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Case Report

Recurrent hemorrhage in hemangioblastoma involving the posterior fossa: Case report

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Abstract

Background: Hemangioblastomas (HGBs) are the most common primary intra-axial posterior fossa tumor in adults. Although spontaneous hemorrhage of these tumors is exceedingly rare, despite their vascular nature, we describe a case of recurrent hemorrhage with associated tonsillar herniation, and demonstrate that a surgical approach can provide a suitable outcome.

Case Description: A 54-year-old female with von Hippel-Lindau (VHL) syndrome presented with acute loss of consciousness and Glasgow Coma Scale (GCS) was 4. Computed tomographic (CT) images demonstrated large volume subarachnoid hemorrhage of the posterior fossa with intraventricular extension and intraparenchymal hemorrhage involving the right cerebellar tonsil. Magnetic resonance imaging (MRI) displayed three lesions in the posterior fossa, two near the hemorrhage site. Patient underwent suboccipital craniectomy with a decent recovery followed by radiosurgery as she refused resection. A second hemorrhage occurred ultimately prompting surgical resection of the three posterior fossa lesions, with a reasonable postoperative course.

Conclusion: Hemorrhage of HGBs of the posterior fossa can present in conjunction of tonsillar herniation. Re-hemorrhage appears to be likely if prior acute hemorrhage has occurred. A stepwise approach of surgical decompression and resection may provide the best outcome.

Key Words: Intracranial hemorrhage, hemangioblastoma, von Hippel-Lindau

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INTRODUCTION

Hemangioblastomas (HGBs) are the most common primary intra-axial posterior fossa tumor in adults.[4] In patients with von Hippel-Lindau (VHL) syndrome, cerebellar HGBs can be found in 44–72% of patients.[13] Complications related to cerebellar HGBs are the leading cause of death in patients with VHL.[28] Spontaneous hemorrhage of cerebellar HGB in VHL is rare and has been infrequently described as a cause of death.[9,10,19,39] Hemorrhages related to HGB have been described to

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occur in the subdural, subarachnoid, intraventricular, or intraparenchymal spaces. Patients with HGBs of the cerebellum often present with a long-standing history of minor neurologic symptoms, such as headache, disequilibrium, and nausea/vomiting that is often followed by a sudden exacerbation (i.e., hydrocephalus).[21] Pathologically, HGBs are composed of a large number of prominent thin-walled capillary vessels and often contain hemosiderin deposits indicating previous intratumoral bleeding.[19] Despite being an intensely vascular tumor, as is often demonstrated angiographically and intraoperatively, massive intratumoral hemorrhage is rare.[19] We present a case of acute cerebellar hemorrhage related to a HGB in a patient with VHL leading to tonsillar herniation and discuss pertinent literature related to this uncommon event.

CASE HISTORY

A 54-year-old female with known VHL presented to the emergency department with acute altered mental status, which was followed by respiratory distress requiring intubation. Her past medical history was significant for sequelae of VHL including breast cysts, retinal detachment causing blindness, multiple pancreatic cysts, a kidney cyst, thoracolumbar resection of a spinal HGB, a right globe prosthesis, left phthisis bulbi, gastrointestinal arteriovenous malformation (AVM), partial nephrectomy for renal cell carcinoma, and hypertension. Her Glasgow Coma Scale (GCS) was 4. Computed tomographic (CT) image [Figure 1] showed a large volume subarachnoid hemorrhage of the posterior fossa with a small amount of intraventricular extension and intraparenchymal hemorrhage involving the right cerebellar tonsil. An emergent ventriculostomy was placed with significant improvement in her neurological function, with the patient following commands the next morning. A magnetic resonance imaging (MRI) was obtained [Figure 2] which showed the presence of three posterior fossa lesions, two of which were in proximity to the site of hemorrhage (fourth ventricular lesion and vermian lesion). The lesions greatest diameter on MRI was 2.9 cm, 1.2 cm, and 1.3 cm. The patient subsequently deteriorated 2 days after ventriculostomy placement and became decorticate, with loss of her gag and cough reflex. Repeat imaging did not reveal a new hemorrhage. The patient was taken emergently to the operating room for a suboccipital craniectomy, evacuation of some subarachnoid clot, and partial resection of the HGB that was visible and duraplasty. Intraoperatively, the margins of the HGB were difficult to separate from the vermis and floor of the fourth ventricle limiting the resection. Postoperatively, the patient slowly recovered her brainstem reflexes and was following commands by postoperative day number 15. The patient required a ventriculoperitoneal shunt, percutaneous endoscopic gastrostomy, and tracheostomy prior to her discharge to a long-term ventilator facility. Patient was discharged home at 2 months and weaned off her gastrostomy and tracheostomy. Discussions were held with the patient to resect the remaining lesions in the posterior fossa, but the patient refused surgery and wanted to proceed with stereotactic radiosurgery. Patient had 24 Gy delivered to three lesions 5 months after her hemorrhage. Patient had an acute headache 1 week later with loss of consciousness requiring intubation. Head CT demonstrated a new hemorrhage. The hemorrhage involved the cerebellum, medulla, and fourth ventricle without significant subarachnoid hemorrhage. The pattern of hemorrhage did not clearly indicate if the same lesion was responsible for the hemorrhage based on MRI [Figure 3]. A cerebral angiogram was performed demonstrating a tumor blush for the two bigger lesions, but embolization was not felt to be necessary [Figure 4]. Patient had surgical resection of three posterior fossa lesions during that admission with return to her baseline neurologic exam postoperatively [Figure 5]. Patient was discharged to a long-term ventilator facility after 3 weeks with a tracheostomy.

DISCUSSION

HGBs are the most common primary intra-axial posterior fossa tumor in adults.[4] They represent 1–2.5% of all

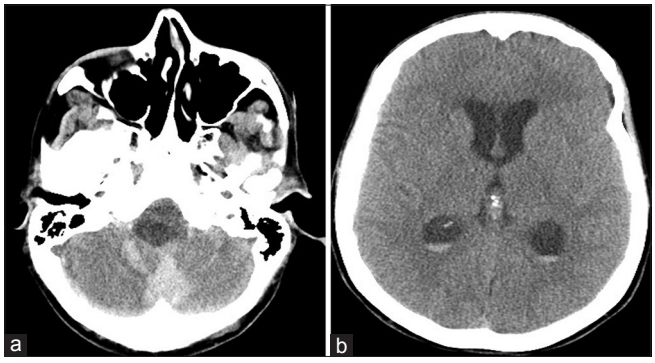

Figure 1: (a and b) Non contrast axial computed tomographic scan suggesting subarachnoid, intraventricular, and intraparenchymal hemorrhage in the posterior fossa

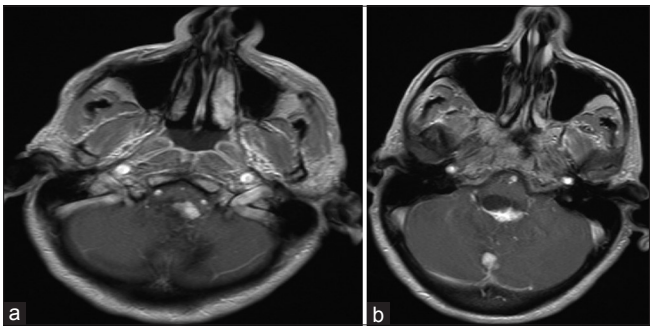

Figure 2: (a) Axial T1 magnetic resonance imaging with contrast showing vermian hemangioblastomas. (b) Axial T1 magnetic resonance imaging with contrast showing 4th ventricular hemangioblastomas and cerebellar hemangioblastomas

intracranial tumors and 7–12% of primary posterior fossa tumors.[21] Because 58–76% of HGBs are found within or near the cerebellum,[30,40] rupture can lead to acute hydrocephalus, brainstem compression, and profound neurological decline. Despite the vascular nature of HGBs, they rarely present acutely in the setting of a catastrophic hemorrhage in the intraparenchymal or subarachnoid space. The presence of significant amounts of subarachnoid hemorrhage could also raise concern for the rupture of an aneurysm or AVM.[9] To this date, 56 cases of acute hemorrhage have been described with an underlying HGB.[1-3,5-7,10,11,15-18,20,22-25,31,33-35,37,41] A total of 19 cases have been described in the posterior fossa. Large hemorrhages related to HGBs have been described more commonly in spinal locations.[7,10,17-19,24]

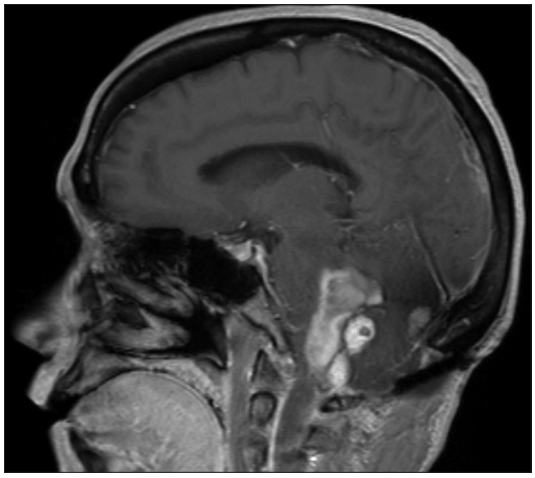

Figure 3: Sagital T1 magnetic resonance imaging with contrast demonstrating the 3 Hemangioblastomas preoperatively with underlying blood in the 4th ventricle

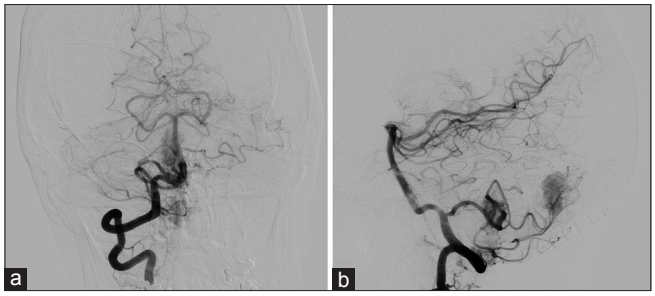

Figure 4: (a) Anteroposterior view of cerebral angiogram with right vertebral artery injection demonstrating two separate areas of tumor blush from the posterior inferior cerebellar artery. (b) Lateral view of cerebral angiogram with right vertebral artery injection demonstrating two separate areas of tumor blush from the posterior inferior cerebellar artery

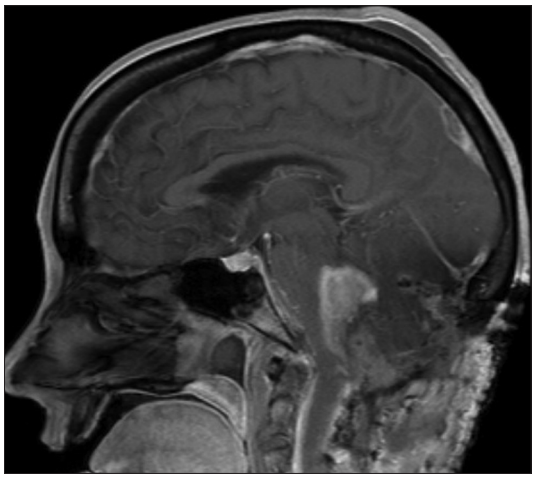

Figure 5: Image b demonstrates a postoperative view after resection of the 3 hemangioblastomas with residual intraventricular hemorrhage in the 4th ventricle

The proposed mechanisms of hemorrhage from a HGB may include hemorrhagic infarction from stenosis or occlusion of vessels by endothelial proliferation or tumor emboli, rupture of thin fragile vessels due to direct invasion of the vessels from tumor cells, vessel rupture due to loss of perivascular support tissue, rupture of fragile neovasculature, venous occlusion, and vascular degeneration due to radiation and/or chemotherapy.[19] A potential contributing factor in HGB-related hemorrhage may be due to upregulation of vascular endothelial growth factor (VEGF) causing fragile tumor vessels.[40] VHL is an autosomal dominant neoplastic disorder associated with a tumor suppressor gene defect on chromosome 3p25. There have also been studies suggesting that HGBs are associated with a loss of chromosomal 6q.[26] This deletion may be associated with a mutation in D6Mit135 that induces vascular dilation and may, in some cases, lead to hemangioblastomal hemorrhaging.[29,40]

The risk of hemorrhage in hemangioblastoma (HGB) has been suggested to be linked to their size. Glasker *et al.* calculated a 0.0024 risk of spontaneous hemorrhage per person per year in patients with HGBs.[17] This retrospective study and literature review concluded that the risk is virtually 0 when the HGB is less than 1.5 cm. Our case suggests the offending lesion was likely the larger lesion (fourth ventricular lesion at 2.9 cm versus vermian lesion at 1.2 cm) given the presence of intraventricular hemorrhage at presentation, but their proximity questions that assumption [Table 1]. Some more recent series have reported some cases of acute hemorrhage with tumors noted to be smaller than 1.5 cm.[12]

Solid HGBs are often more vascular during surgery[32] and more likely to cause massive hemorrhage than variants

Table 1: Location and changes in size of hemangioblastoma lesions over a 2-year period prior to hemorrhage

HGB location	Size 2 years prior to presentation, cm	Size day of presentation, cm
Fourth ventricle	2.2 × 1.1 × 1.5	2.9 × 1.4 × 1.7
Vermis	1.0 × 0.6 × 1.2	1.1 × 0.8 × 1.2
Cerebellar hemisphere	0.7 × 0.5 × 0.3	1.3 × 0.8 × 1.3

with large cystic components.[19] Our case suggests that the culprit for the initial hemorrhage seems to be the lesion involving the fourth ventricle. The significance of its transformation from a purely solid lesion to a partially cystic lesion over the 2 years prior to presentation is unclear. Review of an MRI of our patient [Figures 6–8], taken 2 years prior to presentation, demonstrates significant growth of all lesions, especially in the fourth ventricular lesion, which may be more representative of risk of rupture [Table 1]. The prevalence of HGB-related hemorrhage resulting in herniation is not well elucidated in the literature. HGB-related hemorrhages in the cerebellum resulting in tonsillar herniation are exceptional occurrences and have been reported in two cases, both of which involved the posterior fossa. All patients died from herniation after several episodes of intracranial hemorrhage.[14,23]

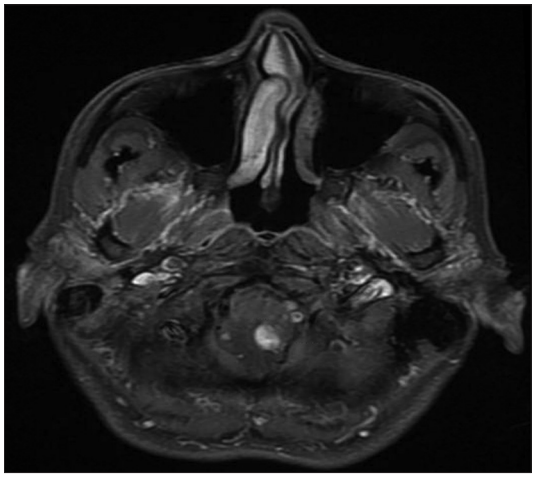

Figure 6: Axial T1 magnetic resonance imaging with contrast showing vermian hemangioblastomas 2 years prior to presentation

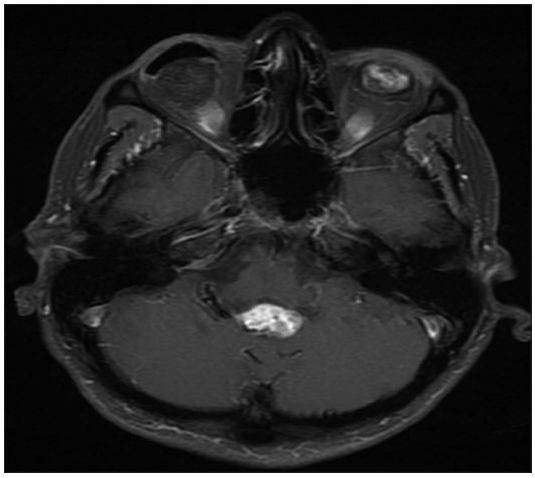

Figure 7:AxialT1 magnetic resonance imaging with contrast showing 4th ventricular hemangioblastomas 2 years prior to presentation

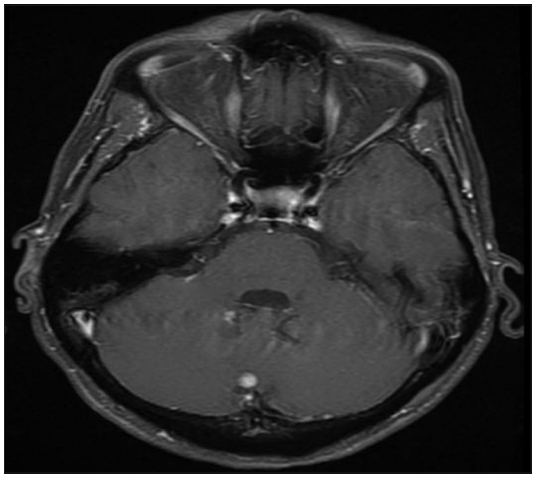

Figure 8:AxialT1 magnetic resonance imaging with contrast showing cerebellar hemangioblastomas 2 years prior to presentation

Partially debulked HGBs may also cause hemorrhage.[20,36] Re-hemorrhage rates are unknown in cases of HGBs when partial debulking has been accomplished. The rate seems high as demonstrated by our case as well as others.[14,23] The causal relationship to radiosurgery and the second hemorrhage is likely fortuitous. Pathological changes in benign brain tumors within months after radiosurgery demonstrate homogenous coagulation necrosis with scattered cell debris and apoptosis.[27] The coagulative necrotic area is surrounded with varying degrees of phagocytic cells and lymphocytes. Radiosurgical series of HGB only describe two cases of hemorrhage after treatment. Chun *et al.* describe a life-threatening hemorrhage 4 years post-radiosurgery for a HGB, despite adequate initial response.[8] A microhemorrhage has also been reported in one other case of a HGB treated with radiosurgery.[38]

We recommend complete resection of the HGB in case of hemorrhage, given their risk of subsequent hemorrhage. A staged approach may be necessary to perform decompression first, followed by a definitive resection once brain swelling has subsided.

CONCLUSION

Despite their vascular nature, symptomatic hemorrhage from HGB is an uncommon event. This case illustrates multiple posterior fossa HGBs leading to an acute event with tonsillar herniation. A staged approach with decompression followed by resection of the HGB can be safe in that situation. Re-hemorrhage of a HGB seems to be likely if a prior acute hemorrhage has occurred, and surgical resection should be emphasized over other treatment options.

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Conflicts of interest
There are no conflicts of interest.

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