Case Report

Two cases of solitary fibrous tumor/hemangiopericytoma with different clinical features according to the World Health Organization classification: case report and review of the literature

Tomoya Nishii[1]^, Yoshitaka Nagashima[1], Yusuke Nishimura[1], Hiroshi Ito[1], Takahiro Oyama[1], Mamoru Matsuo[1], Ayako Sakakibara[2], Satoko Shimada[2], Ryuta Saito[1]

[1]Department of Neurosurgery, Nagoya University, Nagoya, Japan; [2]Department of Pathology, Nagoya University, Nagoya, Japan

Correspondence to: Yoshitaka Nagashima, MD. Department of Neurosurgery, Nagoya University School of Medicine, Nagoya 466-8550, Japan. Email: y-nagashima@med.nagoya-u.ac.jp.

Abstract: Solitary fibrous tumors (SFTs) and hemangiopericytomas (HPCs) have been classified as one entity by the World Health Organization in 2016 due to gene fusion between NAB2 and STAT6. In the Central Nervous System (CNS), a hypocellular, collagenized tumor with a classic SFT phenotype is considered grade I, whereas more densely cellular tumors mostly corresponding to the HPC phenotype are classified as grade II or III (anaplastic) depending in mitotic count (<5 *vs.* >5 mitoses per 10 high-power fields). Herein, we report two cases of targeted SFT/HPC in which pathological differences and WHO grading affect clinical features. A 75-year-old woman presented with headache and had an intradural extramedullary tumor at the C1 to C2 level. The tumor was well-circumscribed and attached only to the dura mater. It was totally removed and diagnosed SFT/HPC grade I. In contrast, a 68-year-old woman presented with numbness in the right upper limb and had an intradural extramedullary tumor at the medulla to C3 levels The tumor was irregularly marginated and strongly adherent to the spinal cord and involved the vertebral artery. It was sub totally removed and diagnosed SFT/HPC grade II. To the best of our knowledge, there are only 12 cases of SFT/HPC at the craniocervical junction, including the present two cases, of which four that were adherent to the spinal cord or involved the vertebral artery were grade II or III. Although the location of the tumor was almost the same, there were significant differences in the intraoperative findings according to the WHO grading.

Keywords: Solitary fibrous tumor/hemangiopericytoma (SFT/HPC); craniocervical junction; spinal tumor, case report

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Introduction

Solitary fibrous tumor (SFT) and hemangiopericytoma (HPC), which were previously treated as different types of tumors, have been defined as combined entities since the 2016 World Health Organization (WHO) reclassification, following the discovery of the NAB2/STAT6 fusion gene in both tumors. In the Central Nervous System (CNS), a hypocellular, collagenized tumor with a classic SFT phenotype is considered grade I, whereas more densely cellular tumors mostly corresponding to the HPC phenotype are classified as grade II or III (anaplastic) depending in mitotic count (<5 *vs.* >5 mitoses per 10 high-power fields) (1,2).

^ ORCID: 0000-0001-9138-9602.

SFT/HPC in the spinal region is less frequently than that occurring in the intracranial region (3,4). The primary goal of treatment for spinal SFT/HPC is total removal of the tumor. The extent of surgical resection is thought to affect the patient's prognosis, and completely resected tumors have been reported to have a better prognosis (5,6); however, the factors that determine the surgical removal rate for spinal SFP/HPC remain unclear.

This case report presents two targeted cases of intradural extramedullary SFT/HPC at the craniocervical junction: WHO grade I tumor with no invasion into the spinal cord or vessels and WHO grade II tumor involving the vertebral artery. These histological diagnoses were confirmed by two pathologists. Since the craniovertebral junction is a complex area with vital neural and vascular structures, infiltration into surrounding tissues may reduce the rate of tumor removal. This study aimed to highlight the difference between the WHO grade of SFT/HPC at the craniocervical junction and the intraoperative findings that determine surgical difficulty. We present the following article in accordance with the CARE reporting checklist (available at https://dx.doi.org/10.21037/jss-21-83).

Case presentation

Case 1

A 75-year-old woman with past medical history of rheumatoid arthritis was referred to our hospital after a tumor was found by a magnetic resonance imaging (MRI) of the head due to headache. She had no other neurological symptoms. MRI of the cervical spine showed an intradural extramedullary tumor at the C1 to C2 levels. The spinal cord was markedly compressed and displaced to the contralateral side. The tumor was isointense on T2-weighted images and showed homogeneous enhancement after administration of gadolinium (*Figure 1*). Thus, the preoperative diagnosis was spinal meningioma.

We performed C1-2 laminectomy. After cutting the dura with a microsurgical technique, an intradural extramedullary tumor mass was visualized on the right side of the spinal canal. The tumor was well circumscribed, smooth-marginated with dural attachment, and scarcely vascularized. The tumor did not adhere to the surrounding structures. Intraoperative frozen section analysis suggested a grade I meningioma. Total removal of the tumor was achieved, and according to the meningioma excision classification, it was Simpson grade II (*Figure 2*).

Hematoxylin-eosin-stained sections revealed patternless architecture of a tumor composed of cells with bland ovoid-to-spindle-shaped nuclei and scant eosinophilic cytoplasm, and stromal collagen deposition. The mitotic figures were scant. Immunohistochemical studies showed that the tumor cells were positive for STAT6, CD 34. The Ki-67 labeling index was 1–3%. This revealed that the pathologic type was SFT/HPC, WHO grade I (*Figure 3*).

The patient's postoperative course was uneventful. At 6-month follow-up, the patient showed no recurrence.

Case 2

A 68-year-old previously healthy woman presented with a 2-month history of numbness in the right upper limb. MRI of the spine revealed an intradural extramedullary tumor in the medulla to the C3 level. The tumor had a speckled high-signal area with a partial flow void on T2-weighted images and partially heterogeneous enhancement after administration of gadolinium. Preoperative left vertebral angiography showed that the posterior meningeal artery was the primary feeding vessel (*Figure 4*).

We performed suboccipital craniotomy and C1-3 laminectomy. After midline dural opening under a microscope, a solid tumor was found on the posterior medulla and spinal cord. The caudal part of the tumor was soft and there were no adhesions to the spinal cord, but the cranial part of the tumor was hemorrhagic and firmly adherent to the pia of the spinal cord and medulla, and involved the vertebral artery, the nerve roots of C1, C2, and lower cranial nerves. Intraoperative frozen section analysis suspected primitive neuroectodermal tumor (PNET), equivalent to grade IV. The tumor was removed in a piecemeal manner, and subtotal removal of the tumor was achieved (*Figure 5*).

Hematoxylin-eosin-stained sections revealed diffuse high cellularity with little intervening stroma. There was no necrosis, but scattered mitotic figures were observed. Immunohistochemical staining was positive for STAT6 and CD 34. The Ki-67 labeling index was 5%. Thus, the pathological diagnosis was SFT/HPC WHO grade II (*Figure 6*).

The postoperative course was uneventful. FDG-PET 1 month postoperatively showed no local recurrence or metastasis. Adjuvant radiotherapy was not performed. At 18 months of follow-up, MRI showed no tumor recurrence.

All procedures performed in studies involving human participants were in accordance with the ethical standards

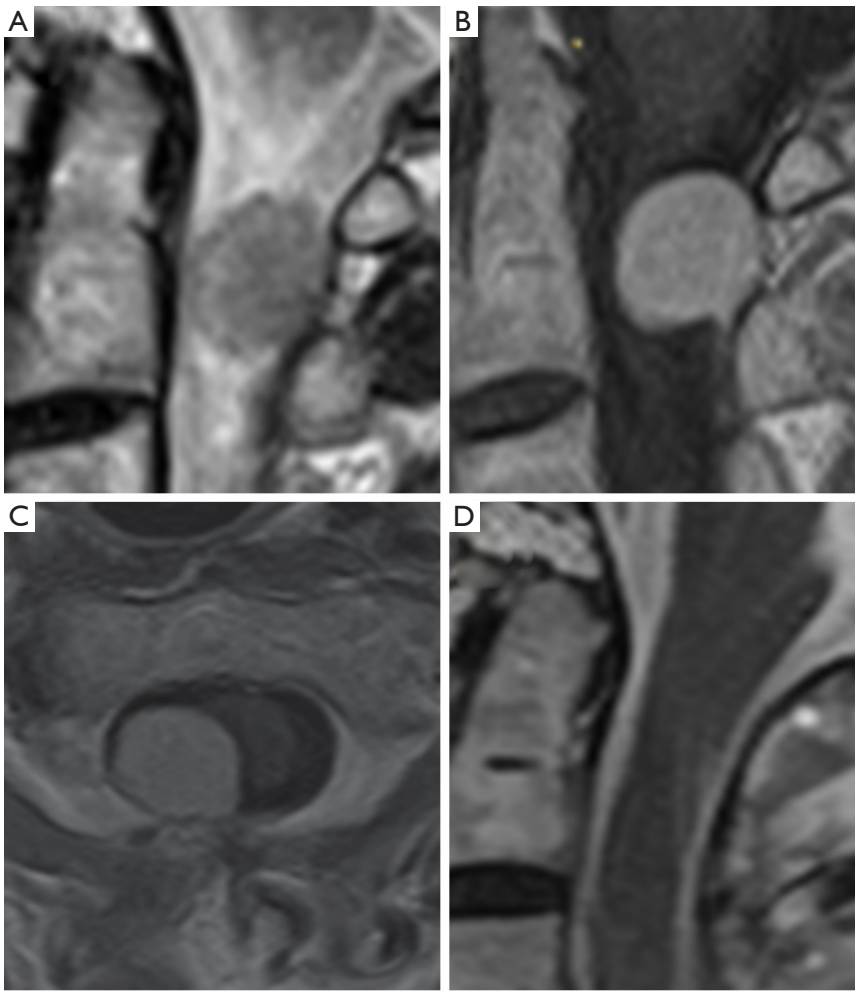

Figure 1 Magnetic resonance imaging (MRI). (A) Preoperative sagittal T2 weighted MRI demonstrating isointensity of the lesion. (B) Preoperative sagittal T1 weighted MRI with contrast demonstrating homogeneous enhancement of the tumor, and (C) axial. (D) T2 weighted MR image demonstrating gross total removal of the tumor.

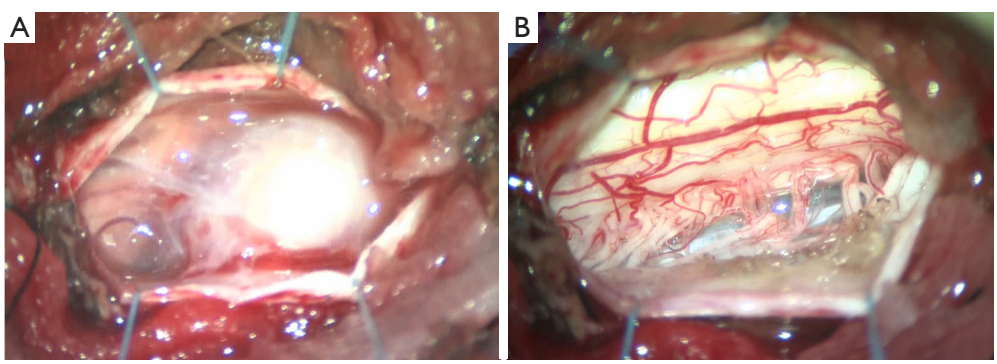

Figure 2 Intraoperative photographs of the tumor resection. (A) Following the dural opening, a mass lesion was visible on right lateral to the dura. (B) The tumor was completely resected.

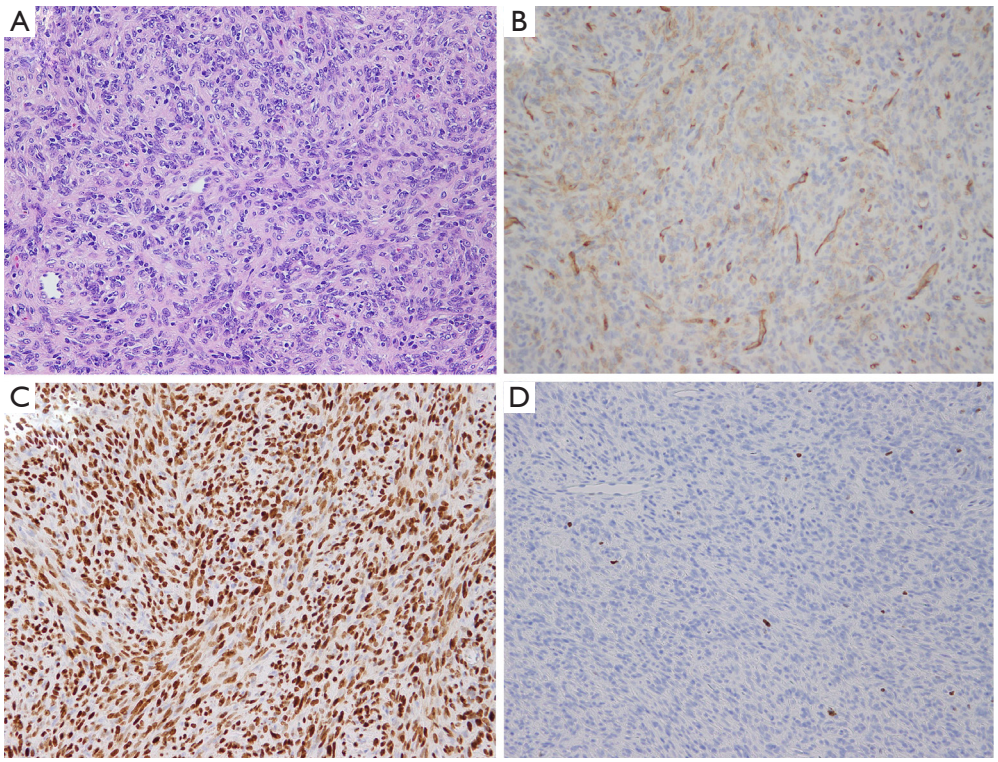

Figure 3 Histopathology of the resected mass. (A) Microscopic findings. Hematoxylin and eosin (H&E) stained section showed patternless architecture of a tumor composed of cells with bland ovoid-to-spindle-shaped nuclei and scant eosinophilic cytoplasm, and stromal collagen deposition. The tumor cells were positive for (B) CD34 and (C) STAT6. (D) The Ki-67 labeling index was 1–3%. Original magnifications ×200.

of the institutional and national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

Previously different entities, SFTs and HPCs of the meninges were combined into a single category of fibroblastic type mesenchymal tumors according to the 2016 WHO classification because of the common NAB2-STAT6 fusion gene (1). SFT/HPC may be found in any soft tissue, but it is rarely found in the CNS, while tumors in the spinal cord are even more rare (3,4). The classical SFT subtype with low cell density, low nuclear fission, and more collagenous fibers has become WHO grade I, the classical HPC subtype with high cell density as grade II, and variable amounts of mitoses and necrosis as grade III. The distinction is based on the level of mitoses, with less than 5 mitosing cells per 10 high-power fields defined as grade II, and five or more mitosing cells per ten high-power fields defined as grade III. These grades have been reported to correlate with clinical prognosis (7).

SFT/HPC did not demonstrate any specific imaging features. Radiological findings such as mixed high and low signal areas (yin-yang sign) on T2-weighted images, less dural tail sign unlike meningioma, less calcification, and bone destruction are reported as key points to consider for differentiation (4,8,9). The tumor often demonstrates marked enhancement with prominent flow voids on MRI due to its hypervascularity (10). However, there have been reports of cases that do not show such imaging features, and these often mimic various tumors, such as meningioma and schwannoma, in imaging findings. Case 1 presented with a dural tail sign and no yin-yang sign or flow void, which led to the preoperative diagnosis of meningioma. Case 2 demonstrated an irregular shape with flow void on T2WI MRI. Differences in these imaging features also

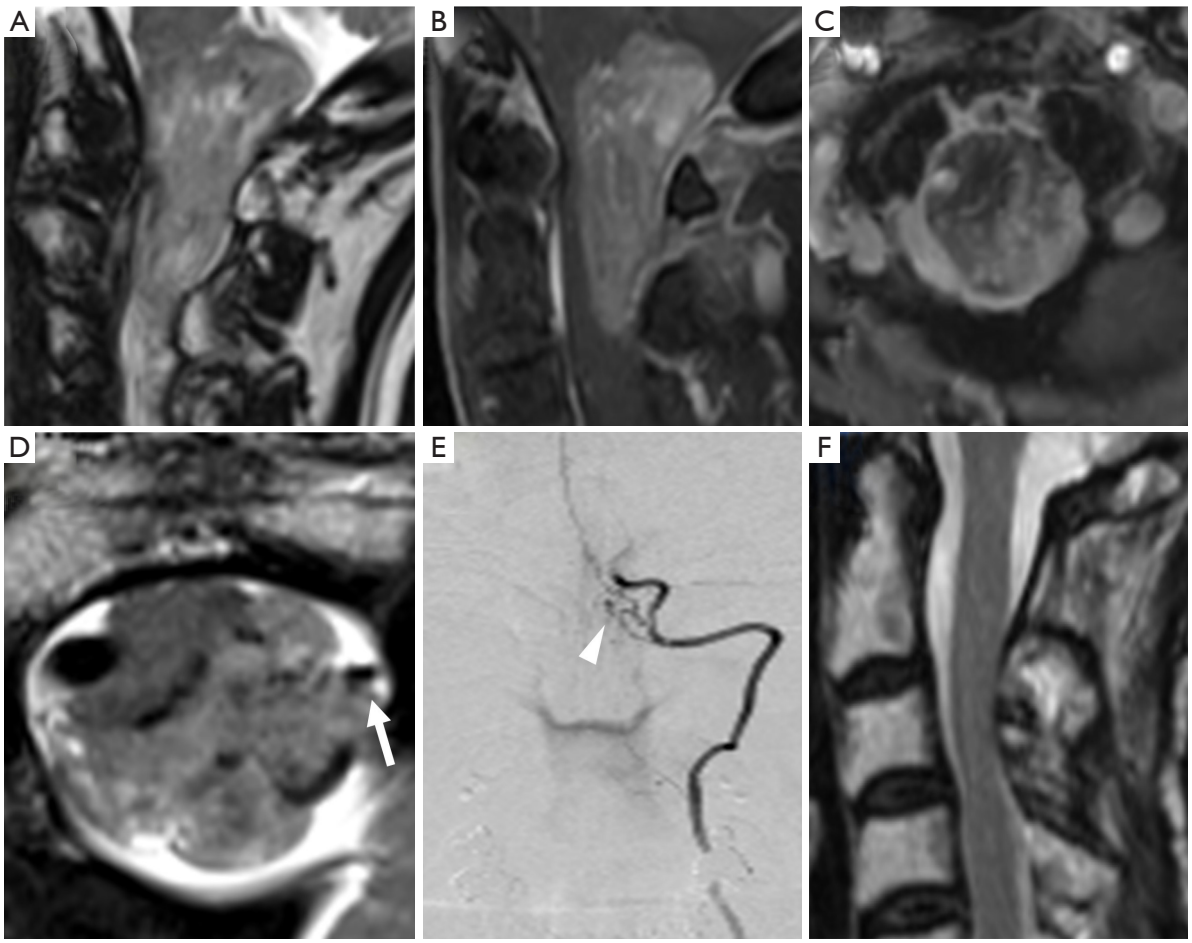

Figure 4 Magnetic resonance imaging (MRI) and angiography. (A) On preoperative sagittal T2 weighted MRI, the tumor demonstrated a speckled high-signal area with a partial flow void. (B) Preoperative sagittal T1 weighted MRI with contrast demonstrating partially heterogeneous enhancement of the tumor, (C) and axial. (D) Preoperative axial T2 weighted MRI demonstrate left vertebral artery (arrow) was passing through the tumor. (E) Preoperative frontal view of left vertebral artery angiography showed that the posterior meningeal artery (arrowhead) was the main feeding vessel, but it did not strongly feed the tumor. (F) Postoperative T2 weighted MRI showed that the tumor was almost entirely removed.

affect intraoperative findings: in case 1, a meningioma was suspected due to the hardness of the tumor with an attachment to the dura; in case 2, a highly malignant tumor was suspected because it was easily hemorrhagic and partially adherent to the surrounding tissue. Intraoperative frozen section diagnosis showed that case 1 was a grade I meningioma and case 2 was a grade IV PNET-like tumor; however, the definitive histopathologic diagnosis was SFT/HPC in both cases because of STAT6 positivity. As shown above, it is difficult to predict the diagnosis of SFT/HPC.

The gold standard treatment for spinal SFT/HPC is total resection, if possible. Partial resection increases the recurrence rate, and pathologic grade is associated with local recurrence and distant metastasis rates (11). However, because of the low incidence of spinal cord SFT/HPCs, there have been no reports on the relationship between tumor grading and removal rate. According to the literature, SFTs that are currently equivalent to grade I are well circumscribed, smooth surfaced, partially or completely encapsulated, and described as firm and mostly solid (3). On the other hand, hemangiopericytoma, which is currently equivalent to grade II or III, sometimes infiltrates the nerve roots or invades the spinal cord (12,13). These differences in tumor characteristics determine the degree of surgical difficulty, especially in the craniocervical junction. To the best of our knowledge, there have been 12 cases

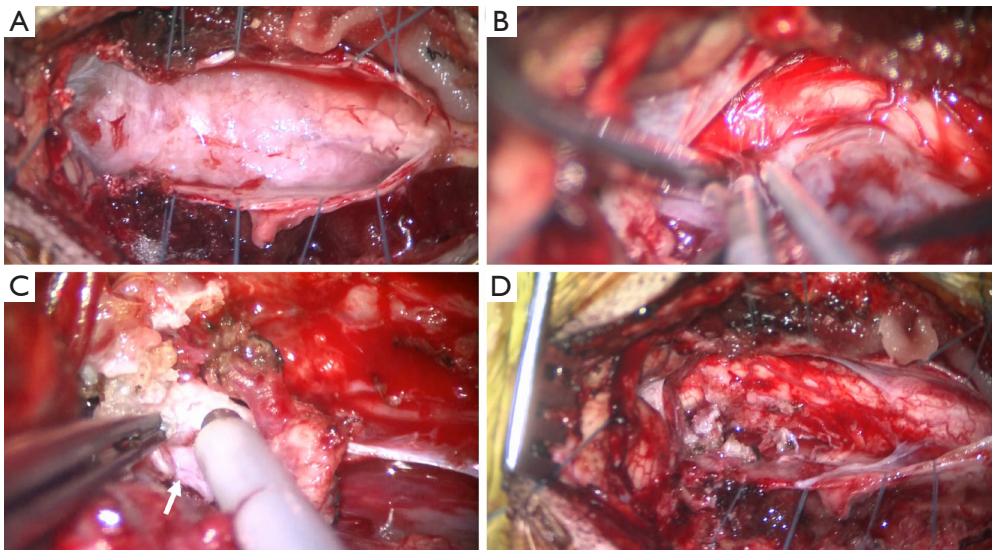

Figure 5 Intraoperative photographs of the tumor resection. (A) Intraoperative photographs of the tumor resection. After dural opening, extramedullary tumors were identified in the medulla and posterior spinal cord. (B) In the area higher levels than C1, it was firmly adherent to the pia of the spinal cord and medulla. (C) The tumor was hemorrhagic and involved the vertebral artery (arrow). (D) Tumor has been almost completely removed.

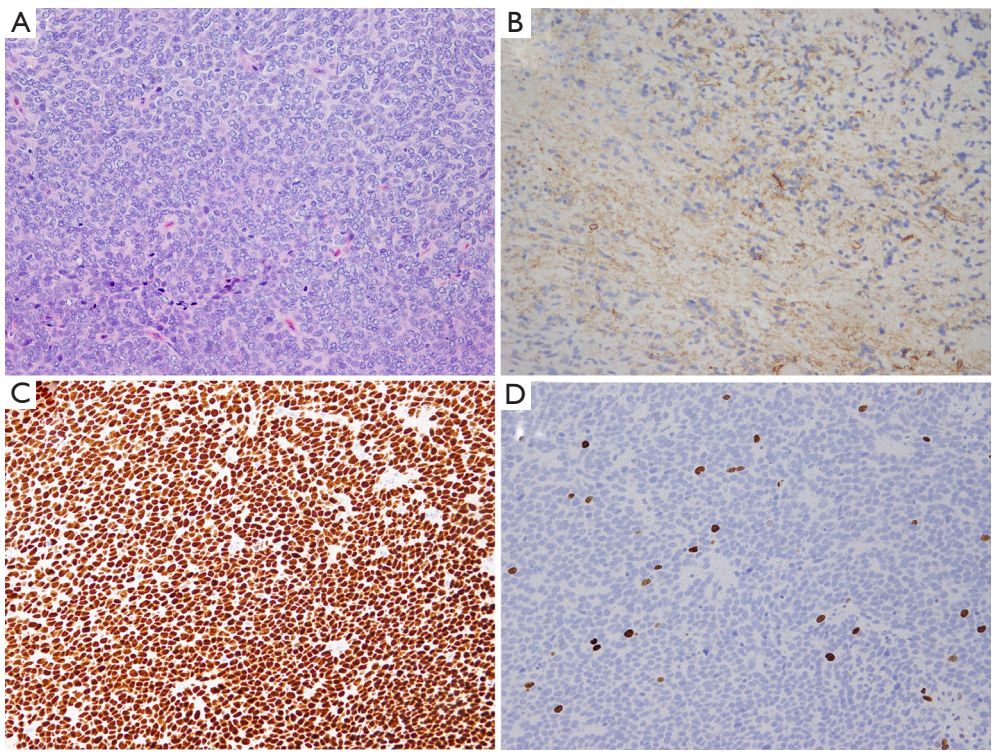

Figure 6 Histopathology of the resected mass. (A) Microscopic findings. Hematoxylin and eosin (H&E) stained section showed diffuse high cellularity with little intervening stroma. There was no necrosis. Mitotic figures were found sparingly. The tumor cells were positive for (B) CD34 and (C) STAT6. (D) The Ki-67 labeling index was 5%. Original magnifications ×200.

Table 1 Literature summary of SFT/HPC cases in the craniocervical junction

Authors & year	Age (yrs)/sex	Level	Compartment	Intraoperative findings	Grade
Brunori *et al.* 1999	18/male	O-C3	ED & IDEM	Attachment to the dura	I
Endo *et al.* 2003	63/female	C2-C4	ED	Attachment to the dura	I
Obara *et al.* 2003	49/female	C2-C4	ED	Detached from dura easily	I
Hirakawa *et al.* 2004	52/male	C1-C2	ED	No involved nerve roots	I
Fargen *et al.* 2011	62/male	C1-C2	IDEM	Arise from the arachnoid	I
Fargen *et al.* 2011	28/female	C2-C3	IDEM & IDIM	Visualized dorsal to the spinal cord	I
Zaldivar-jolissaint *et al.* 2016	33/male	C1-C4	ED	Detached from dura easily	I
Present case 1, 2021	75/female	C1-C2	IDEM	Attachment to the dura	I
Shirzadi *et al.* 2013	56/male	C1-C3	IDEM	Adherent to the spinal cord	II or III
Tsutsumi *et al.* 2014	19/male	C1-C2	ED	Involved vertebral artery	II or III
Gregory *et al.* 2017	10/male	C1-C3	IDEM	Adherent to the spinal cord	II
Present case 2, 2021	68/female	O-C3	IDEM	Adherent to the spinal cord and medulla, and involved vertebral artery	II

ED, extradural; IDEM, intradural extramedullary; IDIM, intradural intramedullary; NR, not reported.

of SFT/HPC occurring at the craniocervical junction, including our cases (6,12,14-19). Of these, 8 were grade I, and 4 were grade II or III, with an average age of 44 years (10–75 years); 7 were male and 5 were female. Clinical symptoms ranged from paralysis and sensory disturbances to headache and neck pain. The minimum follow-up period was 6 months, with a maximum of 7 years. In this series, there was only one case of classic HPC (grade II or III) had tumor recurrence (6), and none had metastasis. According to the surgical findings in the literature describing SFT/HPC grade I, detachment of the tumor and surrounding structures is easy, while for Grade II or III tumors, this is difficult to remove as the vertebral artery is involved or the tumor was adherent to the spinal cord (*Table 1*). As in our two cases, the higher the WHO grade, the more careful the surgical manipulation required to avoid damaging the surrounding vital structures and the higher the risk of residual tumor.

Conclusions

We encountered two targeted cases of SFT/HPC at the craniocervical junction. The more malignant the lesion, the more likely it is to progress to involve the surrounding vital tissues. SFT/HPC at the craniocervical junction with a higher WHO grade might have difficulty achieving gross total removal of the tumor, leading to poor prognosis.

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Footnote

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performed in studies involving human participants were in accordance with the ethical standards of the institutional and national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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