

# Geant Supratentorial Hemangioblastoma, Case Report and Discuss the Radiopathological Correlation

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## Abstract

Hemangioblastomas are sporadic tumors found in the cerebellum or spinal cord. Supratentorial hemangioblastoma are exceedingly rare tumors. We present a case of cerebral hemangioblastoma not associated with von Hippel-Lindau disease. A 35-year-old woman was admitted to a hospital with one month history of headache associated with partial seizures. Magnetic resonance imaging revealed a giant cystic mass occupying much of the right frontal hemisphere, with a hyperdense mural nodule. During surgery, the cyst was evacuated, and the mural nodule was completely excised. The postoperative course was uneventful. The histopathologic findings were considered consistent with hemangioblastoma. The finding of a cyst with a mural tumor nodule in the cerebral hemispheres suggests primarily a benign astrocytoma, but other tumors can present in a similar form. The treatment of choice for supratentorial hemangioblastoma is surgical resection. We report a case of supratentorial hemangioblastoma, review the literature, and discuss the radiopathological correlation and diagnostic difficulties associated with such lesions.

**Keywords:** supratentorial hemangioblastoma; radio pathological correlation.

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## INTRODUCTION

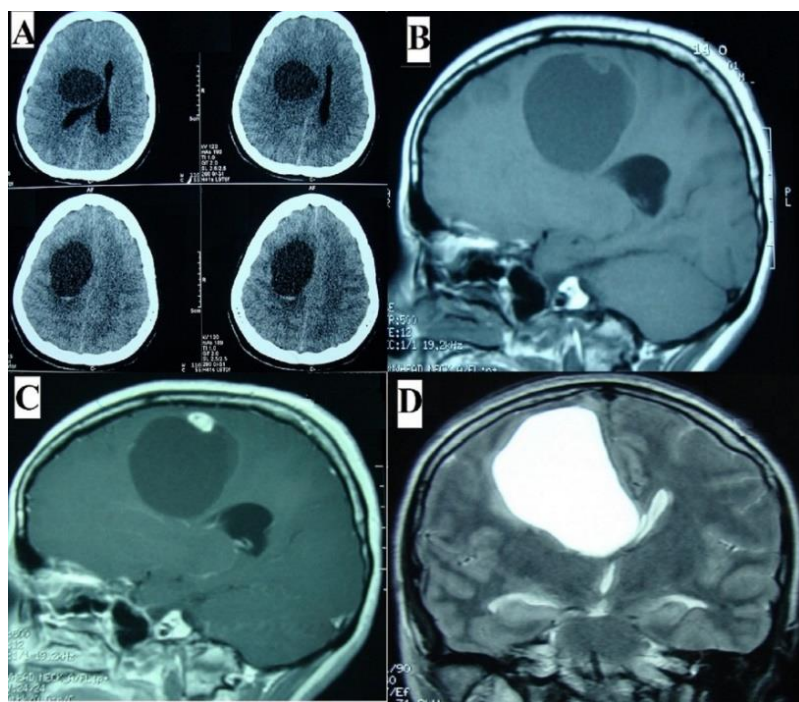
Hemangioblastomas (HBs) are highly vascular benign tumors composed of neoplastic stromal cells with rich capillary networks [1]. Hemangioblastoma of the central nervous system usually occurs in young or middle-aged adults as a cerebellar tumor and represents 1.1% to 2.4 % of all intracranial space-occupying lesions [2]. Supratentorial hemangioblastomas are exceedingly rare tumors and have been found in approximately 131 patients. Here, we present a case of cerebral hemangioblastoma not associated with von Hippel-Lindau disease.

## CASE REPORT

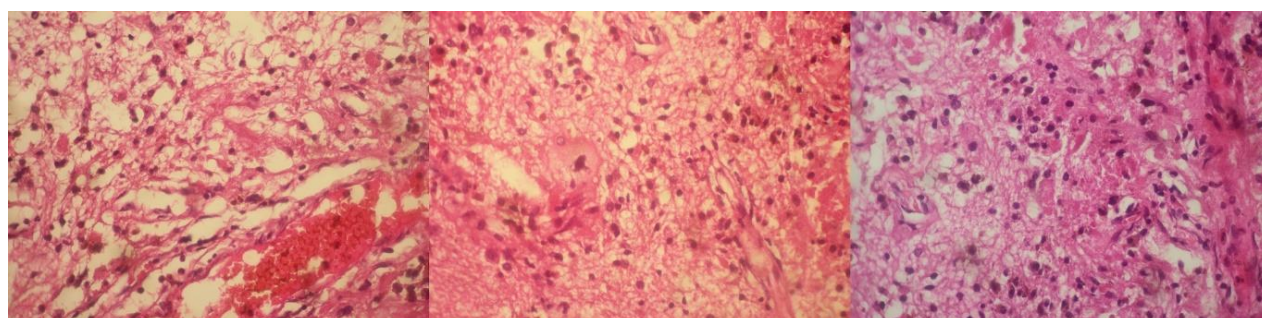
A 35-year-old woman was admitted to a hospital with one month history of headache associated with partial seizures. Neurologic examination on admission was unremarkable. Fundoscopic examination and visual fields were normal. The tomography (CT) scan revealed a cystic right frontal process with small

nodule spontaneously hyperdense enhanced by the contrast. Magnetic resonance imaging revealed a giant cystic mass occupying much of the right frontal hemisphere, with a hyperdense mural nodule (Figure 1).

During surgery, a large cyst with xanthochromic fluid was found. The cystic mass was well circumscribed, and no dural attachment was observed. The cyst was evacuated, and the mural nodule was completely excised. The postoperative course was uneventful. The patient is asymptomatic with down 1 year. Because of the possible association with von Hippel-Lindau disease, a bilateral renal ultrasound examination was performed and the family history was investigated. Ultrasound examination results were normal, and no family history of von Hippel-Lindau disease was apparent. The patient remained in fairly good condition for more than 24 months. The histopathologic findings were considered consistent with hemangioblastoma (Figure 2).



**Figure 1: Brain computerized tomography (CT) scan (A) and Magnetic resonance imaging T1 (B), T1 with contrast enhancement (C), T2 (D): Giant cystic mass occupying much of the right frontal hemisphere, with a hyperdense mural nodule.**



**Figure 2: Histological appearance of the brain tumor. Numerous vacuolated stromal cells are present between several small blood vessels without atypical core (HEx200).**

## DISCUSSION

HBs are benign tumors of vascular origin that develop in the central nervous system. They account for 2% of all intracranial tumors. HBs primarily arise in the cerebellum. Supratentorial occurrence of HBL, it is an extremely rare event. It was first described by Bielschowsky in 1902 [3]. Till date, approximately 131 cases of supratentorial HB have been reported in the literature.

Some hemangioblastomas are associated with von Hippel-Lindau disease. The association between hemangioblastoma of the central nervous system and von Hippel-Lindau disease has been estimated as ranging from 13.5% to 34.3% [2]. Von Hippel-Lindau disease should be considered for patients with more than 1 tumor, for young patients, and for patients with a hemangioblastoma in an unusual location [4]. In the present case, the tumor was located in the right frontal hemisphere. Based on the few cases published, locations are mostly frontal and parasagittal. However, they can

also be found in the parietal lobe as well. The most common reason for admission is headache. Tumor size on presentation varies from 1.5 cm to 7 cm. Hemangioblastomas frequently present as mural nodules projecting into sizable cysts. The tumor tends to be well demarcated from adjacent non neoplastic tissues. The finding of a cyst with a mural tumor nodule in the cerebral hemispheres recommends mainly a benign astrocytoma, but other tumors (such as ependymomas, hemangioblastomas, neuroblastomas, choroid plexus papillomas, and even meningiomas) can present in a similar procedure [5]. Hemangioblastomas (HB) and Pilocytic astrocytomas (PA) can present the same morphological characteristics on conventional MRI sequences, most frequently in the form of a cerebellar cystic mass with a mural nodule that likely improves on post-contrast T1 images [6].

Perfusion MRI showed his interest in the differentiation of these two tumors, The maximum relative cerebral blood volume (rCBV max ), defined as

the ratio between the CBV max in tumor tissue and the CBV in healthy, contralateral white matter, is considered to be indicative of the type of tumor. The HB show a greatly increased rCBV max, as well as disorders of the permeability. The first-pass method of perfusion MRI is a quick and useful way to differentiate between PA and HB [7]. In such cases, some imaging features can suggest a preoperative differential diagnosis, but the correct diagnosis can be achieved only with the addition of histologic findings. The histologic characteristics considered indicative of a hemangioblastoma are the occurrence of great amounts of thin-walled, fairly closely packed blood vessels and dominant polygonal stromal cells [8]. The main histopathologic differential diagnosis of hemangioblastoma contains angioblastic meningioma and metastatic renal cell carcinoma. Yet, the existence of angioblastic meningioma as an entity separate from hemangioblastoma is still controversial [4]. Angioblastic meningioma is generally supratentorial, solid, and attached to the dura, whereas hemangioblastomas are commonly cystic lesions with mural nodules, lack a dural attachment, and are extremely uncommon in supratentorial localization [8]. In this patient, despite the supratentorial localization of the tumor, other features such as the absence of an apparent dural attachment and the characteristic surgical and histologic appearance confirmed the diagnosis of hemangioblastoma.

The treatment of choice for supratentorial HB is surgical resection. Mills *et al.* [9] reported that patients with supratentorial HB undergoing gross total tumor resection (GTR) experienced a significant improvement in progression-free survival (PFS) compared with those receiving subtotal resection (STR) (5-year PFS: GTR, 100% vs. STR, 53%). Total resection of the tumor prevents recurrence and postoperative hemorrhage and radiotherapy may be an alternative or adjuvant therapy for multiple, subtotally resected, and recurrent tumors.

## CONCLUSION

Because of the relative rarity of this tumor, we report a case of cerebral hemangioblastoma, and discuss the radiopathologic correlation and diagnostic difficulties associated with such lesions. Although rare in the supratentorial location, hemangioblastoma must be

considered in the histopathologic differential diagnosis of especially cystic tumors with mural nodule.

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- Conflict of Interest: None declared.

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