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How to differentiate spinal cord hemangiopericytoma from common spinal cord tumor?

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1 **How to differentiate spinal cord hemangiopericytoma from common spinal**
2 **cord tumor?**

3
4 **Abstract**

5
6 **Introduction:** Tumors of the spinal cord are rare and some can be confused with each other.
7 We report a rare spinal cord solitary fibrous tumor/hemangiopericytoma (SFT/HPC), and
8 propose keys to differentiate spinal cord tumors from each other.

9 **Case report:** A 67-year-old man presented weakness with recent diffuse sensory disorders in
10 the right lower limb. Spinal MRI revealed a T8-T9 intradural extramedullary mass with spinal
11 cord compression. Gross total resection of a poorly vascularized intradural tumor was
12 achieved. It was an encapsulated extramedullary tumor, which was difficult to separate from
13 the spinal cord due to the presence of pial adhesions. Definitive diagnosis was grade 1
14 SFT/HPC of the spinal cord. One-year follow-up MRI revealed complete excision without
15 any evidence of residual tumor.

16 **Conclusion:** SFT/HPC is a very rare spinal tumor that can be extramedullary, intramedullary
17 or both. It may perfectly mimic meningioma. The maximal resection is the best treatment, but
18 can be challenging because of the tumor's firm consistency and pial adherences to the spinal
19 cord. Outcome is good in case of gross total resection, but there is a risk of very late
20 recurrence, requiring long-term follow-up.

21
22
23 **Abbreviations:** HPC: Hemangiopericytoma; MRI: Magnetic Resonance Imaging; SFT:
24 Solitary Fibrous Tumor

25
26 **Key-words:**

1 **Introduction**

2 Solitary Fibrous Tumor (SFT) and hemangiopericytoma (HPC) are a rare spinal cord entity.
3 We report a challenging case of spinal cord SFT/HPC and propose some key features to help
4 differentiate this unusual entity from other more frequent spinal cord tumors. For the first
5 time, we describe differential diagnosis between SFT/HPC and common spinal tumors such as
6 meningioma, schwannoma, ependymoma and astrocytoma, based on clinical, MRI, surgical,
7 follow-up and histological characteristics.

8

9 **Clinical Presentation**

10 A 67-year-old man presented with asthenia and weakness with recent diffuse sensory
11 disorders in the right lower limb. He also described a sensation of unstable gait. However,
12 neurological examination did not find signs of focal sensorimotor deficit, and walking was in
13 fact normal without help. Spinal MRI revealed a T8-T9 intradural extramedullary mass in T2-
14 weighted hyposignal with spinal cord compression (Figure 1a and b). There were signs of
15 spinal cord injury (Figure 1a). Surgery was proposed because of the presence of symptomatic
16 spinal cord compression.

17 The patient underwent a posterior approach under electrophysiologic monitoring. Operatively,
18 after opening the dura mater at T8-T9, the spinal cord was pushed back on the side opposite to
19 the tumor. The tumor was poorly vascularized and attached to the right lateral side of the dura
20 mater. It was an encapsulated extramedullary tumor, which was difficult to separate from the
21 spinal cord due to the presence of pial adhesions. It also showed some arachnoid invasion in
22 the medial part. It was very firm, requiring high-intensity ultrasonic debulking. Gross total
23 resection was achieved.

24 Definitive diagnosis of grade 1 spinal cord SFT/HPC according to the 2016 WHO
25 classification [1] was made on histopathological analysis. The tumor was composed of spindle
26 cells in an abundant collagenic stroma (Figure 1c). On immunohistochemistry, the neoplastic cells
27 strongly expressed CD34 and STAT6 (Figure 1d).

28 The last follow-up showed neurological progression without abnormality, complete
29 neurological recovery and normal walking. Follow-up MRI at a year and a half revealed
30 complete excision of the tumor without any evidence of residual tumor.

31

32 **Discussion**

33 SFT/HPC is a rare tumor of the central nervous system, and only a few cases have been
34 reported in the spinal cord. The most common location in the spine is the epidural space The

1 intradural type (type II according to Liu's classification: A, extramedullary; B,
2 intramedullary) [2] is rarer and often difficult to differentiate from more common spinal cord
3 tumors.

4 SFT/HPC is a slowly growing tumor. Symptoms are non-specific, depending on location in
5 the spinal canal. Diagnosis is difficult based only on history-taking, unless the patient has
6 history of SFT/HPC located elsewhere. On MRI, the tumor is brightly enhanced by
7 gadolinium, revealing its highly vascularized character. Hyposignal on the T2-weighted
8 sequence suggests SFT/HPC (which can be confused with calcification). Spinal SFT/HPC can
9 be extramedullary, intramedullary, or both. Some radiological features may help differentiate
10 SFT/HPC from other spinal tumors: 1) thickened dura mater with comet-tail sign suggests
11 meningioma [3]; 2) dumbbell tumor suggests schwannoma; 3) intratumoral or satellite cysts
12 suggests ependymoma [4]; and 4) heterogeneous enhancement after gadolinium injection with
13 infiltrative tumor suggests astrocytoma [5] (Table 1). In the absence of such features, a
14 diagnosis of spinal SFT/HPC may be discussed. Operatively, the tumor is often firm, and
15 removal can be challenging even with ultrasonic aspiration. When resection is very difficult,
16 the best instrument is a laser [6]. Some authors have reported significant bleeding that
17 required stopping the surgery and organizing embolization of the spinal SFT/HPC [7,8],
18 although indications for angiography (and embolization) are a matter of debate.
19 Histopathological examination is the key to diagnosis: SFT/HPC has a strong fibrocollagenic
20 component with very specific immunostaining for STAT6 (Table 1).
21 Concerning treatment, gross total resection is the gold standard. Some authors suggested
22 adjuvant radiotherapy or chemotherapy, particularly in case of partial resection [2,9]. Due to
23 the rarity of spinal cord cases in the literature, no consensual therapeutic management can
24 currently be reached. Follow-up should be very long, even when gross total resection was
25 achieved, as recurrence can occur [10,11].

26

27 **Conclusion**

- 28 • SFT/HPC is a very rare spinal tumor that can be extramedullary, intramedullary, or
29 both; there is significant risk of torquing the cord when en-bloc resection is attempted
30 without taking account of the growth pattern.
- 31 • Maximal surgical resection is the best treatment, but can be challenging because of the
32 tumor's firm consistency and subpial adherences to the spinal cord.
- 33 • Outcome is good in case of GTR gross total resection, but there is a risk of very late
34 recurrence, requiring long-term follow-up.

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The patient gave informed signed consent for the writing and publication of this article.

References

[1] Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. *Acta Neuropathol* 2016;131:803–20. doi:10.1007/s00401-016-1545-1.

[2] Liu H, Yang A, Chen N, Yang J, Qiu X, Zhang J. Hemangiopericytomas in the spine: clinical features, classification, treatment, and long-term follow-up in 26 patients. *Neurosurgery* 2013;72:16–24; discussion 24. doi:10.1227/NEU.0b013e3182752f50.

[3] Maiti TK, Bir SC, Patra DP, Kalakoti P, Guthikonda B, Nanda A. Spinal meningiomas: clinicoradiological factors predicting recurrence and functional outcome. *Neurosurg Focus* 2016;41:E6. doi:10.3171/2016.5.FOCUS16163.

[4] Dauleac C, Messerer R, Obadia-Andre N, Afathi M, Barrey CY. Cysts associated with intramedullary ependymomas of the spinal cord: clinical, MRI and oncological features. *J Neurooncol* 2019. doi:10.1007/s11060-019-03241-9.

[5] Kim DH, Kim J-H, Choi SH, Sohn C-H, Yun TJ, Kim CH, et al. Differentiation between intramedullary spinal ependymoma and astrocytoma: comparative MRI analysis. *Clin Radiol* 2014;69:29–35. doi:10.1016/j.crad.2013.07.017.

[6] Sciubba DM, Liang D, Kothbauer KF, Noggle JC, Jallo GI. The evolution of intramedullary spinal cord tumor surgery. *Neurosurgery* 2009;65:84–91; discussion 91-92. doi:10.1227/01.NEU.0000345628.39796.40.

[7] Walker CT, Amene CS, Pannell JS, Santiago-Dieppa DR, Rennert RC, Hansen LA, et al. Hemorrhagic intramedullary solitary fibrous tumor of the conus medullaris: case report. *J Neurosurg Spine* 2015;23:438–43. doi:10.3171/2015.1.SPINE13915.

[8] Lee C-H, Kim K-J, Jahng T-A, Kim H-J. Spinal Hemangiopericytoma Which Needed Intraoperative Embolization due to Unexpected Bleeding. *J Korean Neurosurg Soc* 2013;54:253–6. doi:10.3340/jkns.2013.54.3.253.

[9] Das A, Singh PK, Suri V, Sable MN, Sharma BS. Spinal hemangiopericytoma: an institutional experience and review of literature. *Eur Spine J* 2015;24 Suppl 4:S606-613. doi:10.1007/s00586-015-3789-1.

[10] Chen H, Zeng X-W, Wu J-S, Dou Y-F, Wang Y, Zhong P, et al. Solitary fibrous tumor of the central nervous system: a clinicopathologic study of 24 cases. *Acta Neurochir (Wien)* 2012;154:237–48; discussion 248. doi:10.1007/s00701-011-1160-9.

[11] Metellus P, Bouvier C, Guyotat J, Fuentes S, Jouvet A, Vasiljevic A, et al. Solitary fibrous tumors of the central nervous system: clinicopathological and therapeutic considerations of 18 cases. *Neurosurgery* 2007;60:715–22; discussion 722. doi:10.1227/01.NEU.0000255418.93678.AD.

Figure 1: Spinal cord SFT/HPC

a: Sagittal T2 MRI showing a T8-T9 intradural mass in hyposignal, without dura mater attachment, associated with intramedullary hypersignal.

b: Axial T1 sequence with gadolinium injection, showing a right mass associated with a cyst in the dorsal portion.

c: The tumor shows hypocellular areas with spindle cells embedded in a thick collagenous stroma (Hematoxylin Phloxine Saffron staining, original magnification x 200).

d: There is a strong nuclear immunoexpression of STAT6 by the neoplastic cells (original magnification x 200).

1 **Table 1:** Distinctive features between spinal SFT/HPC, meningioma, schwannoma,
2 ependymoma, and astrocytoma.
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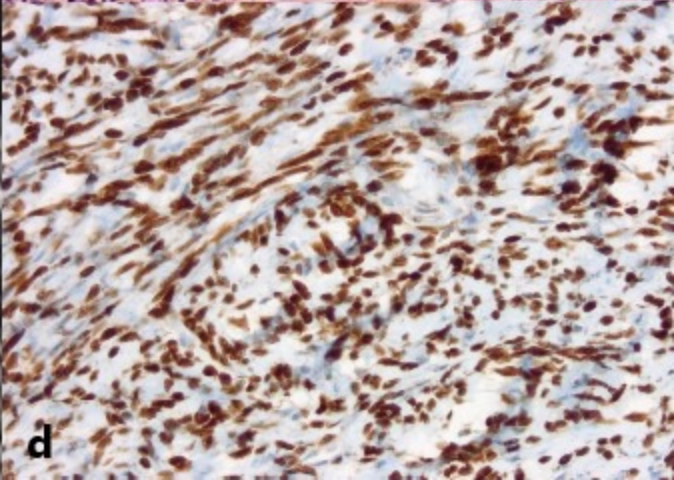
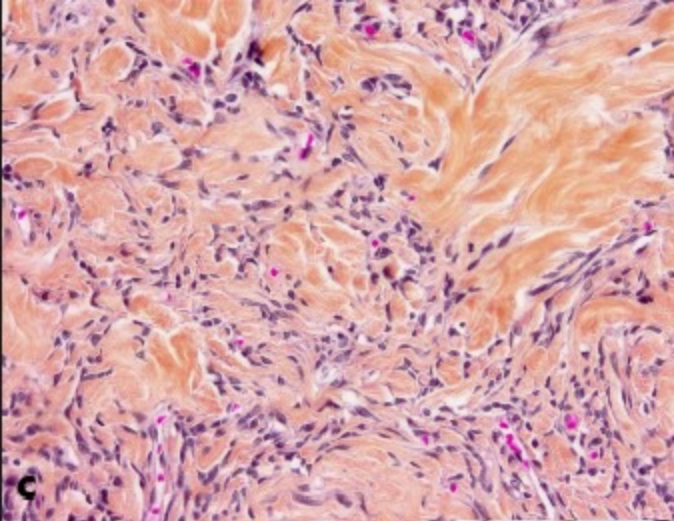
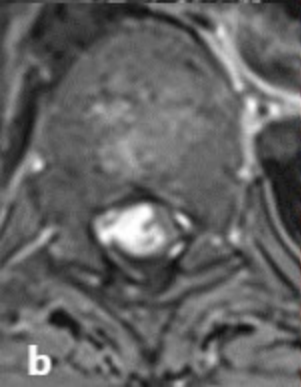


Table 1: Distinctive features between spinal SFT/HPC, meningioma, schwannoma, ependymoma, and astrocytoma.

	SFT/HPC	Meningioma	Schwannoma	Ependymoma	Astrocytoma
Epidemiology					
Frequency	Very rare	30%	24%	23%	6%
Sex ratio, M:F	1.3:1	1:3.5	1:2.1	1.5:1	1.4:1
Median age	Adult	64 years	47 years	44 years	Children
Others	Other location		NF	NF2	NF
Location	EM ± IM	EM	EM	IM + filum	IM
Level	Th > cervical	Th > cervical	Lumbar	Cervical	Th > cervical
MRI					
T1	Iso	Iso	Hypo	Iso	Hypo
T2	Hypo/hyper	Iso/hyper	Hyper	Iso	Hyper
Enhancement	Homogeneous	Homogeneous	Homogeneous	Enhanced	Heterogeneous
Other features		Thickened dura, dural comet-tail	Dumbbell-shaped, enlarged foramina	Intratumoral or satellite cyst	Intratumoral or satellite cyst
Surgical findings					
Capsule	Encapsulated	Encapsulated or in plaque	Encapsulated	Encapsulated or without capsule	Infiltration of spinal cord
Attachment	Subpial	Dura	Nerve root sheath	NA or filum	NA
Outcome	Recurrence or metastasis (sometimes very delayed)	Very low risk of recurrence if GTR is performed	Very low risk of recurrence if GTR is performed, except for NF	Very low risk of recurrence if GTR is performed	Progression because GTR is often difficult to perform
Histology					
EMA	Negative	Positive	Negative	Positive	Negative
STAT6	Positive	Negative	Negative	Negative	Negative
PS100	Negative	Negative	Positive	Positive	Positive
GFAP	Negative	Negative	Negative	Positive	Positive

EM: extramedullary; EMA: epithelial membrane antigen; F: female; GFAP: glial fibrillary acidic protein; GTR: gross total resection; Hyper: hypersignal; Hypo: hyposignal; IM: intramedullary; Iso: isosignal; M: male; NA: not applicable; NF: neurofibromatosis; PS100: S100 protein; STAT6: signal transducer and activator of transcription 6; T1: T1-weighted sequence; T2: T2-weighted sequence; Th: thoracic.