

Vein Of Galen Malformation, Case Report Anugrah Onie w

BACKGROUND

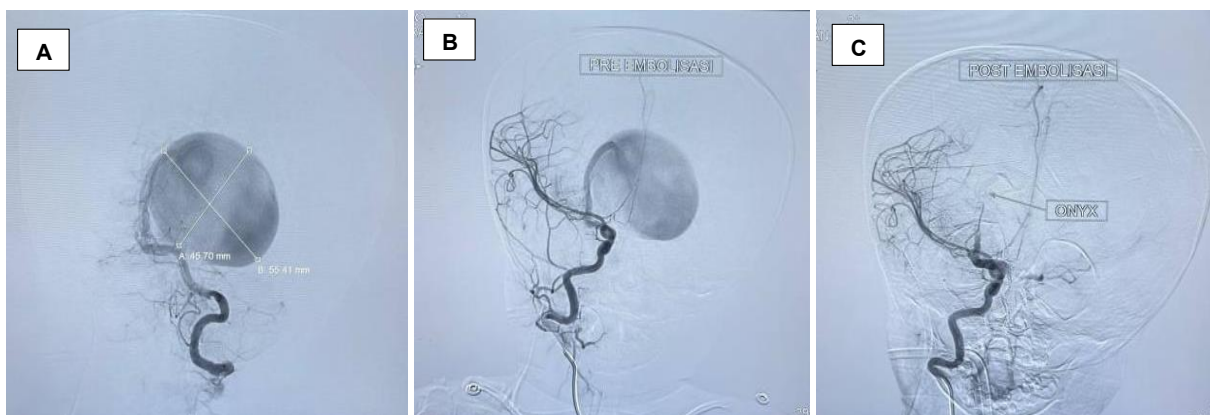
Venous Galen malformation is a rare congenital malformation caused by impaired development of the embryonic precursor vein, namely Markowski's median prosencephalic vein. The incidence of Galen vein malformation is about one in three million population, representing 1% of all arteriovenous malformations in the brain.¹ Rapid and precise diagnosis is very crucial in this case with endovascular embolization as the standard therapy of choice for Galen vein malformation.^{2,3} Regardless of age, the long-term goal in the management of venous Galen malformation is complete closure of the lesion in order to promote normal neurological development without deficits in the patient.^{4,5,6} For infants and children, the short-term goal of intervention in venous Galen malformation is to restore balance. normal hydrovenosa to allow for normal development



Feature 1. MRI Showed macrocephali, dilatation of lateral ventricle and mass effect at the galen vein

Case Report

A 11 months old boy was referred to the neurosurgery department of RSUP Dr. Moh. Hoesin with recurrent general seizures accompanied by loss of consciousness shortly after the seizure, limb weakness and developmental delay, no fever. General physical examination of the head reveals macrocephaly with limb motor strength unable to resist vigorous movement. Following these findings, the neurosurgery department decided to perform digital subtraction angiography (DSA) to confirm the diagnosis of Galen's vein malformation along with embolization.



Feature 2. Angiogram A and B before the embolisation, and C After Embolisation

Results

Angiographic examination revealed that the right internal carotid artery and vertebrobasilar artery supplying the fistula originate from the posterior communicating artery, emphasizing that there is one feeder artery draining the right

posterior communicating artery. The lesion is then treated with two embolic materials: coil and ONYX. Angiography confirmed total exclusion of fistula and circulation in Galen's vein.

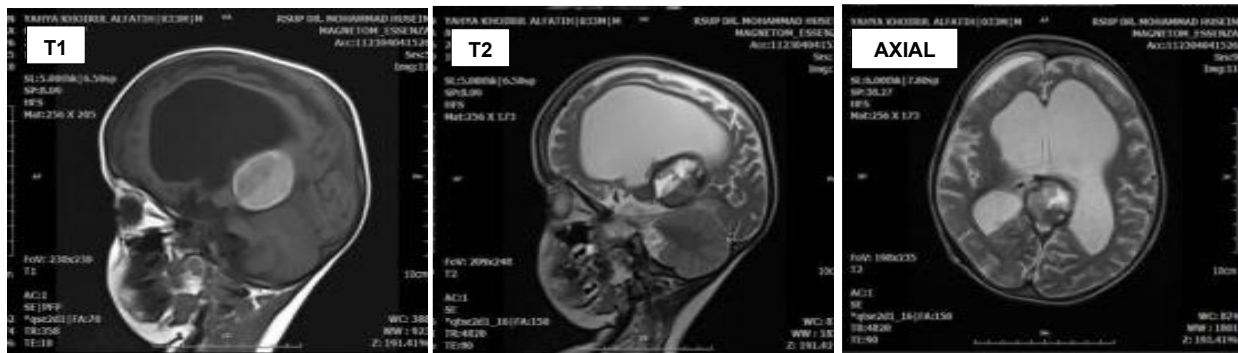


Figure 3. MRI Follow Up 6 months after treatment, showed thrombosis of the vein of Galen, decreased in size.

DISCUSSION

VGA is a vascular abnormality typically found in children⁷. Raybaud et al.⁸ believe it is reminiscent of fetal anatomy produced by frequent occlusions of posterior fossa dural sinus, especially at the sigmoid sinus. Even though the VGA can be asymptomatic in adults^{9,10}, it is typically diagnosed during the neonatal period or in childhood with heart failure signs, macrocephaly and/or cranial murmurs¹¹. Children with slow flow fistulas have a better extra uterine adaptation¹². Adults usually present slow flow fistulas, headaches, seizures, hydrocephalus and rounded calcified masses in the pineal region¹¹. Subarachnoid and intracerebral hemorrhages can occur because of blood flow reorganization to pial veins¹³.

There are many classifications for vein of Galen malformations. The two most used are those proposed by Yasargil¹³ and Lasjaunias et al.¹⁴. Yasargil's lesions types 1, 2 and 3 are direct fistulas between the malformations and the vein of Galen. Lesion type 4 are parenchymal arteriovenous malformations, which drain directly into the vein of Galen. According to Lasjaunias et al.¹⁴, these malformations can be divided into mural and coroidal types depending on the fistula localization. The coroidal type is characterized by multiple fistulas at the anterior and terminal segment of the median prosencephalic vein. This type usually presents at the neonatal period causing serious heart failure unfolding to multiple organ failure and death. The mural type has the fistula at the vascular wall usually at the lateral-inferior wall of the median prosencephalic vein. They are commonly slow flow and asymptomatic⁹. VGA differ from Galen dilatation, which results from an obstruction of the normal vein of Galen. This alteration is mentioned as a dilatation of the persistent median prosencephalic vein, also known as the Markowski vein¹¹.

Cerebral angiography is the gold standard for the diagnosis of VGA. The exam shows the dynamic aspect of the cerebral venous system and vascular relationships to the fistula^{12,13}.

Successful transarterial endovascular treatment for Vein Of Galen Malformation already presented, and improvement of the development during 6 months follow up was reported.

References

1. Puvabanditsin S, Mehta R, Palomares K, Gengel N, Da Silva CF, Roychowdhury S, Gupta G, Kashyap A, Sorrentino D. Vein of Galen malformation in a neonate: A case report and review of endovascular management. *World J Clin Pediatr.* 2017 Feb 8;6(1):103-109.

2. Brinjikji W, Krings T, Murad MH, Rouchaud A, Meila D. Endovascular Treatment of Vein of Galen Malformations: A Systematic Review and Meta-Analysis. *AJNR Am J Neuroradiol.* 2017 Dec;38(12):2308-2314.
3. Mohan R, Nayyar R, Ryder L, Lord D, Athayde N. Vein of Galen aneurysm. *Australas J Ultrasound Med.* 2016 May 20;19(2):75-77.
4. Savage C, Hale AT, Parr MS, et al. Outcomes of endovascular embolization for Vein of Galen malformations: An individual participant data meta-analysis. *Front Pediatr.* 2022.
5. Bhatia K, Mendes Pereira V, Krings T, et al. Factors Contributing to Major Neurological Complications from Vein of Galen Malformation Embolization. *JAMA Neurol.* 2020; 77(8): 992-999
6. Agrawal A, Britz G. *Pediatric Vascular Neurosurgery Part I: Principles and Practice of Neurovascular Disorders.* Springer: New York. 2016; 137-42
7. Paumier A, Winer N, Joubert M, et al. Galen vein aneurysm: review of the literature and report of two cases. *J Gynecol Obstet Biol Reprod (Paris)* 1998;27:814-820.
8. Raybaud CA, Strother CM, Hald JK. Aneurysms of the vein of Galen: embryonic considerations and anatomical features relating to the pathogenesis of the malformation. *Neuroradiology* 1989;31:109-128.
9. Rosenfeld JV, Fabinyi GC. Acute hydrocephalus in an elderly woman with an aneurysm of the vein of Galen. *Neurosurgery* 1984;15:852-854.
10. Mylonas C, Booth AE. Vein of Galen aneurysm presenting in middle age. *Br J Neurosurg* 1992;6:491-494.
11. Gupta AK, Varma DR. Vein of Galen malformations: review. *Neurology (India)* 2004;52:43-53.
12. Sung KB, Chang DI, Kim JH, Kim MH, Hahm CK, Jeon SC. A case of aneurysm of the vein of Galen. *J Trauma* 1994;36:565-567.
13. Yasargil MG. *Microneurosurgery IIIB.* New York: Thieme Medical Publishers, 1988: 323-357.
14. Lasjaunias P, Ter Brugge K, Ibor LL, et al. The role of dural anomalies in vein of Galen aneurysms: report of six cases and review of the literature. *AJNR* 1987;8:185-192.