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Case Report

Spinal intradural solitary fibrous tumor/hemangiopericytoma with intramedullary invasion mimicking a hemangioblastoma

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ABSTRACT

Background: Solitary fibrous tumor/hemangiopericytomas (SFT/HPCs) are rare mesenchymal tumors of nonmeningothelial origin that comprises <1% of all central nervous system tumors.

Case Description: A 45-year-old male presented with sleep apnea (apnea-hypopnea index was 17.1 events/hour) and dysesthesias of the right upper and lower extremities. The magnetic resonance demonstrated a heterogeneous intradural extra-axial C1 mass with syringobulbia and syringomyelia. The right vertebral angiography revealed a hypervascular mass (i.e., intense tumor staining). With the preoperative diagnosis of a spinal hemangioblastoma, the patient underwent tumor removal. However, intraoperative findings demonstrated that the ventral component of the tumor was intramedullary without a dural attachment. Further, the histological diagnosis was consistent with SFT/HPC (HPC phenotype). The postoperative course was uneventful, and the patient's symptoms and the syrinxes spontaneously regressed.

Conclusion: A 45-year-old male presented a rare spinal intradural lesion at C1 appeared to be a spinal hemangioblastoma, but proved to be SFT/HPC (HPC phenotype) with intramedullary invasion.

Keywords: Hemangioblastoma, Hemangiopericytoma, Intramedullary invasion, Solitary fibrous tumor

INTRODUCTION

Hemangiopericytoma (HPC) and solitary fibrous tumor (SFT) are rare intradural mesenchymal tumors that are difficult to differentiate from meningiomas, schwannomas, ependymomas, astrocytomas, and hemangioblastomas, and constitute < 1% of all central nervous system (CNS) tumors.[3,8] Although SFTs are generally considered benign and HPCs are more malignant,[1] in 2016, the World Health Organization (WHO) combined these two as they share common "inversions" at the genetic site 12q13 (i.e., leading to STAT6 nuclear expression).[3] Here, a 45-year-old male presented with a C1 intradural lesion documented on magnetic resonance (MR) with accompanying syringobulbia/syringomyelia that proved to be intramedullary in location and histologically consistent with a SFT/HPC (HPC phenotype).

CASE REPORT

Clinical presentation

A 45-year-old male presented with numbness in his right forearm, hand, and leg of 2 months' duration. He was found to have sleep apnea (i.e., polysomnography [PSG] showed apnea-hypopnea index [AHI] of 17.1 events/hour [normal range: <5/h]) and a history of hypertension, hypercholesterolemia, asthma, and a lumbar disc herniation. On examination, he had dysesthesias in the right occipital region, the right C5–C7 distributions, the medial right lower leg, and the medial sole of the foot.

MR evaluation

The cervical MR imaging (MRI) revealed a heterogeneously enhancing right-sided dorsal intradural/extramedullary mass with pial/subpial invasion most consistent with a spinal hemangioblastoma at the C1 level. It was isointense on T1 MR and moderately hyperintense on T2 MR studies; flow voids surrounded the tumor [Figure 1]. There was accompanying syringomyelia and syringobulbia, and there was no enhancement of the cyst/syrinx walls [Figure 1]. The right vertebral angiography documented a hypervascular C1 lesion fed by the C1/C2 radicular arteries [Figure 2].

Surgery

A suboccipital craniotomy, with laminectomy of the atlas, and cephalad laminotomy of C2 (C0-C2) were performed utilizing neurophysiological monitoring. Once the dura was opened, a bright red vascular lesion was identified on the dorsal aspect of the cord. However, the tumor's ventral component was intramedullary in location. Once the pial feeding vessels were coagulated, the tumor was circumferentially separated and resected *en bloc* [Figure 3].

Histopathology

Histopathology confirmed that the tumor was a WHO Grade II SFT/HPC (HPC phenotype). It demonstrated round/spindle cell proliferation and compact chromatin-stained nuclei that were relatively uniform without mitotic figures (i.e., consistent with a benign lesion). Immunohistochemistry was positive for CD34 and CD31 and negative for Bcl-2; the Ki-67 index was 2%. In addition, many small and large blood vessels with characteristic Staghorn vasculature were evident throughout [Figure 4].

Postoperative course

Postoperatively, the numbness of the right upper and lower extremities improved. The postoperative PSG showed

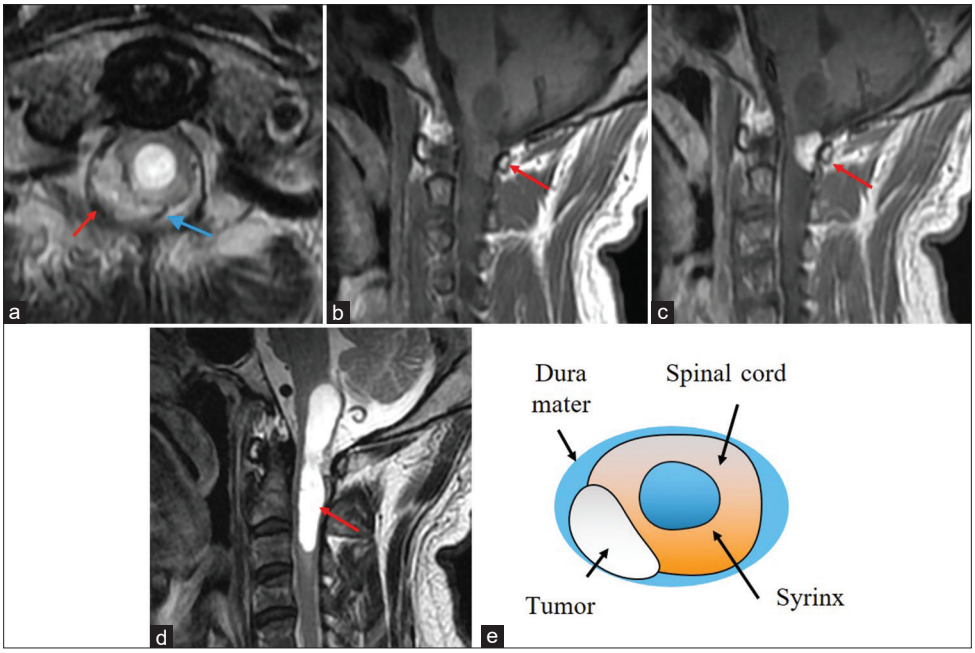

Figure 1: T2-weighted axial (a), T1-weighted sagittal (b), and T1-weighted contrast-enhanced sagittal magnetic resonance imaging (c) showing an intradural extramedullary mass at the C1 level (red arrow in (a),(b),(c)). The mass is hyperintense on T2 and isointense on T1 and is heterogeneously enhanced with gadolinium. Flow voids can be seen around the tumor (blue arrow) (a). T2-weighted sagittal MRI reveals a syringobulbia and syringomyelia (red arrow) (d). A scheme demonstrates the anatomical relationship between the tumor and the spinal cord (e). The tumor is an extramedullary mass with pial invasion or a subpial mass with extramedullary extension.

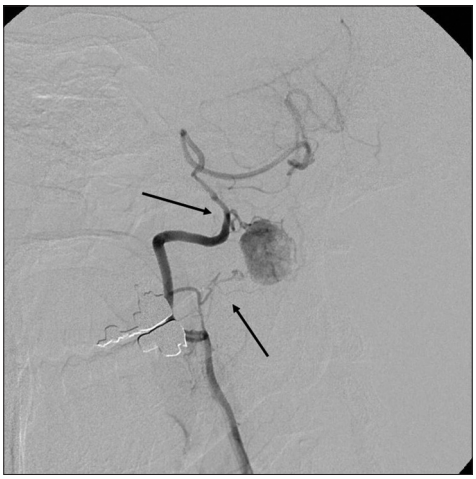

Figure 2: Digital subtraction angiogram of the right vertebral artery showing dense tumor stain supplied by C1 and C2 radicular arteries (arrow).

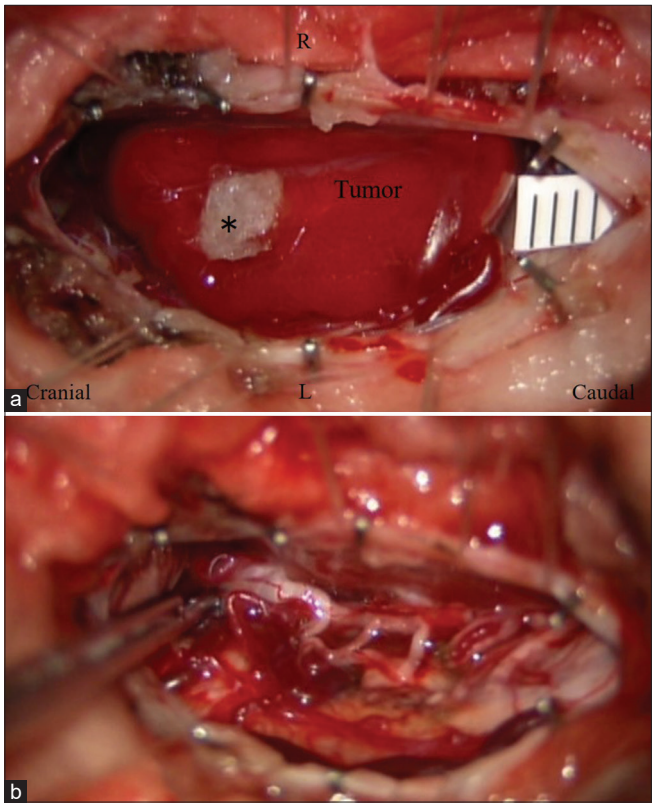

Figure 3: Intraoperative picture showing intradural extramedullary mass with pial invasion. It was red, vascular, and richly supplied by blood vessels (a). Oozing from the tumor was controlled by hemostatic materials (*). Total gross removal was accomplished (b).

improvement as his AHI decreased to 0.4 events/h, and his dysphagia disappeared. No further radiation was scheduled due to the complete tumor excision. Eight years later, he has exhibited no tumor recurrence [Figure 5].

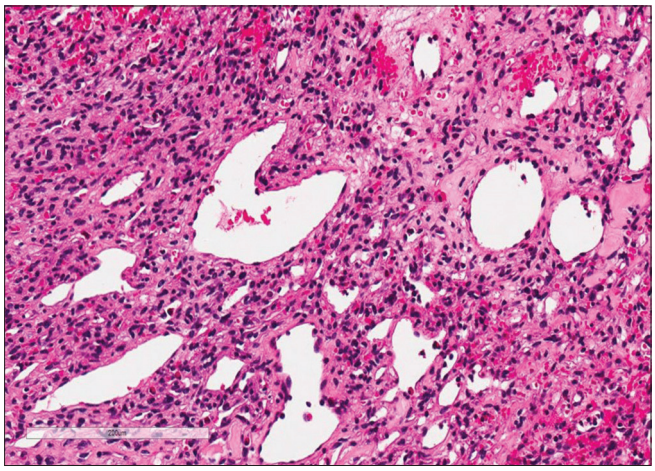

Figure 4: A photograph of the surgical specimen shows large vascular spaces with the typical Staghorn appearance of the vessels.

Table 1: Summary of spinal intradural SFT/HPC with subpial invasion in the literature.

Characteristics	*n*
Total patients[1-8]	34
Age (years)	
Mean	48.1
Range	15–83
Sex	
Male	21
Female	13
Original diagnosis[1-8]	
SFT	23
HPC	11
Location[1-8]	
Cervical	12
Cervicothoracic	2
Thoracic	18
Lumber	2
Preoperative diagnosis[1-8]	
Meningioma	7
Schwannoma	6
Hemangioblastoma	5
Ependymoma	3
Neurofibroma	3
Astrocytoma	2
Others	7

HPC: Hemangiopericytoma, N: Total number of patients, SFT: Solitary fibrous tumor

DISCUSSION

Case summary

SFT/HPC, nonmeningothelial mesenchymal tumors that are rarely intradural/extramedullary account for <1% of all CNS tumors [Table 1].[3] Here, the MRI revealed a well-demarcated and intensely enhancing mass on the dorsal surface of the

Table 2: Radiological characteristics of the spinal intradural SFT/HPC with subpial invasion reported in the literature.

MRI	SFT		HPC		SFT/HPC	
	T1WI	T2WI	T1WI	T2WI	T1WI	T2WI
Isointense (%)	69.6	0	100	18.2	79.4	5.9
Hypointense (%)	26.1	82.6	0	9.1	17.7	58.8
Hyperintense (%)	4.3	13	0	72.7	2.9	32.4
Syrinx/edema (%)	8.7/69.6		27.3/36.4		14.7/58.8	
Homogeneous enhancement (%)	87		63		79.4	

HPC: Hemangiopericytoma, MRI: Magnetic resonance imaging, SFT: Solitary fibrous tumor, WI: Weighted image

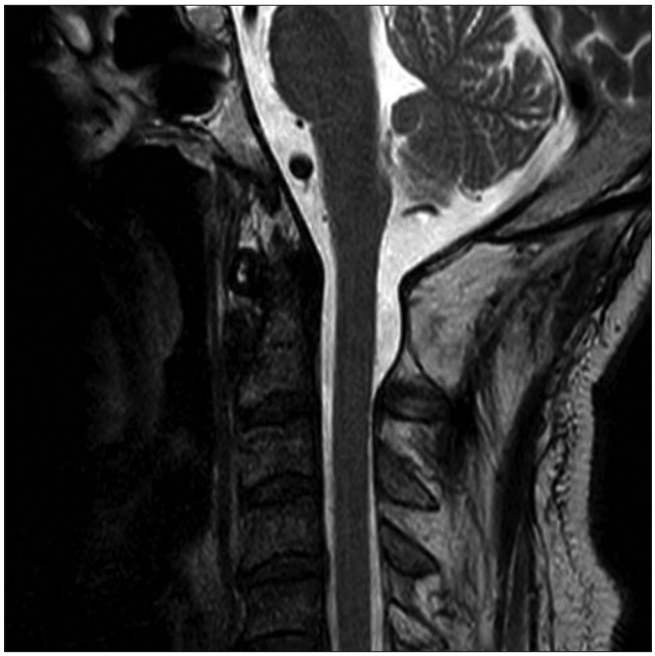

Figure 5: The postoperative T2-weighted magnetic resonance imaging 6 years after the operation shows complete tumor resection with a resolution of syringobulbia and syringomyelia.

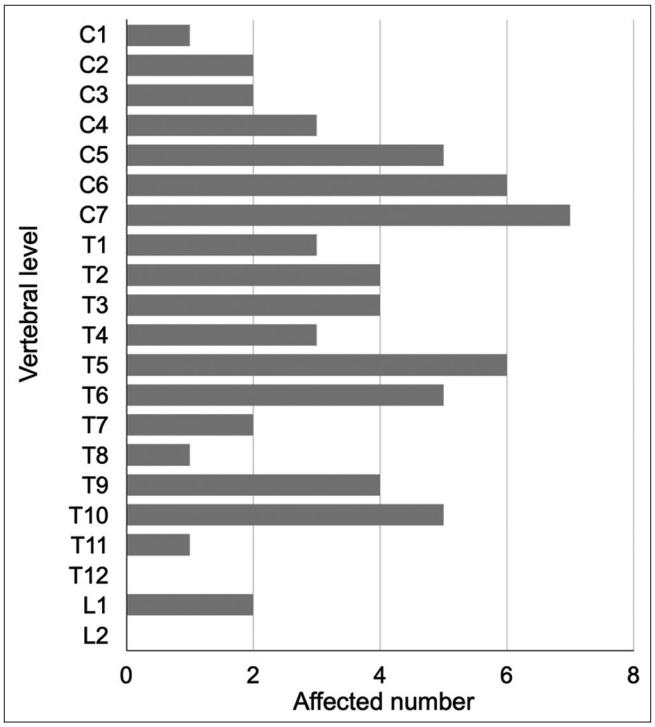

Figure 6: Anatomic location of the tumor. The tumor was most commonly located in the thoracic region followed by the cervical and lumbar levels.

cord surrounded by vascular flow voids (i.e., hypervascular on vertebral angiogram) with accompanying syringobulbia/syringomyelia prompting the preoperative diagnosis of a hemangioblastoma.[4] However, at surgery, this 45-year-old male's C1 lesion was both intradural and intramedullary (i.e., subpial invasion), and pathologically proved to be a SFT/HPC (HPC phenotype).

Literature summary

We found 34 cases of SFT/HPC cases (i.e., including this case) in our review of case studies from the literature [Supplemental Table 1].[1-8] Patients averaged 48.1 years of age, and there was a 1.6:1 male-to-female ratio [Table 1].[1-8] Tumors were typically located in the lower cervical region followed by the upper and midthoracic regions, but were only rarely encountered in the lumbar spine [Figure 6]. T1 MR findings for SFT/HPC were nonspecific. However, the hypointensity on T2 MR best correlated with the collagen-rich content, hypocellularity, and calcification of the SFT [Table 2].[2] The additional findings of syrinx/edema in the spinal cord were also common with intradural SFT/HPC and subpial invasion [Table 2].

Treatment and long-term prognosis

Following WHO 2016 guidelines, gross total resection (GTR) with radiotherapy is the mainstay for the treatment of the CNS SFT/HPC that typically result in good outcomes [Table 3].[3] In contrast subtotal resections usually have high recurrence and metastasis rate and are fatal.[7,8] Due to paucity of strong evidence, the role of postoperative radiotherapy in recurrence-

Table 3: Treatment and outcome in the spinal intradural SFT/HPC with subpial invasion.

	n	Mean follow-up (months)	Recurrence
Treatment			
GTR	28	27	No
GTR+RT	2	49.5	No
STR	3	6	No

HPC: Hemangiopericytoma, GTR: Gross total resection, N: Total number of patients, RT: Radiotherapy, SFT: Solitary fibrous tumor, STR: Subtotal resection

free survival is yet to be established.[2,5,6] Our patient, GTR was made possible by a clear plane between the tumor and cord, allowing for the 8-year follow-up without tumor recurrence.

CONCLUSION

Here, we reported a 45-year-old male with a C1 spinal intradural SFT/HPC (HPC phenotype) with an intramedullary invasion that resulted in MR-documented syringomyelia and syringobulbia. Following GTR, the patient remained asymptomatic 8 years later.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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SUPPLEMENTAL TABLE

Supplemental Table 1: Summary of spinal intradural solitary fibrous tumor/hemangiopericytoma with subpial invasion in the literature

No	Author /Year	Age Sex	Phen -otype	Location	Treatment	Recurrence	FU month	*Preoperative diagnosis*	*Angio-gram*	*MRI T1WI*	*MRI T2WI*	*MRI Gd-DTPA*	*MRI remark*
1	Kanahara /1999	62 M	SFT	C6-C7	NA	NA	NA	NA	NA	Hypo	Hypo	Marginally enhanced	Edema
2	Mordani /2000	33 M	SFT	C5	GTR	No	18	NA	NA	Iso	Hypo	Heterogeneous	Edema
3	Kawamura /2004	64 M	SFT	T2-T3	STR	No	6	Schwannoma, neurofibroma, meningioma, astrocytoma, ependymoma	NA	Hypo	Hypo	Homogeneous	Edema
4	Bohinski /2004	49 F	SFT	C4	GTR	No	10	NA	NA	Iso	Hypo	Homogeneous	Edema
5	Jallo /2005	59 M	SFT	T5	GTR	No	56	Intradural tumor	NA	Iso	Hypo	Homogeneous	NA
6	Jallo /2005	37 F	SFT	T2-3	GTR	No	60	Intradural tumor	NA	Iso	Hypo	Homogeneous	NA
7	Jallo /2005	41 M	SFT	C6-C7	GTR	No	41	Intradural tumor	NA	Iso	Hypo	Homogeneous	Edema
8	Jallo /2005	17 M	SFT	T5-T6	GTR	No	18	Intradural tumor	NA	Iso	Hypo	Homogeneous	Edema
9	Ogawa /2005	63 F	SFT	T11	GTR	No	18	NA	No tumor staining	Hypo	Hypo	Homogeneous	Edema
10	Ogungbo /2005	53 M	SFT	C3-C4	GTR	NA	NA	Hemangioblastoma	NA	Iso	Hypo	Homogeneous	Syrinx
11	Kashiwazaki /2007	31 M	HPC	T4-T6	GTR	No	36	Hemangioblastoma, subpial schwannoma	NA	Iso	Hyper	Heterogeneous	Edema flow void
12	Endo/2008	62 M	HPC	L1	GTR	NA	48 days	NA	Tumor staining+not hypervascular	Iso	Hyper	Homogeneous	Edema
13	Chou/2009	80 M	HPC	T10	GTR	No	36	Primary neurogenic tumor, meningioma	NA	Iso	Hyper	Homogeneous	Edema
14	Ishii/2009	63 F	SFT	C5	GTR	No	14	Schwannoma, neurofibroma, meningioma, astrocytoma, ependymoma	NA	Iso	Hypo	Homogeneous	Edema

(Contd...)

Supplemental Table 1: *(Continued).*

No	Author /Year	Age Sex	Phen -otype	Location	Treatment	Recurrence	FU month	*Preoperative diagnosis*	*Angio-gram*	*MRI T1WI*	*MRI T2WI*	*MRI Gd-DTPA*	*MRI remark*
15	Ciappetta /2010	75 F	SFT	T6-T7	GTR	No	24	NA	NA	Hypo	Hypo	Homogeneous	Edema
16	Ackerman /2011	58 M	HPC	T10	GTR	NA	NA	Meningioma, schwannoma, neurofibroma, ependymoma	-	Iso	Hypo	Homogeneous	Edema
17	Moscovici /2011	20 M	HPC	T9-T10	GTR	No	24	Meningioma, schwannoma	NA	Iso	Iso	Homogeneous	Cyst
18	Mariniello /2012	67 M	SFT	C4-C7	GTR	No	12	NA	NA	Iso	Hypo	Homogeneous	Edema
19	Mariniello /2012	75 M	SFT	T6-T7	GTR	No	12	NA	NA	Hypo	Hypo	Homogeneous	NA
20	Liu/2013	32 F	HPC	T5-T6	GTR+RT	No	79	Hemangioblastoma	NA	Iso	Hyper	Heterogeneous	
21	Liu/2013	24 M	HPC	C5-C7	GTR+RT	No	20	Ependymoma	NA	Iso	Hyper	Heterogeneous	Cyst
22	Hwang /2014	48 M	SFT	C7-T1	STR	No	6	NA	NA	Iso	Hyper	Homogeneous	
23	Robert /2014	49 F	SFT	T9-T10	STR	No	6	NA	NA	Hypo	Hypo	Homogeneous	Edema
24	Turk/2015	19 F	HPC	C2	GTR	NA	NA	NA	NA	Iso	Hyper	Homogeneous	Syrinx
25	Turk/2015	15 F	HPC	T9-T10	GTR	NA	NA	NA	NA	Iso	Hyper	Homogeneous	Syrinx
26	Bruder /2015	83 M	SFT	T8-T9	GTR	No	8	Meningioma	NA	Hyper	Hypo	Homogeneous	Edema
27	Walker /2015	47 F	SFT	L1	GTR	No	12	Ependymoma, cavernoma	No tumor staining	Iso	Hypo	Homogeneous	Cyst Hemorrhage
28	Wang/2016	31 M	SFT	T1-T5	GTR	NA	NA	NA	NA	Iso	Hetero	Heterogeneous	Syrinx
29	Yang/2019	35 F	SFT	C6-T1	GTR	No	23	Hemangioblastoma	Hypervascular on CT angio-gram	Iso	Hyper	Homogeneous	Edema
30	Chungyang /2020	80 F	HPC	T4-T5	GTR	No	10	NA	NA	Iso	Iso	Homogeneous	
31	Rodriguez-Mena /2020	30 M	SFT	C2-C3	GTR	No	24	NA	Tumor staining +	Iso	Hyper	Homogeneous	Edema
32	Rodriguez-Mena /2020	41 M	SFT	C5-C7	GTR	No	36	NA	NA	Iso	Hypo	Homogeneous	Edema
33	Yotsuya /2021	47 F	SFT	T2-T3	GTR	No	6	Intramedullary tumor	NA	Iso	Hypo	Homogeneous	Edema
34	Present case	46 M	HPC	C1	GTR	No	84	Hemangioblastoma	Tumor staining +Hypervascular	Iso	Hyper	Heterogeneous	Syrinx, flow void

HPC, hemangiopericytoma; GTR, gross total resection; MRI; magnetic resonance imaging; NA, not available; OP, operative; SFT, solitary fibrous tumor; WI, weighted imaging