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Case Report

Parafalcine Hemangioblastoma with no Von Hippel Lindau Syndrome Association: A Case Report

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Abstract

Sporadically occurring supratentorial hemangioblastomas are extremely rare tumors. Our aim is to report a detailed analysis of a parafalcine hemangioblastoma case and to contribute to the literature in terms of diagnosis and treatment of such neoplasm. We report a 37-years-old patient with a history of headache and mild forgetfulness. Magnetic resonance imaging revealed 1cm x 1,5cm x1cm lesion, adherent to falx cerebri, and MRI spectrograph confirmed only the presence of neoplasm but could not help with the differentiated diagnosis. After the preparations total excision of the lesion was performed with a preliminary diagnosis of metastasis. Histopathological examinations revealed that the lesion was a capillary hemangioblastoma and the genetic study of the patient was negative for von Hippel Lindau mutations. Sporadic supratentorial hemangioblastomas are rare but curable tumors. They can be easily misdiagnosed because their imaging characteristics are very similar to gliomas or meningiomas. Every detailed report carries important information about the diagnosis and treatment of such neoplasms.

***Keywords:** Hemangioblastoma, parafalcine, von Hippel Lindau, sporadic, supratentorial.*

Introduction

Hemangioblastomas are slow-growing uncommon tumors of the central nervous system (CNS), and they are first described by von Hippel in 1895, classified as Grade I according to WHO classification (1). They are benign tumors of vascular origin consisting of 1.1-2.4% of the intracranial lesions composed of vessels and neoplastic stromal cells. Hemangioblastomas are typically located in the posterior fossa and spinal cord. In 30-35% of hemangioblastoma cases, they are associated with von-Hippel-Lindau (VHL) disease. Until 2022, 168 supratentorial cases have been reported in the literature (2). The available literature mainly consists of case reports and small case series, because of that the epidemiological data is also limited. We believe all information about such tumors is vital, which is why we are conducting this case report.

Clinical Case

A 37-year-old woman was admitted to our hospital with a three-month history of headache and mild amnesia. Her physical and neurological examination was normal. Her medical and family history was unremarkable about any inherited disease. Magnetic resonance imaging (MRI) revealed a 1 cm x1.5 cm x1 cm significantly enhanced mass adjacent to cerebral falx, while it was isointense on T1- and hyper-isointense on T2- weighted non-contrast and fluid-attenuated inversion recovery (FLAIR) images (Figure 1.A-D). There was massive vasogenic edema present around the lesion, suggesting a metastasis or a lymphoma. MRI spectroscopy findings confirmed the neoplasm but could not differentiate glioma, lymphoma, or metastasis.

A well-circumscribed solid lesion was excised totally under a surgical microscope via a left frontal interhemispheric approach. During surgery, the borders of the purple-colored lesion adjacent to the cerebral falx were distinguished from the cerebral edematous parenchyma. Histopathological examination of the surgical specimen showed the characteristic findings of a capillary hemangioblastoma with expansion into the glial tissue. The neighboring edematous cerebral tissue was reported as pyloid gliosis. Immunohistochemical results showed positive staining for the stromal cell markers S100, vimentin, neuron-specific enolase (NSE), and glial fibrillary acidic protein (GFAP) but negative staining for inhibin.

Epithelial membrane antigen was negative for the tumor cells. Neighbouring arachnoid cap cells were positive for the cytoplasmic membrane. Reticuline was positive for the sample (Figure 2). The Ki-67 labeling index was <2%. The genetic study of the patient was negative for VHL mutations. The postoperative course was uneventful. Follow-up MR imaging of the patient three years after surgical removal did not show any signs of recurrence and metastasis (Figure 1.E)

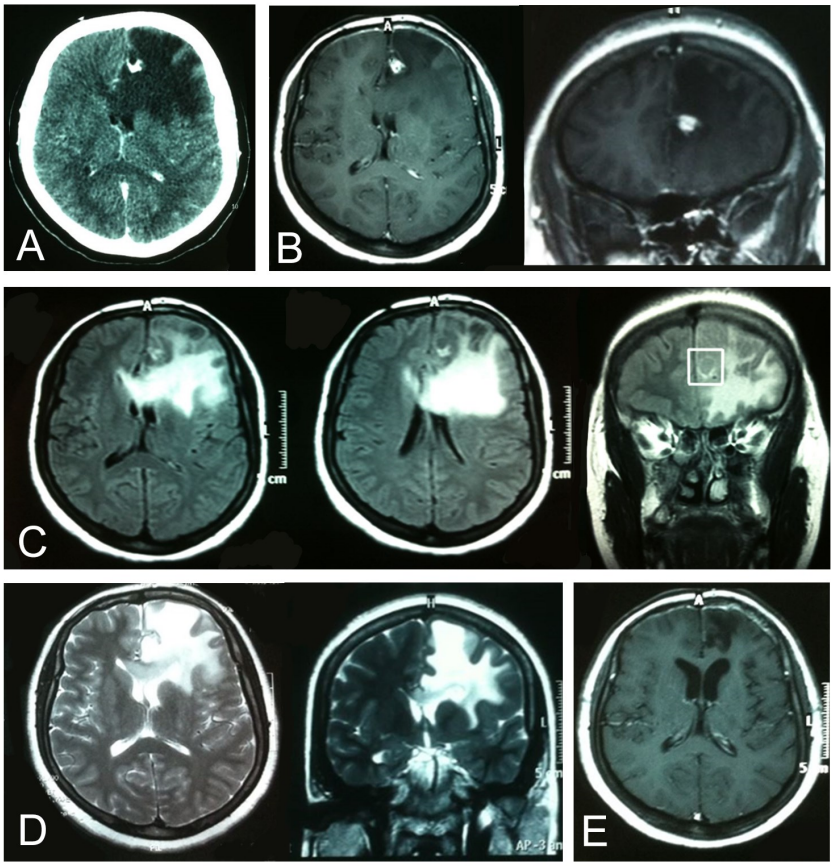

Figure 1: *A: Preoperative CT scan showing large edema on left frontal lobe and a contrast enhanced mass lesion adherent to falx cerebri. B: Preoperative contrast enhanced T1 MRI showing solid component. C and D: Peritumoral "finger-like" edema on FLAIR and T2 MRI. E: Postoperative 3 years follow-up contrast-enhanced MRI with no recurrence.*

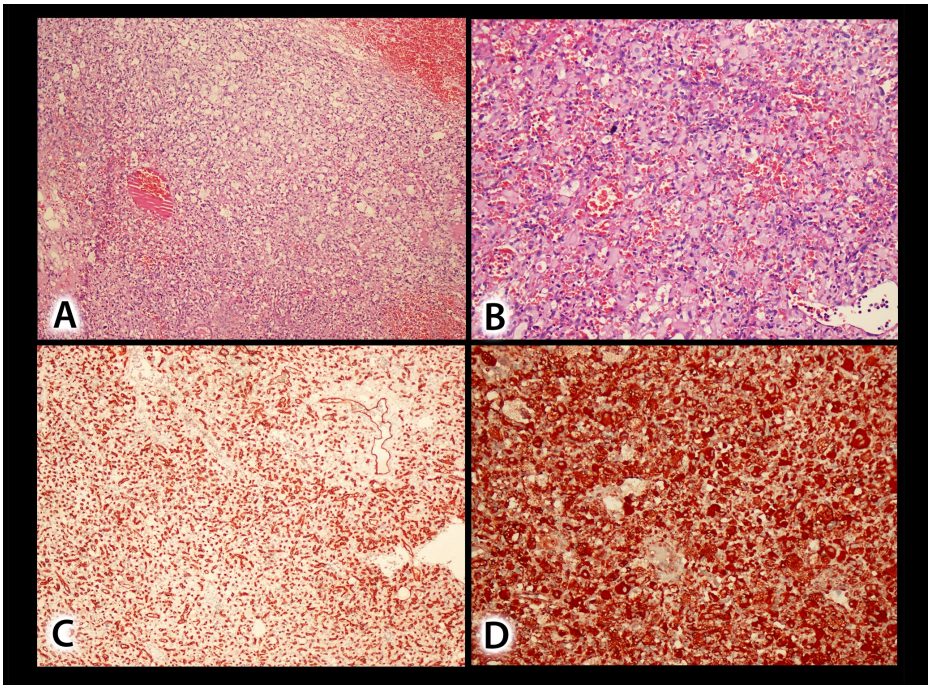

Figure 2: *A and B: (Haematoxylin eosin x100 and x200) Thin-walled blood vessels and foamy cytoplasm cells. C: Immunoreactivity on vessel wall with CD 34. D: Immunoreactivity in stromal cells with vimentin.*

Discussion

Hemangioblastomas are benign neoplasms of CNS consisting of 1.5-2.5% of all intracranial neoplasms (3). The highly vascular well-circumscribed solid or cystic neoplasms of the CNS (or retina) are the most common primary intra-axial tumors of the posterior fossa in adults. They are primarily infratentorial tumors and, they mostly occur in the cerebellum, brainstem, and spinal cord. They rarely occur supratentorial (4-6).

Supratentorial variation of hemangioblastomas is a rare type, first described by Bielchowsky in 1902 (7). They mainly occur in the frontal lobe, followed by the parietal and temporal lobe. They also rarely invade neighboring structures such as meninges, bones, or veins. In the literature total of 168 cases were reported. So far only 33 patients had been reported with dural connection, and 22 were sporadic. Twenty-two of the 33 patients were on the frontal lobe, and only 4 of them had falx connection (Table 1) (2). In addition, half of the frontal tumors were sporadic, like the reported case. The median age of the study was 50 years, and our patient was in the fourth decade at the time of the diagnosis.

With a benign neoplasm, the clinical onset can be slow and asymptomatic (3). In most cases, the clinical characteristics are specific to the location and growth patterns of the lesion. It might appear with a rapid onset of neurological symptoms which requires neurosurgical surgery in rare circumstances. The presented patient had a mild headache and amnesia which started three months ago.

Hemangioblastomas can occur as part of VHL disease, with an inherited VHL gene mutation on the 3p25-26 chromosome, or as a spontaneous tumor, with a somatic VHL gene mutation. In each case, the activation of the VHL-hypoxia-inducible factor-1 (HIF-1) pathway is thought to play an active role (8). In our case, the gene study showed no mutations in the VHL gene. The patient's blood count was also normal.

Histologically, hemangioblastomas are formed of stromal cells and arteries. Small capillaries with a single layer of plump, homogeneous endothelial cells make up the vascular component, whereas big and vacuolated stromal cells constitute the cellular component. It has been subclassified into two forms based on the quantity of the stromal cell component: The more frequent reticular subtype displaying numerous capillaries and stromal cells with positive glial fibrillary acidic protein-immunoreactive, which may lead to an erroneous gliotic diagnosis, and the rarer cellular hemangioblastomas distinguished by zellballen-like cellular clusters of uniform tumor cells (9). Metastatic renal cell carcinoma and angiomatous meningioma are included in the histological differential diagnosis. In our case, the lesion was from the more common reticular subtype.

The use of proton MR spectroscopy in the diagnosis of the lesion is well documented as it reveals a high mobile lipid peak without a lactate peak, a low creatine/phosphocreatine peak, a rise in choline-containing compounds, and absence of acetyl aspartate peak (which indicates nonneurogenic origin) in which when combined with the absence of necrotic component. The MR spectroscopy study before the operation showed no NAA peak, and the characteristic 1.3 ppm lipid peak, however, radiology reports favoured a metastatic lesion. The non-specific imaging features on T1-2 and FLAIR MRI also can be misleading in this tumor. The dural involvement and cystic/solid components usually suggest meningioma or pilocytic astrocytoma but the finger-like edema suggests a malignancy like metastasis or glioma.

Hemangioblastomas are classified as Grade-1 meningeal neoplasm with unknown origin by the World Health Organization in their classification of CNS malignancies for 2021 (1). Although they are benign, they cause a mass effect that leads to cyst formation and peritumoral edema. There was no cyst formation in the presented case, but there was significant peritumoral edema. It is known that pre-surgical endovascular embolization of the lesion can be attempted to reduce bleeding, but the procedure also has its own complications, such as brain ischemia, subarachnoid hemorrhage, and intramural hemorrhage. Considering the lesion's relatively non-eloquent localization, we have decided not to perform embolization before the operation. We had no bleeding complications.

The complete resection of hemangioblastoma is curative and is associated with minimum morbidity and mortality (2, 4). In our case, we have seen no recurrence yet with MRI follow-ups, and the patients have no neurological deficits. Recurrence can be seen even in sporadic cases, and en-bloc resection should be the main goal in surgery. Total resection may not be possible in cases with meningeal or vascular involvement or misdiagnosed. With subtotal resection, the survival rates are decreasing to %53 in five-year follow-ups (5). Regular follow-up is mandatory in these cases.

Table 1: Supratentorial hemangioblastomas with meningeal involvement described from 1942 to 2022.

Reported Cases	Age	Sex	Location	Area	Aspect	VHL
Zeitlin *et al*. (1942)	54	M	Parasagittal	Frontal	Solid	-
Rivera *et al*. (1966)	16	M	Parasagittal	Parietal	Solid	-
Ischwar *et al*. (1971)	62	F	Falx	Occipital	Solid	+

table continued…..

Böckem (1975)	43	M	Tentorium	Occipital	Solid	NA
Lee *et al*. (1978)	46	M	NA	Frontal	Solid	+
Tomaccini *et al*. (1980)	9	F	Falx	Occipital	Solid	NA
Sharma *et al*. (1995)	72	M	Convexity	Parietal	Solid	-
Choi *et al*. (1998)	26	F	NA	Parietal	Solid	-
Kim *et al*. (2001)	45	M	Convexity	Frontal	Solid	+
Agostinelli *et al*. (2004)	10	F	Convexity	Frontal	Solid	-
Iyigun *et al*. (2004)	61	M	Convexity	Frontal	Solid	-
Varsik *et al*. (2004)	38	M	Parasagittal	Frontal	Solid	NA
Zamzuri *et al.* (2004)	46	M	Falx	Frontal	Solid	+
Cosar *et al*. (2006)	50	M	Parasagittal	Parietal	Solid	-
Jang *et al*. (2007)	68	F	Convexity	Frontal	Solid	-
Murali *et al*. (2007)	57	M	Parasagittal	Frontal	NA	NA
Sherman *et al*. (2007)	52	F	Convexity	Frontal	Solid	+
Takeuchi *et al*. (2008)	58	M	Parasagittal	Frontal	Solid	-
Courcoutsakis *et al*. (2009)	53	M	Sellar/parasellar region	-	Solid	+
Elguezabal *et al*. (2010)	67	F	Falx	Frontal	Solid	-
Lozano-Tangua *et al*. (2010)	74	F	Convexity	Frontal	Solid	-
Kaloostian *et al*. (2012)	49	F	Falx	Frontal	Solid	-
Kim *et al*. (2013)	51	F	Convexity	Frontal	Solid	+
She *et al*. (2013)	60	F	Falx	Frontal	Solid	-
Kim *et al*. (2016)	77	F	Convexity	Frontal	Solid	-
Pandey *et al*. (2016)	40	M	Convexity	Parietal	Solid	-
Rocha *et al*. (2017)	62	F	Convexity	Parietal	Solid	-
Tabibkhooei *et al*. (2018)	29	F	Sellar/parasellar region	-	Solid	-
Vicente *et al*. (2019)	64	M	Parasagittal	Frontal	Solid	-
Bian *et al*. (2019)	70	F	Falx	Parietal	Solid	-
Baran *et al*. (2019)	57	F	Convexity	Parietal	Solid	-
Khelifa *et al.* (2020)	20	M	Falx	Occipital	Solid	-
Sánchez Ortega *et al*. (2020)	43	M	Parasagittal	Parietal	Solid	-
Ovalioglu *et al.* (2022)	37	F	Falx	Frontal	Solid	-

Conclusion

Sporadic supratentorial hemangioblastoma is a rare but curable tumor. Those localized to parafalcine are even rarer and more difficult to diagnose preoperatively. We believe it is vital to report every case with detailed analyses to improve our knowledge of such a rare entity.

Conflict of Interest

The authors declare no conflict of interest.

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