

Endovascular Treatment of Vein of Galen Malformation in Kenya, East Africa

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Abstract

Vein of Galen aneurysmal malformations (VGAM) are arteriovenous fistulas between the choroidal or quadrigeminal arteries and the embryonic precursor of the vein of Galen – the median prosencephalic vein. VGAM represents 30% of arteriovenous malformations in the paediatric age group. They are associated with a high morbidity and 90% mortality if untreated but respond well to intervention with mortality reduced to below 40%. There are few documented cases of VGAM worldwide and scarce data about these lesions in sub-Saharan Africa with challenges in diagnosis and management. We present two cases of infants diagnosed with VGAM and treated with endovascular embolisation. We examine the clinical and radiological data at presentation, intervention and follow-up at one year. We report on some of the technical neuro-interventional details. Both patients had satisfactory

radiological results and the interventions resulted notable clinical improvements from baseline. Physicians should maintain a high index of suspicion treating infants and toddlers presenting with macrocephaly, neurocognitive decline and cardiopulmonary symptoms. Upon diagnosis endovascular interventions are feasible and available.

Keywords: Endovascular, Vein of Galen, Coiling, Embolization, Arteriovenous Fistula

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Introduction

Vein of Galen aneurysmal malformations (VGAM) are arteriovenous fistulas between the choroidal and quadrigeminal arteries and the embryonic precursor of the vein of Galen – the median prosencephalic vein (1, 2). VGAM represents 30% of arteriovenous malformations in the paediatric age group. VGAM is associated with 90% mortality if left untreated, and an associated high morbidity rate (3). In sub-Saharan Africa there are very few documented cases of VGAM, with diagnosis and management being cited as a challenge (4). There is paucity of data as to the treatment and outcomes of VGAM in this region. The lesion requires intensive medical management and timely

endovascular treatment to arrest brain parenchymal injury and severe neurodevelopmental compromise. We present two cases of infants diagnosed with VGAM and treated with endovascular embolization with satisfactory outcome.

Case 1

A 1 year 6-month-old male infant was referred because of macrocephaly and progressive neurological decline noted with worsening speech, unsteady gait and spasticity over 6 months. There was no history of convulsions. The child was appropriate for age developmental milestones and had unremarkable past

medical history in infancy. On examination, he was in fair general condition, alert, pupils were bilaterally equal and reactive to light. On CNS examination, of note was

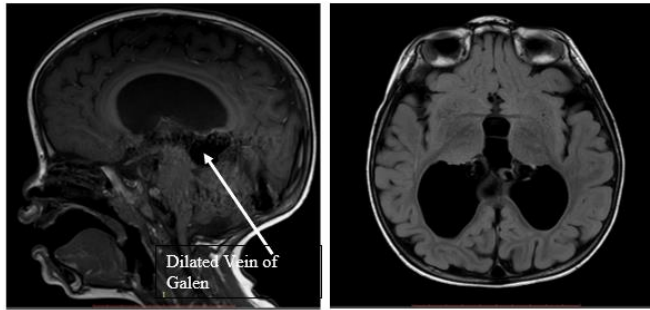


Figure 1. Sagittal (A) and axial (B) T1 MRI of the brain demonstrating aneurysmal dilatation of the vein of Galen with thrombus causing compression of aqueduct of Sylvius with subsequent hydrocephalus

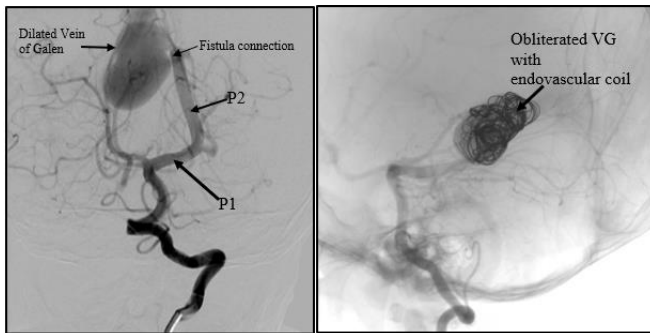


Figure 2. Digital subtraction imaging pre (A) and post coiling of the venous sac (B). Demonstration of fistulas connection from branches of posterior cerebral artery to Vein of Galen

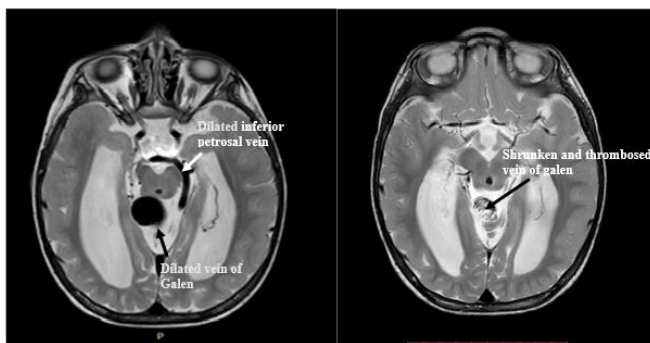


Figure 3. MRI T2 pre-endovascular embolization (A) and 5 months post treatment (B) demonstrating resolution of the dilated vein of Galen and tributaries, reduced ventriculomegaly

the macrocranium with a head circumference of 58.5 cm but had no sunset eyes. (Normal head circumference at

birth is 35cm and 40-45 cm at 18 months of age) (5). Gaze was appropriate without nystagmus or any cranial nerve deficit. He had an ataxic gait, raised muscle tone with hyperreflexia and power grade 5 on all muscle groups. Magnetic Resonance Imaging (MRI) as depicted in image 1 demonstrated a VAGM with ventriculomegaly. Full hemogram, blood biochemistry and liver function assessments were within normal parameters. An echocardiogram was also unremarkable. The child was managed with trans arterial coil embolization in a single session. The Right femoral artery was accessed, the Left Vertebral artery cannulated with 5Fr Multi- Purpose D Envoy guide catheter. DSA runs noted a large fistulous connection of the left Posterior Cerebral artery, PCA, P2 segment with another feeder of the Right PVA as well. It was consistent with a Lasjunias- Mural type of VAGM/ Yasargil Type 2. A Vasco-10MP microcatheter was navigated into the Left P2 and then into the dilated Vein of Galen. Six large coils were deployed with reduced flow of approximately 80%. Magnetic Resonance Angiography (MRA) at five months follow up revealed resolution of venous hypertension and ventriculomegaly. He had much improved speech and gait had become an active young man.

Case 2

A 1-year female infant who was previously well with normal developmental milestones presented with 3-month history of progressive loss of previously achieved motor milestones, with associated three-week history of right lower limb focal seizures, headache and vomiting. At presentation the child was unable to sit up with loss of neck support, unable to stand or walk. On assessment, the child was in poor general condition, with macrocrania with a head circumference of 43 cm, closed fontanelles and generalized hypotonia. An MRI done demonstrated an aneurysmal dilatation of the vein of Galen with associated ventriculomegaly as noted in Image 3. She was managed with trans-arterial coiling embolization over 2 sessions at 3-month intervals. The right femoral artery was accessed with 4 Fr Sheath. A 4Fr C2 guide catheter was used to access bilateral Middle Cerebral Arteries (MCA) and the left Vertebral

artery. DSA runs noted a large fistulous connection of the left MCA Cerebral artery with multiple other feeders from both the right MCA and bilateral PCAs with both

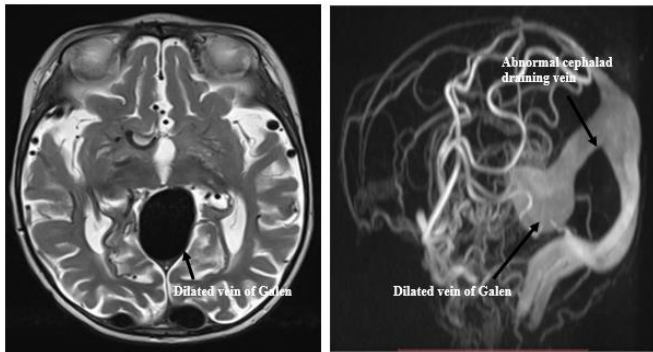


Figure 4. (A) MRI T2 demonstrating dilated vein of Galen, straight sinus and transverse sinus. Note early features of brain atrophy (B.) MRA/MRV demonstrating multiple feeders into the vein of Galen.

choroidal and quadrigeminal supply. She had the persistent abnormal sinus draining high into the lambdoid area of the superior sagittal sinus. The VGAM was consistent with Lasjaunias Choroidal type/ Yasargil Type 1. A Vasco-10MP microcatheter was navigated into the Left P2 and then into the dilated Vein of Galen with single 6mm/20cm standard coil for embolization of the left MCA/PCA feeders at the first session with 60% reduction in flow. Twelve weeks later, we repeated the procedure and coiled the right MCA/PCA with further flow reductions totaling to about 90% of the original flow rates.

At 1 year post procedure patient has had impressive neurological improvement with resumption of sitting, truncal stability and standing. We also have resumption of monosyllabic speech.

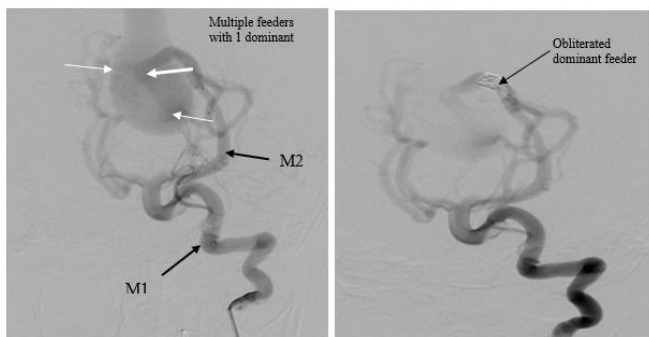


Figure 5. A. Digital subtraction image demonstrating a dilated vein of Galen with multiple choroidal artery feeders (white arrows). B. DSA post coiling with obliteration of dominant arterial feeder.

Discussion

Vein of Galen aneurysmal malformation (VGAM) is a rare condition with a reported incidence of 1 in 25,000 births (2, 6). There is paucity of data on treatment of VGAM in sub-Saharan Africa (6). VGAM typically presents in early childhood and infancy and carries a high morbidity and mortality rate reported to be up to 100% if left untreated and 37% with treatment (6, 7). VGAM occur sporadically with no demonstrable risk of genetic inheritance.

VGAM including the feeding arteries and draining veins are found in the subarachnoid space in the cistern of velum interpositum and quadrigeminal cistern (1). The contemporary classification systems within the literature is that of Lasjunias which morphologically describes two types of VGAM: choroidal VGAM – which have interposing arterial nidus network between feeders and the venous sac and mural VGAM – which have a single or few fistulas on the wall of a dilated draining vein as was the case 1(1, 2). Some VGAMs have mixed archetypical pattern and may have elements of both types, as we see in case number 2. It is paramount to accurately describe the arterial supply, location, structure of arteriovenous connections, pathologic venous anatomy and drainage of the deep infra-tentorial brain and venous outflow obstruction which are

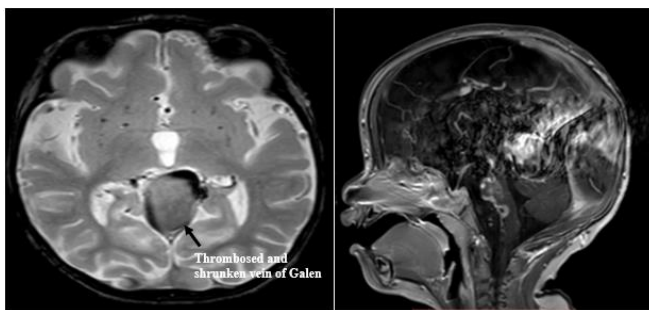


Figure 6. MRI T2 demonstrating changes 5 months post endovascular glue embolization of vein of Galen Malformation. Note the thrombus in the vein of Galen with less prominent tributary veins.

important in prognostication and drafting a safe management strategy (10).

There are three classical presentations of VAGM: neonatal high cardiac output failure, paediatric hydrocephalous with developmental delay, and paediatric and adult neurologic and haemorrhagic complications (11). Both our patients presented with hydrocephalus and developmental delays as described in literature. Neonatal VAGM presents in 0 -2 months with features of high output cardiac failure in 94% of the cases (12). After the first month VAGM typically present with paediatric hydrocephalus and developmental delay as was the case with our two patients. These children typically present with macrocrania, increased intracranial pressure, ventriculomegaly, and developmental delay with systemic symptoms that are secondary to venous hypertension (12). The pathophysiology of this presentation is the hydrodynamic disorder created by increased venous hypertension secondary to the AV fistula. A reversed gradient between the CSF and venous system is created with resulting impaired gradient for CSF absorption secondary to venous hypertension, with accumulation of CSF within the white matter (12). The brain also suffers a brain melting disorder first described by Lasjaunias and colleagues which is as a result of progression of the hydrodynamic dysfunction characterized by progressive brain atrophy (12) and is worsened by VP shunting.

Without treatment the mortality is estimated at 75 – 90%, however endovascular treatment has reduced this to 10 -37% with moderate or severe neurologic morbidity in 26% (10, 12). The goals of treatment are: cure, normal neurologic development or prevention of morbidity and mortality (11). Treatment involves medical optimization of heart failure and endovascular embolization of the fistulous connections. Challenges with endovascular embolization include obstruction of deep normal cerebral vein, diversion of arterial blood from the malformation to the surrounding pial tissue resulting in haemorrhage and pulmonary embolus (8,11). These complications have not yet been observed in our patients at 24 months follow-up.

Conclusion

VAGM is a rare condition which if left untreated has dismal survival rates. Early recognition, intensive medical treatment with timely endovascular embolization profoundly modifies the outcome in these children. These effective treatment options should be made available to selected patients.

Conflict of interest

None to disclose

Author contributions

Both authors contributed equally to writing and editing the original draft.

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