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Review Article

Sporadic supratentorial hemangioblastoma with meningeal affection: A case report and literature review

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ABSTRACT

Background: Hemangioblastomas are vascular tumors, of benign behavior, that originate in the central nervous system. Supratentorial hemangioblastomas are extremely rare and are generally associated with Von Hippel-Lindau disease (VHL). The involvement of structures by contiguity, such as blood vessels or meninges, is something exceptional. There are few references in the literature of supratentorial hemangioblastomas with meningeal involvement and most of them are described in reports or small case series.

Methods: We performed a systematic review of the literature to observe the characteristics of patients with supratentorial hemangioblastomas with meningeal involvement. In addition, we present the case of a 43-year-old male patient with a sporadic supratentorial hemangioblastoma with both, meningeal and vascular involvement that recurred years after treatment.

Results: The patients presented supratentorial tumors with meningeal involvement, we had a 1.2:1 ratio male-female distribution. The mean age was 50 years. Most tumors were located in the cerebral hemispheres, the lobe affected most frequently was the frontal lobe. About 67% of the cases were sporadic and only 21% were related to VHL disease. There were three cases of recurrence after surgery.

Conclusion: Supratentorial hemangioblastomas are extremely rare lesions. It is rare for supratentorial hemangioblastomas to invade adjacent structures such as blood vessels or meninges, however, when this happens, it is recommended a preoperative vascular imaging study, especially in parasagittal hemangioblastomas where superior longitudinal sinus may be involved. In these cases, *en bloc* surgical resection is difficult and the probability of recurrence is higher. Although clinical trials and studies with a greater casuistry are necessary to establish guidelines for the management of supratentorial hemangioblastomas, nowadays, contribution of new cases is useful for understanding this pathology.

Keywords: Meninges, recurrence, supratentorial hemangioblastoma, Von Hippel-Lindau disease

INTRODUCTION

Hemangioblastomas are benign tumors of vascular origin that develops in the central nervous system. They represent approximately 2% of all intracranial tumors and the most frequent nonmalignant intra-axial tumor in the posterior fossa in adults.^[7] Hemangioblastomas can arise in any region of the central nervous system; however, they are more frequently located in the posterior fossa, accounting for up to 5–15% of tumors in this location.^[10] The next most frequent location is the spinal cord and other less frequent locations include the brain stem and the

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supratentorial compartment.^[15] Most hemangioblastomas are sporadic, but up to 30–35% occur in the context of Von Hippel-Lindau disease (VHL).^[32,39] These tumors rarely invade contiguous structures such as the meninges or blood vessels, with few cases published in the literature so far.

The present study presents the case of a 43-year-old male patient with a sporadic supratentorial hemangioblastoma, with meningeal involvement, which recurs 10 years after the first surgery. A review of the characteristics of these tumors described in the literature is performed.

MATERIALS AND METHODS

A comprehensive systematic review of literature was performed. Articles were identified through PubMed search using the key words “supratentorial hemangioblastoma” and “spontaneous hemangioblastoma” and in combination with the key words “VHL disease” and “meningeal involvement.” All references of these articles were classified to extract data about patients that presented a supratentorial hemangioblastoma and had meningeal involvement whom underwent surgery. A total of 168 articles on supratentorial hemangioblastomas were identified, of which, only 32 had meningeal involvement. All references contained individual patient data or purely supratentorial aggregated data sets of either histologically confirmed tumors. Both patients with sporadic hemangioblastomas and VHL disease-associated hemangioblastoma were included in the study.

Data from individual and aggregated case series were extracted from each article. The epidemiological characteristics of the patients, the location of the tumor, the appearance of the tumor, the association with VHL disease, and postoperative relapses were reviewed.

Finally, we present the case of a 43-year-old male patient who had previously undergone an epididymal cyst 18 years earlier and a right parasagittal parietal hemangioblastoma 12 years ago [Figure 1]. The genetic study of VHL disease was negative. During the 1st year of follow-up, the patient presented clinical-radiological stability. Ten years after surgery, he was admitted with drug-resistant somatosensory seizures. The magnetic resonance imaging (MRI) showed a tumor recurrence with invasion of the superior longitudinal sinus (SLS), reducing the blood flow of its posterior portion [Figure 2]. A subtotal resection was performed, showing a cystic, pink looking tumor that invaded the SLS and the adjacent dura. The histological study was compatible with hemangioblastoma [Figure 3]. Postoperative MRI showed a tumor remainder closely attached to the SLS. The patient was treated with stereotactic radiosurgery and after 6 months remains asymptomatic and free of somatosensory seizures, maintaining radiological stability of the residual tumor. Our case represents the twentieth-second case that meets

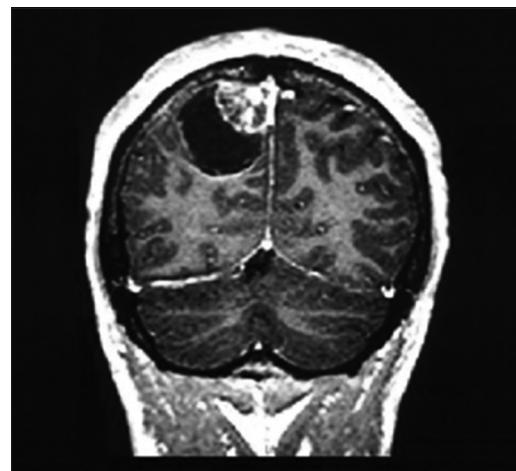


Figure 1: T1 sequenced MRI with paramagnetic contrast showing a right parietal tumor lesion with a peritumoral cyst in a coronal section.

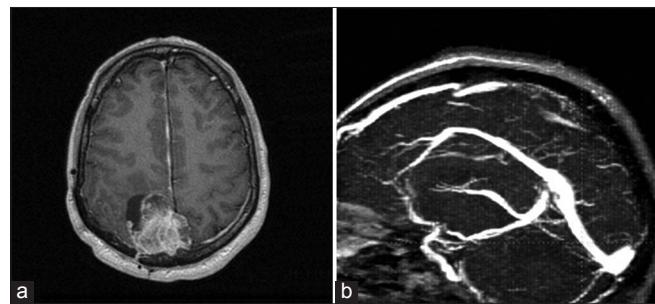


Figure 2: Left: T1 sequenced MRI with paramagnetic contrast showing tumor recurrence invading the SLS. Right: Venous phase angioresonance showing a signal void in the posterior region of the SLS.

these characteristics, the third case where a recurrence was detected during the 1st year from the initial treatment and the first one that causes involvement of the SLS, more rarely, reducing its blood flow.

RESULTS

The literature search yielded a total of 168 references that contained data on patients with supratentorial hemangioblastomas. However, only 32 patients had a supratentorial hemangioblastoma with meningeal involvement.^[1,3-9,11-14,16-19,21,25,27,29,33,35-38,40,42-44,46-48] Of the 33 published cases of supratentorial hemangioblastomas with meningeal involvement, including our case, 17 (52%) were male and the other cases were female, and the median age was 50 years. In relation to the intracranial location, 18 (55%) were affecting the frontal lobe, 9 (27%) the parietal lobe, 4 (12%) the occipital lobe, and only 2 (6%) in the sellar/parsellar region. There were not described supratentorial hemangioblastomas with meningeal involvement located in the temporal lobe

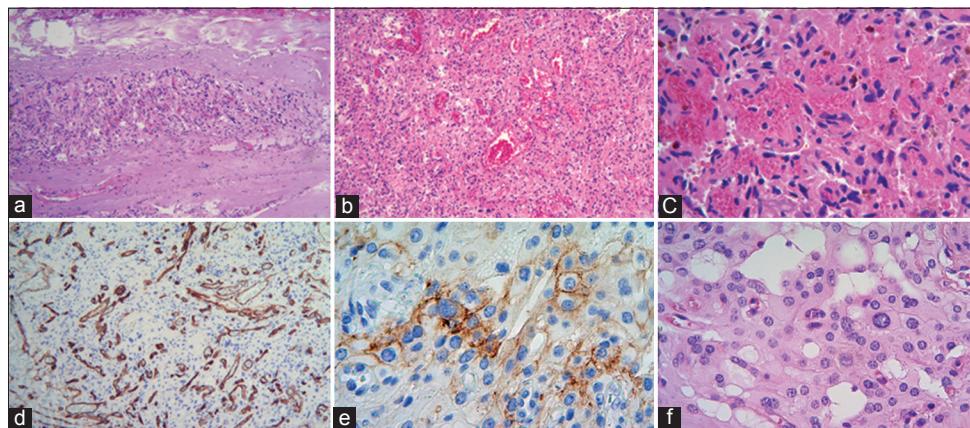


Figure 3: (a) Hematoxylin-eosin (H-E) staining showing the tumor invading meningeal structures. (b) H-E staining showing tumor tissue with numerous blood vessels. (c) H-E staining showing stromal and capillary cells with hemosiderin deposits due to bleeding. (d) Stromal cells showing immunoreactivity for CD34. (e) Vascular endothelial proliferation showing immunoreactivity for podoplanin D2-40. (f) H-E staining showing neoplastic stromal cells.

neither in the insular region. The situation of the lesions presented 12 cases (36%) on the cerebral convexity, 8 (25%) in the parasagittal region, closely related to the SLS, but not invading it, 9 (27%) adjacent to the brain sickle or tentorium, 2 (6%) in the sellar/parasellar region, and in 2 cases (6%), their intracranial location was not accurately described. The appearance of the tumors was described as a solid mass in all cases of supratentorial hemangioblastoma with meningeal involvement, except in one article, in which the tumor appearance was not specified. In relation to the association of VHL disease, 22 cases (67%) were not related to VHL disease, 7 cases (21%) had the disease, and in 4 cases (12%), the VHL gene mutation was not determined, neither personal nor family stigmas were described related to the clinical case [Table 1]. It is possible that this figure is not true to reality and in clinical practice, there are many more cases treated.

Finally, only three cases of supratentorial hemangioblastoma with meningeal involvement presented recurrence 2 years after surgery.

DISCUSSION

Supratentorial hemangioblastomas are rare tumors and account for 1–2% of all hemangioblastomas.^[7] To date, 168 cases have been described in the literature of supratentorial hemangioblastomas. They generally affect men between the third and the fifth decade of life and most of them appear in the context of VHL disease. According to Mills *et al.*,^[26] 60% of patients with supratentorial hemangioblastomas are diagnosed with VHL disease or have a positive genetic study for the VHL gene mutation, which is located on the short arm of chromosome 3, position 3p25–26.^[39] Regarding the intracranial location, most are located in the cerebral hemispheres (58%), the sellar and parasellar region (26%), the cerebral ventricles (10%), and the other

less frequent locations (6%), among which stand out two cases located in the Meckel's cavum^[20,34] and another in the choroidal fissure.^[36] Hemangioblastomas located in the sellar and parasellar region are of special interest, since 87% of them are related to VHL disease.^[2] According to the studies by Peyre *et al.*^[30] and Lonser *et al.*,^[23] the most frequent location of supratentorial hemangioblastomas associated with VHL disease is the pituitary stalk, the hippocampus/parahippocampal region, and the optic nerve, which supports the hypothesis that hemangioblastoma progenitor cells can originate from diencephalic structures.

Supratentorial hemangioblastomas that develop in adjacent areas or involving meningeal structures are even rarer. According to Rocha *et al.*^[35] in 2017, a total of 20 cases of supratentorial hemangioblastomas with meningeal involvement were described, but between 1942 and 2017, we collected seven cases more than. In 2018, Tabibkhooei *et al.*^[40] described a case in the sellar/parasellar region with cavernous sinus invasion. In 2019, Vicente *et al.*^[45] described another case, a 64-year-old woman with a left frontal parasagittal supratentorial hemangioblastoma adjacent to the dura. That year, Bian *et al.*^[4] and Baran *et al.*^[3] described two supratentorial hemangioblastomas located in the parietal lobe. Finally, Khelifa *et al.*^[16] described a new case, a 20-year-old man with an occipital hemangioblastoma in the relation with the falx cerebri. The case described in our work represents the thirtieth-third case described of the literature regarding a supratentorial hemangioblastoma with involvement of the meningeal membranes.

Most supratentorial hemangioblastomas are located in the cerebral hemispheres. Of these 33 supratentorial hemangioblastomas with meningeal involvement cases described in the literature, 31 (94%) were located in the cerebral hemispheres, with the frontal lobe being the most

Table 1: Supratentorial hemangioblastomas with meningeal involvement described from 1942 to 2020.

Cases	Age	Sex	Localization	Aspect	VHL	
Zeitlin <i>et al.</i> (1942) ^[48]	54	M	Parasagittal	Frontal	Solid	-
Rivera <i>et al.</i> (1966) ^[33]	16	M	Parasagittal	Parietal	Solid	-
Ischwar <i>et al.</i> (1971) ^[11]	62	F	Falx	Occipital	Solid	+
Böckem (1975) ^[5]	43	M	Tentorium	Occipital	Solid	NA
Lee <i>et al.</i> (1978) ^[21]	46	M	NA	Frontal	Solid	+
Tomaccini <i>et al.</i> (1980) ^[43]	9	F	Falx	Occipital	Solid	NA
Sharma <i>et al.</i> (1995) ^[36]	72	M	Convexity	Parietal	Solid	-
Choi <i>et al.</i> (1998) ^[6]	26	F	NA	Parietal	Solid	-
Kim <i>et al.</i> (2001) ^[19]	45	M	Convexity	Frontal	Solid	+
Agostinelli <i>et al.</i> (2004) ^[1]	10	F	Convexity	Frontal	Solid	-
Iyigun <i>et al.</i> (2004) ^[12]	61	M	Convexity	Frontal	Solid	-
Varsik <i>et al.</i> (2004) ^[44]	38	M	Parasagittal	Frontal	Solid	NA
Zamzuri <i>et al.</i> (2004) ^[47]	46	M	Falx	Frontal	Solid	+
Cosar <i>et al.</i> (2006) ^[7]	50	M	Parasagittal	Parietal	Solid	-
Jang <i>et al.</i> (2007) ^[13]	68	F	Convexity	Frontal	Solid	-
Murali <i>et al.</i> (2007) ^[27]	57	M	Parasagittal	Frontal	NA	NA
Sherman <i>et al.</i> (2007) ^[38]	52	F	Convexity	Frontal	Solid	+
Takeuchi <i>et al.</i> (2008) ^[42]	58	M	Parasagittal	Frontal	Solid	-
Courcoutsakis <i>et al.</i> (2009) ^[8]	53	M	Sellar/parasellar region		Solid	+
Elguezabal <i>et al.</i> (2010) ^[9]	67	F	Falx	Frontal	Solid	-
Lozano-Tangua <i>et al.</i> (2010) ^[25]	74	F	Convexity	Frontal	Solid	-
Kaloostian <i>et al.</i> (2012) ^[14]	49	F	Falx	Frontal	Solid	-
Kim <i>et al.</i> (2013) ^[18]	51	F	Convexity	Frontal	Solid	+
She <i>et al.</i> (2013) ^[37]	60	F	Falx	Frontal	Solid	-
Kim <i>et al.</i> (2016) ^[17]	77	F	Convexity	Frontal	Solid	-
Pandey <i>et al.</i> (2016) ^[29]	40	M	Convexity	Parietal	Solid	-
Rocha <i>et al.</i> (2017) ^[35]	62	F	Convexity	Parietal	Solid	-
Tabibkhooei <i>et al.</i> (2018) ^[40]	29	F	Sellar/parasellar region		Solid	-
Vicente <i>et al.</i> (2019) ^[45]	64	M	Parasagittal	Frontal	Solid	-
Bian <i>et al.</i> (2019) ^[4]	70	F	Falx	Parietal	Solid	-
Baran <i>et al.</i> (2019) ^[3]	57	F	Convexity	Parietal	Solid	-
Khelifa <i>et al.</i> (2020) ^[16]	20	M	Falx	Occipital	Solid	-
Sánchez Ortega <i>et al.</i> (2020)	43	M	Parasagittal	Parietal	Solid	-

Updating of the tables described by Lozano-Tangua *et al.*, Kim *et al.*, Rocha *et al.*, and Khelifa *et al.*

frequent location; while 2 (6%) were located in the sellar/parasellar region.

As described previously, supratentorial hemangioblastomas usually appear correlated with VHL disease. In relation to supratentorial hemangioblastomas with meningeal involvement, 22 cases (67%) were not related to VHL disease, 7 cases (21%) presented the disease, and in 4 cases (12%), the VHL gene mutation was not identified, neither personal nor family stigmas were described in relation to the clinical case. Our case, despite having suffered from an epididymis cyst, had no other stigmata of VHL disease and the genetic study was negative. At the moment, it is unknown why some hemangioblastomas invade the meningeal membranes and whether or not there is a genetic mutation that predisposes these tumors to have a certain affinity for invading adjacent tissues. Therefore, in patients with intracranial hemangioblastomas, it is recommended to carry out a genetic

and extension study to determine whether or not the patient suffers from this disease.

The pathogenesis of the disease is unknown, although tumor secretion of certain factors, such as vascular endothelial growth factor (VEGF), platelet-derived growth factor, transforming growth factor type alpha (TGF- α), or erythropoietin, is associated with progressive tumor growth and erythrocyte production, respectively. The most accepted hypothesis for this process is a mutation of the VHL gene that encodes the VHL protein, a protein involved in proteasomal degradation of the hypoxia-inducible factor (HIF-1 alpha).^[28] In the presence of the mutation, the VHL protein is truncated and overproduction of HIF-1 alpha and its angiogenic products occurs, resulting in increased vascular cell recruitment and the formation of hypervascular tissue characteristic of the tumor.

The most frequent clinical manifestations of patients with supratentorial hemangioblastomas are headache and seizures.

These can be partial or partial with secondary generalization and their expression depends on the location of the lesion. The case we present suffered from partial sensory seizures in the left extremities in relation to a supratentorial hemangioblastoma located in the parietal lobe and with involvement of the right somesthetic cortex. Other symptoms such as focal deficits or cognitive deterioration are less common and are related to tumors with a significant cystic component that causes compression of the underlying brain parenchyma.^[46] Infiltration of the nervous tissue is rare and the debut in the form of intracranial hemorrhage is exceptional.

Radiological diagnosis is based on MRI. In most cases, a solid nodule and an adjacent cystic component are identified. In T1-weighted sequences and after gadolinium administration, the nodule appears homogeneous and hyperintense; while in the T2-weighted sequences, the cystic component is hyperintense. Most infratentorial hemangioblastomas are cystic, whereas supratentorial hemangioblastomas are usually solid cystic with a smaller cystic component. This could be because the brain offers less resistance to fluid diffusion than the cerebellum.^[30] Likewise, supratentorial hemangioblastomas with meningeal involvement predominantly have a solid nodule attached to the meningeal membranes, leaving the cystic component without contact with the meninges. Furthermore, parasagittal supratentorial hemangioblastomas can be related to or even invade vascular structures such as SLS. In these cases, it is recommended to perform an MRI in the venous phase or an angiography to assess the degree of patency of the SLS and visualize the arteries that feed the tumor. In this way, it is possible to define a surgical strategy, including endovascular embolization before surgery. The vascular study becomes more important in the parietal or occipital lobe, where the SLS involvement can cause serious complications in the patient.

Blood tests can be useful, since the presence of polyglobulia supports the diagnosis of the disease. Likewise, the presence of other lesions, in particular, hemangioblastomas in other locations, renal or pancreatic tumors, cystadenomas of epididymis (men) or broad ligament (women), and/or pheochromocytomas can guide the diagnosis of VHL disease.^[23] In these cases, it is necessary to carry out a genetic study to confirm the disease and do a family screening.

The differential diagnosis of supratentorial hemangioblastomas includes other tumors such as meningiomas, hemangiopericytomas, and metastases from renal carcinoma. However, supratentorial hemangioblastomas show many radiological similarities with angioplastic meningiomas,^[15] so their diagnostic orientation is not always easy and confirmation of the tumor is only possible through histological study.

The definitive diagnosis is made by histological confirmation. Hemangioblastomas are benign tumors

(Grade I of the World Health Organization) and highly vascularized.^[24] Macroscopically, they are made up of a constant solid component and a variable cystic component. The solid component consists of a mural nodule, closely related to the pial surface and reddish in appearance, given its high vascularity. The cystic component has a yellowish color, due to its high-protein concentration. It develops as a consequence of fluid loss through the fine and irregular tumor vessels. Hemangioblastomas can be classified according to their cystic pattern: single peritumoral cyst (51%), single intratumoral cyst (17%), multiple peritumoral and intratumoral cysts (4%), and noncyst-associated hemangioblastomas (28%). Microscopically, hemangioblastomas are composed of a network of capillaries surrounded by a layer of endothelium and reticulin fibers. This capillary framework is made up of two cellular components: vascular cells and stromal cells, the latter of polygonal morphology and with a clear cytoplasm and abundant lipid deposits. With immunohistochemical techniques, hemangioblastomas are highly positive for inhibin-alpha, podoplanin D2-40, glucose transporter 1 (GLUT-1), and vimentin. Positivity for the latter supports the hypothesis that hemangioblastomas have a mesenchymal origin. On the other hand, these tumors show negative reactivity for cytokeratin, gliofibrillar acid protein, and epithelial membrane antigen (EMA).^[41] Reactivity is variable for protein S-100 and neuron-specific enolase. In addition, there is an overexpression of VEGF. Likewise, macrophages positive for Schiff's periodic acid staining are found. In this way, it is possible to distinguish hemangioblastomas from angioplastic meningiomas and metastases from renal carcinoma, since the latter are positive for EMA and negative for inhibin-alpha, contrary to what happens in hemangioblastomas.

The treatment of hemangioblastomas is radical *en bloc* resection. In cases where the resection of the tumor is not complete, there is an increased risk of recurrence, something that seems more trending in supratentorial hemangioblastomas that invade structures such as SLS. In our case, the patient underwent a subtotal resection of the hemangioblastoma 12 years earlier, with only some remaining tumor attached to the SLS. After 10 years, the patient develops the recurrence that is presented in this work. In the majority of cases of supratentorial hemangioblastomas with meningeal involvement described in the literature, the removal of the tumor was complete and the follow-up time was not more than 5 years. Only the cases described by Rivera *et al.*^[33] and Ishwar *et al.*,^[11] they presented tumor recurrence, at 8 years and at 2 years, respectively. Although there are no clinical trials or reviews with a greater number of cases in the literature, it can be concluded that in patients treated by partial or subtotal resection, close follow-up should be carried out, even beyond 5 years; as well as attributing

possible new-onset neurological symptoms to the possibility of tumor recurrence.

Treatment with radiosurgery in small focal lesions or in tumor remains after incomplete resection, seems to delay tumor progression, and, in some cases, keeps the course of the disease stable.^[22] There are no clinical trials determining the benefit or not of stereotactic radiosurgery in the treatment of supratentorial hemangioblastomas. Mills *et al.*^[26] in their systematic review of the literature explained that the disease-free survival at 5 years for supratentorial hemangioblastomas, both sporadic and associated with VHL disease, treated with stereotactic radiosurgery is 63–85%, although the scarcity of cases limits the conclusions of the studies. On the other hand, radiotherapy and chemotherapy have not shown effectiveness in these tumors. In most published series, complete tumor resection was superior to radiation therapy in terms of disease-free survival. In recent years, adjuvant therapy has been tried with sunitinib, a VEGF inhibitor, with little result.^[31]

CONCLUSION

Supratentorial hemangioblastomas are extremely rare lesions. Despite the fact that the majority of hemangioblastomas are sporadic, those that occur in the supratentorial compartment are more related to VHL disease, so it is necessary to carry out a genetic and systemic study to rule out extracranial lesions in these patients. It is rare for supratentorial hemangioblastomas to invade adjacent structures such as blood vessels or meninges, however, when this happens, it is advisable to perform a preoperative vascular imaging study, especially in parasagittal hemangioblastomas where SLS may be involved. In these cases, *en bloc* surgical resection is difficult and the probability of recurrence is higher. Despite adjuvant treatment with radiotherapy or stereotactic radiosurgery, it is recommended to closely monitor these patients to detect possible tumor recurrences.

Authors' contributions

All authors have participated in the preparation of the manuscript and concur with the findings and results.

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.

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