Vein of Galen Aneurysmal Malformation in Children: Challenges of Imaging in An African Setting

Godwin Inalegwu Ogbole, Amos Olufemi Adeleye[1], Richard Busayo Olatunji

Department of Radiology, University College Hospital, Ibadan, [1]Department of Surgery, Division of Neurosurgery, College of Medicine, University of Ibadan, Ibadan, Nigeria

Correspondence: Dr. Godwin Inalegwu Ogbole, Department of Radiology, University College Hospital, Ibadan, PMB 5116, Nigeria. E-mail: gogbole@yahoo.com

ABSTRACT

Aneurysmal malformations of the vein of Galen (VGAM) are a rare congenital intracranial vascular disorder. It is known to occur in two main forms. We present two children with classic demonstrations of broadly recognized categories. It typically results in high-output congestive heart failure or may present with developmental delay, hydrocephalus and seizures. Our aim is to demonstrate the challenges of imaging and management in an African setting of a rare congenital intracranial vascular disorder presenting in older African children with infrequently recognized associations using limited imaging techniques. Computed tomography (CT) showed extensive gyriform hyperdensities at the grey-white matter junction, dilatation of a portion of the superior sagittal sinus with erosion of the inner table of the calvarium, multiple abnormal tortuous vessels in the suprasellar region, and a markedly dilated vein of Galen and ventricular system. Magnetic resonance imaging (MRI) showed a tortuous dilated pontomesencephalic vein in the quadrigeminal cistern, dilated vein of Galen and straight sinus. A network of vessels in the quadrigeminal cistern draining into dilated vein of Galen thinning of the corpus callosum is seen. Chest radiograph and electrocardiographic chart showed evidence of cardiomegaly and cardiac failure in one patient. We review the literature and discuss the challenges of imaging and management in our African setting. The VGAM is a recognized rare anomaly, however, knowing the true prevalence and improving its management in our African environment is limited due to the challenges of available imaging technology, lack of personnel in interventional neuroradiology, and inaccessibility to care of a large poor population.

Key words: Africa; aneurysm; arteriovenous malformation; children; vein of Galen malformation

Introduction

The vein of Galen is a midline venous structure in the quadrigeminal cistern that drains the internal cerebral veins, the basal veins of Rosenthal, and some posterior fossa veins into the straight sinus. Vein of Galen dilatation is a rare congenital intracranial vascular disorder.[1] It is known to occur in two main forms, either as a true aneurysmal malformation or as a variceal dilatation secondary to a cerebral parenchymal arteriovenous malformation (AVM) upstream.[2] Aneurysmal dilatation of the vein of Galen constitute 1% of intracranial vascular abnormalities and 30% of all pediatric vascular malformations with an incidence of <1/25000 deliveries.[3] Steinheil in 1895 made the first reference to it as a varix aneurysm but the vein of Galen aneurysm was first described by Jaeger *et al*., in 1937 and about 250 cases have been reported since then.[4,5] However, majority of the reported cases have been of neonates and infants. Reports in older children and adults are few and far in between globally.[6-8] Documented cases in Africans are even rarer still, probably due to the fact that the required diagnostic imaging technology (computed tomography (CT), magnetic resonance imaging (MRI), and angiography), as well as competent personnel in neuroradiologic diagnosis, is up till now not widely available in this region. We present the clinical and radiological findings of two children who presented 6 months apart at a tertiary hospital in Nigeria.

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Case Reports

Case 1

A 5-year-old boy presented with 16-month history of recurrent afebrile generalized tonic-clonic seizures, right

sided weakness, slurred speech, and poor school performance. He had spastic quadriparesis worse on the right with bilateral extensor plantar responses.

The evaluation of a contrast enhanced cranial CT [Figure 1] revealed, extensive areas of mildly enhancing gyriform and parenchymal hyperdensities (HU 116) consistent with calcifications in the grey-white matter junction of all the lobes of both cerebral hemispheres. Similar but punctate hyperdensities were also seen in both basal ganglia and the brainstem. The CT examination also showed extensive abnormal multiple tortuous vessels at the base of the skull around the midbrain, in the suprasellar cistern and extending within the cerebral and cerebellar hemispheres with marked distortion of the normal anatomy of the vessels of the circle of Willis. The vein of Galen, straight sinus, superior sagittal sinus, and the transverse sinuses were all markedly dilated with associated interrupted rim calcification of their walls. The torcula herophili was similarly dilated and ballooned with resultant pressure erosion of the adjacent inner table of the occipital bone. There was moderate dilatation of all components of the ventricular system, except the 4th ventricle, which appeared normal in size, consistent with a noncommunicating obstructive hydrocephalus.

The diagnosis of an extensive AVM involving the vessels of the circle of Willis and the vein of Galen was entertained. The possibility of congenital Herpes simplex type 2 or Cytomegalovirus infection was also considered in view of the widespread intracranial calcifications. A reasonable control of patient's seizures was achieved with Carbamazepine with remarkable improvement in overall neurological status. The required follow up imaging with CT angiographic evaluation and possible referral for endovascular treatment were hampered by parent's financial constraints as the hospital operates an out-of-pocket payment system. The patient's parents consequently received counseling and the child is being managed conservatively at the out-patient clinic.

Case 2

An 8-year-old boy presented with 7 months history of recurrent headaches that were worse in the morning and associated with blurring of vision. The headaches were preceded by a 1-week history of irrational speech and behavior. On physical examination, he had macrocephaly with an occipitofrontal circumference (OFC) of 62 cm (normal for age ≈ 50-54 cm) and bifrontal bossing of the skull. He also had anisocoria, left strabismus, right hemiparesis, and dysdiadokokinesia. Fundoscopy revealed bilateral papilloedema. He had tachycardia, bounding peripheral pulses, a third heart sound, and left basal coarse crepitations in the chest.

A cranial MRI revealed a network of signal void vessels in the quadrigeminal cistern inferior to the splenium of the corpus callosum at the base of the third ventricle. The

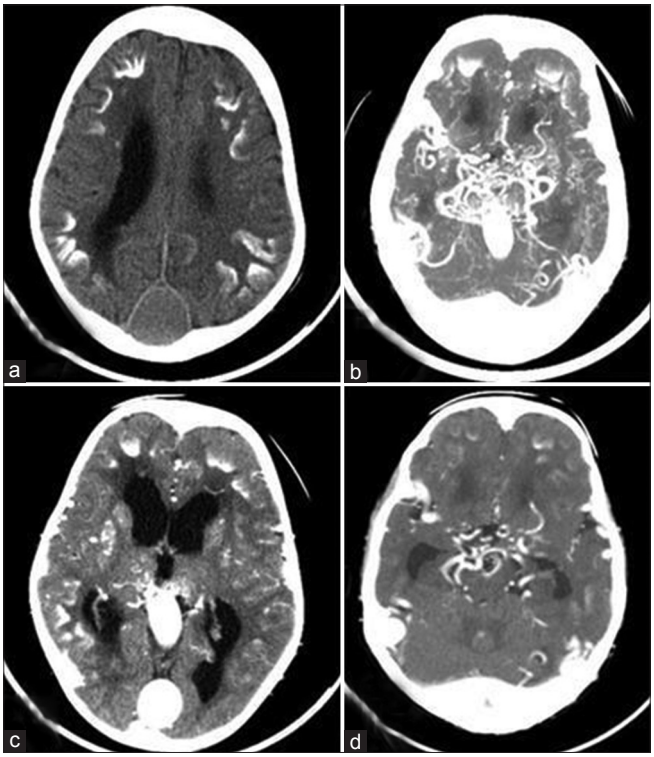

Figure 1: (a) NCCT showing extensive gyriform hyperdensities at the grey-white matter junction in keeping with calcifications. Isodense dilatation of a portion of the superior sagittal sinus with erosion of the inner table of the calvarium is demonstrated. (b) CECT showing multiple abnormal tortuous vessels in the suprasellar region. A markedly dilated vein of Galen is located posteriorly. (c) CECT shows early filling and aneurysmal dilatation of the vein of Galen and the torcula herophili. The ventricular systems are also dilated. (d) CECT shows peripheral location of multiple abnormal tortuous dilated veins

vein of Galen was ballooned with marked dilatation of the straight sinus, torcula herophili, and the transverse sinuses. The pontomesencephalic veins were similarly dilated but to a lesser extent. All components of the ventricular systems, except the fourth ventricle, were moderately dilated in keeping with noncommunicating obstructive hydrocephalus. The splenium of the corpus callosum appeared thinned out [Figure 2]. Based on the MRI findings a diagnosis of an AVM involving the posterior choroidal vessels and the vein of Galen was made. The patient was scheduled to have a further work up with a Computed Tomographic Angiography (CTA) but this was not done due to the breakdown of the hospital's CT facility at the time of this report. Patient's chest radiograph revealed an enlarged heart with an engorged pulmonary vasculature consistent with congestive cardiac failure [Figure 3]. The electrocardiographic chart showed voltage criteria in keeping with left ventricular hypertrophy [Figure 4]. The parents of this child also received counseling and the child is being followed up with conservative management. At the time of this report, the child has remained stable.

Discussion

Dilatation of the vein of Galen is a long recognized type of cerebral AVM. The most appropriate terminology for the vein of Galen anomaly has long been the center of considerable debate regarding the correct use of the term

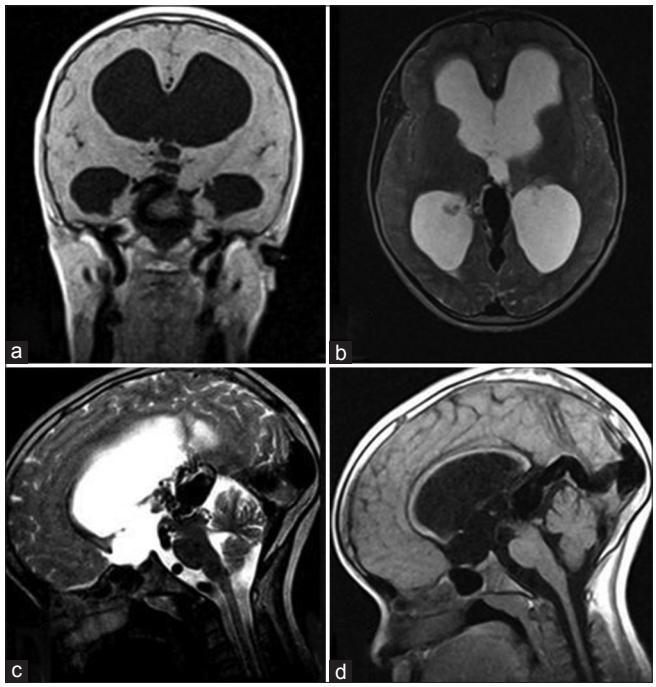

Figure 2: (a) Coronal T2W MRI shows a tortuous dilated pontomesencephalic vein in the quadrigeminal cistern. (b) Axial T2W image showing dilated vein of Galen and straight sinus. The third ventricle is also dilated. (c and d) Sagittal T2 and T1W MRI shows dilated torcula herophili and a signal void ball of vessels in the quadrigeminal cistern draining into dilated signal void vessels including vein of Galen, straight sinus, and torcula herophili. A dilated lateral and third ventricles as well as thinned splenium of the corpus callosum are demonstrated

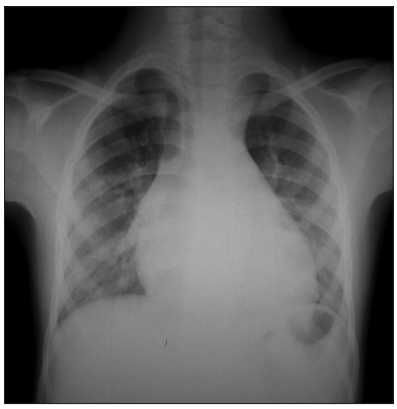

Figure 3: Chest radiograph showing cardiomegaly and prominent pulmonary vasculature markings suggestive of cardiac failure

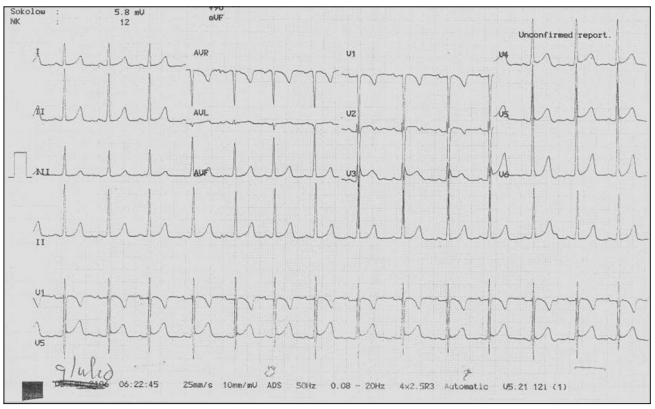

Figure 4: Electrocardiographic chart showing voltage criteria indicative of left ventricular hypertrophy

'vein of Galen aneurysmal malformation (VGAM). Various schools of thought backed by embryological correlation appear in support of the use of VGAM to describe those entities that have persistence of the primitive venous system and a characteristic absence of a true vein of Galen as their hallmark.[9] These have in addition abnormal venous channels such as falcine and occipital sinuses as prominent drainage pathways.[5,10] Vein of Galen aneurysmal dilatation is thus appropriate when the vein of Galen, though dilated, is in its normal anatomical position. This sort of variceal dilatation invariably arises from the increased venous drainage from AVM upstream and normal drainage from the brain parenchyma.[11-13]

The patients we have presented distinctly fall into the class of vein of Galen aneurysmal dilatation, which some have grouped as a subset of VGAM.[12] These cases are archetypal of cerebral AVMs with drainage mainly into the vein of Galen. The rarity ascribed to VGAM may be whittled down when Vein of Galen Aneurysmal Dilatations (VGADs) are considered as a separate entity. Also, typical presentation of VGAMs is in the neonatal life and early childhood with rarer presentation in later life.[9] This stems from the usual severe and early symptom-producing nature of the accompanying cardiovascular compromise associated with the VGAMs.

In contrast, VGADs may well survive into later childhood and adulthood owing to fair compatibility with relatively normal life. Only one of the two patients presented showed cardiovascular compromise. The clinical features in the second case conform to the well documented pattern of presentation in later childhood. The first patient had heavy cerebral parenchymal calcifications, which were presumed to have been due to the fairly familiar congenital infections by the TORCHES group in Nigeria and other parts of Africa.[14-16] While this is a logical angle to interpret the finding given the background relative endemicity of TORCHES congenital infection in sub-Sahara Africa,[17] Lasjuania *et al*., and Jayakumar *et al*., posited that parenchymal calcifications are consequences of cerebral damage resulting from the diffuse chronic hypoxic changes due to chronic elevation of the cerebral venous pressure.

Their documented predominance in the grey-white matter region[18,19] is further confirmed and beautifully demonstrated in the first patient. Chronic elevation of cerebral venous pressure is also supposed to account for the hydrocephalus seen in these patients[20,21] though mechanical obstruction at the aqueduct of Sylvius may also be contributory as seen in one of our patients. Peripheral calcification of the dilated vascular channels seen on the precontrast axial CT images may represent calcified mural thrombi.[22] This may be the pathological basis for previously documented episodes of spontaneous thrombosis in sufferers. Although Chapman *et al*., did report that rim calcifications as seen are common in patients with thrombosis;[23] follow up of these patients will be needed to validate the observed poor predictive value of rim calcification as a marker for subsequent thrombosis.

Nonetheless, the documentation of calcification at 5 years of age in one of our two patients may challenge their (Chapman *et al*.'s) earlier claim that it is rare before the age of 15 years.[23]

The other well documented plausible explanation for these rim calcifications is chronic endothelial injury resulting in thrombophlebitis from the high blood flow in the dilated veins.[24,25] Absent intracranial hemorrhage in our cases is also a pertinent negative finding. Parenchymal and subarachnoid hemorrhages are known complications of both forms of Galenic vein abnormality and their occurrence could have worsened the prognosis.

Further studies will be needed to determine if the low frequency of hemorrhagic complications reported in literature is not due to the rapid fatality that may likely attend bleeding from high flow channels in a location as vital as the brainstem where these anomalies occur predominantly. Inability to perform detailed angiographic study casts a pall on the classicality of the imaging findings, albeit, sufficient insight was derived from the reported findings to guide the clinical management of the patients.

This is a vital improvement for us in this clime. It is hoped that sufficient technological infrastructure and skill will soon become more available to us enabling our ability to truly determine the prevalence of vein of Galen and sundry other vascular anomalies in our practice. It is also hoped that the required personnel and facilities for interventional neuroradiology would soon be available for the definitive treatment of these anomalies.[4] The images presented in the article are sufficient for fairly conclusive diagnosis and scheduling for therapy.

Nevertheless neonatal cranial ultrasound if performed in any neonate with cardiomegaly of unrecognized cardiac origin may assist in the selection of patients that may benefit from further neuroimaging as ultrasound is certainly cheaper and less invasive and has been shown to be invaluable in low income settings in experienced hands.[26]

The cases presented were unfortunately not treated due to lack of expertise and facility for noninvasive vascular neurointervention. These cases highlight some of the challenges of radiology practice in a third world setting. The increasing availability of neuroimaging modalities in Nigeria has improved the capacity to identify brain abnormalities that hitherto presented significant diagnostic challenges in the past. There is what appears to be a distressing delay in developing and increasing the capacity for neurointerventional radiology practice in the West African sub-region. The present state of radiology training in Nigeria has greatly improved but appears to still be majorly focused on general diagnostic radiology.[27] This trend is also seen

in many other African countries. The need for subspecialty training cannot be overemphasized in order to effectively address the management of neurovascular lesions with minimal complications, which is the current global practice. The neuroradiologists and interventionists in the developed world need to assist those in the developing world to build their capacity and bridge the knowledge and technology gap, which continually affect the quality of care available to African populations.

Conclusion

The VGAM is a recognized rare anomaly, however, knowing the true prevalence and improving its management in our African environment is limited due to the challenges of limited available imaging technology, lack of personnel in interventional neuroradiology, and inaccessibility to specialized healthcare of our largely impoverished patient population. It is hoped that in the near future, the necessary facilities and expertise will be readily available at affordable cost for the majority of patients.

References

1. Nicholson AA, Hourihan MD, Hayward C. Arteriovenous malformations involving vein of Galen. Arch Dis Child Fetal Neonatal Ed 1989;64:1653-5.
2. Campi A, Rodesch G, Scotti G, Lasjaunias P. Aneurysmal malformation of the vein of Galen in three patients: Clinical and radiological follow up. Neuroradiology 1998;40:816-21.
3. Casasco A, Lylyk P, Hodes JE, Kohan G, Aymard A, Merland JJ. Percutaneous transvenous catheterization and embolization of vein of Galen aneurysms. Neurosurgery 1991;28:260-6.
4. Ciricillo SF, Edwards MS, Schmidt KG, Hieshima GB, Silverman NH, Higashida RT, *et al*. Interventional neuroradiological management of vein of Galen malformations in the neonate. Neurosurgery 1990;27:22-8.
5. Gupta AK, Varma DR. Vein of Galen malformations: Review. Neurol India 2004;52:43-53.
6. Amacher AL, Shillito J Jr. The syndromes and surgical treatment of aneurysms of the great vein of Galen. J Neurosurg 1973;39:89-98.
7. Carson LV, Brooks BS, El Gammal T, Massey CE, Beveridge WD, Allen MB. Adult arteriovenous malformation of the vein of Galen: A case report with pre- and postoperative computed tomographic findings. Neurosurgery 1980;7:495-8.
8. Rosenfeld JV, Fabinyi GC. Acute hydrocephalus in an elderly woman with an aneurysm of the vein of Galen. Neurosurgery 1984;15:852-4.
9. Jones BV, Ball WS, Tomsick TA, Millard J, Crone KR. Vein of Galen aneurysmal malformation: Diagnosis and treatment of 13 children with extended clinical follow-up. AJNR Am J Neuroradiol 2002;23:1717-24.
10. 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;31:109-28.
11. Berensten A, Lasjaunias P. Arteriovenous fistulas in the brain. Surgical Neuroangiography 4. Endovascular treatment of cerebral lesion. Berlin: Springer-Verlag; 1992. p. 267-317.
12. Yarsagil MG. Microneurosurgery IIIB. New York: Thieme Medical Publishers; 1998. p. 323-57.
13. Garcia-Monaco R, Lasjaunias P, Berenstein A. Therapeutic management of vein of Galen aneurysmal malformations. In: Vinuela F, Halbach VV, Dion JE, editors. Interventional Neuroradiology: Endovascular Therapy of the CNS. New York: Raven Press; 1992. p. 113-27.
14. Olusi T, Gross U, Ajayi J. High incidence of toxoplasmosis during pregnancy in Nigeria. Scand J Infect Dis 1996;28:645-6.
15. Jellife DB. Congenital toxoplasmosis in an African child. Arch Dis Child 1951;26:258.
16. Okwori AE, Olabode AO, Emumwen EG, Lugos MD, Okpe ES, Okopi JA, *et al*. Sero-epidemiological survey of human cytomegalo virus infection among expectant mothers in Bida, Nigeria. Internet J Infect Dis 2008;6.
17. Cutts FT, Robertson SE, Diaz-Ortega JL Samuel R. Control of rubella and congenital rubella syndrome (CRS) in developing countries, part 1: Burden of disease from CRS. Bull World Health Organ 1997;75:55-68.
18. Lasjaunias P, Garcia-Monaco R, Rodesch G, Ter Brugge K, Zerah M, Tardeiu M, *et al*. Vein of Galen malformation: Endovascular management of 43 cases. Childs Nerv Syst 1991;7:360-7.
19. Jayakumar PN, Sathish Chandra P. Cerebral parenchymal calcifications in a child with vein of Galen malformation-role of medullary veins. Neurol India 1997;45:194-76.
20. Zerah M, Garcia-Monaco R, Rodesch G, Terbrugge K, Tardieu M, de Victor D, *et al*. Hydrodynamics in vein of Galen malformations. Childs Nerv Syst 1992;8:111-7.
21. Di Rocco C. Vein of Galen aneurysm and hydrocephalus. Childs Nerv Syst 1991;7:359.
22. Siqueira EB, Murray KJ. Calcified aneurysms of the vein of Galen: Report of a presumed case and review of the literature. Neurochirurgia (Stuttg) 1972;3:106.
23. Chapman S, Hockley AD. Calcification of an aneurysm of vein of Galen. Pediatr Radiol 1989;19:541-2.
24. Brunelle F. Arteriovenous malformation of the vein of Galen in children. Pediatr Radiol 1997;27:501-13.
25. Beltramello A, Perini S, Mazza C. Spontaneously healed vein of Galen aneurysms. Childs Nerv Syst 1991;7:129-34.
26. Nzeh DA, Erinle SA, Saidu SA, Pam SD. Transfontanelle ultrasonography: An invaluable tool in the assessment of the infant brain. Trop Doct 2004;34:226-7.
27. Adeyekun AA. Residents' perception of postgraduate radiology training in Nigeria. West Afr J Med 2010;5:314-7.

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