Case Report

Parasagittal meningeal hemangiopericytoma/solitary fibrous tumor: Two case reports and a literature review

Asen Hristov Cekov, Donika Ivova Vezirska, Christo Tzekov Tzekov, Vladimir Stefanov Nakov

Department of Neurosurgery, Acibadem City Clinic University Hospital Tokuda, Sofia, Bulgaria.

E-mail: Asen Hristov Cekov - cekovmd@gmail.com; *Donika Ivova Vezirska - d.vezirska@gmail.com; Christo Tzekov Tzekov - tzekovchr@abv.bg; Vladimir Stefanov Nakov - vladimir_nakov@yahoo.com

*Corresponding author:
Donika Ivova Vezirska,
Department of Neurosurgery,
Acibadem City Clinic
University Hospital Tokuda,
Sofia, Bulgaria.

d.vezirska@gmail.com

ABSTRACT

Background: Solitary fibrous tumor/meningeal hemangiopericytoma (SFT/M-HPC) is a rare neoplasm which accounts for around 1% of the intracranial masses. This pathology has a high risk for recurrence and metastasis to distant locations such as the liver, lungs, and bones. Precise diagnosis necessitates detailed histopathological examination.

Case Description: We present two case reports of SFT/M-HPC. The first case is a 44-year-old female who presented with headache, nausea, vomiting, and frontal ataxia for several months. Imaging findings showed a large parasagittal extra-axial mass with compression of the frontal horns of both lateral ventricles. She underwent gross total resection with an uncomplicated postoperative period. The patient had no recurrent tumors or distal metastases in the follow-up period of 5 years. The second case is a 48-year-old male who presented with right-sided hemianopsia and hemiparesis. Computed tomography (CT) scans revealed a large parieto-occipital extra-axial mass with superior sagittal sinus engulfment and dislocation of the interhemispheric fissure. He underwent gross total resection with an uncomplicated postoperative period. Six years later, he presented with right-sided weakness. CT scan showed a multifocal recurrent mass at the previous location. He underwent subtotal resection with an uncomplicated postoperative period.

Conclusion: SFT/M-HPC should be considered when presented with a meningioma-like tumor mass on preoperative imaging. Immunohistochemical study is crucial for the correct diagnosis. Strict long-term follow-up examinations and regular magnetic resonance imaging scans are key to preventing the appearance of metastases and large recurrent masses.

Keywords: Meningeal hemangiopericytoma, Meningioma, Parasagittal, Solitary fibrous tumor

INTRODUCTION

Solitary fibrous tumor/meningeal hemangiopericytoma (SFT/M-HPC) is a rare tumor which accounts for around 1% of the intracranial masses[1] and 0.4% of all central nervous system (CNS) pathology.[15] The name of this pathology changed over the years – the meningeal hemangiopericytoma (HPC) was initially considered to derive from the pericytes of Zimmermann. At present, mentioned in the 2016 World Health Organization (WHO) update of the CNS tumors classification as one single entity called solitary fibrous tumor and hemangiopericytoma (SFT/HPC).[11]

The SFT/M-HPC closely mimics meningioma both clinically and radiologically. However, this pathology is much more aggressive and tends to high recurrence rate and metastasis to distant locations both intra- and extracranially.[8,14]

Here, we report two cases of parasagittal SFT/M-HPC and shortly review diagnosis tips, imaging and pathology findings, and current trends in patient management.

CASE PRESENTATION

Case 1

The first case is a 44-year-old female who presented with headache, nausea, vomiting, and frontal ataxia. Magnetic resonance imaging (MRI) scans showed a large extraaxial (65 × 60 mm) parasagittal mass with compression of the frontal lobe and the frontal horns of both lateral ventricles [Figure 1]. The falx cerebri and the anterior cerebral arteries (ACAs) were dislocated to the right.

She underwent bifrontal craniotomy with ligation of the anterior third of the superior sagittal sinus. The depth and borders of the lesion were explored through ultrasound (US). The excised tumor was partially aspirable and formed a well-demarcated arachnoid plane – the feeding vessels were identified to be direct branches of the ACA. Profuse intraoperative bleeding occurred, and blood loss was estimated at around 500 mL – however, this was managed with bipolar coagulation and hemostatic agents. The dura over the lesion was prophylactically excised, and duraplasty was achieved with 7.5 × 7.5 mm Lyoplant® Onlay (Aesculap, BBraun, Melsungen, Germany). Standard layered closure was performed.

On postoperative day 1, a head computed tomography (CT) scan was performed – it revealed subtotal resection of the lesion [Figure 2]. Postoperative anosmia was the only complication in the postoperative period. The patient was discharged on postoperative day 3.

Histopathological examination revealed a malignant tumor with diffuse structure and high cellularity, which consisted of cells with poor cytoplasm and large ovoid vesicular nuclei. Small parts of the probe showed pseudorosettes. Additional immunohistochemical processing showed more than five mitoses/ten high-power fields, as well as EMA-, NSE-, CD34-, and vimentin-positive staining and strongly positive
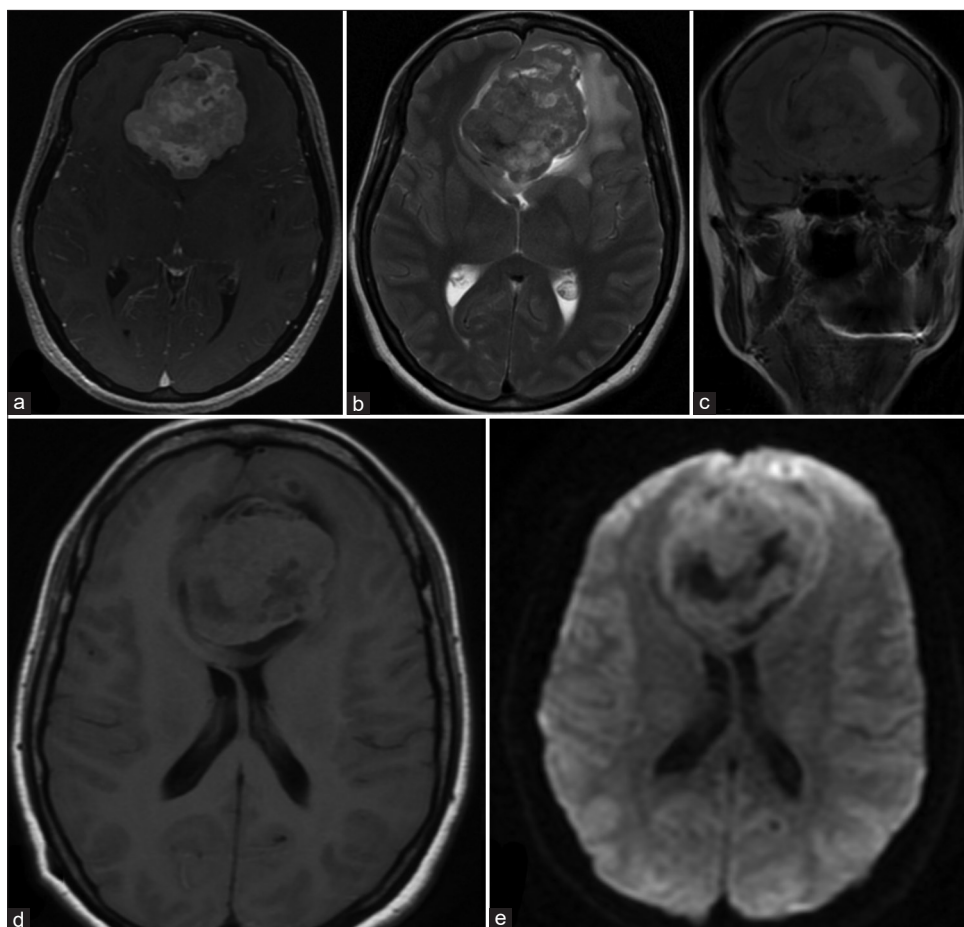

Figure 1: Appearance of the lesion on (a) Axial three-dimensional fast spoiled gradient-echo sequence with contrast matter; (b) Axial T2 PROPELLER sequence; (c) Coronal T2 FLAIR PROPELLER sequence; (d) Axial T1 FLAIR sequence; (e) Axial diffusion-weighted imaging sequence with apparent diffusion coefficient map. FLAIR: Fluid-attenuated inversion recovery.

staining for Bcl2 and CD99. Finally, the lesion was diagnosed as the WHO grade III meningeal HPC.

A multidisciplinary committee agreed on postoperative radiotherapy. Adjuvant field radiotherapy with 60 Gy in 2 Gy/fraction was delivered through three-dimensional conformal radiation therapy. Only mild side effects, such as erythema and alopecia, were observed.

Follow-up head MRI scans were performed at seven months, 15 months, 20 months, and 31 months post-surgery [Figure 3].
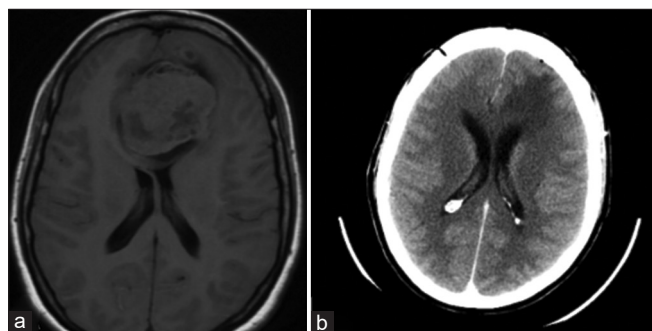

Figure 2: (a) The lesion on an axial section of the preoperative magnetic resonance imaging scan; (b) Postoperative computed tomography head scan showing the subtotal resection of the lesion with decompression of the frontal horns of both lateral ventricles.
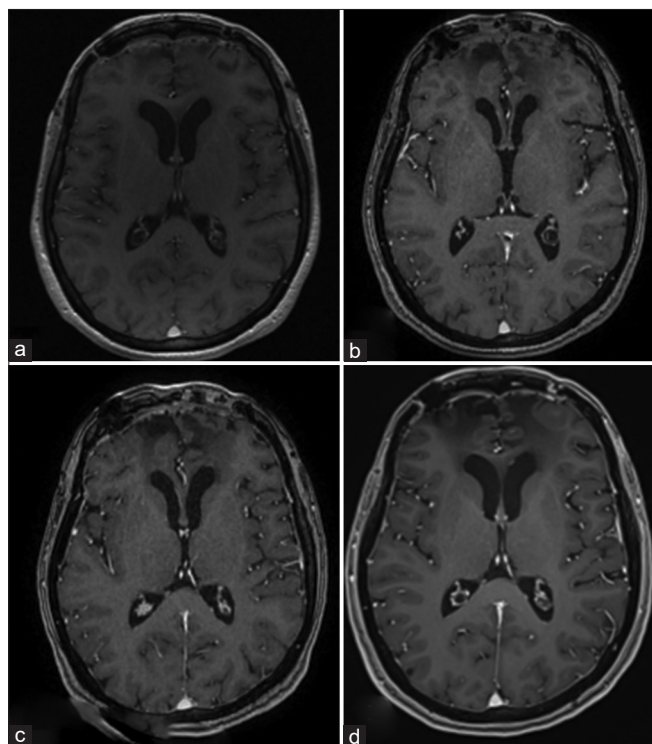

Figure 3: Follow-up head magnetic resonance imaging scans were performed at (a) 7 months; (b) 15 months; (c) 20 months; (d) 31 months post-surgery.

The patient had no recurrent tumors or distant metastases nor any neurological worsening in the follow-up period of 5 years.

Case 2

The second case is a 48-year-old male with hepatitis B who presented with sensory aphasia, right-sided hemianopsia, and 3/5 right-sided hemiparesis. CT scans revealed a large extra-axial (73×58 mm) parasagittal mass in the left parietooccipital region, encasing the superior sagittal sinus and dislocating the interhemispheric fissure 7 mm to the right [Figure 4a].

He underwent a left parietal craniotomy with a preoperatively placed lumbar drain. Mannitol was applied before the durotomy and cerebrospinal fluid was drained through the lumbar drain because of the severely increased intracranial pressure. The depth and borders of the lesion were explored through the US, and the dura was opened in a C-shaped fashion based on the superior sagittal sinus. The excised tumor infiltrated the underlying cortex, the falx, and the superior sagittal sinus. Intraoperative neuromonitoring was used to verify that no functionally active zones of the motor cortex were affected. The tumor was debulked using an ultrasonic aspirator. The middle third of the superior sagittal sinus was ligated, and the infiltrated part of the falx was excised. Gross total resection was achieved. Again, intraoperative bleeding occurred, and blood loss was estimated at around 500 mL. The dura over the lesion was excised, and duraplasty was achieved with an autologous periosteal flap. Standard layered closure was performed.

Postoperatively, the patient was transferred to the intensive care unit for intensive monitoring – he was transferred back to the neurosurgical ward on postoperative day one after a head CT scan showing gross total resection of the mass [Figure 4b]. The remaining part of the postoperative period was uncomplicated – the right-sided hemiparesis
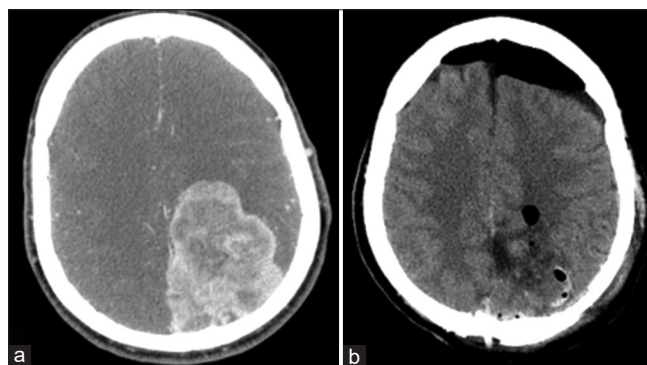

Figure 4: (a) The lesion on an axial section of the preoperative computed tomography (CT) scan with contrast matter; (b) Postoperative CT head scan showing the subtotal resection of the lesion and postoperative pneumocephalus, which was asymptomatic.

progressively improved to 5/5 with occasional episodes of 4/5 muscle power. He was discharged on postoperative day 5.

Histopathological examination revealed a benign mesenchymal tumor with high cellularity which consisted of small monomorphic cells around vessels with HPC-like structure. Additional immunohistochemical processing showed no mitoses/10 high-power fields, as well as NSE-, Bcl2-, CD99-, CD34- and vimentin-positive and EMA-, progesterone receptor-, and CD56-negative staining. Finally, the lesion was diagnosed as a WHO grade I solitary fibrous tumor.

Six years later, the patient presented with 3/5 right-sided weakness again. CT scan showed a multifocal recurrent mass at the previous location, with the largest focus having a diameter of 49 mm [Figure 5a].

He underwent left parasagittal recraniotomy based on the previous surgical intervention. The depth and borders of the lesion were explored through the US, and the dura was opened in a C-shaped fashion based on the superior sagittal sinus. The tumor was debulked using an ultrasonic aspirator. The excised tumor was located over the convexity – it was richly vascularized from branches of the falcine arteries. Gross total resection was achieved. Again, intraoperative bleeding occurred, and blood loss was estimated at around 500 mL. The dura was excised and duraplasty was achieved with an autologous periosteal flap. Standard layered closure was performed.

Postoperatively, the patient was transferred to the neurosurgical ward. A head CT scan was done on postoperative day 1, showing subtotal resection of the mass – a small residual tumor with a diameter of 8 mm remained on the falx [Figure 5b]. The postoperative period was uncomplicated. He was discharged on postoperative day 4.

Histopathological examination revealed a benign mesenchymal tumor with high cellularity, which consisted of cells with poor cytoplasm and large ovoid vesicular nuclei. 5 mitoses/8 mm^2 were present. Again, the lesion was diagnosed as a WHO grade I solitary fibrous tumor.
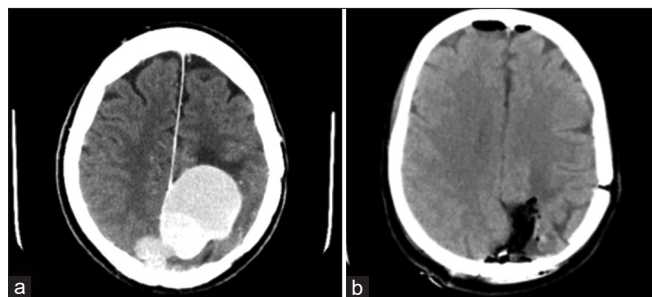

Figure 5: (a) The lesion on an axial section in the late phase of the preoperative computed tomography (CT) scan with contrast matter; (b) Postoperative CT head scan showing the subtotal resection of the lesion.

DISCUSSION

SFT/M-HPC is a rare pathology which is very difficult to diagnose initially.[8] Historically, it has been a challenge to classify and treat because of its complex histological nature, rapidly progressing course, and poor prognosis in comparison to its most frequent imitator, namely meningioma.[19] The most frequent location of this pathology is supratentorial along the falx and the dural sinuses,[20] but some case reports show that posterior fossa localization, as well as spinal and cranial co-existence of meningeal HPCs, is also possible.[13] The clinical presentation of SFT/M-HPC is not specific and has poor diagnostic value – most patients present with headache, vomiting, motor weakness, and seizures.[4] Liu *et al.* outline several important differences in the initial differential diagnosis between meningeal HPC and meningioma [Table 1].[10]

Table 1: Differences between meningeal hemangiopericytoma and meningioma.

	Meningeal hemangiopericytoma	Meningioma
Sex	Predominantly male	Predominantly female
Age	Early 40s	Early 50s
The interval between initial symptoms and the diagnosis	4–12 months	1–2 years

Classification

The etiology of meningeal HPC is a controversial topic which has led to changes in the terminology – first, they were considered to be an angioblastic subtype of meningioma.[3] In 1942, Stout and Murray proposed that HPCs are derived from smooth muscle perivascular pericytes of dural capillaries, also known as pericytes of Zimmerman.[3] In 2016, the new update on the WHO classification of tumors of the CNS classified SFT/HPC as one entity because of their shared inversions at 12q13 which led to STAT6 nuclear expression. A three-grade system for the grading of this tumor based on the number of mitoses/10 high-power fields were designed with grade I being what was deemed solitary fibrous tumor, grade II is what was previously called HPC, and grade III being what was before termed anaplastic HPC. The 2021 WHO classification of the tumors of the CNS renders the term "hemangiopericytoma" obsolete and replaces it fully with "solitary fibrous tumor" – the grading is decided according to a 3-tiered scale.

Radiological findings

Meningeal HPC is very difficult to differentiate from meningiomas radiologically. CT imaging reveals both

tumors as iso-dense with intense contrast enhancement, but meningiomas cause hyperostosis, while HPC may cause erosion of the overlying bone.[17]

MRI imaging reveals that atypical meningeal HPCs present with lobulated and/or irregular cross-leaf-shaped masses associated with prominent brain edema and frequent bone destruction, as well as a narrow base of the "dural tail."[3] Several authors propose the use of magnetic resonance diffusion-weighted imaging for the identification of intracranial HPCs – they use apparent diffusion coefficients (ADC) to observe the heterogeneity of the tumors.[2,10,18] The study shows that HPCs tend to be more heterogeneous than meningiomas. However, the higher the grade of the meningioma, the more heterogeneous it tends to become. The latter does not change the treatment strategy for both atypical high-grade meningiomas or HPC – radical surgical resection is indicated in both cases. Besides ADC, Chen *et al.* considered susceptibility-weighted imaging (SWI) as another modality to identify HPCs, which can map tumor microvasculature and different blood products by their intratumoral susceptibility signals.[2] Their study concluded that the ADC ratio showed superior diagnostic accuracy in differentiating SFT/HPC from high-grade meningioma, and SWI was better for setting apart SFT/HPC from low-grade meningioma. However, calcifications in SWI sequences were not considered, and only nine subtypes of meningiomas were included in this study. In addition, advanced MRI imaging is not always possible, especially in low-income countries.

Histopathological findings

Tumor size was found not to be linked to the overall survival or recurrence rate.[5] Staghorn vessels may indicate SFT/HPC.[3] Electronic microscopy shows that gap junctions and desmosomal attachment, a key feature of the pathological image of meningiomas, are not present in meningeal HPCs – a highly electron-dense basal membrane-like substance encircles the cells. Meningiomas reportedly have less extracellular space and more intracellular space, restricting net water diffusion compared to hemangiopericytomas.[18] This may be the explanation for the difference in the ADC maps.

Immunohistochemical staining is key for the labeling of the tumor as meningeal HPC. The 2016 WHO guidelines for the diagnosis of HPC/SFT utilize the NAB2-STAT6 fusion gene and STAT6.[5] However, the relevant molecular assays can be quite costly which renders them difficult to utilize in lower-income countries.[8] According to Han *et al.*, markers that can be utilized in most pathology laboratories include CD34, Bcl-2, and CD99, along with histoimmunochemical staining.[7] Negative markers include SSTR2A and EMA, especially if they are co-expressed, indicating meningioma. For our cases, we acted in accordance with this recommendation.

Management

Current management strategies mostly include surgery, radiotherapy, and chemotherapy.[3] Gross total resection remains the most important part of the treatment.[12,15] Rutkowski *et al.* reported that the group of patients who underwent gross total resection had a median survival of 13 years, compared to the subtotal resection group of patients who had a median survival of 9.75 years.[16] If a multimodal approach was used, the median time to local recurrence was reported to be between 12 and 96 months with lower recurrence rates. Melone *et al.* observed the survival rates in a group of patients who underwent microsurgical resection at 5 and 10 years to be 94% and 72%, respectively.[12] However, gross total resection is sometimes impossible because of the risk of injury to important neurovascular structures[3] and the subsequent possibility of neurological deficits and venous thrombosis.[9] Preoperative embolization may be helpful in preventing massive intraoperative blood loss, similar to meningioma surgery – nevertheless, this is sometimes not possible because of the possibility of vessel occlusion.[3] This leaves subtotal resection as the only viable option without compromising either the patient's quality of life or the success rate of the treatment. In such cases, radiotherapy may prove to be a useful adjuvant therapeutic tool but studies are not unanimous regarding the matter.[1] Haas *et al.* present a retrospective study of 48 patients that shows the combination of subtotal resection with radiotherapy is superior to subtotal resection only regarding local recurrence control, but both methods have no statistically significant difference when it comes to the occurrence of distant metastases.[6] Reportedly, stereotactic radiosurgery can also be utilized in small HPC (<2 cm).[3,15] However, it is also suggested that radiotherapy may increase the risk of distant metastases.

Contrary to some studies, Rutkowski *et al.* demonstrate a large-scale study which concludes that postoperative adjuvant radiotherapy does not greatly influence the overall survival rate.[16] They propose that a radiation dose of >50 Gy is associated with higher mortality – however, the poor survival rate may also be linked to an overall more malignant tumor, thus worsening the prognosis as an independent factor. Melone *et al.* suggest that adjuvant radiotherapy does not affect the overall survival rate but prolongs the recurrence-free period.[12]

Management strategies for recurrent masses, such as our second case, are yet to be established – despite the high recurrence rate of HPC, the studies for this topic have a very limited sample size and cannot provide physicians with guidelines. However, Rutkowski *et al.* propose the use of surgical resection aided by adjuvant radiotherapy – their study shows that the time to second recurrence among patients who received adjuvant radiotherapy was 10.3 years compared to 5.3 years in patients who did not.[14] We

estimated that radiotherapy was unnecessary in our case because of the favorable histological result.

CONCLUSION

SFT/HPC should be considered when presented with a meningioma-like tumor mass on preoperative imaging. Initial gross total resection of the tumor provides a viable management strategy, and radiotherapy may be a good option for adjuvant therapy in selected cases. Strict long-term follow-up examinations and regular MRI and/or CT scans are key to preventing the appearance of metastases and large recurrent masses, even if pathological findings show WHO grade I with low chances for recurrence and malignization, as in our second case. However, management of this pathology remains complex, and further research is necessary to establish treatment guidelines, especially regarding recurrent cases.

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Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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