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Staged surgical treatment of a hypervascular cerebellar hemangioblastoma and saccular superior cerebellar artery aneurysm using preoperative embolization with a low viscosity non-adhesive liquid embolic agent

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ABSTRACT

This report presents a patient with a cerebellar hemangioblastoma associated with obstructive hydrocephalus and a saccular superior cerebellar artery (SCA) aneurysm who was successfully treated using a three-stage approach that included ventriculoperitoneal shunting, preoperative endovascular embolization of the tumor vasculature and aneurysm with a low viscosity non-adhesive liquid embolic agent, and total microsurgical tumor removal. Removal of these tumors is associated with a high risk of profuse intraoperative blood loss, cerebral edema, and other equally dangerous complications. Our case was complicated by a tumor feeder aneurysm, which increased the risk of spontaneous subarachnoid hemorrhage and intraoperative aneurysm rupture. A three-stage approach allowed radical and safe removal and resulted in improved neurological symptoms.

1. Introduction

Hemangioblastomas (HABs) are rare benign central nervous system tumors. They most frequently develop below the tentorium and affect the cerebellum, brain stem, and spinal cord [1]. HABs account for 1.5% to 2.5% of all intracranial tumors and 7%–8% of posterior cranial fossa mass lesions. Up to 76% of PCF HABs are located in the cerebellar hemispheres, which makes them the most common cerebellar tumor in adults [2,3]. Most HABs develop sporadically; however, approximately 33% are associated with von Hippel–Lindau (VHL) syndrome, a genetic disorder associated with tumor formation in multiple organs [2]. Sporadic HABs often develop in the fifth decade of life and mainly occur in men [2,4,5]. In addition to the cerebellar hemispheres, HABs can develop in the brain stem [6], craniovertebral junction, cerebellopontine angle [2], and spinal cord [7]. According to the 2016 World Health Organization (WHO) central nervous system tumors classification, HABs are considered a mesenchymal, non-meningeal tumor (9161/1) [12].

Although a few reports have described treatment of HABs with embolization followed by surgery [8,11], none have described three-stage treatment with combined ventriculoperitoneal shunting, embolization of tumor feeding vessels and associated saccular aneurysm, and microsurgical tumor removal, which is presented here. Our case is relevant considering the lack of an established approach to complex PCF HABs as well as reliable prognostic data to guide surgical decision making.

2. Case report

A 57-year-old male presented with a 7-month history of gradually worsening symptoms of increased intracranial pressure (headache, general weakness, dizziness, unsteady gait, nausea, and vomiting). A series of neuroimaging examinations, including computed tomography (CT), contrast-enhanced magnetic resonance imaging (MRI), and computed tomography angiography (CTA), showed a hypervascular right cerebellar hemisphere mass and obstructive hydrocephalus caused

Abbreviations: HAB, hemangioblastoma; PCF, posterior cranial fossa; VHL, von Hippel–Lindau; WHO, World Health Organization; MRI, magnetic resonance imaging; CT, computed tomography; CTA, computed tomography angiography; PHIL, precipitating hydrophobic injectable liquid; LV, low viscosity.

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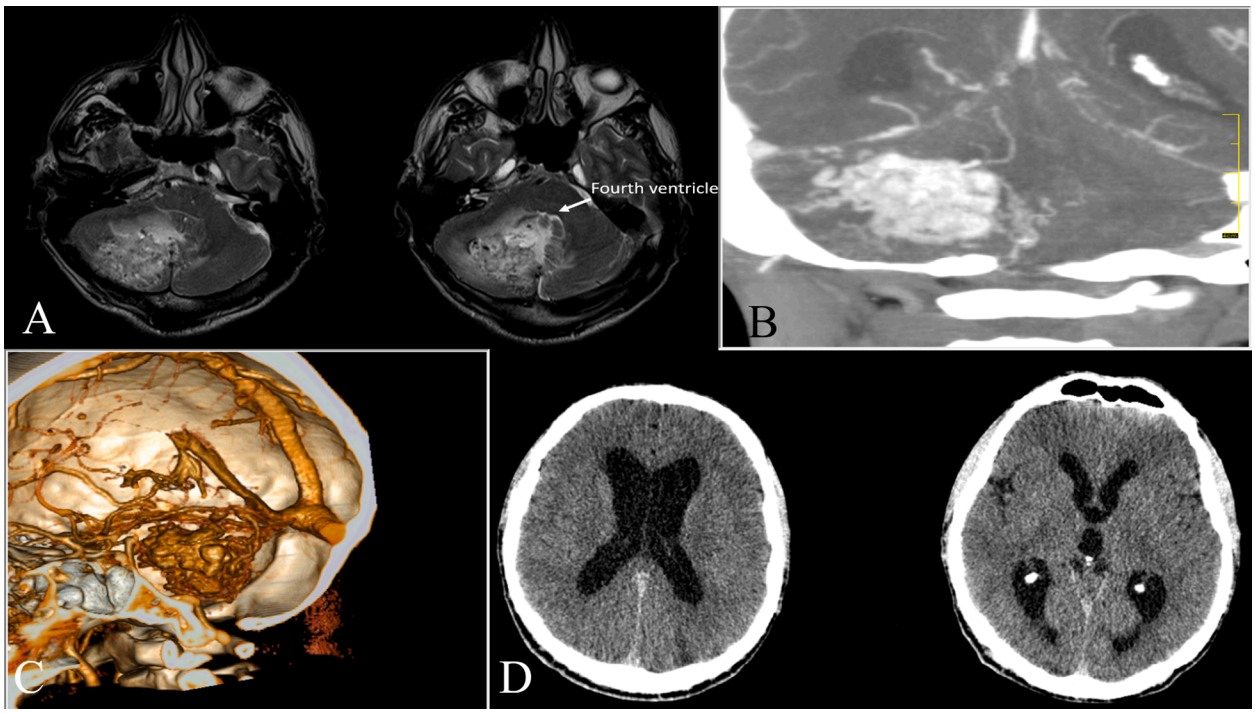

Fig. 1. A, Preoperative axial T2-weighted imaging with gadolinium; B, preoperative coronal computed tomography angiography (CTA); C, three-dimensional CTA reconstruction. D, preoperative axial CT. A hypervascular right cerebellar hemisphere mass and fourth ventricle compression with obstructive hydrocephalus are shown.

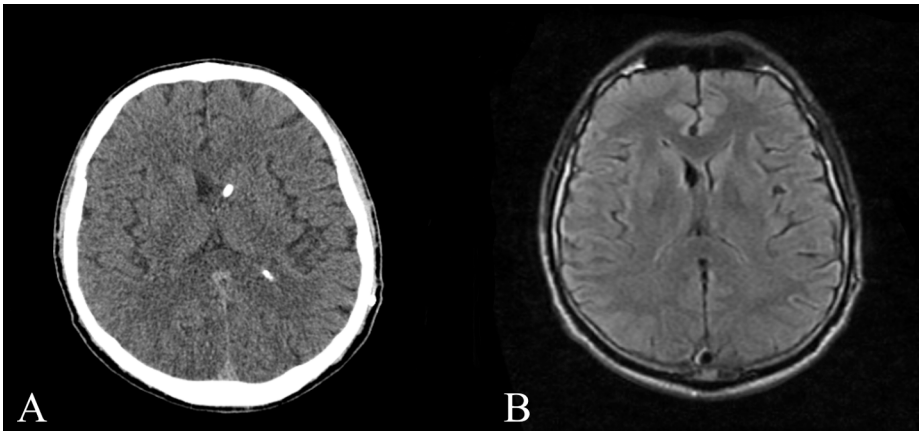

Fig. 2. A, Axial computed tomography after shunt placement; B, postoperative axial T1-weighted imaging. Intraventricular catheter location in the anterior horn of the left lateral ventricle and resolution of hydrocephalus are shown.

by occlusion of the fourth ventricle (Fig. 1).

After analyzing the clinical data, neuroimaging findings, and literature regarding treatment of hypervascular HABs with surgical removal, endovascular embolization, radiation therapy, or various combinations, we formulated the following staged treatment approach:

Stage I. Ventriculoperitoneal shunting using a Medtronic (Dublin, Ireland) medium-pressure CSF shunt system (CFS)
Stage II. Preoperative endovascular embolization of tumor feeding vessels using precipitating hydrophobic injectable liquid (PHIL) 25% low viscosity (LV) embolic agent
Stage III. Radical microsurgical tumor removal

The shunt was placed first to treat the patient's obstructive hydrocephalus, prevent hydrocephalus worsening that could result from post-embolization venous outflow impairment and cerebellar edema, and create comfortable operating conditions during microsurgical tumor removal. His general and neurological state had improved by the day after stage I surgery. Follow-up CT showed signs of satisfactory shunt functioning and resolution of hydrocephalus (Fig. 2).

Stage II (preoperative embolization) was performed using PHIL 25% LV, an extra low viscosity non-adhesive liquid embolic agent. Compared to standard liquid embolic agents (Onyx 18, Squid 18, PHIL 25%), it has greater penetrability into the distal vascular bed and the least reflux [9,10], which were regarded as advantageous for embolization in our patient. Sonic 1.2F detachable tip microcatheters were used to deliver the embolic agent because of their low profile and ability to access distal arterial branches. After injection of PHIL 25% LV, the arterial vasculature of the entire lesion was filled deeply without significant reflux. Although use of PHIL 25% LV for arteriovenous malformations embolization often requires proximal penetration depth control, in our patient, deep penetration was considered advantageous. Minimum risk of

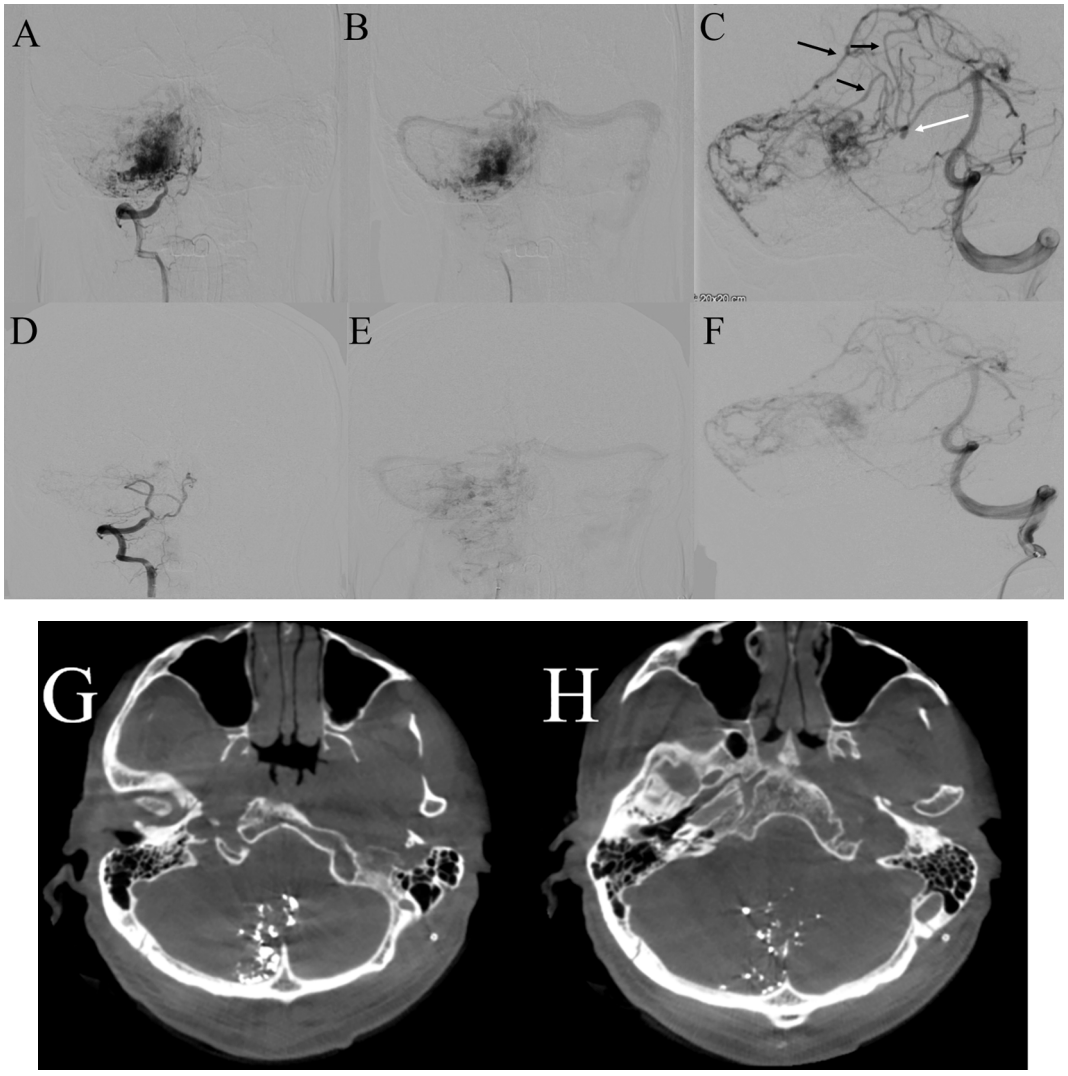

Fig. 3. A–C, Pre-embolization angiography; D–F, postembolization angiography; G and H, postembolization axial flat-detector computed tomography. Pre-embolization angiography shows a hypervascular right cerebellar hemisphere mass with feeders from the right posterior inferior cerebellar artery (PICA) in the arterial phase (A) and venous phase (B). The V4 segment of the right vertebral artery is aplastic. Feeders from the right superior cerebellar artery (SCA) include secondarily recruited cortical branches (short black arrows) and one with a small saccular aneurysm (long white arrow) (C). Postembolization angiography shows subtotal tumor embolization. The main tumor compartment was completely embolized from the right PICA while preserving the artery itself and its branches (D). The entire PICA tumor vasculature was filled with embolic agent (E). The smaller tumor compartment was partially embolized from the right SCA, including a feeder with a small aneurysm and the aneurysm itself. Postembolization computed tomography shows multiple hyperdense foci representing the embolic agent with no evidence of hemorrhage (G, H).

reflux, which significantly reduces the risk of ischemic complications, and maximum depth of penetration into the distal vascular bed are the very properties of PHIL 25% LV that led to its selection.

As the patient had a disconnection of the vertebrobasilar junction (aplasia of the V4 segment of the right vertebral artery [VA] above the origin of the right posterior inferior cerebellar artery [PICA]), the feeders arising from the right PICA were embolized from the right VA and those from the right superior cerebellar artery (SCA) were embolized from the left VA. A saccular aneurysm was detected on a feeder from the right SCA and also embolized using PHIL 25% LV (Fig. 3). A small portion of the tumor vasculature was secondarily recruited via right SCA cortical branches, these were not embolized because of the risk of ischemic complications.

Follow-up angiography showed subtotal embolization of the lesion vasculature. Intraoperative flat-detector CT showed multiple hyperdense foci (PHIL 25% LV) within the vessels of the lesion and no acute intracerebral hemorrhage (Fig. 3).

Stage III was radical microsurgical removal of a giant (5.5 cm diameter) hypervascular right cerebellar hemisphere mass. A bilateral suboccipital craniotomy with resection of the occipital ring and C1 lamina was performed. The dura mater was tense and nonpulsatile. After infusion of 200 mL of 15% mannitol, the brain relaxed and pulsations were visualized. The dura mater was opened in a Y-shaped manner with the base toward the torcula. The tumor node was then dissected, first inferolaterally, then medially from the cerebellar vermis. It was bright red, hemorrhagic, and tense. Arterial feeders and veins were isolated and coagulated as the dissection proceeded. The tumor drained into the tentorial sinus, vein of Galen, and dilated cerebellar tentorium lacunae. After the tumor was isolated, it was removed *en bloc*. To decompress the craniovertebral junction, the dura mater was additionally opened over the cisterna magna inferiorly to the C2 level. Tumor removal was greatly facilitated by the stage II embolization. Intraoperatively, the embolic agent appeared as whitish rubbery densities within the vasculature (Fig. 4).

There were no surgical complications. Postoperative head CT showed no evidence of hemorrhage (Fig. 5). The patient was discharged

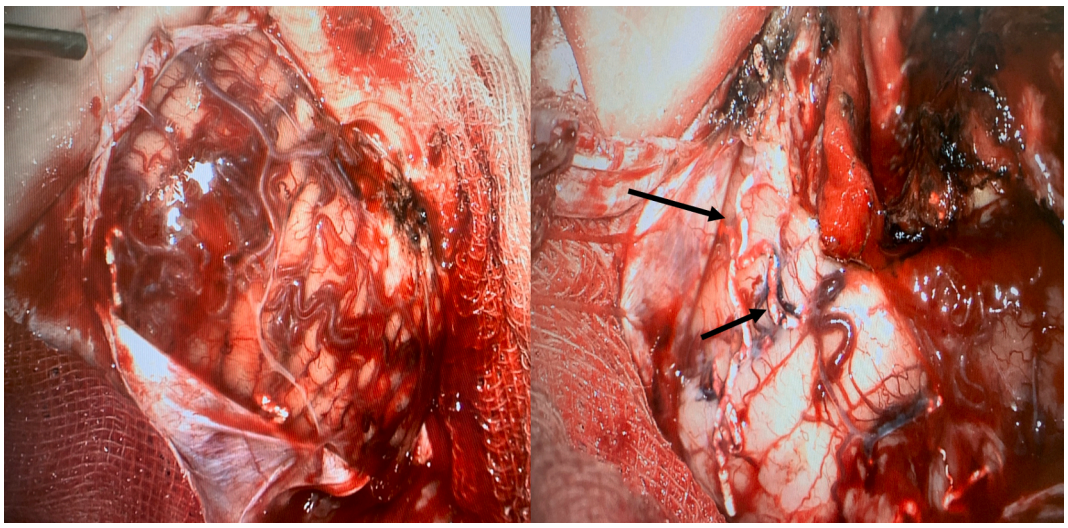

Fig. 4. Intraoperative photography. After opening of the dura mater, the cerebellar hemisphere was tense. The tumor was bright red and clearly delineated from brain tissue. Feeding vessels filled with embolic agent (black arrows) are visualized in the cerebellar cortex. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

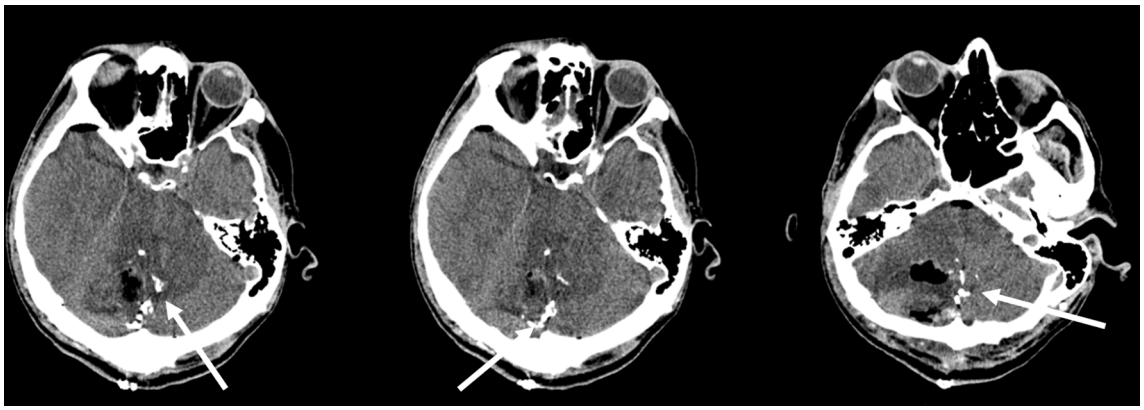

Fig. 5. Postoperative axial computed tomography. The tumor cavity contained air without hemorrhage. High-density embolic agent is visualized in the vessels surrounding the tumor cavity (white arrows).

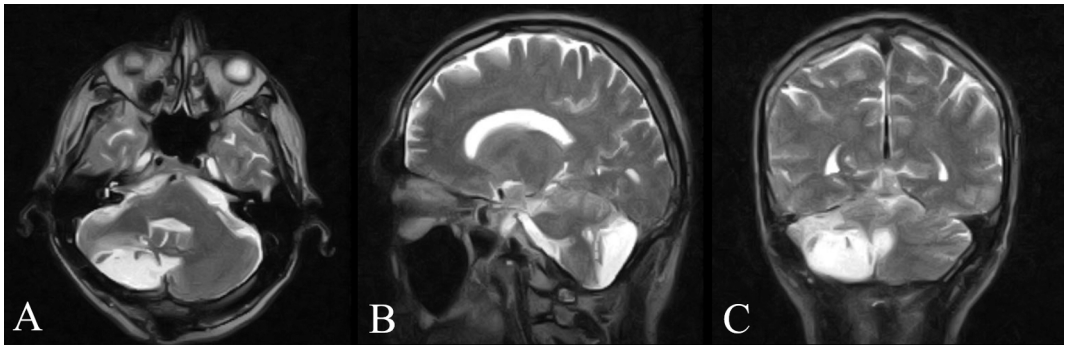

Fig. 6. Postoperative axial (A), sagittal (B), and coronal (C) T2-weighted imaging shows no evidence of tumor recurrence.

in satisfactory condition with improved neurological symptoms (modified Rankin Scale score 1). Follow-up brain MRI 6 months after surgery showed no evidence of tumor recurrence (Fig. 6).

2.1. Histopathological evaluation

On histopathological examination, the tumor was predominantly solid and consisted of densely spaced, thinly walled capillaries of various size with cells containing transparent, foamy cytoplasm between the vessels. No mitotic figures or necrosis were visualized. These parameters were consistent with a diagnosis of HAB.

Table 1
Previously reported cases of aneurysm associated with hemangioblastoma [16].

Authors and year	Age (years)/ gender	Hemorrhage	Feeding vessel	Aneurysm location	Aneurysm classification	Tumor location	Clinical onset
Yoshii *et al.*, 1976 [18].	50F	SAH before tumor removal	Rt PICA	BA bifurcation Lt ICA bifurcation	Unrelated	Rt cerebellum	Tumoral, SAH
Ueno *et al.*, 1977 [19].	50F	No	Lt PICA	Lt ICA	Unrelated	Lt cerebellum	Tumoral
Guzman *et al.*, 1999 [20].	53M	No	Lt PICA Lt AICA	Lt distal AICA	Distal flow -related	Lt cerebellum	Tumoral
Menovsky *et al.*, 2002 [21].	52F	SAH before tumor removal	Lt AICA	Lt BA–AICA	Proximal flow -related	Lt cerebellum	Tumoral, SAH
Zager *et al.*, 2002 [17].	53M	No	Rt AICA	Rt distal AICA	Distal flow -related	Rt cerebellum	Tumoral
Murai *et al.*, 2006 [22].	72M	No	Rt SCA	Rt ICA–PPTA	Unrelated	Vermis	Tumoral
Suzuki *et al.*, 2014 [16].	37F	SAH before tumor removal	Lt PICA	Lt distal PICA	Distal flow -related	Vermis	IVH, SAH
Our case	57M	No	Rt PICA Rt SCA Rt AICA	Rt distal SCA	Distal flow -related	Rt cerebellum	Tumoral

Unrelated, aneurysm on artery unrelated to the hemangioblastoma feeder; proximal flow-related, aneurysm on the proximal portion of a major artery located on the ICA, circle of Willis, or the vertebrobasilar arteries feeding the hemangioblastoma; distal flow-related, aneurysm on the distal portion of the artery feeding the hemangioblastoma; AICA, anterior inferior cerebellar artery; BA, basilar artery; ICA, internal carotid artery; Lt, left; PICA, posterior inferior cerebellar artery; PPTA, persistent primitive trigeminal artery; Rt, right; SAH, subarachnoid hemorrhage; F, female; M, male
Suzuki M, Umeoka K, Kominami S, Morita A. Successful treatment of a ruptured flow-related aneurysm in a patient with hemangioblastoma: Case report and review of literature. Surg Neurol Int 26-Sep-2014;5: https://doi.org//10.4103/2152-7806.141887.

Table 2
Case descriptions of preoperative hemangioblastoma embolization [11].

Age & sex	Presentation	Aborted previous surgery (blood loss)	Material	Tumor vascularity after EVE	Timing of surgery	Extent of excision	Blood loss (mL)	Outcome, mRS	Adjuvant treatment
49M	Ataxia, Dysmeteria	No	PVA	Near total occlusion	15	Total	850	Improved, 1	None
18M	Ataxia, Long tract signs	No	PVA	Incomplete	15	Total	700	Improved, 2	None
48M	Ataxia, Dysmeteria, Long tract signs	Yes (6 L)	PVA	Total	15	Total	640	Improved, 2	None
20M	Ataxia, Long tract signs	Yes (4 L)	NBCA	Near total occlusion	15	Total	590	Improved, 1	None
55M	Ataxia, Dysmeteria	Yes, twice (NA)	Onyx	Total occlusion	7	Total	370	Improved, 4	VP shunt
49F	Ataxia, Dysmeteria	No	Onyx	Near total occlusion	21	Subtotal	200	Improved, 3	VP shunt
32F	Ataxia, Long tract signs	No	Onyx	Total occlusion	4	Total	170	Improved, 2	None
36M	Ataxia, Long tract signs	No	Onyx	Total occlusion	10	Total	340	Improved, 2	None
40F	Ataxia, Dysmeteria	No	Onyx	Total occlusion	21	Total	130	Improved, 0	None
*57M	Ataxia, Dysmeteria	No	PHIL 25% low viscosity	Near total occlusion	2	Total	500	Improved, 1	VP shunt (before embolization)

*our case
EVE, endovascular embolization; mRS, modified Rankin Scale; NA, not available; NBCA, N-butyl cyanoacrylate; PVA, polyvinyl alcohol; PHIL, precipitating hydrophobic injectable liquid; VP, ventriculoperitoneal
Sultan, A., Hassan, T., Aboul-Enein, H., Mansour, O., & Ibrahim, T. (2016). The value of preoperative embolization in large and giant solid cerebellar hemangioblastomas. Interventional Neuroradiology, 22(4), 482–488. https://doi.org//10.1177/1591019916633244 (https://doi.org/10.1177/1591019916633244).

3. Discussion

Although HABs are rare benign brain tumors, they are the most common primary cerebellar tumor in adults [1,2]. HABs develop sporadically or in association with VHL syndrome [2]. Safe and complete removal is challenging due to the relatively small size, vascular anatomy, and cranial nerves of the PCF. Surgical removal is associated with a high risk of significant blood loss because of tumor hypervascularity as well as other various postoperative complications because of the unpredictable nature of the operation. Therefore, our approach in this patient was based on reducing tumor blood flow using preoperative endovascular embolization to ensure the safest possible complete removal (Table 1) [11].

Neurological deficit in HAB patients is caused by tumor mass effect or hemorrhage [15]. Although signs of obstructive hydrocephalus, such as ataxia, dizziness, headache, and nausea are the most common clinical manifestations of PCF HABs [15], they can also present with spontaneous subarachnoid hemorrhage, which typically occurs because of a ruptured saccular aneurysm on a tumor feeder or direct tumor hemorrhage (Table 2) [16–18]. A few cases similar to ours have been previously reported; however, our patient also had a saccular aneurysm on a feeder from the right SCA. We agree with previous authors who have reported that a saccular aneurysm in the tumor or on the feeders significantly increases the risk of massive intraoperative bleeding [16–18].

Currently, HAB treatment options include surgery, endovascular embolization, radiosurgery, and radiation therapy [23]. Radiation therapy was not considered in our patient as it is reserved for recurrent HAB, partially resected tumors, or patients with surgical contraindications [13]. Preoperative endovascular embolization of HABs can significantly reduce intraoperative blood loss and facilitate safe microsurgical removal, the same as for arteriovenous malformations [14].

4. Conclusion

Three-stage surgical treatment of cerebellar hemisphere HAB allows radical and safe tumor removal. After "preparation" with embolization, the tumor can be removed atraumatically while preserving local anatomic structures without significant blood loss or postoperative complications. Preoperative embolization in patients with HAB associated with saccular aneurysm is necessary because of the increased risk of massive intraoperative bleeding. Use of a LV embolic agent significantly reduces the risk of neurological complications and allows complete embolization of the tumor vasculature.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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