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A rare case of giant arteriovenous malformation of the vein of galen in an adult

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Abstract

Vein of Galen aneurismal malformations (VGAMs) arise from persistent arteriovenous shunting from primitive choroidal vessels into the median prosencephalic vein of Markowski, the embryonic precursor of the vein of Galen. VGAMs rarely present past infancy, and their natural history in adults is unknown. We report a case of a VGAM in an adult male patient. Though these lesions are extremely uncommon, they are of special interest to the interventional neuroradiologist because endovascular therapy has proved itself to be an effective, and often the only safe therapeutic modality available to treat these patients.

Keywords

VGAMs; neuroradiology; vein; endovascular therapy

Case Presentation

A 22-year-old male presented to radiology department with left sided pulsatile neck swelling since 15 years. The patient had no intellectual and developmental disabilities. His past medical history was unremarkable, and there was no family history of neurologic disease. There was no history of trauma. General and cardiovascular examinations as well as routine biochemical analysis were within normal limits. There was no evidence of skin lesions to suggest capillary malformation neither there was any limb hypertrophy.

Ultrasound Examination was performed using high frequency probe which showed dilated tortuous vascular channels along the left side neck predominantly below the angle of left mandible. These tortuous vascular channels showed mainly venous triphasic flow with significant aliasing, raising the possibility of AV malformation left side neck vessels. Right-sided neck vessels appeared unremarkable.

No other pathological or nodular focus identified in neck or supraclavicular region on either side. For further workup CT angiography was recommended.

The patient underwent CT angiogram, which showed multiple dilated enhancing vessels centered in the midbrain, third and lateral ventricles. There was vascular hypertrophy with multiple dilated veins opening into the vein of Galen. Calcifications identified in the wall of vein of Galen. The vein of Galen drained posteriorly into large sigmoid sinus sinus which ultimately drained into left internal jugular vein.

There was aneurysmal dilatation of the left external jugular vein. Findings were consistent with vein of Galen aneurysmal malformation. Unfortunately, this patient was lost to follow up and therefore the treatment and management could not be documented.

Discussion

Vein of Galen malformations (VOGMs) are rare anomalies of intracranial circulation that constitute 1% of all intracranial vascular malformations. However, they represent 30% of vascular malformations presenting in the pediatric age group [1].

The symptoms in older children include headache that may or may not be associated with subarachnoid hemorrhage. Gold et al.'s survey found that subarachnoid hemorrhage occurred in 10 of the 13 patients belonging to adult age category. Although the discovery of VGAM at the adult age is still considered rare, late diagnosis in pauci- or asymptomatic patients may become more common with increased availability and use of MR imaging. The median age of diagnosis has not been documented in available literature. Because of its rarity in adults, and despite what is known from published studies, there is still insufficient information about this disease during adult age. Use of oral contraceptives, postpartum status, sickle cell anemia, and aseptic meningitis were risk factors related to thrombosis of the vein of Galen [2].

It has been associated with poor clinical outcome with a reported mortality of up to 76.7% if left untreated [3].

Steinheil in 1895, made the first reference to a Galenic malformation-referring to it as a 'varix aneurysm' [4]. The nomenclature is imprecise because the dilated venous structure that is characteristic of these malformations has been demonstrated to represent the embryonic median prosencephalic vein, and not the vein of Galen. These lesions are characterized by the presence of an aneurysmally dilated midline deep venous structure, fed by abnormal arteriovenous communications.

Although the etiological basis of Vein of Galen malformation is presently unknown, arteriovenous shunts are thought to develop during the 6th to 11th weeks of gestation. The lesion is located midline and generally receives a bilateral, symmetrical blood supply.

There are two types of angioarchitecture associated with Vein of Galen malformations VGAM: choroidal and mural. The choroidal type receives a contribution from all choroidal arteries before emptying into a venous pouch, and is a very primitive condition associated with poor. clinical outcomes in neonates. The mural type is better tolerated than the choroidal condition, and therefore is found in infants who do not experience severe cardiac symptoms and have better clinical prognoses .[5] This type is characterized by direct AVFs within the wall of the median prosencephalic vein of Markowski. Whereas the mortality rate for Vein of Galen malformations was close to 100% in the past, recent advances in endovascular techniques have greatly improved the survival rate. In this paper we will describe the clinical presentation of VGAM, review the history of treatment options, and present recent advances in endovascular management.

Patients with a VGAM most commonly present with cardiac and neurologic complications. The clinical presentation depends on the age of presentation. Neonates tend to present with high-output cardiac failure, pulmonary hypertension, and, in more severe cases, multiorgan system failure. Infants

commonly present with hydrocephalus, seizures, or neurocognitive delay.

Gold et al in 1964 provided a classification scheme taking into account the age at which the patient presented [6]. According to their clinical classification system for VOGMs, the older children and adults usually have low-flow fistulae. These patients usually present with headache and seizures. A small number of patients may also present with developmental delay, focal neurological deficits, proptosis and epistaxis. Subarachnoid hemorrhage and intracerebral hemorrhage can occur in this age group due to rerouting of blood into the pial veins [7]. Venous hypertension coupled with impaired CSF reabsorption may also lead to hydrocephalus and associated complications [8].

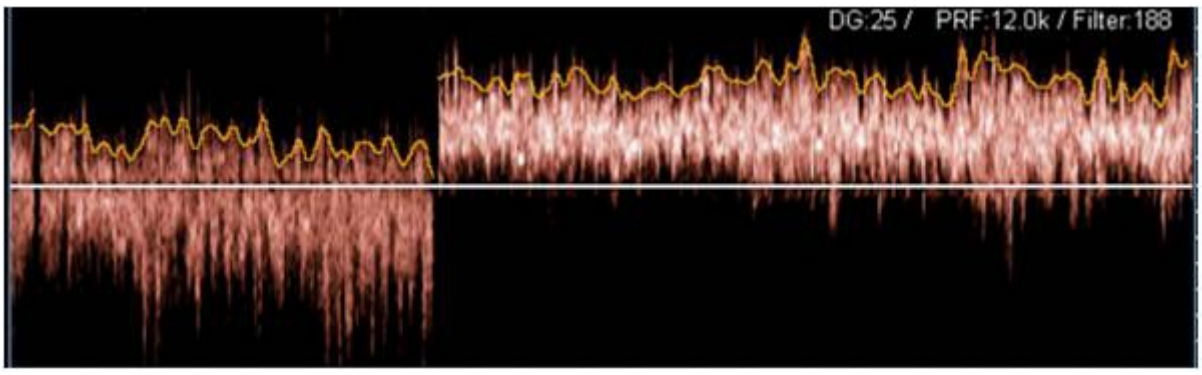
Magnetic resonance imaging (MRI) of the brain can help confirm the diagnosis of VGAM and detect cerebral changes such as infarcts, atrophy, and hydrocephalus [9,10]. An electroencephalogram should be performed on patients in the intensive care unit setting to rule out seizure activity. Computed tomography (CT) is a useful screening tool and is often the imaging modality that detects a mass in older patients [8]. CT angiography is a useful, noninvasive imaging modality that provides a clear map of the arteries and veins. It has better spatial resolution and can be obtained faster than magnetic resonance angiography (MRA) [11]. Conventional angiography is the gold standard imaging modality to evaluate the VGAM angioarchitecture.

Treatment depends on the structure of the malformation [12]. Endovascular therapy is the first-line treatment of a VGAM and its associated sequelae. Medical management is used to stabilize the patient until endovascular intervention can be performed. A ventriculoperitoneal shunt may be required if there is hydrocephalous. To reduce the blood flow into the vein, the feeding fistulous arteries into the Vein of Galen must be blocked [13]. Surgery is used as an adjunct to treat hydrocephalus, intracranial hemorrhages, and as a last resort when endovascular interventions have failed.

Most noted complication is intracranial hemorrhages [14] . Incidence of mortality is about 77% in untreated cases. The mortality rate is about 39.4% even after surgery [15].

Given the complex nature of treating patients with a VGAM, a multidisciplinary approach is recommended. Endovascular therapy combined with a multidisciplinary management strategy to treat a VGAM has significantly lowered mortality and can result in normal neurologic development in surviving patients. Clinicians should be given awareness about this condition for proper management and for better prognosis.

Figures



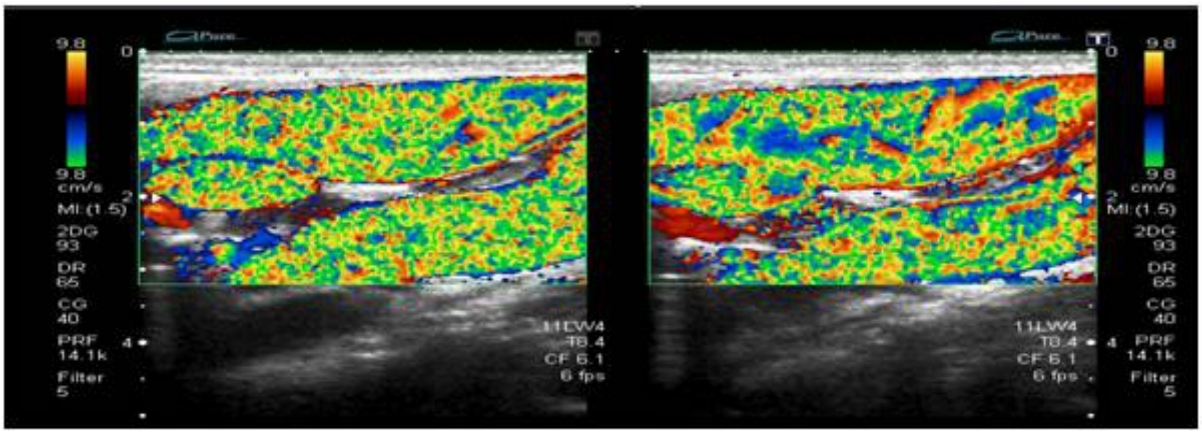


Figure 1: Ultrasound Examination showing tortuous vascular channels with venous flow and significant aliasing, raising the possibility of AV malformation in neck vessels

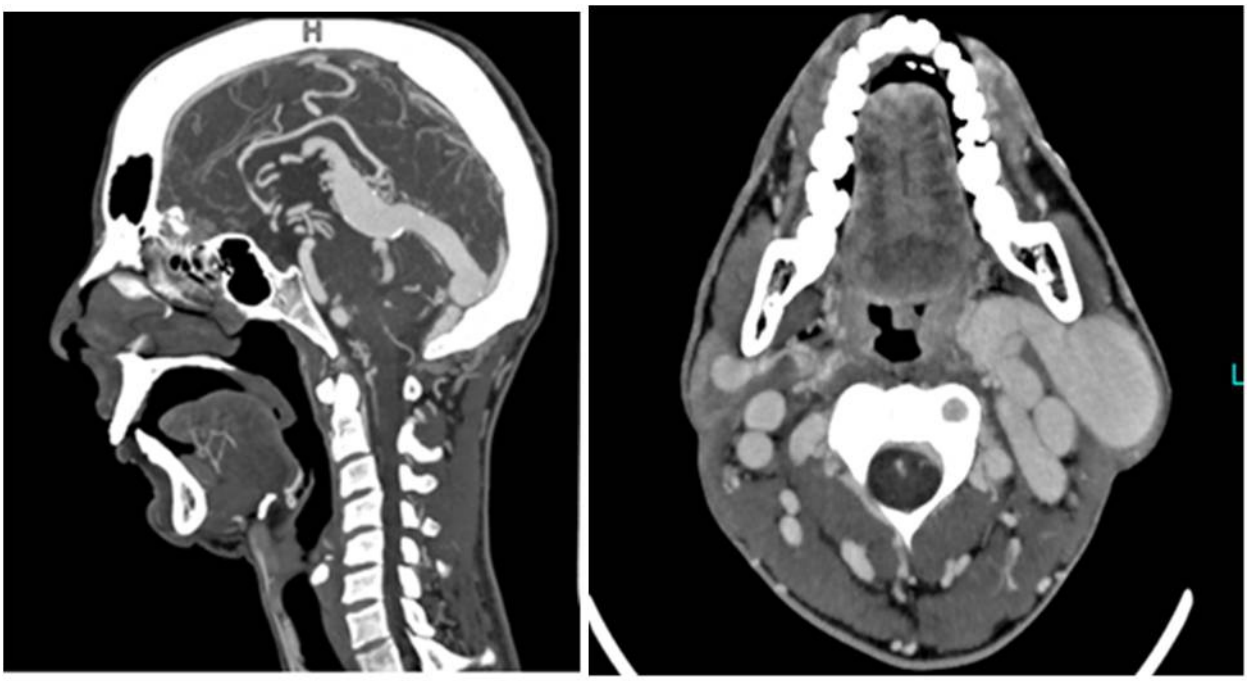


Figure 2: Computed tomography of malformation with contrast-medium enhancement.

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