

Case Report

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Case Report on Vein of Galen Malformation with Intranidal Aneurysm and Diverticulum

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Abstract

The case report consists of a mixed pathology in the vein of Galen complex - a Galenic malformation with its own aneurysm. Furthermore, the aneurysm was seen to have its own diverticulum. Investigations included multiple imaging modalities to yield the final diagnosis of the patient. A carefully curated approach to treatment and a safe recovery of the patient has been followed in the report. Differential diagnoses may include but are not limited to cerebral venous thrombosis and intracranial hemorrhage. The age group in which the malformation arises is important with respect to the outcome. Infants and children with the diagnosis have a more optimistic prognosis compared to neonates as the latter have a higher likelihood of developing heart failure. In conclusion, the rarity of the case stems from the presence of a mixed pathology and warrants a very intricate understanding of neuroanatomy as a whole.

Keywords: Vein of galen; Malformation; Intranidal; Aneurysm; Diverticulum; Endovascular; Embolization; Mixed pathology

Abbreviations: VoGM: Vein of Galen Malformation; VoGAM: Vein of Galen Aneurysmal Malformation; AVMs: Arteriovenous Malformations; HR: Heart Rate; BP: Blood Pressure; Temp: Temperature; RR: Respiratory Rate; CT: Computed Tomography; MRA: Magnetic Resonance Angiography; DSA: Digital Subtraction Angiography

Introduction

Brain AVMs are a leading cause of hemorrhage in children and adults. Vein of Galen Malformations are usually diagnosed in infancy or early childhood and differ from Vein of Galen Aneurysmal Malformations. But, the case at hand is neither a VoGAM nor a simple VoGM. Here we see a saccular aneurysm arising on a Galenic malformation; complicated with an aneurysmal diverticulum - the most likely source of the bleed. The case is a culmination of different vascular pathologies - a so called 'mixed pathology'. The choice of treatment in such cases is of great importance and interest to not just neurosurgeons, but doctors in general; globally. The overall mortality in brain AVMs is 0.7 to 2.9% per year [1].

Background

Brain arteriovenous malformations (AVMs) are comprised of a complex tangle of abnormal blood vessels called the nidus, not clearly an artery or a vein and lacking a true capillary bed [1]. There is usually high flow through the feeding arteries, nidus, and draining veins (with increased pressure on the venous side), which may result in rupture and intracranial hemorrhage [1]. The hallmark feature is shunting of blood directly from the arterial to venous circulation [1]. In Galenic malformations, the cardiovascular system takes a toll; and clinically, its relevance is seen when the patient presents with signs of heart failure [2] due to higher-than-

normal pressures and flow draining into the jugular systems.

Case Presentation and Management

A 9 y/o Georgian male presented to the ER with sudden, unbearable headache and blurry vision; positive meningeal signs were noted along with nuchal rigidity. No past relevant history was noted. Vital signs: HR – 99 bpm, BP – 100/70 mmHg, RR- 23 breaths/min, Temp – 37.1oC. At imaging, a head CT showed an intraparenchymal hemorrhage. MRA confirmed the presence of a vascular pathology, followed by a DSA; which showed a vein of Galen malformation with an intranidal aneurysm. Furthermore, the

aneurysm was found to have a diverticulum – the most likely source of the bleed. Images of all imaging modalities are attached below. Currently, three treatment options exist for brain AVMs, namely – microsurgery, endovascular embolization and radiosurgery. An endovascular surgical approach was preferred and the aneurysm was treated using platinum coils. The AVM remnants were embolized using Histoacryl glue. Hypotensive drugs were used for the first 24 hours post-op in order to maintain normovolemia and normotension. On recovery, the patient was seen to have a modified Rankin Score of 0.

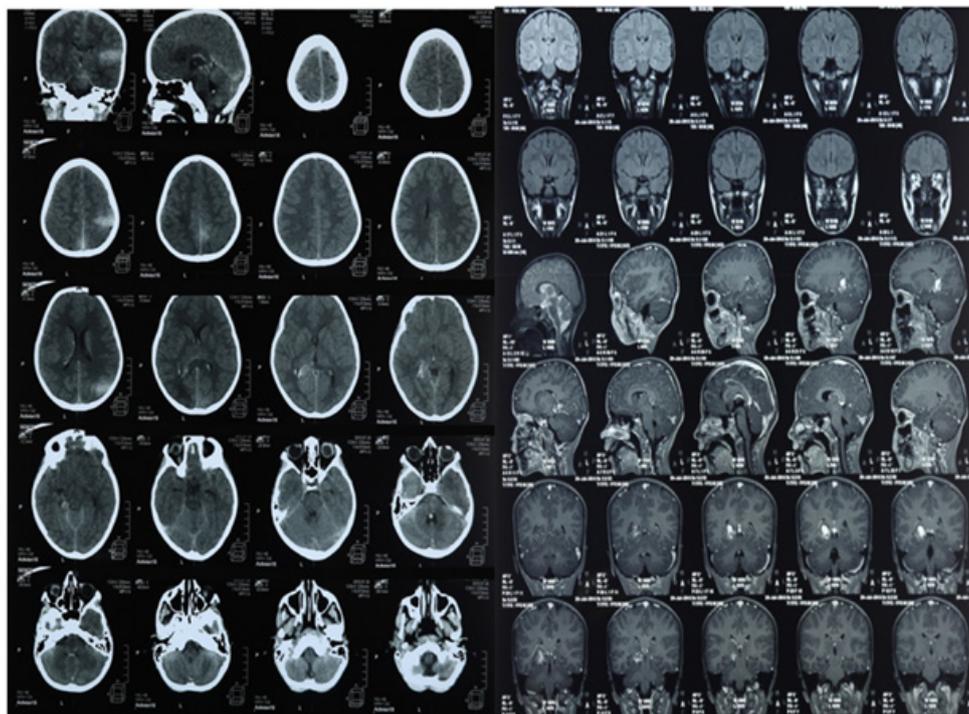


Figure 1: CT with contrast showing an intraparenchymal hemorrhage.

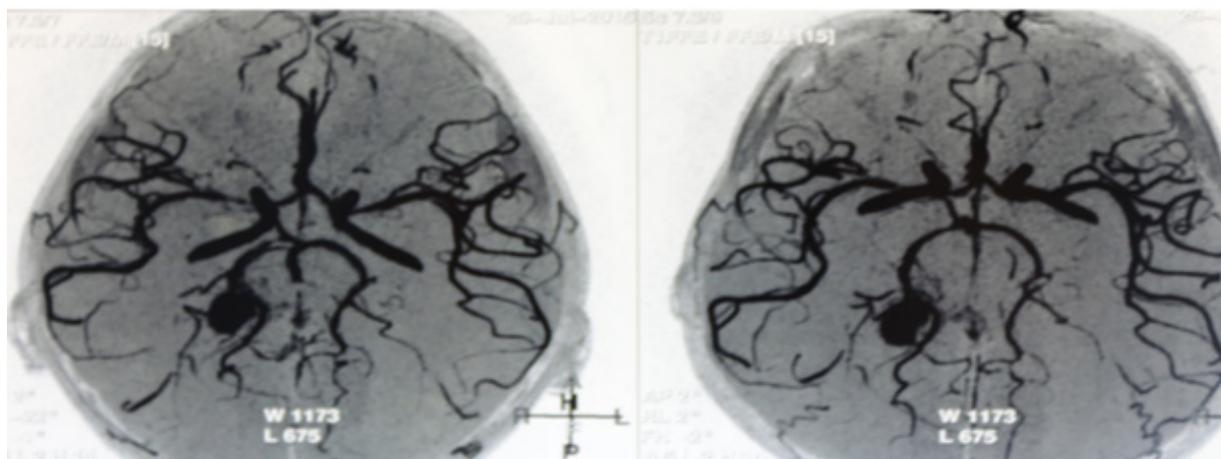


Figure 2: MRA showing a vascular pathology.

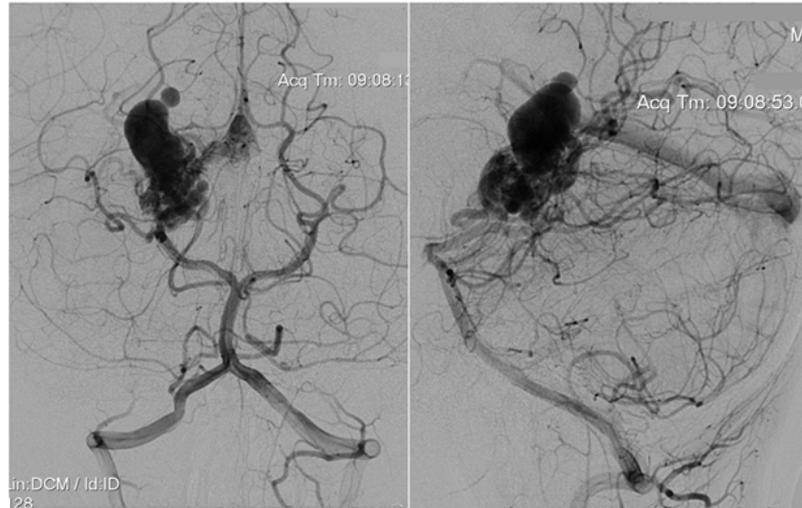


Figure 3: DSA showing vein of Galen malformation, intranidal aneurysm and its diverticulum.



Figure 4: Endovascular coiling of the aneurysm with platinum coils.

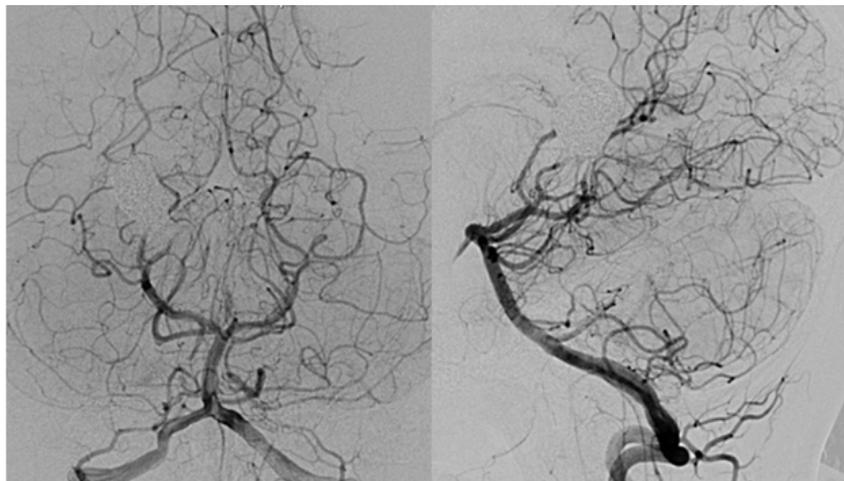


Figure 5: DSA showing anterior and lateral views after treatment.

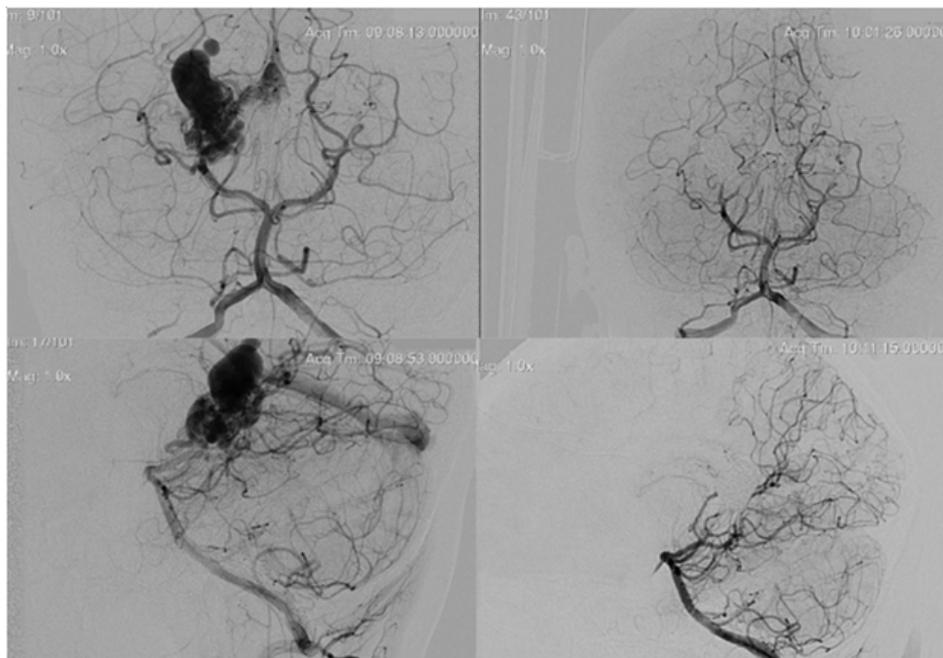


Figure 6: DSAs comparing anterior and lateral views before and after treatment.

Table 1: Spetzler-Martin Scale.

Spetzler-Martin Grading	Points
Size, cm	
<3	1
3 - 6	2
>6	3
Venous drainage	
Superficial	0
Deep	1
Eloquence*	
No	0
Yes	1
Total	5

*Eloquent locations: areas of sensorimotor, language, visual, thalamus, hypothalamus, internal capsule, brainstem, cerebellar peduncles and/or deep cerebellar nuclei

Lower score (1-3) = Better outcome

Higher score (4-5) = Worse outcome and warrants least invasive procedures.

Discussion

According to Martin M Mortazavi and co-authors, "Offering the best treatment option is dependent on an understanding of the aberrant anatomy and pathophysiology of these entities, and tailored therapy is recommended" [3]. In accordance with the above statement, the patient was treated using an endovascular embolization not just due to the presence of a VoGM, but also the complex angioarchitecture of the pathology involving an intranidal

aneurysm (with a diverticulum); highlighting the rarity of the case. Patients are advised to have MRA and DSA scans done at the 1st, 3rd and 5th years post-op.

Acknowledgements

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Conflict of Interest

Kartik Bhagat and Rhea Potdar declare that they have no conflict of interest.

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