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Case Reports & Case Series

Solitary fibrous tumor/hemangiopericytoma expanding the superior and inferior cerebellar tentorium: A case report

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ABSTRACT

Background: A rare case of a solitary fibrous tumor/hemangiopericytoma (SFT/HPC) expanding the superior and inferior cerebellar tentorium is described.
Case description: A 28-year-old male presented with convulsions. Magnetic resonance imaging revealed a tumor around the right cerebellar tentorium. The tumor was completely removed after embolization of the feeding arteries, and the pathological diagnosis was World Health Organization grade III SFT/HPC. The patient's post-operative course was uneventful, and there was no recurrence or extracranial metastasis at 1.5 years following the operation.
Conclusions: It was possible to fully remove the SFT/HPC tumor; however, strict observation of the whole body is needed because of the possibility of recurrence and extracranial metastasis.

1. Introduction

Given the histological and immunohistochemical similarities between solitary fibrous tumors and hemangiopericytoma, in 2016 the World Health Organization (WHO) created the combined term solitary fibrous tumor/hemangiopericytoma (SFT/HPC). SFT/HPCs are rare, and it is even rarer that they occur in the central nervous system. As per our knowledge, although many SFT, HPC, and SFT/HPC cases have been reported to date, only 11 SFT cases and 1 SFT/HPC case expanding the superior and inferior cerebellar tentorium have been reported (Table 1) [1–11]. Herein, we describe a rare case of a SFT/HPC expanding the superior and inferior cerebellar tentorium.

2. Case report

A 28-year-old male presented with sudden onset of tonic convulsion. He had no medical or family history of note. On arrival, he had no neurological deficits. Computed tomography (CT) revealed a right high-density mass lesion expanding the superior and inferior cerebellar tentorium with peritumoral edema (Fig. 1A). Magnetic resonance imaging (MRI) indicated a mass lesion of 3.7 × 3.2 × 5.4 cm with clear margins that was isointense on T1-weighted imaging (WI), isohyperintense on fluid attenuated inversion recovery, and well-enhanced on gadolinium-enhanced T1WI (Fig. 1B–D). There were no apparent tumor lesions in other parts of the patient's body. Digital subtraction angiography revealed feeding arteries from a mastoid branch of the right occipital artery and right posterior meningeal artery (Fig. 2A, B). In addition, the right transverse sinus and sigmoid sinus were partially occluded. The right vein of Labbe drained into the partially residual right transverse sinus (Fig. 2C).

Based on the above results, we presumed the tumor to be a tentorial meningioma. We planned to remove the tumor after transcatheter arterial embolization of the feeding arteries (mastoid branch of the right occipital artery and right posterior meningeal artery) with Embosphere® (Nippon Kayaku, Tokyo, Japan) and a coil the day before the operation day.

3. Operation

The operation started with the patient in the lower left park bench position. We made an incision in the skin surrounding the right auricle in a reverse J shape and performed the right occipital and suboccipital craniotomy. First, we removed the tumor under the cerebellar tentorium, which was red, soft, and partly hemorrhagic with clear margins

Abbreviations: CD, clusters of differentiation; CNS, central nervous system; CT, computed tomography; EMA, epithelial membrane antigen; GTR, Gross total resection; HPC, hemangiopericytoma; HPF, high-power fields; MRI, magnetic resonance imaging; OS, overall survival; PFS, progression free survival; RT, radiation treatment; SFC, solitary fibrous tumor; WHO, world health organization; WI, weighted imaging

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Table 1
Reported cases of solitary fibrous tumor extending the superior and inferior cerebellar tentorium.

Case no.	Author, year	Age (years), gender	Tumor size (cm)	Treatment	Immunohistochemical findings	Outcome
1	Carneiro, 1996(1)	62, F	8	GTR	CD34+, MIB-1 < 4%	No Rec at 7 M
2	Carneiro, 1996(1)	51, F	N/A	GTR	CD34+, MIB-1 < 4%	No Rec at 20Y
3	Suzuki, 2000(2)	54, F	N/A	STR	CD34+, vimentin+, EMA-, S-100-, MIB-1 6.2%	Rec at 15y No Rec at 7Y after 2nd surgery
4	Hori, 2007(3)	72, M	Large	STR	CD34+, Bcl-2+, EMA-, S-100-, MIB-1 2.2%	Died at 12Y
5	Secer, 2008(4)	76, M	6.5 × 5.2 × 3.7	GTR	CD34+, vimentin+, Bcl-2+, EMA-, S-100-, MIB-1 < 1%	No Rec at 6 M
6	Okamoto, 2009(5)	29, F	6 × 5 × 5	STR +RT 40Gy	CD34+, vimentin+, Bcl-2+, EMA-, S-100-	2nd surgery (GTR) at 12D after RT →No Rec at 18 M
7	Bisceglia, 2011(6)	59, F	4	GTR	CD34+, vimentin+, Bcl-2+, EMA-, S-100-, MIB-1 < 2%	No Rec at 3.5Y
8	Sun, 2011(7)	57, F	N/A	GTR +RT 50Gy	CD34+, vimentin+, Bcl-2+, EMA-, S-100-, MIB-1 25%	No Rec at 11.5 M
9	Vassal, 2011(8)	44, F	3 × 3 × 2.5	STR	Scarce mitoses CD34+, MIB-1 < 2%	No Rec at 2Y (Residual lesion+)
10	Alapatt, 2012(9)	52, F	6 × 6	GTR	CD34+, vimentin+, MIB-1 low	No Rec at 2Y
11	Uneda, 2016(10)	49, F	5.5 × 5 × 4.6	STR	CD34+, Bcl-2+, EMA-, S-100-, MIB-1 < 1%	No Rec at 9 M
12	Sung, 2018(11)	51, M	N/A	STR+ RT + GKS	N/A	2 surgeries, 1RT, 4GKS →11.6Y alive after the 1st operation
13	Present case	28, M	3.7 × 3.2 × 5.4	GTR	11 Mitoses/10HPF CD34+, vimentin+, Bcl-2+, EMA-, S-100-, MIB-1 6%	No Rec at 1Y

F, female; M, male; N/A, not available; F, female; GTR, gross total removal; STR, subtotal removal; RT, radiotherapy; Rec, recurrence; M, month; Y, year; D, day; GKS, gamma knife surgery.

(Fig. 3A). Next, we removed the tumor over the cerebellar tentorium while preserving the right vein of Labbe and the right transverse sinus (Fig. 3B). We completely removed the tumor, including a part of the cerebellar tentorium (Fig. 3C). Lastly, we performed electrocoagulation on the dural wall of the right transverse sinus where the tumor had been attached.

4. Pathological findings

Short spindle cells proliferated densely and deer horn-like vessels had developed (Fig. 4A). Furthermore, 11 mitoses per 10 high-power fields (HPF) and infiltration into the cerebrum were recognized (Fig. 4B, C). On immunostaining, cluster of differentiation (CD) 34 was partially positive (Fig. 4D), and vimentin and Bcl-2 were positive. Cytokeratin, epithelial membrane antigen (EMA) (Fig. 4E), S-100, and factor VIII were negative. Ki-67 MIB-1 labeling index was 6% (Fig. 4F). According to the above results, we made a diagnosis of WHO grade III SFT/HPC.

5. Postoperative course

Postoperative MRI revealed no apparent residual tumor. There were no postoperative complications or seizures; therefore, the patient was discharged on day 14 after the operation. On the 1.5-year postoperative follow-up, neurological signs were not observed and imaging did not show any apparent lesions (Fig. 5A–C). No metastatic lesions were observed extracranially, including in the spinal cord.

6. Discussion

Intracranial SFT is a rare mesenchymal neoplasm first described by Carneiro et al. [1] characterized by fibrous features arising predominantly from thick collagen bands. HPC is also a rare tumor type, comprising only 0.4% of all intracranial tumors [12], that is characterized by arising from pericytes surrounding capillary walls. In 2016, WHO created the combined term SFT/HPC because of the similar histological and immunohistochemical features between SFT and HPC. The specific features of SFT/HPC include well-defined tumor borders, spindle-to-oval cells, biphasic hypo- and hypercellular areas, well-developed branching vasculature, frequent expression of CD34, negativity of EMA, and NAB2-STAT6 fusion [13]. WHO also created grades of SFT/HPC, where a grade III SFT/HPC has 5 or more mitoses per 10 HPF [14]. We were unable to examine NAB2-STAT6 fusion in the current case because of our hospital system, but we could confirm CD34, 11 mitoses per 10 HPF, and negativity of EMA and therefore diagnosed our case as WHO grade III SFT/HPC.

As per our knowledge, to date, there have been 11 reported SFT cases and 1 reported SFT/HPC case expanding the superior and inferior cerebellar tentorium [1–11]. The 11 SFT cases originated from the cerebellar tentorium, and 7 of them were completely removed. Only 1 of 7 cases recurred and the patient died 12 years after the initial operation [3]. In the present study, we considered the tumor to have originated from the cerebellar tentorium because it adhered strongly to the tentorium.

Kim et al. reported the prognosis of 47 cases of SFT/HPC [13]. They reported that the average progression free survival (PFS) for grade II and III was 89.7 months and the average overall survival (OS) for grade III was 194.8 months. Recurrence rate in grade III is 73.7%. Gross total resection (GTR) was significantly associated with longer PFS and OS. In addition, patients undergoing any form of adjuvant radiation treatment (RT) had longer mean PFS than those who did not. Furthermore, grade III was strongly correlated with the occurrence of extracranial metastasis (42.1%), with an average occurrence time of 208.7 months. Sung et al. reported the prognosis of 60 cases of SFT/HPC [11]. In grade II cases, GTR or adjuvant RT was associated with longer PFS. The PFS, OS, and time to extracranial metastasis were shorter for patients in the grade III group than those in the grade II group (111.3 vs 31.9 months). Based on these reports, treatment for SFT/HPC is GTR in principle, although RT is also effective when GTR has not been achieved.

In the present case, we were able to completely remove the tumor and found no recurrence or extracranial metastasis at 1.5 years after the operation. However, we must continue to strictly observe the whole body of the patient, including extracranial areas, because recurrence or extracranial metastasis is likely in grade III cases.