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Unexpected CT Scan Discovery of a Giant Galen Vein Aneurysm in An Infant: A Rare Case Report

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Abstract **Case Report**

Aneurysmal malformation of the Galen vein is an uncommon abnormality, comprising less than 1% of all congenital cerebral vascular malformations. However, it accounts for approximately 30% of such malformations in the pediatric population. This condition represents a cerebral arteriovenous malformation that develops prenatally, initiating prior to ten weeks of gestation. It arises from the formation of arteriovenous fistulas connecting the choroidal circulation and the median prosencephalic vein, an embryonic precursor of the Galen vein, which subsequently undergoes dilation. Here, we present an exceptional case of this malformation that was incidentally detected during a Brain CT scan.

Keywords: Vein of Galen aneurysmal malformation, VGAM, Markowski Vein.

INTRODUCTION

Vein of Galen aneurysmal malformation (VGAM) is an infrequent cerebrovascular malformation that was initially described by Steinhel and cited by Dandy. It constitutes a minority, accounting for less than 1%, of all intracranial arteriovenous malformations, but interestingly, it represents a substantial proportion, approximately 30%, of such cases in the pediatric population. VGAM is an intricate anomaly characterized by the atypical persistence of Markowski's median porencephalic vein, which, nourished by the primitive embryonic meningeal arteries, assumes the function of a blood collector within this malformation. Essentially, this leads to a pseudoaneurysmal dilation of the ampulla of Galen along with the presence of one or more arteriovenous fistulas [1].

The diagnosis of VGAM predominantly occurs during the neonatal or postnatal period, while occurrences in adulthood are exceedingly rare [2]. The severity of the vein of Galen arteriovenous malformation (VGAM) can exhibit a wide range, and the clinical manifestations may vary depending on the age of the patient at the time of disease presentation. It is important to note that the diverse clinical features and progression of AAG necessitate thorough evaluation and comprehensive management approaches tailored to individual patients.

CASE REPORT

We report the case of a 3 years old girl with a medical history of psychomotor retardation, which presented to the pediatric emergency department with aperytic convulsive seazures and in whom a brain computed tomography (CT) scan using spontaneous contrast showed an aneurysmal dilatation of the Galen vein, which appears isodense and exhibits localized wall calcifications. After the administration of contrast material, there is normal enhancement without detectable thrombus. The dimensions of the dilatation measure 40 x 43 x 36.5 mm (anterior-posterior x transverse x cranio-caudal). This dilatation is situated in the pineal region posterior to the third ventricle, protruding into the left lateral ventricle. It is associated with minimal biventricular hydrocephalus. The dilatation appears to communicate with the dilated right anterior choroidal artery.

In addition, there is dilation observed in the inferior right sinus, torcula, transverse sinuses, sigmoid sinuses, inferior and superior petrosal sinuses, and cavernous sinuses. These structures exhibit spontaneous isodensity and localized wall calcifications, which demonstrate normal enhancement after contrast

administration. Multiple dilated and tortuous vascular structures are observed within the brain tissue, involving the cortex, subcortex, and both supra- and subtentorial regions. The majority of these vessels exhibit calcified walls, indicating peri-aneurysmal collateral vessels. Refer to Figures 1, 2 and 3 for visual representation.

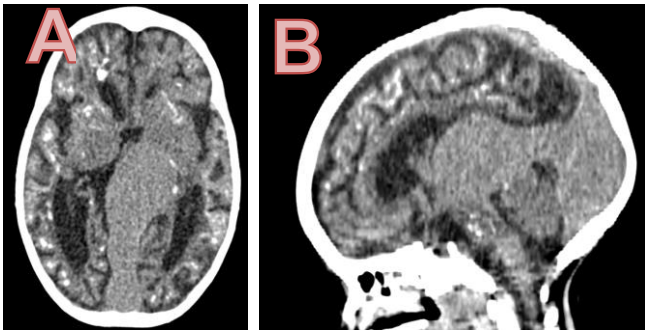

Figure 1: A non-contrast-enhanced cerebral computed tomography (CT) scan in axial (A) and sagittal (B) planes.

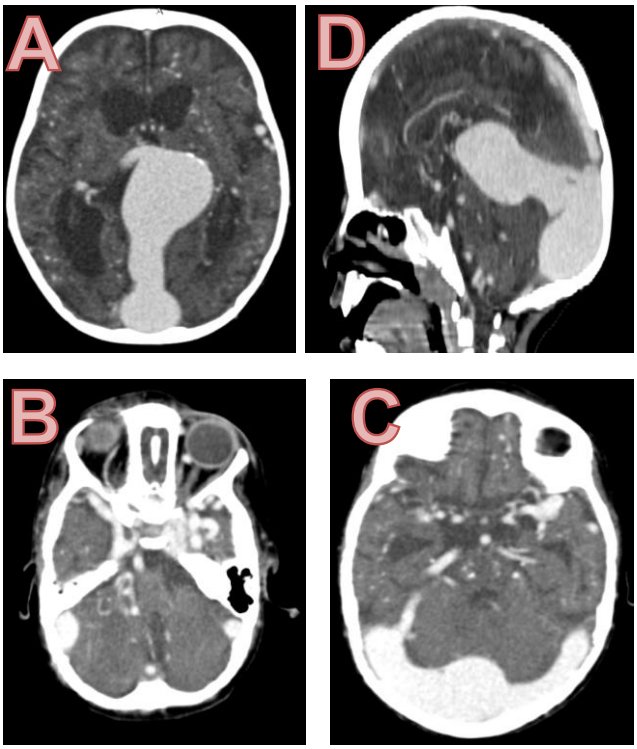

Figure 2: A contrast-enhanced cerebral computed tomography (CT) scan in axial (A, B, C) and sagittal (D) planes

DISCUSSION

Vein of Galen aneurysmal malformation (VGAM) was initially documented by Steinhel in 1895 [3]. It is an uncommon congenital cerebral arteriovenous abnormality, with an incidence of approximately 1 in 25,000 individuals. VGAM typically develops during the 6th to 11th weeks of gestation and can potentially be identified prenatally through the use of ultrasound. However, in most cases, the detection of VGAM occurs after birth. The aneurysmatic enlargement of the large unpaired cerebral vein, known as Vena Galeni, is caused by multiple shunts with brain arteries [4].

A vein of Galen aneurysm (VGA) is characterized by the presence of a solitary midline venous sac accompanied by direct arteriovenous fistulas within its wall. These fistulas typically receive bilateral blood supply from both the prosencephalic and mesencephalic arterial systems. Notably, this anatomical arrangement deviates from the normal vein of Galen and instead corresponds to the transient embryonic structure known as the median prosencephalic vein of Markowski. The arterial distribution of the feeders within the malformation aligns with the relatively stable pattern established by the sixth week of embryonic development. Furthermore, the transient median prosencephalic vein normally regresses by the eleventh week. These observations lead to two important inferences: the vein of Galen malformation is acquired rather than congenital and the insult that gives rise to this malformation occurs during

the critical period between the 6th and 11th weeks of gestation when the brain vesicle relies on the prominent choroid plexus for nourishment and prior to the establishment of intrinsic brain vascularization [5].

The clinical presentation of VGAM can exhibit variations based on factors such as the patient's age and the specific anatomy and angioarchitecture of the malformation. In neonates with the choroidal type of VGAM (characterized by multiple arteriovenous shunts due to the contribution of all choroidal arteries before draining into a venous pouch), volume overloading is often the dominant manifestation. This can manifest as increased cardiac output, tachycardia, cardiomegaly, cardiac insufficiency (high-output failure), pulmonary hypertension, respiratory distress syndrome, pulmonary edema, and even multiorgan failure. In infancy, the mural type of VGAM (involving fewer but larger shunts as direct arteriovenous fistulas within the wall of the median prosencephalic vein of Markowski) is associated with hydrocephalus, macrocrania, and developmental retardation. Epileptic seizures, indicating brain damage, are rarely observed in VGAM. It should be noted that clinical presentations of VGAM can differ across different regions of the world [1, 3, 4].

The diagnosis of vein of Galen aneurysm (VGAM) relies on various imaging modalities, including ultrasonography, angiography, computed tomography (CT), and magnetic resonance imaging (MRI). Ultrasonography is commonly employed for intrauterine detection, with grey-scale sonography primarily identifying cystic dilatations and peripheral manifestations. To further characterize these cystic malformations as vascular lesions, color Doppler sonography is utilized. Additionally, 3D power Doppler sonography offers multiple perspectives of the lesion's nature and anatomical structure during or after examination. Prenatal or postnatal MRI provides a detailed assessment of the lesion's configuration, aiding in the confirmation of the diagnosis and identification of complications such as hemorrhagic injuries within the brain's white matter. Postnatal angiography can also be beneficial for diagnostic purposes and predicting appropriate treatment strategies [6-8].

If left untreated, the mortality rate of newborns with severe cardiac insufficiency due to VGAM is 100%, while in infancy, it amounts to 72%. The safest and preferred treatment for VGAM is endovascular embolization, primarily performed through a transarterial approach using a special type of glue. Other therapeutic methods such as transvenous embolization or surgical treatment are associated with higher rates of complications and mortality. The timing and method of endovascular embolization depend on the patient's clinical signs and symptoms. The Bicêtre score, which is based on the research group's experience with over 300 VGAM patients, can be utilized to assess the therapeutic management of neonates with VGAM. This scoring system evaluates the cardiac, cerebral, respiratory, hepatic, and nephrological status of the patient. If cardiovascular and neurological symptoms remain stable (Bicêtre score > 12), it is recommended to delay treatment until the age of 5 to 6 months. In cases where the Bicêtre score falls between 8 and 12 points, emergency endovascular embolization should be performed. If profound neurological deficits or medically uncontrollable cardiac insufficiency are present (Bicêtre score < 8), invasive procedures are not indicated [9, 10]

Historically, the implementation of embolization procedures for the management of vein of Galen aneurysm (VGAM) resulted in unfavorable outcomes, characterized by significant mortality and morbidity rates in both neurosurgical and conservatively treated patients. However, recent studies conducted by various research groups have shown improved survival rates and favorable outcomes in VGAM patients who underwent timely therapeutic embolization. These positive outcomes are associated with normal growth trajectories and, at most, mild neurological deficits in the affected individuals [9].

CONCLUSION

In conclusion, the aneurysmal malformation of the Galen vein is an uncommon congenital cerebral vascular anomaly, accounting for a small proportion of such malformations in general, but exhibiting a higher occurrence in pediatric cases. Our case report details a distinctive instance of this malformation that was fortuitously identified during a Brain CT scan. These findings underscore the significance of comprehensive diagnostic imaging in detecting and characterizing rare cerebral vascular abnormalities, even when they are incidentally encountered. Further research and clinical investigation are essential to enhance our comprehension of the clinical implications and optimize management strategies for this unique condition.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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