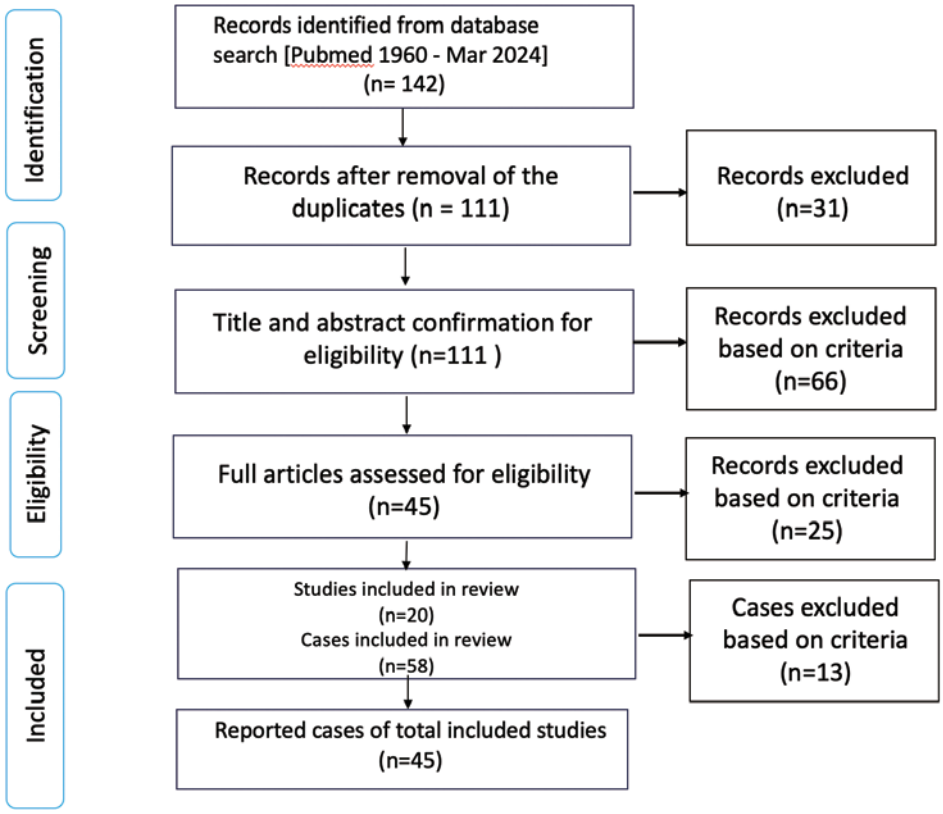

Fig. 3 PRISMA flow diagram.
PRISMA, Preferred Reporting Items for Systematic Reviews and Meta-Analyses.

(complete/gross total/subtotal/partial removal), (3) absence of information on adjuvant radiation therapy following initial surgery, (4) failure to mention recurrence-free survival (RFS) and follow-up time, and (5) articles not written in English. Search results were meticulously screened for duplicates and filtered based on eligibility criteria. As the definitions of GTR and subtotal removal (STR) were not necessarily consistent among previous reports, we decided to follow the author's descriptions and definitions of the extents of resection, such as GTR and STR. In our case, based on intraoperative findings, GTR was defined as no visible residual tumor observed under the microscope. STR was defined as the presence of visible residual tumor under the intraoperative microscope. Titles and abstracts were assessed for suitability, followed by a thorough review of each article in its entirety (Fig. 3). Statistical analysis of the data was performed using Easy R (Saitama Medical Center, Jichi Medical University) to assess whether factors such as patient age, sex, extent of tumor resection, and postoperative radiation therapy influenced RFS. The Mann-Whitney U test was used to evaluate differences in median RFS, whereas Spearman's rank correlation test was used to investigate the correlation between patient age and RFS.

Discussion

Originally, SFTs and hemangiopericytoma (HPC) were considered separate entities. HPC was classified as a low-grade sarcoma (WHO grade II), whereas SFT was classified as a benign tumor (WHO grade I) with a favorable prognosis following GTR.[2] In 2016, the WHO unified these tumors as SFT/HPC and delineated three grades within this group: grade I representing the SFT phenotype, grade II corresponding to the HPC phenotype, and grade III characterized by mitotic activity ≥ 5 /10 high power field (HPF).[3] Furthermore, in 2021, the WHO Classification of Tumors of the CNS integrated the terms SFT and HPC under the umbrella term SFT, being based on a disease-defining NAB2-STAT6 fusion.

There have been no randomized controlled trials for adjuvant therapy in spinal SFTs due to the rarity of this tumor in the spinal cord. The current standard of care for this disease is maximal safe resection; however, the post-resection tumor behavior remains unpredictable, and the roles of adjuvant radiation therapy (RTX) and chemotherapy (CTX) remain controversial.

Our systematic literature review identified 46 cases of recurrent spinal SFT/HPC following GTR or STR (Table 1).[4-18] The average age at initial diagnosis was 40 years (± 15.6 standard deviations), with a slight male preponder-

Table 1 Characteristics of the cases included in the systematic review

Case	Age	Sex	Extent of tumor resection	WHO Classification at first surgery	RFS (month)	Follow-up (month)	RT	Case report, year
1	39	M	GTR	NA	108	204	No	Pitlyk et al., 1965[4]
2	30	M	STR	NA (HPC)	132	145	Yes	Muraszko et al., 1982[5]
3	11	F	STR	NA (HPC)	39	72	Yes	Muraszko et al., 1982
4	35	M	GTR	NA (SFT)	78	102	No	Muñoz et al., 2008[7]
5	60	M	STR	NA (HPC)	96	276	No	Wu et al., 2009[8]
6	42	M	STR	NA (HPC)	36	60	No	Wu et al., 2009
7	56	M	GTR	NA (HPC)	6	36	No	Shirzadi et al., 2013[9]
8	31	F	STR	II	72	300	Yes	Liu et al., 2013[10]
9	23	F	GTR	III	28	72	Yes	Liu et al., 2013
10	31	F	STR	III	6	19.2	Yes	Liu et al., 2013
11	51	M	GTR	III	26	72	No	Liu et al., 2013
12	53	M	GTR	II	53	153.6	Yes	Liu et al., 2013
13	18	M	GTR	III	5	64.8	Yes	Liu et al., 2013
14	2	M	STR	III	4	8.4	No	Liu et al., 2013
15	56	F	STR	II	60	129.6	Yes	Liu et al., 2013
16	14	M	STR	III	24	48	Yes	Liu et al., 2013
17	25	F	GTR	III	4	55.2	Yes	Liu et al., 2013
18	37	M	GTR	II	36	72	Yes	Liu et al., 2013
19	73	F	STR	II	240	264	Yes	Liu et al., 2013
20	44	F	STR	II	84	153.6	Yes	Liu et al., 2013
21	44	M	STR	III	48	54	Yes	Liu et al., 2013
22	25	F	STR	III	6	33.6	Yes	Liu et al., 2013
23	53	M	STR	II	24	57.6	Yes	Liu et al., 2013
24	19	M	GTR	II	24	30	Yes	Liu et al., 2013
25	14	M	STR	III	6	24	Yes	Liu et al., 2013
26	36	F	GTR	II	54	120	Yes	Liu et al., 2013
27	40	M	STR	I	211	276	No	Kobayashi et al., 2014[11]
28	36	F	GTR	II	73	75	No	Jia et al., 2018[12]
29	42	M	STR	III	37	46	Yes	Jia et al., 2018
30	49	F	STR	III	17	24	Yes	Jia et al., 2018
31	57	M	STR	III	12	18	No	Jia et al., 2018
32	40	M	GTR	NA	36	36	No	Zhang et al., 2019[13]
33	37	F	GTR	III	22	62	No	Wang et al., 2019[14]
34	43	F	GTR	II	49	85	Yes	Wang et al., 2019
35	38	M	STR	III	26	59	No	Wang et al., 2019
36	45	F	STR	III	35	110	No	Wang et al., 2019
37	66	M	STR	III	25	47	No	Wang et al., 2019
38	35	F	STR	III	32	66	No	Wang et al., 2019
39	48	M	STR	III	21	80	No	Wang et al., 2019
40	46	F	STR	III	19	58	No	Wang et al., 2019
41	49	F	GTR	III	12	31	No	Murata et al., 2020[15]
42	50	M	GTR	III	52	62	Yes	Singla et al., 2020[16]
43	38	F	GTR	NA	165	213	No	Apra et al., 2022[17]
44	44	M	GTR	II	12	37	No	Apra et al., 2022
45	74	F	STR	III	84	120	No	Tomomatsu et al., 2023[18]
46	31	M	GTR	I	234	237	No	Current case

GTR, gross total resection; STR, subtotal resection; WHO, World Health Organization; NA, not applicable; HPC, hemangiopericytoma; SFT, solitary fibrous tumor; RFS, recurrence-free survival; RT, radiation therapy

ance (56.5%). The mean RFS from the first GTR/STR was 53.8 months (median, 35.5 months; interquartile range (IQR), 19.5-69), and the mean follow-up time was 95 months (median, 65.4 months; IQR, 8.4-120). Notably, there was no significant correlation between age of disease onset and RFS ($r = 0.20$, $n = 46$, $p = 0.19$).

For intracranial cases, Lee et al. demonstrated the significant impact of GTR on controlling progression-free survival (PFS) and local control (LC).[19] Conversely, in spinal cases, Liu et al. found no notable benefit in terms of time to recurrence and survival time regardless of the extent of resection in their study of 26 cases of spinal SFTs.[10] Building upon this research, our systematic review identified 46 reported cases of recurrent spinal SFTs following either GTR or STR.[4-18] Of 46 cases, 20 cases (42.6%) initially underwent GTR, whereas 26 cases (56.5%) involved STR. Despite these differences in initial surgical approaches, our analysis revealed no significant disparity in RFS between the GTR group (median, 36 months; IQR, 19.5-58.8) and the STR group (median, 33.5 months; IQR, 19.5-69.0) ($p = 0.89$).

Although the efficacy of adjuvant radiotherapy remains controversial, it has been considered beneficial due to the heightened risk of recurrence in SFTs. Piscopo et al. observed a trend toward reduced disease recurrence and improved survival in a cohort of 21 cases, including both intracranial and spinal SFTs.[20] On the other hand, Liu et al. noted lower metastasis rates associated with radiotherapy, although they did not observe a significant enhancement in local recurrence control.[10] Of 45 reported spinal SFT cases, 23 received radiation therapy, including stereotactic radiosurgery (SRS), following either GTR or STR. However, there was no significant difference in RFS between the 23 cases with radiotherapy (median, 37 months; IQR, 20.5-81.0) and the 23 cases without radiotherapy (median, 32 months; IQR, 20.0-53.5) ($p = 0.84$). Of 20 cases (45%) where GTR was achieved, 9 received radiation therapy, while 11 (55%) did not. However, radiation therapy again did not demonstrate a significant difference in RFS between patients who underwent RTX after GTR (9 patients, median RFS, 36 months; IQR, 24-52 months) and those who did not receive RTX after GTR (11 patients, median RFS, 36 months; IQR 17-93 months) ($p = 0.40$). One might speculate whether the WHO grade influenced the decision for radiation therapy. However, there was no apparent correlation between WHO grades and the decision to administer radiation therapy. Of the nine patients who underwent RTX after GTR, five had WHO grade II tumors, and four had WHO grade III tumors. Conversely, among the 11 patients who had GTR without RTX, 2 had grade I tumors, 2 had grade II tumors, and 3 had grade III tumors. The remaining patients' pathological grading could not be specified. This variability in radiation therapy decisions based on histological grades suggests a lack of consensus on the appropriate indications for radiotherapy in spinal SFT cases.

Ghia et al. demonstrated that a combination of GTR and radiation therapy with a dosage exceeding 60 Gy was associated with enhanced LC in intracranial SFTs.[21] However, due to the spinal cord's lower tolerance to higher radiation doses compared with the brain, the maximum tolerable therapeutic radiation dose for spinal cord lesions is estimated to be between 45 and 50 Gy.[22] This limited radiation dosage to the spinal cord lesion partially explains our review's inability to highlight the benefits of radiation therapy in terms of RFS. Although this systematic review failed to demonstrate evidence supporting the efficacy of RTX after GTR or STR of spinal SFTs, we still believe that it is beneficial for the patient in the present case report to undergo RTX, given the recurrent nature of the tumor after 19 years of silence and its high Ki-67 proliferative index. Considering the uncertain roles of GTR and RTX (radiation therapy) in our review, the importance of long-term follow-up for this disease persists even after GTR.

Our case report highlights the recurrence of a spinal cord SFT 19 years after GTR, emphasizing the challenge of long-term disease management. GTR remains the primary treatment option for spinal SFTs, provided that it is feasible. However, the efficacy of adjuvant radiotherapy following gross or STR remains controversial, as suggested by our review.

Conclusions

We have reported a case of recurrent spinal cord SFT 19 years after the primary total resection. Our findings emphasize the importance of lifelong follow-up for patients with spinal SFTs, even after successful tumor resection. Continuous monitoring is essential for early detection of recurrence and optimizing long-term outcomes in these cases.

Abbreviations

SFT, solitary fibrous tumor; HPC, hemangiopericytoma; GTR, gross total resection; STR, subtotal resection; WHO, World Health Organization; RFS, recurrence-free survival; PFS, progression-free survival; LC, local control; RTX, radiation therapy

Informed Consent

Informed consent was obtained from all patients included in the study.

Conflicts of Interest Disclosure

The authors declare no conflicts of interest.

References

1) Page MJ, McKenzie JE, Bossuyt PM, et al.: The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. *BMJ* 372: n71, 2021
2) Shidoh S, Yoshida K, Takahashi S, Mikami S, Mukai M, Kawase T: Parasagittal solitary fibrous tumor resembling hemangiopericytoma. *Brain Tumor Pathol* 27: 35-38, 2010
3) Louis DN, Perry A, Wesseling P, et al.: The 2021 WHO Classification of Tumors of the central nervous system: a summary. *Neuro Oncol* 23: 1231-1251, 2021
4) Pitlyk PJ, Dockery MB, Miller RH: Hemangiopericytoma of the spinal cord: report of three cases. *Neurology* 15: 649-653, 1965
5) Muraszko KM, Antunes JL, Hilal SK, Michelsen WJ: Hemangiopericytomas of the spine. *Neurosurgery* 10: 473-479, 1982
6) Pakasa NM, Pasquier B, Chambonnière ML, et al.: Atypical presentations of solitary fibrous tumors of the central nervous system: an analysis of unusual clinicopathological and outcome patterns in three new cases with a review of the literature. *Virchows Arch* 447: 81-86, 2005
7) Muñoz E, Prat A, Adamo B, Peralta S, Ramón Y Cajal S, Valverde C: A rare case of malignant solitary fibrous tumor of the spinal cord. *Spine* 33: E397-E399, 2008
8) Wu W, Shi JX, Cheng HL, et al.: Hemangiopericytomas in the central nervous system. *J Clin Neurosci* 16: 519-523, 2009
9) Shirzadi A, Drazin D, Gates M, et al.: Surgical management of primary spinal hemangiopericytomas: an institutional case series and review of the literature. *Eur Spine J* 22(supple 3): S450-S459, 2013
10) Liu HG, Yang AC, Chen N, Yang J, Qiu XG, Zhang JG: Hemangiopericytomas in the spine: clinical features, classification, treatment, and long-term follow-up in 26 patients. *Neurosurgery* 72: 16-24, 2013
11) Kobayashi K, Imagama S, Ito Z, et al.: Recurrence of solitary fibrous tumor of the cervical spinal cord. *Nagoya J Med Sci* 76: 217-223, 2014
12) Jia Q, Zhou Z, Zhang D, et al.: Surgical management of spinal solitary fibrous tumor/hemangiopericytoma: a case series of 20 patients. *Eur Spine J* 27: 891-901, 2018
13) Zhang YW, Xiao Q, Zeng JH, Deng L: Solitary Fibrous Tumor of the lumbar spine resembling schwannoma: case report and review of the literature. *World Neurosurg* 124: 121-124, 2019
14) Wang J, Zhao K, Han L, et al.: Solitary fibrous tumor/hemangiopericytoma of spinal cord: a retrospective single-center study of 16 cases. *World Neurosurg* 123: e629-e638, 2019
15) Murata K, Endo K, Aihara T, et al.: Salvage carbon ion radiotherapy for recurrent solitary fibrous tumor: A case report and literature review. *J Orthop Surg (Hong Kong)* 28: 2309499019896099, 2020
16) Singla R, Singh PK, Khanna G, et al.: An institutional review of 10 cases of spinal hemangiopericytoma/solitary fibrous tumor. *Neurol India* 68: 448-453, 2020
17) Apra C, El Arbi A, Montero AS, Parker F, Knafo S: Spinal solitary fibrous tumors: an original multicenter series and systematic review of presentation, management, and prognosis. *Cancers* 14, 2022
18) Tomomatsu Y, Takasawa E, Shiba S, et al.: Separation surgery and adjuvant carbon ion radiotherapy for a recurrent solitary fibrous tumor/hemangiopericytoma: a case report. *Spine Surg Relat Res* 7: 402-405, 2023
19) Lee JH, Jeon SH, Park CK, et al.: The role of postoperative radiotherapy in intracranial solitary fibrous tumor/hemangiopericytoma: a multi-institutional retrospective study (KROG 18-11). *Cancer Res Treat* 54: 65-74, 2022
20) Piscopo AJ, Chowdhury AJ, Teferi N, et al.: Surgical management of craniospinal axis solitary fibrous tumors: a single-institution case series and comprehensive review of the literature. *Neurosurgery* 94: 358-368, 2024
21) Ghia AJ, Chang EL, Allen PK, et al.: Intracranial hemangiopericytoma: patterns of failure and the role of radiation therapy. *Neurosurgery* 73: 624-630, 2013
22) Kirkpatrick JP, Kogel AJ van der, Schultheiss TE: Radiation dose-volume effects in the spinal cord. *Int J Radiat Oncol Biol Phys* 76: S42-S49, 2010

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