Recurrent Spontaneous Thalamic Hemorrhage in the Setting of Vein of Galen Malformation

Timothy Hake MD, Mark Tornero MD
The Ohio State University Wexner Medical Center, Columbus Ohio

THE OHIO STATE UNIVERSITY
WEXNER MEDICAL CENTER

Introduction

- Vein of Galen Malformations (VoGM) are rare embryonic vascular malformations that are due to persistent shunting of primitive choroidal arteries into the median prosencephalic vein, the embryonic precursor to the great cerebral vein of Galen. The median prosencephalic vein fails to regress and becomes aneurysmal. Clinical manifestations are dependent on the age of presentation.
- Neurologic complications of thalamic hemorrhage can include neuropsychological disturbances, contralateral weakness, visual-field deficits, and aphasia.
- Lesions to the dominant thalamus (typically left) can lead to thalamic subcortical aphasia.

Case Description

- A 31-year-old male with a history of vein of Galen malformation with associated intracranial hemorrhages, was admitted to inpatient rehabilitation for recurrent spontaneous left thalamic hemorrhage.
- He was initially diagnosed with VoGM in late childhood after presenting with intracranial hemorrhage that was treated with multiple rounds of gamma knife radiation. Despite this, he had multiple hemorrhagic events, within the left thalamus, that were treated with partial embolization. Fortunately, he made good functional recovery and was able to return to normal life activities after each prior hemorrhage.
- Then, in the month prior to admission, he began to experience new headaches. Evaluation with a CT head demonstrated a repeat left thalamic hemorrhage (Image 1).
- He underwent a repeat embolization of the right PCA and left ACA, MCA, and ICA supply to the malformation with neurosurgery. Post-operative MRI showed chronic left thalamic hematoma, small acute cortical infarcts in left parietal and occipital lobes, and small subcortical infarcts in the left parietal lobe (Image 3).
- Once medically stable, he was admitted to inpatient rehabilitation with somnolence, right hemiparesis, right visual field cut, hemi neglect, speech apraxia, aphasia, and cognitive changes.
- Speech therapy noted that his presentation was most consistent with a thalamic subcortical aphasia with islands of preserved fluency, anomia, alexia, agraphia, and frequent semantic paraphasias. He often perseverated on “yah” or “I’m sorry” when attempting to speak with therapies. He required maximum assistance in order to state his name.
- He made steady gains during his rehabilitation admission and at the time of discharge, he was able to communicate more efficiently especially when given additional prompts. He was modified independent to requiring contact guard for completion of activities of daily living (ADLs) and able to ambulate independently (though difficulties with visual scanning due to right inferior quadrantanopia).
- He continued to follow up with outpatient speech therapy and occupational therapy.
- He later followed up with neuro-ophthalmology who indicated that the right visual field cut had resolved and that his visual acuity was sufficient to resume driving.

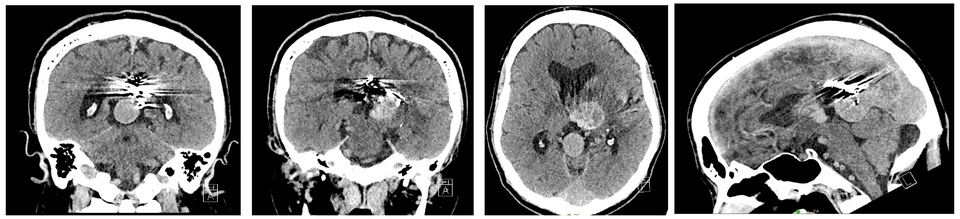

Image 1: Non-contrast computed tomography shows the dilated median prosencephalic vein, left thalamic hemorrhage with edema and mass effect on the 3rd ventricle, and streak artifact from prior embolization material.

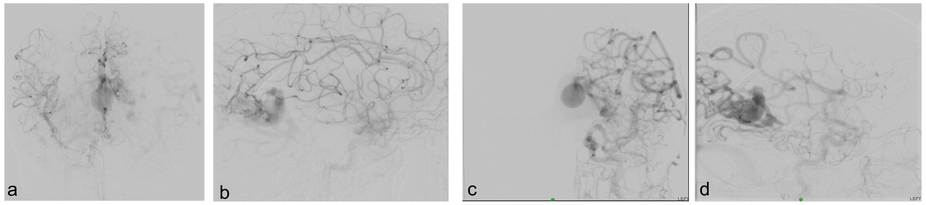

Image 2: Cerebral angiogram with injection into the right internal carotid artery (a and b) and left internal artery (c and d) shows choroidal arteries supplying the dilated median prosencephalic vein via right PCA and left ACA, MCA, and ICA feeders.

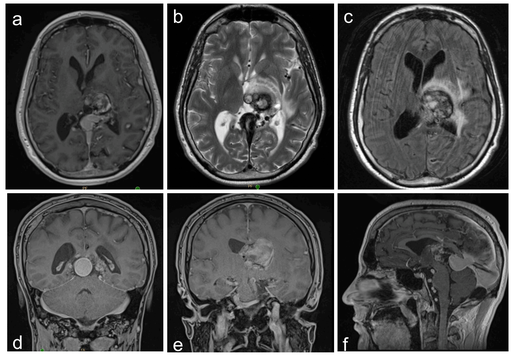

Image 3: Contrast-enhanced magnetic resonance imaging shows a choroidal-type VoGM with newly embolized arterial feeders, small acute cortical infarcts within the left parietal and occipital lobes, and chronic left thalamic hemorrhage. a. axial T1W, b. axial T2W, c. axial FLAIR, d and e. coronal T1W, f. sagittal T1W.

Discussion

- Vein of Galen Malformations (VoGM) are rare embryonic vascular malformations, occurring between the 6th-11th week of gestation. They constitute 1% of all intracranial vascular malformations.
- VoGMs are due to the persistence of the median prosencephalic vein (MPV), which is a venous structure that drains the embryonic choroid plexus. Normally, by the 11th week gestation, the development of the basal ganglia results in the formation of paired internal cerebral veins, which take over the venous drainage from the chorioid plexus and leads to the regression of the MPV, except for the most caudal part that joins the internal cerebral veins to become the vein of Galen. In VoGM, abnormal arteriovenous fistulas with high-flow prevent the normal regression of the MPV.
- There are 2 types of VoGMs depending on the fistula location -- mural and choroidal types.
- The clinical manifestations of VoGMs depend on the age of presentation and are due to the hemodynamics of the fistula.
  - Neonates present with high-output cardiac failure and pulmonary hypertension due to high-velocity shunting and have a poor survival rate.
  - Infants present with hydrocephalus, macrocrania, seizures, developmental delay, and mild cardiac symptoms.
  - Older children and young adults present with headaches or intracranial hemorrhage. They typically have smaller arteriovenous shunts that are low-velocity.
- Untreated VoGM have a poor prognosis. Treatment options include microsurgical techniques, stereotactic radiosurgery, and endovascular surgery. Endovascular embolization is the preferred treatment with overall survival and technical success approaching 80%.
- Complications of endovascular treatment includes intracerebral hemorrhage due to venous hypertension. This can be largely avoided by staging the embolization procedure.
- Lesions affecting the dominant (typically left) thalamus can result in thalamic subcortical aphasia, which is characterized by decreased spontaneous speech, hypophonia, limited grammatical errors, perseveration, word-finding difficulties, and prominent paraphasic errors. Transcortical aphasia with intact repetition is commonly observed.
- A particularly interesting aspect of thalamic aphasia has been described as a “dichotomous state” or a “quasiaphasic disturbance of vigilance”. That is, when fully awake the patient can speak clearly, but when sleepy they become paraphasic.
- The underlying mechanism of thalamic aphasia is controversial but has been proposed to be caused by disconnecting thalamic projections to language-related cortical regions which impairs the ability of the thalamus to monitor, control, and integrate cortical activity.

References

- Berenstein A, Fifi JT, Niimi Y, Presti S, Ortiz R, Ghatan S, Rosenn B, Sorscher M, Molofsky W. Vein of Galen malformations in neonates: new management paradigms for improving outcomes. Neurosurgery. 2012 May;70(5):1207-13; discussion 1213-4. doi: 10.1227/NEU.0b013e3182417be3. PMID: 22089754.
- Gupta AK, Varma DR. Vein of Galen malformations: review. Neurol India. 2004 Mar;52(1):43-53. PMID: 15069238.
- Maeshima S, Osawa A. Thalamic Lesions and Aphasia or Neglect. Curr Neurol Neurosci Rep. 2018 May 23;18(7):39. doi: 10.1007/s11910-018-0844-4. PMID: 29789957.
- Raybaud CA, Strother CM, Hald JK. Aneurysms of the vein of Galen: embryonic considerations and anatomical features relating to the pathogenesis of the malformation. Neuroradiology. 1989;31(2):109-28. doi: 10.1007/BF00698838. PMID: 2664553.
- Recinos PF, Rahmathulla G, Pearl M, Recinos VR, Jallo GI, Gailloud P, Ahn ES. Vein of Galen malformations: epidemiology, clinical presentations, management. Neurosurg Clin N Am. 2012 Jan;23(1):165-77. doi: 10.1016/j.nec.2011.09.006. PMID: 22107867.
- Samara A, Berry B, Ghannam M. Thalamic aphasia secondary to glioblastoma multiforme. J Clin Neurosci. 2020 Apr;74:234-238. doi: 10.1016/j.jocn.2020.01.063. Epub 2020 Jan 21. PMID: 31973919.