Gnoscience Group

Case Reports and Reviews: Open Access

Case Report| Vol 4 Iss 2

Galen Vein Aneurysm: Case Report

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Received: July 07, 2023; **Accepted:** August 10, 2023; **Published:** August 15, 2023

Abstract

The term "Galena vein aneurysm" encompasses various vascular abnormalities characterized by the dilation of the vein of Galen. This condition is considered a rare vascular malformation, with experts estimating an incidence of approximately 1 in 25,000 births. Despite accounting for only 1% of all intracranial abnormalities, it affects up to 30% of the pediatric population. In this report, we present a case of prenatal diagnosis of Galen vein aneurysm using sonography and magnetic resonance imaging techniques.

Keywords: ***Galen vein, Aneurysm, Vascular malformation, Sonography, MRI.***

1. Introduction

Vein of Galen aneurysmal malformations (VGAM) account for less than 1% of all congenital cerebral vascular abnormalities, but they disproportionately affect pediatric populations, constituting approximately 30% of cases [1], [2]. VGAM is a prenatal cerebral arteriovenous malformation that develops prior to the tenth week of gestation, resulting in the formation of arteriovenous shunts between the choroidal circulation and the precursor to the vein of Galen, known as the median prosencephalic vein of Markowski. This anomalous connection leads to the dilation of the vein of Galen in VGAM cases [3]. The severity of VGAM can vary, as can the clinical manifestations, depending on the age at which the condition is diagnosed [4].

Prenatal indicators of the disease encompass heart failure and hydrocephalus caused by venous congestion, as well as oligohydramnios resulting from inadequate renal blood flow due to heart failure [5].

Citation: Zakaria A, Soufiane K, Chaimae ES, et al. Galen vein aneurysm: Case report. Case Rep Rev Open Access. 2023;4(2):130.

2. Case Report

We present a case involving a 25-year-old primigravida who presented at our department for a routine ultrasound checkup during her 28th week of gestation. During the examination, a vein of Galen malformation was identified in the fetus. Prenatal ultrasound revealed the presence of an anechoic structure located superior to the thalami, extending posteriorly. This characteristic appearance is commonly referred to as the "keyhole" sign (Fig. 1).

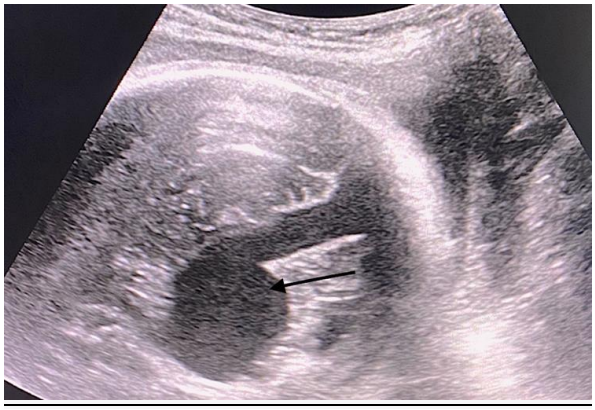

Fig. 1. Gray scale sonographic images in an axial plane of the fetal head at 28 weeks of gestation show a large VGAM (arrow). Evaluation of the hemispheric gray and white matter is limited.

As part of the diagnostic process, an MRI scan of the fetal head was performed using T2-weighted sequences in the coronal, sagittal, and transverse planes, along with diffusion sequences. The MRI examination revealed a well-defined, markedly T2 hypointense, oval-shaped mass measuring 35x31mm along the midline. This finding corresponded to an aneurysmal dilatation of the vein of Galen (Fig. 2).

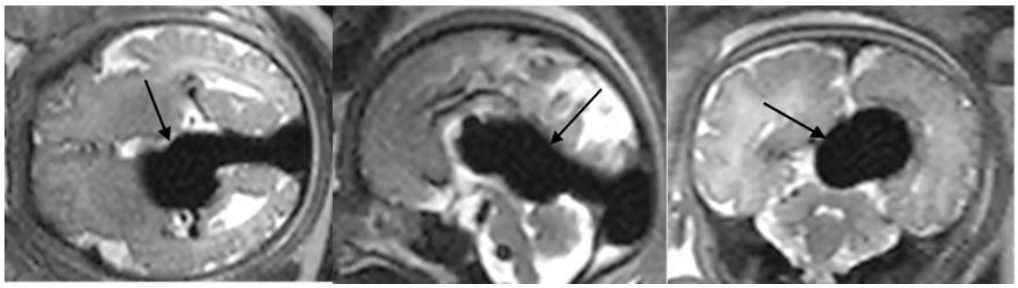

Fig. 2 A-C: Multiplanar T2-weighted images at 28 weeks of gestation show a well-defined, markedly T2 hypointense, oval-shaped mass measuring 35x31mm along the midline, corresponding to an aneurysmal dilatation of the vein of Galen (Arrows).

Additionally, the straight sinus and the torcular were observed to be enlarged. The dilation of the vein of Galen resulted in the compression and collapse of the third ventricle. No evidence of ventricular system modeling was observed in the supra- and infratentorial spaces. Furthermore, a slight midline shift was noted. Notably, no abnormalities were detected on diffusion-weighted images (Fig. 3).

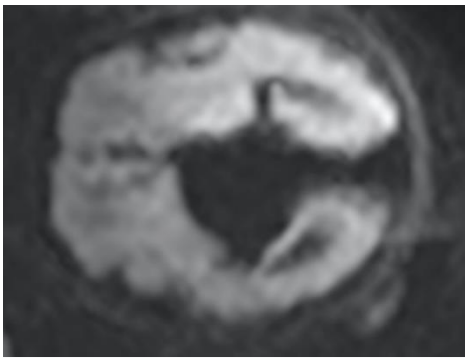

Fig. 3. Diffusion-weighted imaging (DWI)shows no ischemic signal in the brain parenchyma, and the mass was a low signal indicating no thrombus and hemorrhage.

3. Discussion

Vein of Galen malformations (VGMs) are a rare occurrence among cerebral vascular anomalies, accounting for only 1% of cases. However, they represent one of the most prevalent arteriovenous abnormalities in children, with diagnoses made both prenatally and postnatally [4].

In the literature, this congenital malformation is referred to by various names, including "Vein of Galen aneurysms," "arterovenous vein of Galen aneurysms," "arterovenous malformations in the vein of Galen," and "vein of Galen aneurysmal malformations" [6].

The nomenclature surrounding this abnormality can indeed be confusing, as it does not directly involve the vein of Galen itself but rather its embryonic precursor, the median prosencephalic vein (MProsV) [8]. The MProsV, also known as Markowski's vein, undergoes involution as the middle cerebral veins develop during the natural progression of intracranial vasculature. The proximal portion of the MProsV completely regresses, while the distal segment transforms into the vein of Galen [4].

In the presence of vein of Galen malformations (VGMs), the formation of arterovenous connections with the MProsV not only inhibits its regression but also leads to significant dilation of the preserved vessel. This dilated structure becomes distinctly visible in radiological images, positioned in the middle of the brain [7].

Additionally, it is likely that certain structures characteristic of early embryonic development, such as the falcine sinus, remain preserved, potentially impeding the formation of other sinuses, including the straight sinus [8].

Several classification systems have been proposed for vein of Galen aneurysmal malformations (VGAM), including the Yasargil classification and the Lasjaunias classification, among others. The Lasjaunias classification is commonly used and categorizes VGAM into two types: choroidal and mural.

The choroidal type is characterized by numerous feeders, including the thalamoperforating, choroidal, and pericallosal arteries. It is more prevalent in neonates and often results in high-output cardiac failure. The mural type of vein of Galen malformation is characterized by the presence of fistulae in the subarachnoid space within the wall of the median prosencephalic vein. This type is often associated with the absence or stenosis of dural sinuses.

The mural type typically manifests later in life, usually during infancy, and commonly presents with symptoms of hydrocephalus [9].

When it comes to diagnostic imaging, ultrasound examination is considered the preferred modality for detecting vein of Galen malformations. Ultrasound offers advantages such as its non-invasive nature, safety, and cost-effectiveness. In the third trimester of pregnancy, an ultrasound examination can raise suspicion of a vein of Galen malformation. [10]

Magnetic resonance imaging (MRI) serves as a valuable complementary technique in the evaluation of vein of Galen malformations. It is particularly useful in situations where ultrasound images are unclear due to factors such as advanced pregnancy, extreme obesity, or oligohydramnios. MRI offers the advantage of providing detailed visualization of the fetus in various planes, with a wide field of view [10].

One of the notable advantages of MRI is its objective nature, allowing for accurate assessment of the malformation. Furthermore, it plays a crucial role in determining the prognosis and planning appropriate treatments for the postnatal period [19], [20]. MRI provides valuable information that aids in making informed decisions regarding the management of vein of Galen malformations [11].

In the postnatal period, transarterial embolization is the recommended treatment approach for vein of Galen malformations. The efficacy of this treatment largely depends on the extent of the abnormalities present and the development of any associated complications.

Transarterial embolization involves the selective delivery of embolic agents via catheterization of the feeding arteries to block the abnormal arteriovenous shunts within the malformation. The goal is to redirect blood flow and reduce the vascular abnormalities, ultimately improving the overall prognosis and minimizing potential complications. The specific treatment plan should be determined based on a thorough evaluation of each individual case, taking into account the characteristics and severity of the vein of Galen malformation [12].

4. Conclusion

Vein of Galen aneurysm is a rare congenital vascular malformation that arises from the maldevelopment of the embryonic precursor, the median vein of Markowski. Prenatal screening using fetal ultrasonography plays a crucial role in detecting developmental abnormalities, including vein of Galen malformation [4].

Additionally, magnetic resonance imaging (MRI) serves a dual purpose in both the diagnosis and periodic follow-up of vein of Galen malformation. The combined use of fetal ultrasonography and fetal MRI provides a clear and accurate diagnosis during pregnancy, enabling effective planning for the delivery of the fetus and subsequent postnatal therapy. This comprehensive approach ensures appropriate management and treatment for vein of Galen aneurysm cases, optimizing outcomes for affected individuals [10].

5. Conflict of Interest

The authors are contributed equally and declare no competing interest.

6. Guarantor of Submission

The corresponding author is the guarantor of submission.

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