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INCIDENTAL POST-MORTEM FINDING OF A VEIN OF GALEN ANEURYSM FOLLOWING A ROAD TRAFFIC ACCIDENT DEATH WITH REVIEW OF LITERATURE
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# INCIDENTAL POST-MORTEM FINDING OF A VEIN OF GALEN ANEURYSM FOLLOWING A ROAD TRAFFIC ACCIDENT DEATH WITH REVIEW OF LITERATURE

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## ABSTRACT

***Background*: Vein of Galen aneurysmal malformations are rare childhood intracranial vascular anomalies. They account for less than 1% of cerebral vascular malformations.**

***Case Report:* A 7-year-old male presented with post traumatic altered sensorium and quadriparesis. Head and neck examinations showed macrocephaly, occipital scalp abrasions. Glasgow Coma Score was 9/15. Both pupils were 3mm bilaterally and reacted briskly to light. Cranial computed tomography (CT) scan was suggestive of obstructive hydrocephalus with a round spherical mass lesion in the pineal region.**

**Clinical diagnosis of traumatic brain injury with incidental vascular malformation was made. He succumbed to his condition twelve hours into admission.**

***Autopsy findings:* At autopsy, the brain was enlarged with bilaterally symmetrical cerebral hemispheres. There was a fusiform aneurysmal dilatation of the left vein of Galen measuring 3.5cm in length, causing a depression in the inferior surface of the temporal region of the left cerebral hemisphere. Both cerebellar tonsils were prominent. Coronal sections through the left cerebral hemisphere showed expansion of white matter with a markedly dilated lateral ventricle. Focal areas of contusional hemorrhages within grey and white mater of the cerebral hemispheres were seen.**

***Conclusion:* Vein of Galen aneurysms may appear relatively asymptomatic with diagnosis dependent on imaging or a post-mortem examination.**

## INTRODUCTION

Vein of Galen aneurysmal malformations (VGAM) are rare malformations found predominantly in the paediatric population. The incidence has been reported to be less than 1% of all vascular malformations[1], notwithstanding the true nature of the lesion. In the paediatric population, vein of Galen malformations account for 30% of all brain vascular malformations[2].

In the African population, documented cases are rare[3]. This has been previously attributed to paucity of equipment and expertise in diagnostic imaging[3], which is still largely unavailable in most regions today.

Vein of Galen malformations are typically identified by multiple arteriovenous shunts which drain into a median cerebral venous vein derived from a persistent embryonic channel termed "Median Prosencephalic Vein of Markowski". Normally this vein is absent in the adult[4].

Although this vein is erroneously termed, vein of Galen – It was first recognized by "Raybaud and Strother"[5]. Arterial supply can arise from the anterior or posterior circulation. Vein of Galen malformations are predominantly seen in males, with a male: female ratio of 3:1. The first reported case was in 1937[6].

The aetiology of vein of Galen aneurysmal malformations (VGAMs) is relatively unknown. However, different postulates suggest somatic mutations in neural crest cells excluding/including surrounding cephalic mesoderm in the early embryo.

## CASE PROFILE

We like to report an incidental finding of a vein of Galen malformation in a 7-year-old male who presented with post traumatic altered sensorium and quadriparesis following a road traffic accident. Head and neck examinations showed macrocephaly and occipital scalp abrasions. Glasgow Coma Score was 9/15. Both pupils were 3mm bilaterally and reacted briskly to light. Cranial computed tomography (CT) scan was suggestive of obstructive hydrocephalus with a round spherical mass lesion in the pineal region.

A Clinical diagnosis of traumatic brain injury with incidental vascular malformation on Cranial Imaging was made. He unfortunately succumbed to his condition twelve hours into admission.

## AUTOPSY FINDINGS

At autopsy, the brain was moderately enlarged with bilaterally symmetrical cerebral hemispheres showing moderate cerebral oedema. There was a fusiform aneurysmal dilatation of the left vein of Galen measuring 3.5cm in length, causing a depression in the inferior surface of the temporal region of the left cerebral hemisphere. Both cerebellar tonsils were prominent and grooved suggesting raised intracranial pressure. Coronal sections through the left cerebral hemisphere showed expansion of the white matter with markedly dilated lateral ventricles. Focal areas of contusional hemorrhages within grey and white mater of the cerebral hemispheres were seen.

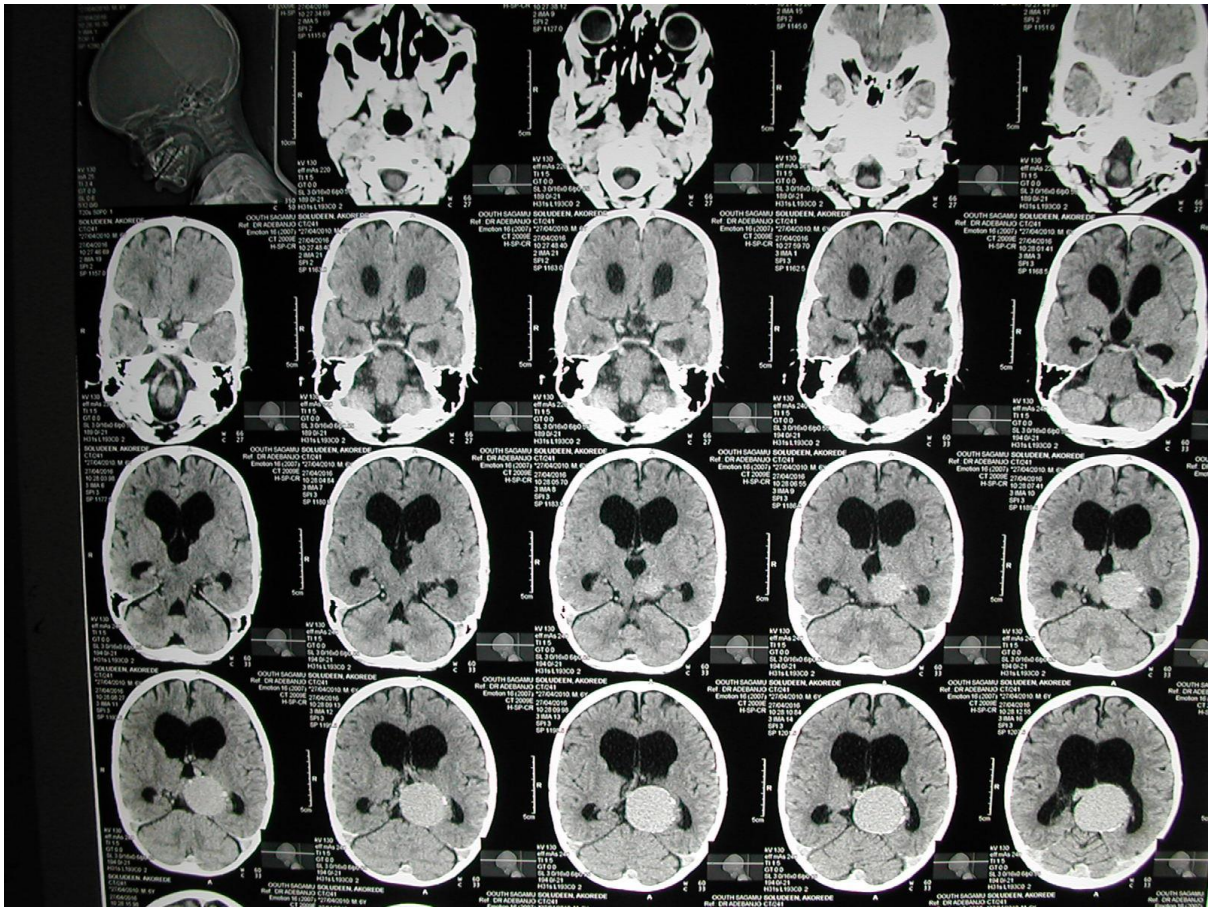

*Figure 1- CT Scan showing dilated lateral ventricles along with an aneurysmal mass located in the pineal region*

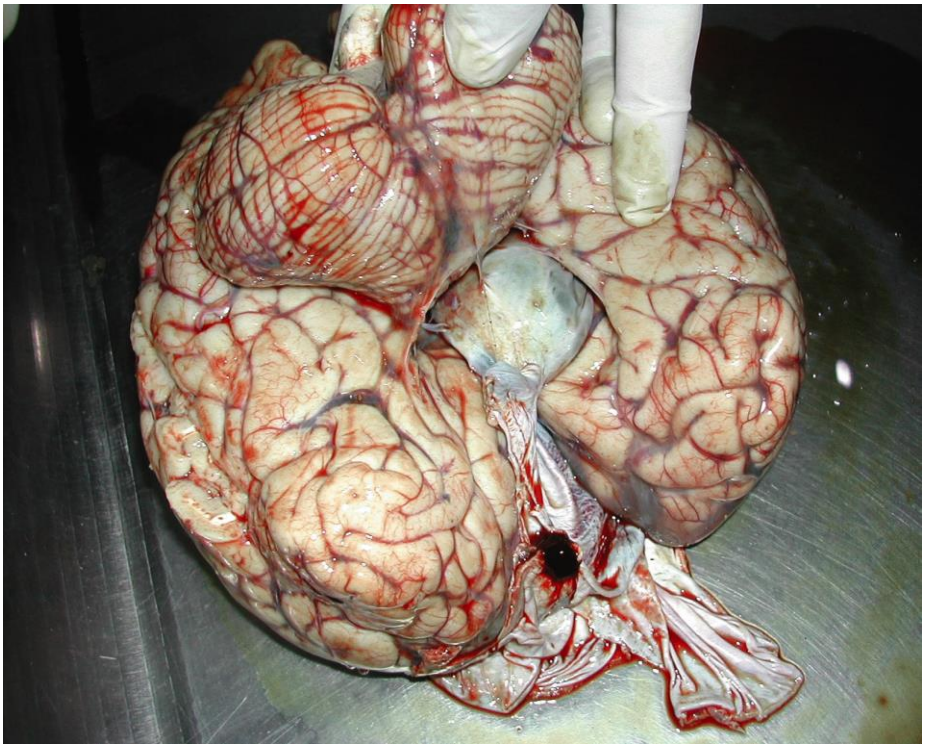

*Figure 2 – Autopsy showed cerebral oedema, congestion of the surface vessels and an aneurysmal vein of galen located in the pineal region*

## DISCUSSION

Various classification systems have been proposed for the vascular malformations seen in the region of the vein of Galen[1]. These classifications are based on the nature of connection between the supplier vessels, the prosencephalic vein and the degree of shunting[1]. They have been broadly classified into primary and secondary vein of Galen malformations. Unlike primary malformations, secondary malformations are misnomers as the true vein of Galen and not the embryonic precursor is enlarged[1]. This secondary enlargement may be due to an adjacent vascular malformation or venous outlet malformation.

Litvak et al[7] proposed a clinicopathologic classification of primary vein of Galen aneurysms into three categories namely;

Category A- involving singular dilation of the great cerebral vein of Galen contigous with a dilated straight sinus and torcula, fed by anomalous branches of the anterior and posterior circulation. Category B – involving conglomerates of blood vessels in the deep cerebral structures with dilated deep venous structures. These drain centripetally into adjoining dilated deep veins and sinuses. Category C – named the transitional type belonging to neither of the above-mentioned categories. Lasjaunia in contrast proposed a classification of primary vein of Galen malformations based on the supply to the median prosenchephalic vein[8]. Lasjaunia type A is composed of feeders from mainly the anterior/posterior choroidal arteries while Lasjaunia type B is composed of single or multiple fistulas connecting to the lateral margin of the wall of the median prosencephalic vein.

Recently, the Yasargil classification represents a broader attempt to classify these primary malformations. The Yasargil classification shows a few similarities with the initial Litvak classification. Yarsagil classification divides these malformations into four types (Types I – IV). Types I-III are malformations of a purely fistulous origin while types IV show malformations with or without fistulas. Further classification within the different Yasargil types is dependent on the origin/territory of the feeder arteries.

These classifications are important in deciding neurosurgical procedures to be attempted[1].

Clinically vein of Galen malformations have been associated with headaches, developmental delays, hydrocephalus, macrocephaly and even high output cardiac failure. The course of clinical presentation often varies with the age on presentation. Presentation with heart failure carries the greatest risk of a poor outcome. Younger patients have a poorer outcome as the venous system matures with age. Some authors agree on age at 5 months to predict clinical prognosis[1]. In the index case, no symptomatology was elicited in the clinical history as the patient was reported to otherwise be in good health prior to the traumatic incident.

Diagnostic imaging is quite useful in detection of these malformations. CT angiography/ Magnetic Resonance angiography being the modality of choice. Trans-fontanellar ultrasonography may prove to be useful in critically ill neonates, but this is dependent on operator experience. In our case, this diagnosis was picked on routine CT scan while assessing for possible head trauma. The obstructive (non-communicating) hydrocephalus seen was due to compression of the aqueduct and 4$^{th}$ ventricle by the venous aneurysm. No history of psychomotor disturbances was given in our patient. A review of other imaging findings previously documented by one of the authors also shows extensive gyriform hyperdensities at the grey-white matter junction, dilatation of a portion of the superior saggital sinus with erosion of the inner table of the calvarium[3]. Multiple

abnormal vessels were also seen in the suprasellar region as well as surrounding the quadrigeminal cistern[3].

Autopsy confirmed the radiologic findings of an aneurysmal vein of galen malformation. However, in this index case, there was no remnant of the promesencephalic vein seen. The true vein of Galen was present, dilated and associated with sub-dural and pial shunts. This presentation is in keeping with Secondary vein of Galen dilation which occur at a low frequency in neonates but much later in late childhood with intra-cranial haemorrhage and focal neurological deficits along with delayed psychomotor development. This may explain the relatively "symptomatic" state before presentation. The cause of death was attributed to severe head injury with Grade 2 diffuse axonal damage secondary to a pedestrian motor vehicle road traffic accident.

**REFERENCES**

1. Mortazavi, Martin, Griesswnauer, Christoph, Foreman, Paul, Shahripour Reza, Shoja M. Vein of Galen aneurysmal malformations: critical analysis of the literature with proposal of a new classification system. *J Neurosurg Pediatr*. 2013;12(September):293-306.

2. French LA, Peyton WT. Vascular malformations in the region of the great vein of Galen. *J Neurosurg*. 1954;11:488-498.

3. Ogbole G, Adeleye A, Olatunji R. Vein of galen aneurysmal malformation in children: Challenges of imaging in an African setting. *West African J Radiol*. 2015;20(1):45.

4. Gailloud P, O'Riordan DP, Burger I, et al. Diagnosis and management of vein of Galen aneurysmal malformations. *J Perinatol*. 2005;25(8):542-551.

5. Raybaud CA, Strother CM. Persisting abnormal embryonic vessels in intracranial arteriovenous malformations. *Acta Radiol Suppl*. 1986;369:136—138.

6. JAEGER, JR. Bilateral congenital cerebral arteriovenous communication aneurysm. *Trans Am Neurol Assoc*. 1937;63:173-176.

7. Litvak J, Yahr MD, Ransohoff J. Aneurysm of the Great Vein of Galen. *J R Soc Med*. 1967;60(5):450-452.

8. Lasjaunias P, Terbrugge K, Piske R, Lopez Ibor L, Manelfe C. [Dilatation of the vein of Galen. Anatomoclinical forms and endovascular treatment apropos of 14 cases explored and/or treated between 1983 and 1986]. *Neurochirurgie*. 1987;33(4):315—333.