Vein of galen malformation: The role of the radiologist

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Learning objectives

To remind the anatomy, describe and illustrate the spectrum of imaging findings of Vein of Galen Aneurysmal Malformation using the different Imaging technics (US, CT, MR, ...) available.
Background

Vein of Galen aneurysmal malformations (VOGM) are uncommon congenital vascular malformations characterized by multiple arteriovenous connections draining into a enlarged median draining vein, the median prosencephalic vein of Markowski (embryonic precursor of the vein of Galen). Although this is a rare entity, both paediatric and general radiologists should know the differences between the true vein of Galen malformation (VGAM) and vein of Galen aneurysmal dilatations (VGAD), choroidal or mural subtypes of VGAM and the specific anatomical and radiological characteristics of it. All of these aspects have very important clinical and prognosis implications.

Vein of Galen aneurysmal malformation (VOGM) is a rare congenital intracranial vascular malformation predominantly diagnosed at the pediatric age (1% of all and 30% of pediatric vascular malformations). Depending on the degree of arteriovenous shunting, the condition is often associated with life-threatening congestive heart failure in the first weeks of life [Figure 1] on page . Later in childhood patients typically present with macrocephaly, hydrocephalus and development delay or focal neurological deficits.

EMBRIOLOGY AND ANATOMY

The vein of Galen is the largest of the deep cerebral veins formed posterior to the pineal body by confluence of the internal cerebral veins and the basal veins, so it drains the deep medially located veins of the diencephalon, basal ganglia, and midbrain. [Figure 2] on page . A damage during the development of this part of cerebral vasculature (between sixth and eleventh gestation week) is thought to be the cause of VGOM.

The true VGAM is often identified immediately after birth and it is usually drained through an embryonic vein. This temporary vein is the median prosencephalic vein of Markowski, and it drains the choroid plexus to a dorsal dural plexus (interhemispheric) called falcine sinus. The development of the basal ganglia and the choroid plexus (between the sixth and the eleventh gestation week) makes this vein return and lead the development of the vein of Galen and other deep brain veins like permanent internal cerebral veins.

If an abnormal arteriovenous fistula formation between embryologic choroidal arteries and the median prosencephalic vein occurs, the persistently elevated blood flow prevents from normal involution of this embryonic vein and precludes the development of the vein of Galen. If this vein doesn't regress normally, a fistulous connection with the primitive choroidal arteries will persist.
According to the Berenstein and Lasjaunias classification, two groups of vein of Galen aneurysmal malformations have been proposed: 1. True vein of Galen aneurysmal malformations (VGAM); 2. Vein of Galen aneurysmal dilatations (VGAD), in which a intraparenchimatous arterio-venous malformation drains trough the already developed vein of Galen, that drains the normal brain parenchyma too.

Two different angioarchitectural variants must be distinguished: a choroidal variant, the most common, composed of numerous feeding arteries (choroidal, pericallosal, and thalamo-perforating vessels) joining the dilated midline vein; and a mural variant, composed of a single or a few arterial feeders which join the dilated vein near a single location. [Figure 3] on page...
In spite of the fact that the number of cases of VGAM diagnosed with prenatal US or MR are increasing, these are techniques not commonly widespread and the most usual form in which radiologist identify this entity is during the neonatal period. So the initial diagnosis is often made with ultrasound, CT, or MRI, more typically with US. Obviously the radiologist must be acquainted with the spectrum of findings of this entity and its complications in the moment of the diagnose, and with the findings after treatment in the follow-up too, using the modality of imaging technique more suitable in which case.

A compilation of the principal findings that every radiologist should know, using the different imaging modalities available, are detailed below:

---**ULTRASOUND (US):**

Cranial ultrasonography is an essential part of the routine standard of care in high-risk neonates, so it is usually the first imaging modality in patients with suspected VGAM.

We routinely perform cranial ultrasonography through the anterior fontanel with 5-8 MHz convex and 5-12 MHz linear array transducers. If settings are optimized for neonatal brain imaging, this provides an excellent view of most supratentorial structures, but visualization of structures located far away from the transducer, such as the cerebellum, may also be less optimal. So the use of supplemental acoustic windows, such as the mastoid fontanels, the posterior fontanel and the temporal windows, can enable a better view of structures located further away from the anterior fontanel and a better visualization of the posterior fossa, brainstem, and the occipital regions of the brain.

It is a well-known fact that major advantages of cranial ultrasonography are that it is safe, relatively inexpensive, and can be performed at the bedside, with little disturbance to the infant. Other important advantage is that it can be initiated at an early stage and repeated as often as necessary, so we can do a very close follow-up of these patients and the complications of this disorder. Although in our experience it is possible to do an additional and more sensitive imaging technique like CT or MR, despite the radiation risk in order to avoid overlook serious complications.
-B-mode US: vein of Galen malformation is usually detected as anechoic, tubular midline structure located superior to the cerebellum or an hypoechoic midline mass indenting the posterior aspect of the third ventricle, in the quadrigeminal plate cistern. [Figure 4] on page ... Hypoechoic tubular structures near the choroid plexus can be visualized too.

-Color Doppler US: color Doppler images demonstrate turbulent flow in the anechoic mass and the tubular structures. The feeding arteries are difficult to analyze, but a tortuous network of dilated arteries is usually visible in the region of the malformation [Figure 5] on page ... Power Doppler sonography can contribute to best characterize the lesion, it gives better definition of vessel courses and identification of smaller and deeper vessels, but it does not provide information on flow direction.

Although the advantages of cranial US are numerous, there are some limitations. So we must always do an additional and more sensitive and specific imaging technique in order to best characterize the lesion.

-->COMPUTED TOMOGRAPHY (CT):

CT scanning parameters protocol of the children's head in our centre is a multi-slice helical low dose scan, which allows posterior multiplanar and MIP reconstructions.

The radiation dosage is the main disadvantage of CT scans, but in this kind of patients the assessment of the risks and benefits of each study have been worth it.

CT scan is the most manageable technique in severe ill patients and the easiest available technique in the emergency centre of most hospitals. For this reason, despite of it is not the best technique to properly define the lesions in this kind of children, it may be the most widespread technique in order to rule out serious complications associated to this entity in the emergency.

CT scan is a good technique to evaluate postsurgical and post-embolization VGM too. MRI and MR angiography are the best primary follow-up tools for these patients, but in many cases a quick evaluation is necessary after the intervention and it is easier with CT.

-Non-enhanced CT: typically a venous pouch mildly hyperdense to brain is visualized [Figure 6a] on page ... MIP reconstruction algorithm can help to define the lesion or to get better images [Figure 6b] on page ... The lesion can have a calcification wall (not frequently) in older children, or if the aneurysm is thrombosed. If we do a contrast-
enhanced CT, we'll be able to see strong enhancement of feeding arteries and veins, but it is not necessary routinely.

Other common findings on CT studies are hydrocephalus, parenchymal calcifications, edema and atrophy in different degrees. We can also see ischemic lesions like low attenuation images and hemorrhagic lesions, intraventricular and parenchimatous, which are described after embolization and surgery too. [Figures 7] on page [Figure 8] on page .

We also carry out CT scans to evaluate changes soon after surgery, like parenchymatous hemorrhagic lesions; or after other procedures like intracranial-pressure sensor or hydrocephalus valves placing [Figure 9] on page .

- **CT angiography**: it gives an excellent delineation of VGM, it is a good adjunct for understanding the anatomic relationships of the vessels previous to angiography but VGM is not a very widespread indication because of the use of contrast agents and the elevated radiation dosage, so for now MR and MR angiography are preferred previous to angiography. In cases of newborn with unstable condition in which it may be difficult to perform a MR, CT angiography is a very good option to describe the lesion previous embolization.

**--->MAGNETIC RESONANCE (MR) AND MR ANGIOGRAPHY AND VENOGRAPHY (MRA/MRV):**

MRI and MRA/MRV are the best non-invasive techniques to describe and evaluate the anatomy of this lesion. US and CT can help us to approach the diagnose, but they can not describe with precision the characteristics of this lesion. MR allows a best characterization and classification in its accurate subtype. Pretreatment evaluation of a VGAM morphology should in our opinion include MR images, high-resolution MR imaging with gadolinium enhancement and MR angiography/venography focusing on the characterization of the abnormal vessels and connections of the lesion and its relationship with the normal cerebral venous drainage too.

MR imaging of the neonatal brain includes in our centre the following sequences: sagittal and axial T1WI, axial and coronal T2WI, axial Flair and gradient-echo and diffusion B800, B1500 and B3000.

- **T1 weighted imaging**: the midline venous pouch and the arterial feeders look like flow voids or mixed intensity due to fast or turbulent flow. We can see hyperintense foci within
the pouch that correspond with thrombus, and hyperintense foci within the parenchyma correspond with calcifications or ischemic lesions. [Figure 10] on page

-T2 weighted imaging: the midline venous pouch and the arterial feeders have the same appearance that in T1WI, but we can see them more accuracy because of the contrast with hyperintense CSF. [Figure 11 a] on page [Figure 11b] on page

-Diffusion weighted imaging: this fast sequence that we do routinely in all neonatal MR, can show restriction in areas of acute ischemia /infarction, so it is a more precise technique for this than CT.

-MR angiography and venography:

The protocol we use in our centre to evaluate the malformation and the venous sinuses includes 2D Time-of-flight sequence (2DTOF), and a volumetric 3D T1 sequence with intravenous gadolinium (3DT1-c).

Using these sequences we can depict with a lot of precision the specific characteristics of the VGAM, and classify according to the angioarchitectural variants that have been described:

• Choroidal: multiple feeders from pericallosal, choroidal and thalamoperforating arteries. [Figure 12] on page [Figure 13] on page [Figure 14] on page [Figure 15] on page

• Mural: few feeders from uni or bilateral collicular or posterior choroidal arteries.

It is very common to find dural venous sinus anomalies, like:

-Persistence of embryonic falcine sinus drains midline venous pouch associated with absent straight sinus. [Figures 12] on page [Figure 14] on page

- Variable absence and stenosis of other sinuses.

Both the MR and MRA/MRV obtain very accurate information and well-delineated anatomy of arterial feeders in the malformation and the venous pouch drainage, and it is an excellent pre-embolization or pre-surgery study to planning these posterior procedures.

-DIGITAL SUBSTRATION ANGIOGRAPHY:
Although this fluoroscopy technique is used by interventional radiologists and it is not a field for general radiologists, we must know that it is the gold standard technique in the diagnose of VGM. It provides the biggest amount of precise information about this entity and it is the best therapeutic technique too, so ideally it should be performed with first embolization (about six months of age of later). [Figure 16] on page [Figure 17] on page

TREATMENT

Treatment is not the purpose of this paper, so only a few words about it.

The universally poor natural history of untreated VGAM at all ages and limited benefits of surgical treatment mean that current treatment relies primarily on endovascular approaches. We can distinguish two groups:

- Neonatal presentation (choroidal subtype): medical therapy for heart failure until 5-6 months of age, although failure warrants early neuro-intervention (bad prognosis); transcatheter at 5-6 months only in absence of intractable cardiac failure, multi-system organ failure and brain damage (arterial embolization best than venous).

- Infant presentation (mural subtype): transcatheter embolization can be performed later and it is technically easier.

Treatment for hydrocephalus is controversial, because shunt placement can exacerbate brain damage and increase risk of intraventricular hemorrhage.

Nevertheless, optimal management requires adjunctive intensive care and pediatric neurology input to ensure the best outcome. Several studies have reported that approximately 60% of patients of all ages treated endovascularly had minimal to no developmental delay at follow-up. However, prognosis differs by age. Patients who present early in life with cardiac failure are particularly difficult to treat.
Conclusion

Knowledge of VOGM anatomy and its spectrum of imaging findings is important to both general and pediatrics radiologists, for doing precise diagnostics because of its notable clinical and prognostic implications.
References


