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Supratentorial hemangioblastoma without von Hippel-Lindau syndrome in an adult. A rare case report

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Keywords
supratentorial, hemangioblastoma, von Hippel-Lindau syndrome

ABSTRACT

Hemangioblastomas (HBLs) are highly vascular and cystic benign neoplasms. They form very small part of intracranial tumours and are often localized in the posterior fossa. Although most of them are sporadic, a significant group is accompanied with von Hippel-Lindau (VHL) syndrome.

This case report presents a 57-year-old woman treated with total resection using micro-surgical technique and was diagnosed as HBL based on histopathologic findings. Contrast-enhanced cranial MRI of the patient with the complaints of spasms in the right side of body showed a right paracentral mass that caused midline shift.

In literature, the previously reported cases of supratentorial HBL unaccompanied with VHL syndrome were searched in PUBMED, compiled and presented. It should be borne in mind that rare HBLs manifesting with various neurological symptoms may occur in the supratentorial region, and may not accompany with VHL syndrome.

INTRODUCTION

Hemangioblastoma (HBL) is a benign vascular tumour of the central nervous system consisting of veins and neoplastic stromal cells (1). It usually occurs after the third decade (2). It may arise sporadically (66-80%) or along with von Hippel-Lindau (VHL) syndrome (20-33%), a familial neoplasia syndrome. Although HBLs are located in infratentorial site, rare supratentorial cases are present. (3). The most common infratentorial localizations in order of decreasing frequency are cerebellum, brain stem, and spinal cord, and less frequently in the cerebral hemispheres along the optical pathways in supratentorial region (4). Although they have benign histology, HBLs can cause symptoms such as peritumoral edema, cyst formation, and as a result of the mass effect occurring in adjacent structures due to tumour growth (5).

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Our case was a 57-year-old female patient and was worth of reporting due to the presence of HBL manifesting supratentorial localization without an accompanying VHL syndrome. The current literature mostly consists of case reports and series of HBL associated with VHL disease. Due to its rarity, detailed information on the clinical features, treatment, and prognosis of supratentorial HBL is limited.

Case Report

A 57-year-old female patient was admitted to our clinic with complaints of spasms in the left side of her body that began about a month ago, frequent awakenings that lasted for about ten minutes. Neurological examination revealed left 3/5 hemiparesis. Her medical history revealed no known comorbidities and medication use, but a previous surgery for varicose veins. Upon conducting contrast-enhanced cranial MRI, a mass lesion in the right paracentral region with dense contrast enhancement of 6x5.5x5.5cm causing a midline shift (Figures 1 and 2). Patient was operated upon a provisional diagnosis of meningioma, and lesion was observed to be hypervascularized. The lesion was totally resected without any complication utilizing microsurgical technique. Histopathologic examination showed a tumour rich in vascular framework harbouring granular cells in where some nuclei revealing "degenerative atypia. Immuno-histochemical examination, showed certain pathognomonic findings with specific antibodies verifying HBL. The reactivities for EGFR and inhibin were significant, while the GFAP reactivity was dubious. Although it is not a general rule, mast cell tryptase activity used to reveal the presence of mast cells embedded in tumour tissue for supporting the diagnosis. Furthermore, rare nuclear progesterone reactivity is a finding that helps with the diagnosis of HBL (Figure 3). In the post-operative examination of the patient, neurological deficit diminished. In the early post-operative period, contrast-enhanced cranial MRI revealed total removal of the lesion. Additional tests showed no stigmata consistent with VHL syndrome after the discharge of the patient.

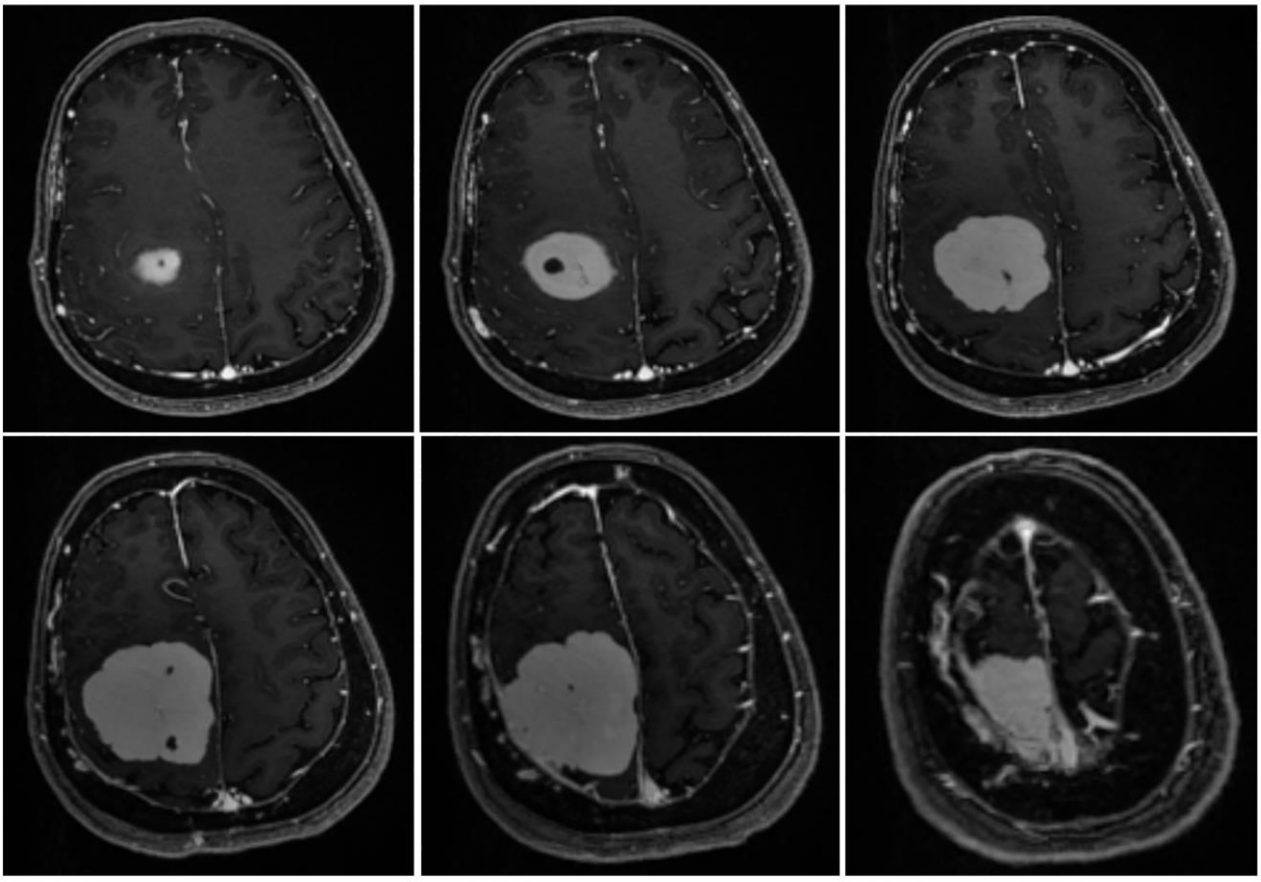

Figure 1. In preoperative axial contrast enhanced cranial MRI sections, a mass lesion with dense contrast enhancement that caused edema in the periphery of paracentral region and a shift in midline is observed.

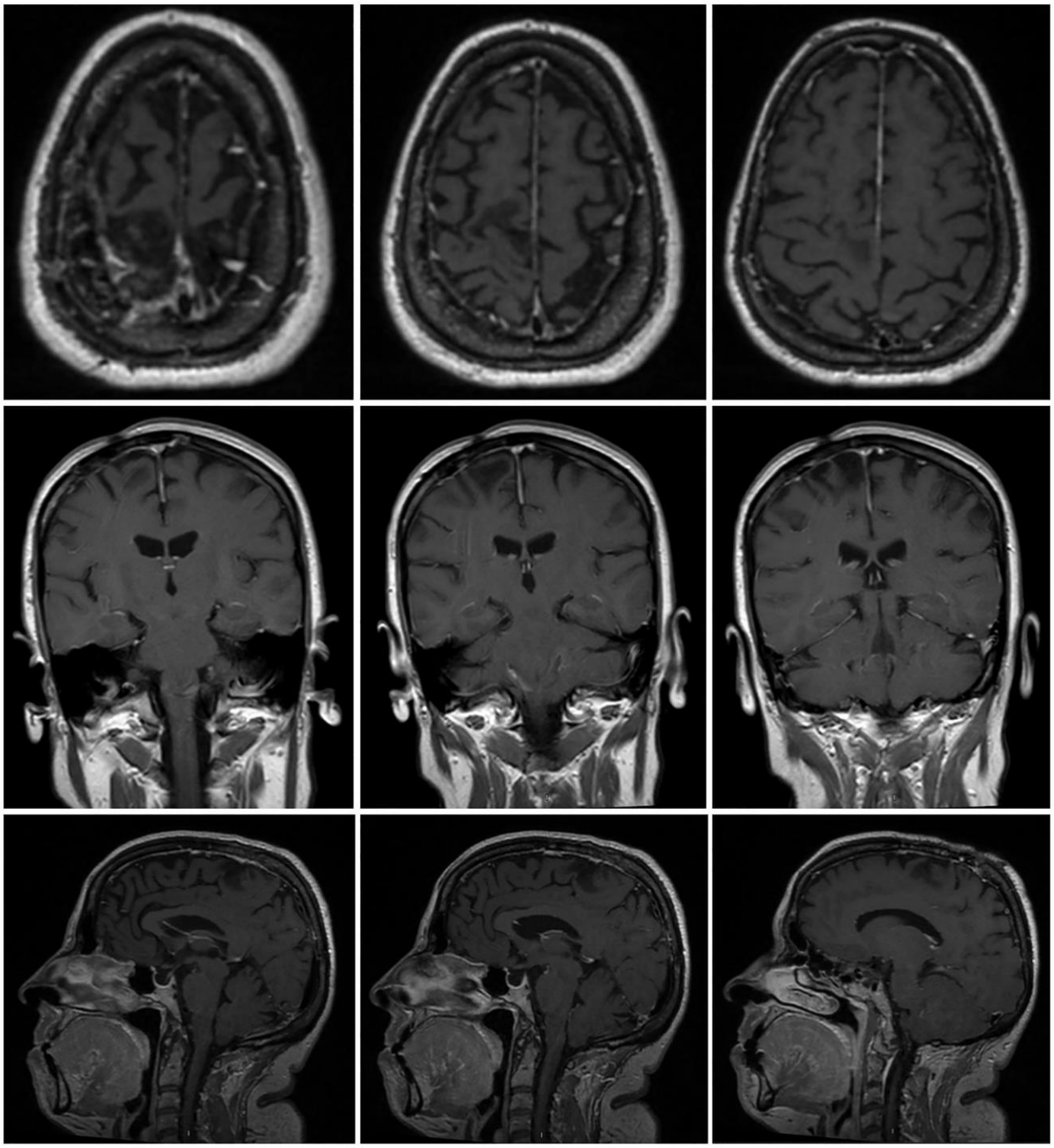

Figure 2. In cranial MRI sections with postoperative contrast, the lesion is observed to have been totally resected.

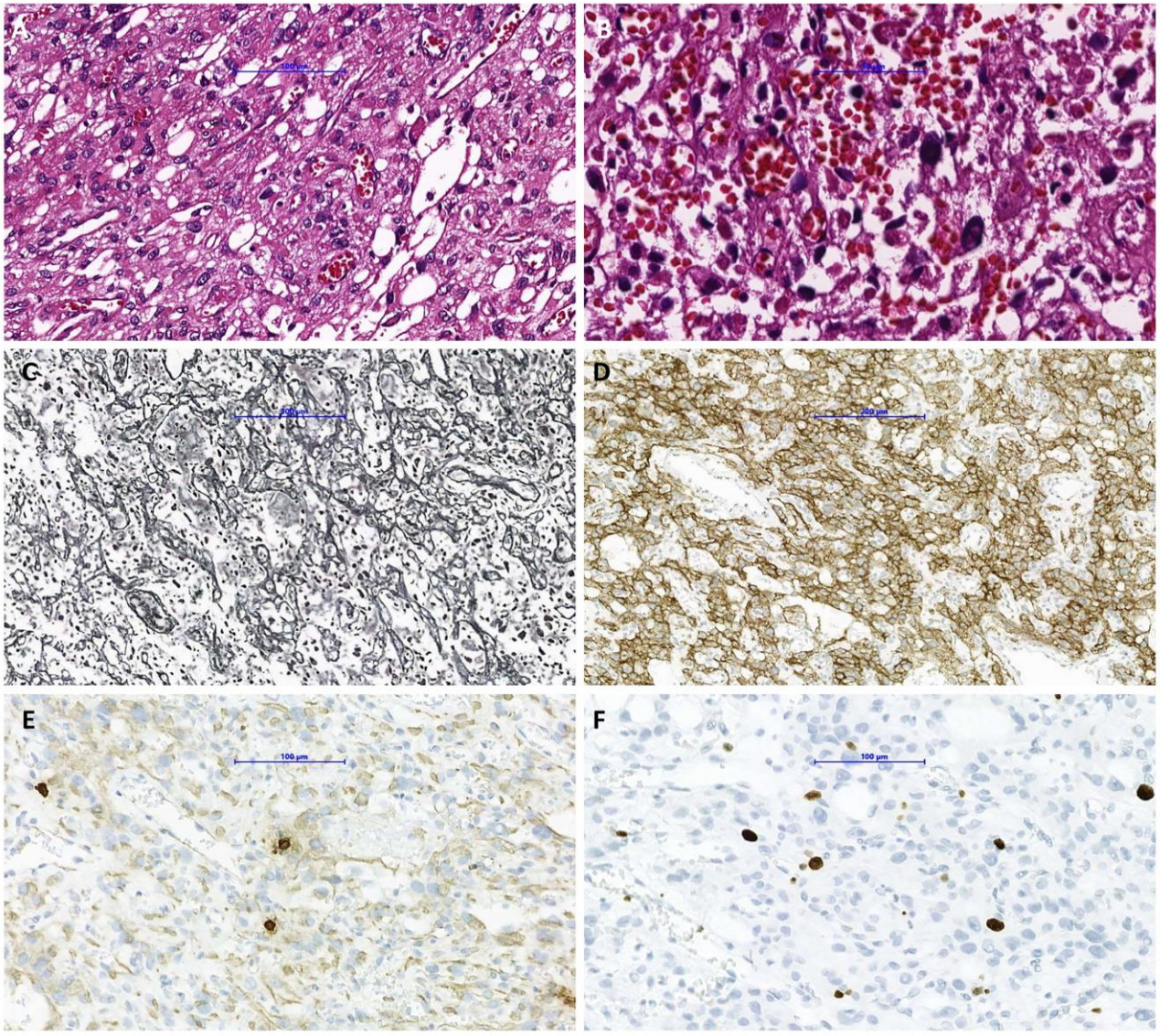

Figure 2. (**A**) Tumour consisting of capillaries surrounded by atypical cells bearing nuclei with rough chromatin pattern and granular cytoplasm typical for hemangioblastoma. (Hematoxylin-eosin, x200).
(**B**) Higher magnification reveals cells with "atypical nuclei" consistent with "degenerative atypia" with no effect on prognosis. (Hematoxylin-eosin, x400).
(**C**) A dense reticulin framework surrounding tumour cells as clusters typical for hemangioblastoma, reticular variant. (x100, reticulin stain).
(**D**) An intense cytoplasmic EGFR reactivity. (EGFR, streptavidin biotinylated complement, x100).
(**E**). Eye-catching mast cells randomly distributed in tumour tissue (Mast cell tryptase [MTC], streptavidin biotinylated complement, x100).
(**F**). Ki-67/MIB-1 labeling index is low (4%). (Ki-67/MIB-1, streptavidin biotinylated complement, x200).

Discussion

HBLs of the central nervous system are rare, benign and vascularized neoplasms of unknown origin (6). Most of the HBLs that are generally macroscopically well limited, are cystic in nature, however, can be 20% solid. For this reason, they mostly show cystic mural nodule adjacent to tumour tissue (7). HBL is generally considered as a non-metastatic tumour, and in many cases, diagnosis is delayed due to the absence of clinical findings (8). They account for approximately 1.5-2.5% of all primary intracranial tumours and 7-10% of primary posterior fossa tumours (9). This is observed in both sexes at approximately equal ratios and most often at the age

of 35-45 years. They are most commonly observed in the cerebellum, less frequently in the brain stem and in the spinal cord, and rarely in the supratentorial region (2,4).

The uncommon supratentorial HBL was first defined by Bielschowsky in 1902 (10). The frontal lobe of the cerebrum is followed by the parietal and temporal lobes according to the incidence (8). The clinical presentation of supratentorial HBLs is anatomically dependent on the site and growth pattern. In some cases, the endothelia of the vascular component of the tumour secrete erythropoietin-like substance, and correspondingly, such paraneoplastic syndromes as polycythemia may be observed (11). In general, supratentorial HBLs have long-term minor symptoms or symptoms may not appear at all. In most cases, sudden exacerbations requiring urgent surgical intervention may occur (12).

It is reported that 5-31% of cerebellar HBL cases, 11% of supratentorial cases, and 80% of spinal cord cases are associated with VHL disease. Rare supratentorial HBLs account for 1-6% of HBLs that accompany VHL disease (6). HBLs are macroscopically well-circumscribed neoplasms with solid and varying sizes containing cystic components. Due to its ability to manifest cystic mural nodules, a differential diagnosis specifically from pilocytic astrocytomas is mandatory (13).

HBL is usually observed frequently in the form of enhancing mural nodule accompanied by cystic component in computed tomography. MRI is the gold standard imaging method in the differential diagnosis. In contrast-enhanced T1-weighted sections, the tumour nodule is prominent and hyperintense in homogeneous form However, in T2-weighted images, the cystic area is displayed as hyperintense. HBL is characteristically of a highly vascular tumour and is located in the avascular cyst, and is directly fed from the vessels originating from dural arteries (14).

Histological features of HBL are distinctive. Vessels of various sizes are important components of tumour tissue and are furnished with a single row of endothelial cells. Stromal cells in the interstitial area contain lipid droplets and glycogen in varying proportions (6). Stromal cells constituting the main component of the tumour show pleomorphism and diverse levels of nuclear hyperchromasia. Mastocytes may be found and mitosis is very rare. In “reticular” variant the stromal cells are dominantly located around the veins whereas wider groups are formed in “cellular” variant (15).

If there is evidence of the progression of the lesion, due to bleeding or mass effect, then surgery is primary choice of treatment. Removal of nodules in cystic lesions during surgical resection is vital (9). For solid lesions, procedures similar to the management of arteriovenous malformation should be performed. Surgical treatment is considered certainly curative (16).

In the relevant literature, there are infrequent supratentorial HBL cases those of which are unaccompanied VHL. Concisely, Table 1 shows a compilation of HBL cases reported in the medical literature.

Author (year)	Age	Sex	Supratentorial location	Gross
Bielschowsky (1902) (10)	24 y	F	Frontal	Solid
Berger and Guleke (1927) (17)	24 y	M	Parietal	Cystic
Schley (1927) (18)	48 y	F	Occipital	Cystic
Marrioti (1936) (19)	N/A	N/A	Posterior part of the corpus callosum	Solid
Zeitlin (1942) (20)	54 y	M	Meningeal parasaggital	Solid
Kautzky and Vierdt (1953) (21)	55 y	M	Right cerebrum-occupied thalamus, globus pallidus, basal surface of brain	Solid
Floris et al. (1954) (22)	32 y	M	Frontal	Solid
Grattarola (1955) (23)	18 y	M	Temporal	Cystic
Morello and Bianchi (1958) (24)	10 y	M	Temporal	Solid
Stein et al. (1960) (25)	49 y	M	Temporal	Solid
Stein et al. (1960) (25)	12 y	F	Frontal	Cystic
Morello and Bianchi (1960) (24)	27 y	M	Parieto-occipital	Solid
Papo et al. (1961) (26)	N/A	N/A	Frontal	N/A

Morello and Bianchi (1960) (24)	27 y	M	Parieto-occipital	Solid
Rivera and Chason (1966) (27)	16 y	M	Meningeal parietal	Solid
Ishwar et al. (1971) (28)	62 y	F	Meningeal falx, occipital	Solid
Perks et al. (1976) (29)	21 y	F	Frontal	Highly vascular
Grisoli et al. (1984) (30)	28 y	F	Pituitary stalk	N/A
Katayama et al. (1987) (31)	N/A	N/A	Third ventricle	N/A
Neuman et al. (1989) (32)	35 y	F	Pituitary stalk	N/A
Black et al. (1991) (33)	15 y	M	Third ventricle	Solid
Sharma et al. (1995) (34)	72 y	M	Meningeal Parietal	Solid
Kachhara et al. (1998) (35)	57 y	F	Sella sphenoid sinus	N/A
Choi et al. (1998) (36)	26 y	F	Meningeal parietal	Solid
Isaka et al. (1999) (5)	47 y	F	Third ventricle	Solid
Tarantino et al. (2000) (37)	N/A	F	Cerebral	N/A
Yamakawa et al. (2000) (38)	17 y	M	Parietal	Cystic
Kim et al. (2001) (39)	45 y	M	Meningeal convexity, frontal	Solid
Ikeda et al. (2001) (16)	62 y	M	Suprasellar	N/A
Ozveren et al. (2001) (40)	40 y	F	Right supratentorial lesion near the splenium	Solid-cystic
Acikalin et al. (2003) (4)	43 y	M	Frontal	Cystic
Rumboldt et al. (2003) (14)	60 y	M	Sellar suprasellar	N/A
Agostinelli et al. (2004) (41)	10 y	F	Meningeal convexity, frontal	Solid
Iyigun et al. (2004) (13)	61 y	M	Meningeal convexity, frontal	Solid
Peker et al. (2005) (42)	54 y	M	Suprasellar	N/A
Tekkök and Sav (2006) (43)	18 m	F	Lateral ventricle	Cystic
Cosar et al. (2006) (44)	50 y	M	Meningeal parasaggital, parietal	Solid
Ohata et al. (2006) (11)	27 y	F	Hippocampus	Solid
Murali et al. (2007) (12)	57 y	M	Meningeal parasaggital	Solid
Sherman et al. (2007) (3)	52 y	F	Meningeal convexity, frontal	Solid
Jang (2007) (45)	68 y	F	Meningeal convexity, frontal	Solid
Takeuchi et al. (2008) (8)	58 y	M	Meningeal parasaggital, frontal	Solid
Jaggi et al. (2009) (46)	30 y	M	Third ventricle	Solid
Peyre et al. (2009) (47)	3 m	M	Lateral ventricle	Cystic
Elguezabal et al. (2010) (48)	67 y	F	Meningeal falx frontal	Solid-cystic
Crisi et al. (2010) (49)	N/A	N/A	Hippocampus	N/A
Schär et al. (2011) (50)	80 y	F	Pituitary	N/A
Yang et al. (2011) (2)	19 y	F	Temporal-occipital lobe	Solid-cystic
Kaloostian and Taylor (2012) (7)	49 y	F	Meningeal falx frontal	Solid
Sarkari and Agrawal (2012) (51)	45 y	F	Midline basifrontal	Solid
She et al. (2013) (9)	60 y	F	Cerebral falx	Solid-cystic
She et al. (2013) (9)	24 y	M	Temporal, choroidal fissure	Solid
She et al. (2013) (9)	21 y	M	Frontal	Cystic
Kishore et al. (2013) (15)	50 y	M	Parietal	Solid-cystic
Al-Najar et al. (2013) (52)	N/A	N/A	Lateral ventricle	N/A
Xie et al. (2013) (53)	64 y	F	Suprasellar	Solid

Raghava et al. (2014) (1)	50 y	M	Frontal	Solid
Pandey et al. (2016) (6)	39 y	M	Parietal	Cystic
Baran et al. (2019)	57 y	F	Right paracentral region	Solid

Table 1. Reported cases of supratentorial hemangioblastoma without VHL.

Conclusions

Infratentorial hemangioblastoma cases are inherently observed at high rates along with von Hippel-Lindau disease. However, it should be borne in mind that a small percentage of supratentorial hemangioblastoma cases may be presented with various neurological symptoms due to mass in imaging studies. Hemangioblastoma should be considered as an entity among other possible tumorous masses occupying supratentorial area.

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