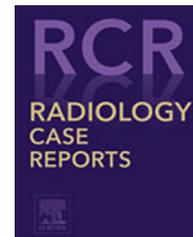


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## Case Report

## Giant vein of Galen malformation in an adult

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## ABSTRACT

Vein of Galen malformations (VoGMs) are rare vascular malformations resulting from persistent shunting of primitive choroidal vessels into the median prosencephalic vein of Markowski. VoGMs are associated with poor clinical outcome with a reported 76.7% mortality if left untreated. We present an exceedingly rare case of a giant, untreated VoGM measuring  $7.8 \times 5.5 \times 7$  cm in a 42-year-old man. The embryologic origin, classification, clinical manifestations, and treatment options of VoGMs are discussed with a review of pertinent literature.

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## Introduction

Vein of Galen malformations (VoGMs) are rare vascular malformations arising from persistent shunting of primitive choroidal vessels into the median prosencephalic vein of Markowski [1–6]. Patients with VoGMs usually present with high-output cardiac failure and hydrocephalus during infancy, with other delayed sequelae such as developmental delay and seizures. It has been associated with poor clinical outcome with a reported mortality of up to 76.7% if left untreated [7].

We present an exceedingly rare case of a giant, untreated VoGM measuring  $7.8 \times 5.5 \times 7$  cm in a 42-year-old man. The

embryologic origin, classification, clinical manifestations, and treatment options of VoGMs are discussed with a review of pertinent literature.

## Case report

A 42-year-old man with a history of seizures, developmental delay, and ventricular peritoneal shunt (VPS) was admitted to our institution for evaluation and treatment of sepsis. Given the history of VPS, a computed tomography (CT) of the brain was obtained which revealed marked calvarial hyperostosis likely due to his antiepileptic use and a large aneurysmal

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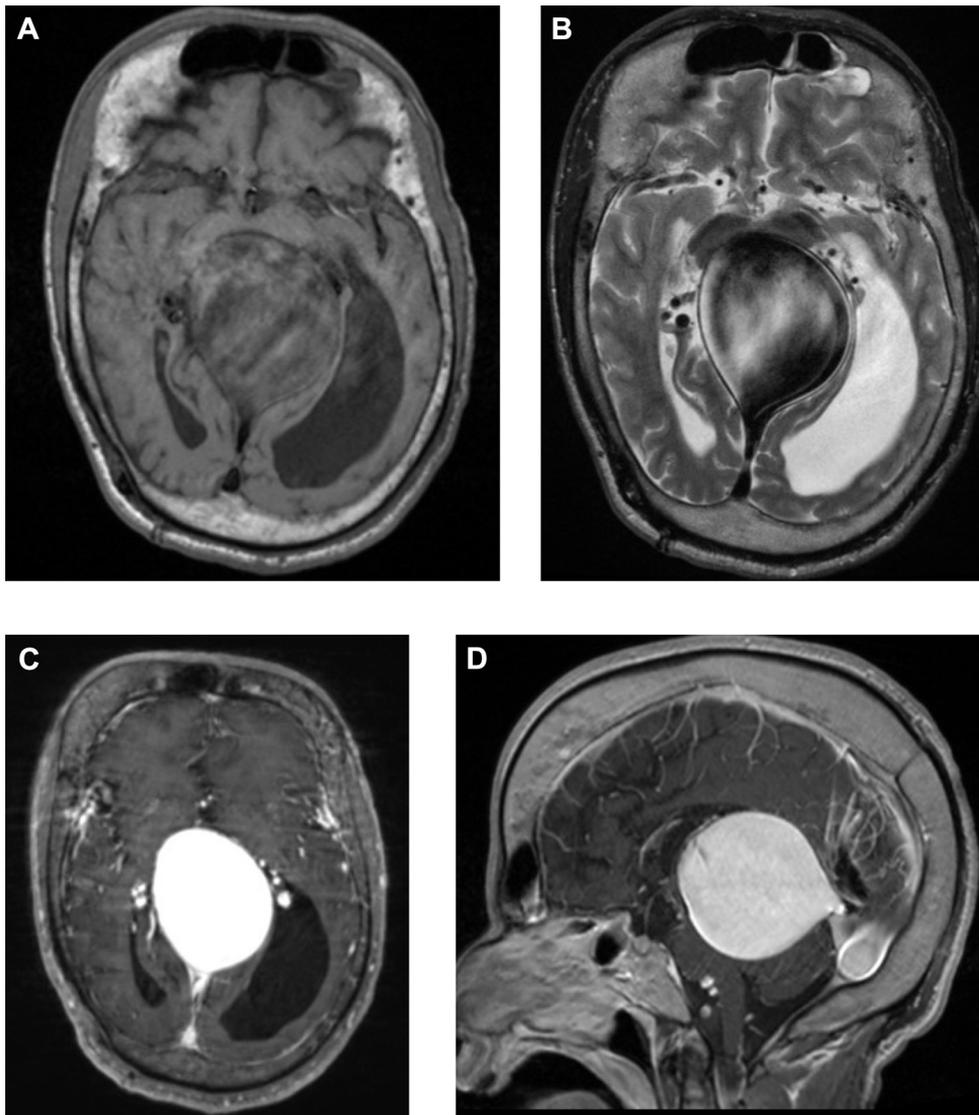
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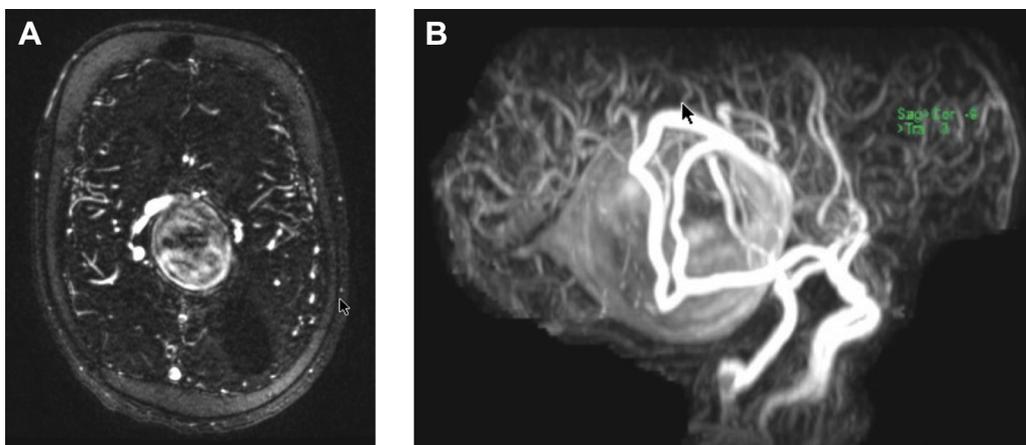
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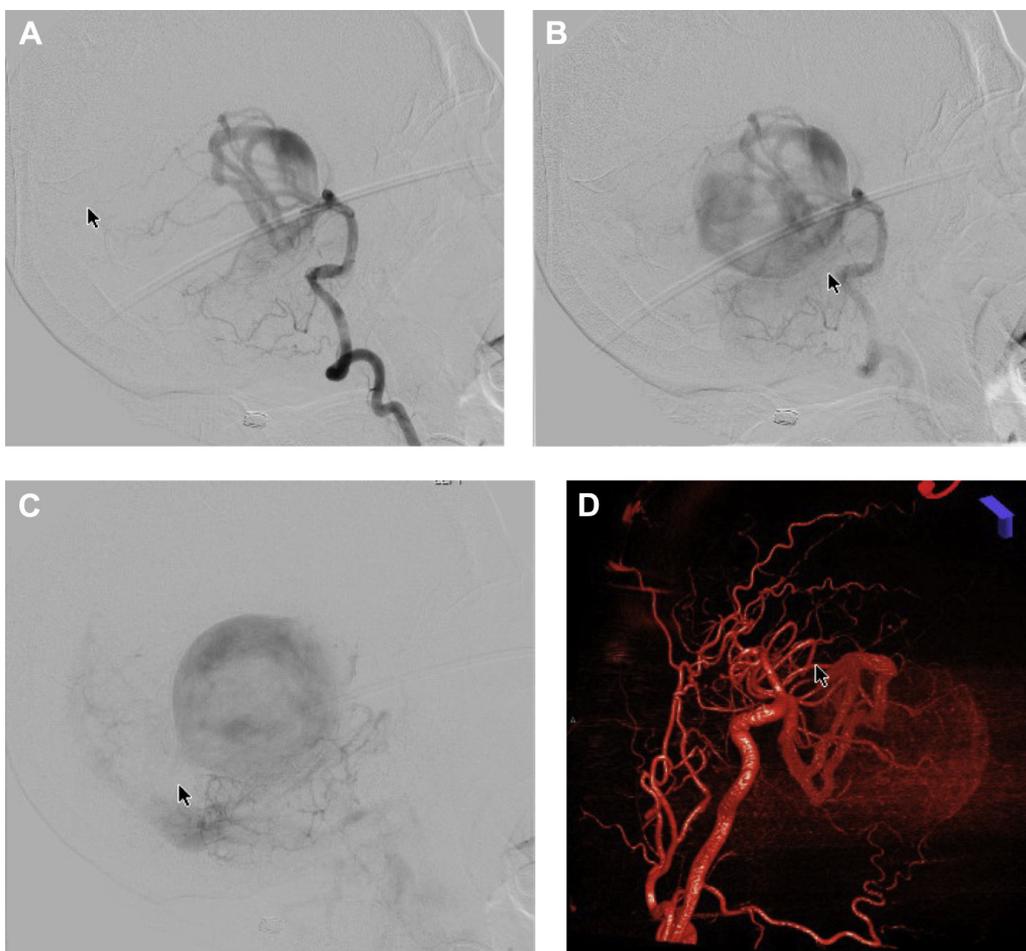
**Fig. 1 – (A)** A 42-year-old male with history of developmental seizures and ventricular peritoneal shunt with a giant VoGM. Noncontrast axial CT scan shows a giant midline venous dilation in the region of the quadrigeminal plate. Note the calcified rim in the wall of the venous malformation. **(B)** Noncontrast sagittal CT scan demonstrating a giant midline venous dilation and faint outline of its vascular communication with the straight sinus. There is also marked calvarial hyperostosis. **(C)** Lateral skull x-ray showing a calcified rim characteristic of a VoGM. CT, computed tomography; VoGM, vein of Galen malformation.



**Fig. 2 – (A)** Axial T1W, **(B)** axial T2W, **(C)** T1W postcontrast, and **(D)** sagittal T1W postcontrast MRI demonstrating a giant VoGM. MRI, magnetic resonance imaging; VoGM, vein of Galen malformation.



**Fig. 3 – (A)** Axial T1W postcontrast MRA demonstrating the arterial feeders to the giant VoGM. Note the large right dilated posterior cerebral artery and left posterior cerebral artery feeding the venous aneurysm. **(B)** 3D MRA demonstrating the main arterial feeders of the VoGM. 3D, three dimensional; MRA, magnetic resonance angiography; VoGM, vein of Galen malformation.



**Fig. 4 – (A)** DSA of the left vertebral artery injection demonstrating the main arterial feeders of the VoGM from bilateral PCA and left PCOM during the early arterial, **(B)** midarterial, and **(C)** early venous phase demonstrating contrast flow through the VoGM. **(D)** 3D DSA of the right vertebral artery demonstrating a large VoGM fed by bilateral P2 branches and left PCOM. 3D, three dimensional; PCA, posterior cerebral artery; PCOM, posterior communicating artery; VoGM, vein of Galen malformation.

dilation with mass effect on the quadrigeminal plate, consistent with a giant VoGM (Figs 1A and B). The patient's family member noted that the patient was diagnosed with a VoGM in early childhood and a VPS was placed at that time. The lateral skull x-ray from shunt series also revealed a giant, round mass with a thin, calcified rim (Fig. 1C).

After resolution of his sepsis and CSF involvement was ruled out, magnetic resonance imaging (MRI) with contrast of the brain was obtained and again demonstrating a giant VoGM measuring  $7.2 \times 5.5 \times 7$  cm with mass effect on the dorsal brainstem and superior cerebellum (Figs 2A–D). Magnetic resonance angiography revealed arterial feeders from bilateral P2 segments of the posterior cerebral artery, predominantly on the right (Figs 3A and B). The vertebrobasilar arteries were also dilated and torturous. The venous aneurysmal dilation drained in to the straight sinus and normal dural sinus tract. The left transverse sinus was dilated and the right hypoplastic. Both internal jugular veins were patent. The anterior and middle third of the superior sagittal sinus was hypoplastic.

Further investigation with a 4-vessel DSA with additional three-dimensional reconstructions of the right ICA was performed. Angiography findings confirmed that the VoGM was primarily supplied by bilateral P2 arteries and right fetal posterior communicating artery appreciated filling during the early arterial (Fig. 4A) and late arterial phases (Figs 4B and C). Given the patient poor clinical status at baseline and potential morbidity associated with further intervention, the family elected to continue observation of the VoGM.

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## Discussion

VoGMs, also known as vein of Galen aneurysmal malformations, are neither a true aneurysm nor does it arise in the vein of Galen. It actually arises from persistent shunting of primitive choroidal vessels into the median prosencephalic vein of Markowski, which is the embryonic precursor of the vein of Galen.

During the early development, the neural tube is nourished by the meninx primitive, a cellular plexus rich in vascular channels. As the anterior and posterior neuropore close, the choroid plexus forms and choroidal arteries supply the developing brain [2]. A single large central midline vein, the median prosencephalic vein of Markowski, also known as a primitive internal cerebral vein, drains the developing brain, diencephalon, and choroidal arteries [3]. During the 10th week, the bilateral internal cerebral veins form and preferentially drain the choroid plexus, rather than the anterior median prosencephalic vein of Markowski, which subsequently occludes. The internal cerebral veins drain into the posterior median prosencephalic vein of Markowski hence forming the vein of Galen [4,5]. Several vascular anomalies can occur with VoGMs including the absence of dural sinuses and dual straight sinus—one of which is actually a falcine sinus draining into the superior sagittal sinus and absent jugular veins [6].

VoGMs either arise from the pericallosal artery, the basilar or posterior cerebral arteries, or the anterior and posterior choroidal arteries [8]. Along with these findings are stenosis or

occluded dural sinuses along with aneurysms in the arterial vessels. Outflow impairment leads to venous hypertension and increased risk of hemorrhage [2]. Yaragil initially described four types of VoGMs, and Lasjuanias later divided VoGMs into two main types—choroidal and mural based on the location and nature of the shunt. The choroidal type consists of multiple high-flow fistulas draining into the tela choroidea, which subsequently drains into the anterior portion of the median vein of prosencephalic vein of Markowski. This is more commonly seen in newborns and infants. The mural type has a single arteriovenous fistula that feeds directly into the inferolateral wall of the median vein of Markowski. The arterial feeders are typically from posterior choroidal, and collicular arteries are a low-flow fistulas presenting in later childhood [9].

Clinical presentation and severity of disease depend on age at the time of diagnosis. Neonates have severe congestive heart failure and respiratory disease and have a poor survival rate. Infants do have cardiac symptoms but are often milder, present with hydrocephalus, and have seizures [8]. Seizures, hydrocephalus, and early diagnosis have been associated with poor outcomes [10]. Children and adults present with subarachnoid hemorrhage and typically have smaller arteriovenous shunting. The patient in this case presented in early childhood with hydrocephalus, seizures, and hemosiderin staining from prior subarachnoid hemorrhage, all of which are common clinical and radiographic findings in patients with VoGMs.

Treatment options include microsurgical techniques, stereotactic radiosurgery, and endovascular surgery. As new management paradigms and treatment strategies have evolved, the overall survival and technical success of endovascular treatment for VoGMs are now approaching 80% [10,11]. Microsurgical approaches have much higher morbidity and often fatal outcomes [12]. The optimal treatment method at this time for addressing VoGMs is endovascular therapy via transarterial or transvenous approaches, which is dependent based on age and clinical symptoms. Although the goal is complete obliteration of the malformation, partial occlusion may halt the progression of cardiac manifestations and promote self-thrombosis preventing the need for further embolization and if needed stage embolization may be used [8].

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## Conclusion

Untreated giant VoGMs in adults are extremely rare. Given the technological advancement in recent years, endovascular treatment has become the treatment of choice and offers the best clinical outcome in well-selected patients.

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