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VEIN OF GALEN MALFORMATION- A RARE LIFE-THREATENING DISEASE

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ABSTRACT

The scope of this review article is to provide information on vein of galen malformation. Vein of Galen aneurysmal malformation (VGAM) is a rare but potentially life-threatening cerebral vascular malformation with an estimated incidence of 1 in 25,000. VGAM represents nearly 30% of all pediatric cerebrovascular malformations. Survival rate is very low in the cases of this disease. In the more severe cases, the blood overload goes almost directly from the vein to the heart and rapidly overwhelms it, leading to congestive heart failure. VGM is a rare life-threatening disease and even after surgical treatment, the mortality rate remains as high as 39.4%. This article also highlights latest cases on VGM and their life expectancy.

KEYWORDS: VGAM, aneurysm, heart failure, blood.

INTRODUCTION

A vein of Galen malformation starts during early prenatal development, as early as the first trimester. In normal prenatal development, a large vein forms at the base of the brain and is eventually replaced by a true vein of Galen. When a child has a vein of Galen malformation, high-pressure, oxygen-rich blood from the arteries flows directly into the initial vein, preventing development of the actual vein of Galen.[1]

Without the vein of Galen, oxygen-rich blood is unable to flow slowly through capillaries that deliver this blood to surrounding tissues. Since there is also no slowing of the blood flow, it rushes quickly away through the vein, causing a constant rush of blood to the heart and lungs. The heart and lungs have to work extra hard to keep up with the volume of blood, which may result in congestive heart failure or pulmonary hypertension. These conditions are often life-threatening.[2]

Vein of Galen aneurysm is a rare form of arteriovenous malformation in which the embryonic precursor to the vein of Galen, a vein at the base of the brain, dilates causing too much blood to rush to the heart. This can lead to rapid heart failure. Other features may include increased head circumference resulting from hydrocephalus, unusually prominent veins on the face and scalp, developmental delay, persistent headache, and other neurological findings. Vein of Galen aneurysm is often recognized on an ultrasound late in pregnancy. In other cases, it is diagnosed after birth. Although the exact cause remains unknown, this condition appears to result from a defect in early fetal development. VGAM consist of a tangled mass of dilated vessels supplied by an enlarged artery.[3] The malformation increases greatly in size with age, although the mechanism of the increase is unknown. Dilation of the great cerebral vein of Galen is a secondary result of the force of arterial blood either directly from an artery via an arteriovenous fistula or by way of a tributary vein that receives the blood directly from an artery. There is usually a venous anomaly downstream from the draining vein that, together with the high blood flow into the great cerebral vein of Galen causes its dilation. The right sided cardiac chambers and pulmonary arteries also develop mild to severe dilation.

In a patient with a vein of Galen malformation, some of the tiny capillaries that normally distribute oxygen-rich blood throughout the brain are missing. Since those capillaries intercept blood coming in from an artery and slow down the flow of blood back to the heart, when they're missing or malformed the blood rushes into the vein of Galen with too much force and little resistance to flow. This causes a rush of high-pressure blood into the veins. This relative lack of resistance sometimes leads to overworking of the heart, with the potential of heart failure. The increased blood flow into the veins can also disrupt the normal balance between cerebrospinal fluid (CSF) production and absorption, resulting in hydrocephalus. Other complications might include brain dysfunction, with developmental delay, spontaneous bleeding in the brain, or seizures. This extra pressure in the veins can cause a number of problems:

The rush of blood toward the heart and lungs forces the heart to work overtime to get blood to the rest of the body. This can lead to congestive heart failure in some infants.

Blood pressure in the arteries from the heart to the lungs may rise, causing a serious condition called pulmonary hypertension.

The high pressure in the veins can prevent the infant's brain from draining adequately. This can lead to widespread brain injury and sometimes causes severe loss of tissue in the brain.

Some infants can develop hydrocephalus (an enlarged head) if the VOGM blocks the normal flow of fluid in the brain. [4]

Vein of Galen aneurysmal malformations (VGAM) and Vein of Galen aneurysmal dilations (VGAD) are the most frequent arteriovenous malformations in infants and fetuses.[5]

Vein of Galen is important because without the vein of Galen, oxygen-rich blood is unable to flow slowly through capillaries that deliver this blood to surrounding tissues. Since there is also no slowing of the blood flow, it rushes quickly away through the vein, causing a constant rush of blood to the heart and lungs. Now the question is how the VGAM affect the body. The malformation causes oxygen-rich blood to flow directly through this vein away from the brain instead of delivering blood to surrounding brain tissues. This rush of blood away from the brain puts pressure on the heart and lungs, often causing congestive heart failure or pulmonary hypertension.[6]

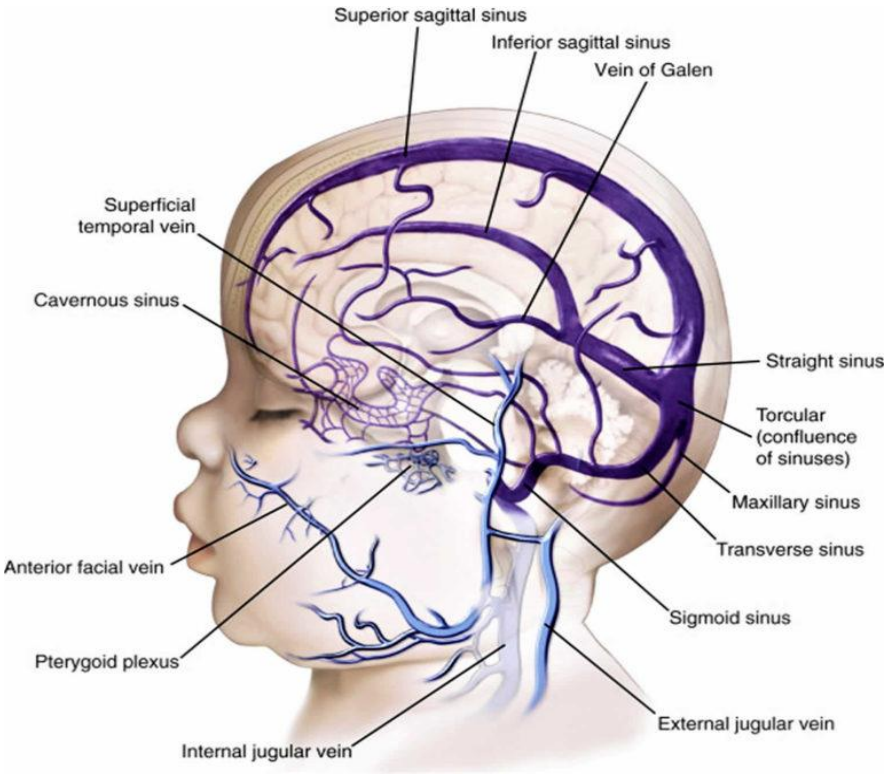

Fig. 1: Vein of galen malformation.

Causes Vein of Galen Malformations[7]

Vein of Galen malformations are not inherited. There is currently no known cause of these malformations, but children with VOGMs are more likely to suffer from other types of arteriovenous malformations (AVMs), or blood vessel malformations.

A vein of Galen malformation develops in a fetus in the first trimester of pregnancy for reasons that are not clear, but it is likely to be genetic in origin. Regardless of the cause, familial transmission or inheritance does not happen. Vein of Galen malformation sometimes, but not always, occurs with vascular anomalies that affect other parts of the body. They generally are not inherited.

Vein of Galen malformation can sometimes be seen on an ultrasound later in the pregnancy, but it's more commonly diagnosed in a newborn or young child.

Diagnosis of VGAM

A definite diagnosis can be made with imaging tests:

A cranial ultrasound is a noninvasive imaging test that allows a neurosurgeon to see the location of an abnormality in the brain.

Computerized tomography (CT) and magnetic resonance imaging (MRI) scans, which are also noninvasive procedures, can produce detailed images of the brain. CT scans use X-rays and MRI scans use magnetic fields and radio-frequency waves to create images of the brain, but

they cannot by themselves see the details of a vein of Galen's structure, which are best seen on an angiogram.

A catheter angiogram (also known as cerebral angiography or cerebral arteriography) is a minimally invasive procedure that can provide a detailed image of blood flow in the brain. To perform an angiogram, a specialist called an interventional radiologist inserts a catheter into a blood vessel and feeds it gently toward the location of the suspected arteriovenous malformation. A special dye called a contrast agent is released through the catheter and makes its way to the malformation. The path of the contrast agent, when viewed on an x-ray, produces a detailed image of the malformation's structure.

An angiogram may be performed along with a CT or MRI scan for greater detail. CT angiography combines the minimally invasive catheter angiogram with the noninvasive CT scan to provide detailed pictures of the malformation. A magnetic resonance angiogram (MRA) is an MRI combined with an angiogram, which allows the radiologist and neurosurgeon to examine the blood vessels as well as the structures of the brain. CT angiograms and magnetic resonance angiograms provide excellent diagnostic views of a vein of Galen malformation.

Treatment of VGM

The goal of treating a vein of Galen malformation is to eliminate or decrease as much of the blood flow through the vein of Galen malformation as possible, while maximizing the blood supply to the brain. Treatment for a vein of Galen malformation depends on the severity of the condition. Some of the treatments may be:

Anti-seizure medications to manage convulsions
A shunt to manage hydrocephalus (see vein of Galen malformation surgery)
Cardiac management of congestive heart failure

The most common treatment is endovascular embolization, a minimally invasive procedure, is available to treat vein of Galen malformation itself, greatly improving the prognosis. For this procedure, the neurosurgeon inserts a catheter (a thin, flexible tube) into an artery in the child's groin through a tiny incision. The surgeon guides the catheter up into the brain and injects a special material (such as a type of glue or soft metal coils) into the blood vessels of the vein of Galen malformation to close off the blood flow.[8]

Most children have no pain or other symptoms with embolization, and most are able to leave the hospital within a few days. Some children will need to spend several days in the ICU for observation.

Case studies

A cross-sectional study of a United Kingdom cohort of neonatal vein of galen malformation shown that Neonates with angiographically confirmed vein of Galen malformation presenting to 1 of 2 UK treatment centers (2006-2016) were included; those surviving were invited to participate in neurocognitive assessment. Results in each domain were dichotomized into "good" and "poor" categories. Cross-sectional and angiographic brain imaging studies were systematically interrogated. Logistic regression was used to explore potential outcome predictors. 85 children with neonatal vein of Galen malformation, 51 had survived. Thirty-four participated in neurocognitive assessment. Outcomes were approximately evenly split between "good" and "poor" categories across all domains, namely, neurological status, general cognition, neuromotor skills, adaptive behavior, and emotional and behavioral development. Important predictors of poor cognitive outcome were initial Bicêtre score ≤ 12 and presence of brain injury, specifically white matter injury, on initial imaging; in multivariate analysis, only Bicêtre score ≤ 12 remained significant. Despite modern supportive and endovascular treatment, more than one-third of unselected newborns with vein of Galen malformation did not survive. Outcome was good in around half of survivors. The importance of white matter injury suggests that abnormalities of venous as well as arterial circulation are important in the pathophysiology of brain injury.[9-11]

A surgery is performed called endovascular embolization, a neurosurgeon inserts a catheter through an artery or vein in the groin and guides it up the circulatory system to the site of the vein of Galen malformation, where it delivers a kind of liquid material (known as "glue") and/or specially designed platinum wires ("coils") that occlude the abnormal connections between arteries and veins and strop abnormal blood flow to the malformed vessel, thus restoring normal circulation. This relatively new technique is a challenging procedure to perform on infants and small children, and it should be performed by a highly trained, experienced expert in cerebrovascular surgery.[12-13]

In rare cases, a neurosurgeon may also place a shunt in the head to drain away blocked or excess cerebrospinal fluid (CSF) to relieve hydrocephalus.[14] The shunt redirects CSF (cerebrospinal fluid) into the abdomen, where it is reabsorbed into the body. The majority of individuals with vein of Galen malformations do not require this type of treatment for their hydrocephalus. It is preferred to treat the malformation first in the hopes of avoiding any need for a shunt.[14-16]

Life expectancy in VGM cases

A vein of Galen malformation is a rare intracranial vascular lesion affecting the pediatric population. Its poor prognosis has been significantly improved with the development of endovascular embolization. Most cases occur during infancy when the mortality rates are at their highest. Vein of Galen malformations are a relatively unknown condition, attributed to the rareness of the malformations. Babies with prenatally diagnosed vein of

Galen malformation have unexpectedly poor outcomes in the presence of cardiac or cerebral anomalies, while those with strictly isolated vein of Galen malformation tend to have more favorable outcomes. Of 21 cases of prenatally diagnosed vein of Galen malformation, 4 (19.0%) cases were isolated and 17 (81.0%) were associated with other anomalies. There were nine terminations (42.9%) and six neonatal deaths (28.6%).[17]

Few studies described the long-term outcomes of children by the time they reached school age with vein of Galen aneurysmal malformation (VGAM). Long-term outcome of patients with VGAM appears to be less favourable than outcome described at the short- and medium-term, even in the absence of encephalomalacia at birth. Even patients with good outcome often have neuropsychological disorders that may have repercussions on learning and requiring appropriate rehabilitation or medical management.

The complications that are usually associated with vein of Galen malformations are usually intracranial hemorrhages. Over half the patients with VGAM have a malformation that cannot be corrected. Patients frequently die in the neonatal period or in early infancy. Vein of Galen malformations are devastating complications. Studies have shown that 77% of untreated cases result in mortality. Even after surgical treatment, the mortality rate remains as high as 39.4%.[18]

Conflict of Interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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