

# Intracranial Pial Arteriovenous Fistula Mimicking a Vein of Galen Aneurysm with Hydrocephalus Managed with Endovascular Method: Case Report

Jorge Valderrama <sup>1</sup>, Ricardo Garcia <sup>1</sup>, Guru Satyarthee <sup>2</sup>, Willem Guillermo Calderon-Miranda <sup>3</sup>, Samer Hoz <sup>4</sup>, Alexis Narvaez-Rojas <sup>5</sup>, Luis Rafael Moscote-Salazar <sup>6\*</sup>

<sup>1</sup> National Institute of Neurology and Neurosurgery Manuel Velasco Suarez, Mexico City, Mexico

<sup>2</sup> Department of Neurosurgery, Jai Prakash Narayan Apex Trauma Center, All India Institute of Medical Sciences, New Delhi, India

<sup>3</sup> Hospital General Dr.Manuel Gea González, Mexico City, Mexico

<sup>4</sup> Neurosurgeon, Neurosurgery Teaching Hospital, Baghdad, Iraq

<sup>5</sup> The National Autonomous University of Nicaragua, Managua, Nicaragua

<sup>6</sup> Department of Neurosurgery, University of Cartagena, Colombia

\*Corresponding Author Address: Department of Neurosurgery, University of Cartagena, Colombia. Telephone: +57 5 6646183. E-mail:rafaelmoscote21@gmail.com

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## Abstract

**Background and Importance:** Intracranial pial arteriovenous fistula is a recently described matter as a rare vascular malformation occurring in children. It is usually located in the subpial space in the intracranial cavity with a high-flow shunt.

**Case Presentation:** Authors report on a 2-year-old child with intracranial pial arteriovenous fistula and progressive enlargement of head and seizures mimicking progressive obstructive hydrocephalus for eight months. Cranial computed tomography showed a hyperdense midline vein of Galen and enlarged straight sinus associated with obstructive hydrocephalus. Presence of an aneurysm of the vein of Galen was suspected.

**Conclusion:** Authors believe that cranial screening with computed tomography for suspected early diagnosis is of high importance.

**Keywords:** Pial; Arteriovenous; Fistula; Galen; Aneurysm; Pediatric; Embolization; Hydrocephalus

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## Background and Importance

Intracranial pial arteriovenous fistula (IPAVF) is a group of infrequently occurring vascular malformation in children [1-7]. IPAVF represents a subpial vascular malformation with high-flow shunt fed by mostly one or more pial arterial feeders and drainage by a solitary vein and lacking nidus [1,3,4]. Characteristically, these pediatric cases become symptomatic with catastrophic presentations. Thus, early treatment is advised for good neurological outcome. Intracranial pial arteriovenous fistulas can be managed by either endovascular method or direct attack of craniotomy with surgical resection or rarely a combination of procedures [5-7].

## Case Presentation

A 2-year-old boy was referred to the emergency department with macrocrania and seizures.

Eight months before that, his mother had noticed signs which suggested psychomotor development retardation (PDR). His mental status was postictal. The patient had normal vital signs with the physical examination otherwise being unremarkable. Axial computed tomography (CT) scan of the brain without contrast showed hydrocephalus, a hyperdense vein of Galen in the midline and associated enlarged straight sinus. The probable diagnosis could be an aneurysm of the vein of Galen (Figure 1).

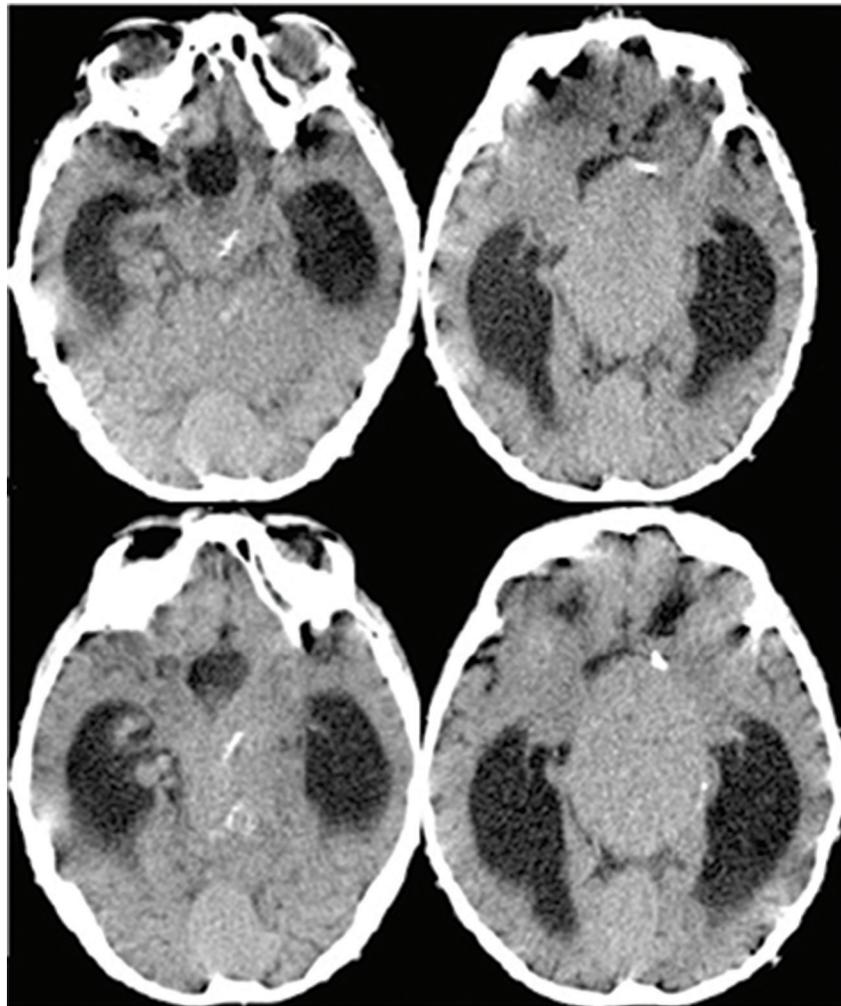
Magnetic resonance imaging (MRI) was

performed in which an enlargement of the right posterior cerebral artery at the level of the circle of Willis and a large dilated vascular structure in the upper portion were found (Figure 2). To further delineate the architecture of vascular lesion, a digital subtraction angiography was carried out, right internal carotid and vertebral injection angiograms demonstrated the presence of a high-flow fistula being supplied by the right posterior cerebral artery (PCA) and the fistula was draining into the straight sinus (Figure 3). However, no intervening nidus could be identified. The child underwent endovascular embolization in order to reduce high-flow arteriovenous shunt (Figure 4). He tolerated the procedure well and was discharged with a good clinical condition.

### Conclusion

An IPAVF is characterized by the presence of a direct arteriovenous connection with high-flow shunt located in the subpial space [6]. Most of the arteries feeding the IPAVF are drained into a single vein, which becomes dilated, elongated and tortuous due to high flow. They also become arterialized and often associated with a giant varicose vein [10,12]. These IPAVFs have single or multiple arterial feeders [6,7,10-12]. An IPAVF is usually congenital in nature in most cases, but may occasionally occur following head trauma, cranial surgery, or brain ischemia and after an episode of cortical vein thrombosis [8].

Intracranial pial arteriovenous fistula accounts for about 4.7% of pial arteriovenous



**Figure 1. Noncontrast Compute Tomography Scan Showing Head Enlargement with Hydrocephalus with Presence of Large Venous Varix**

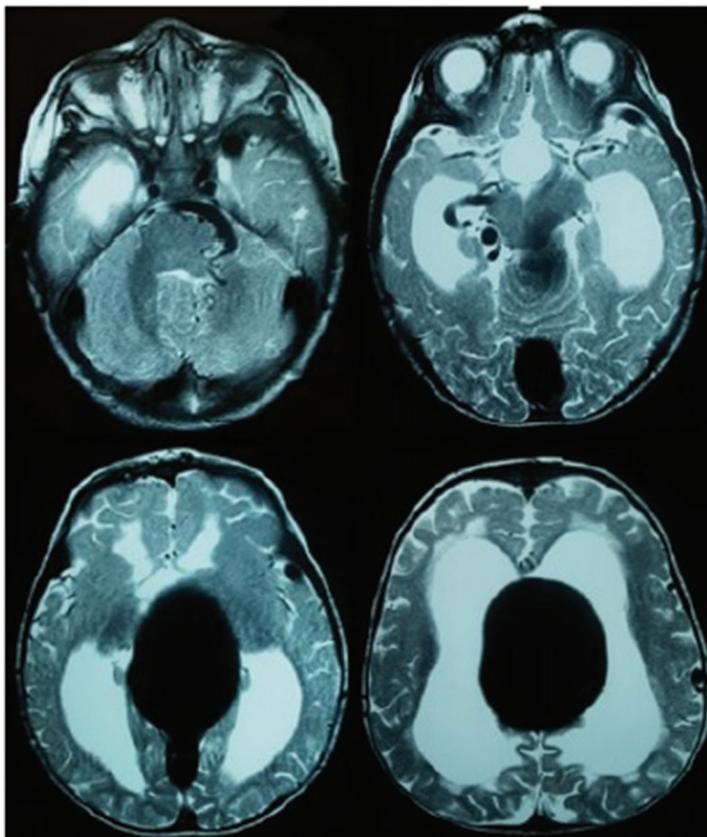


Figure 2. Magnetic Tomography Scan, Axial Section, T2-weighted Image Showing Hydrocephalus with Presence of Large Venous Varix

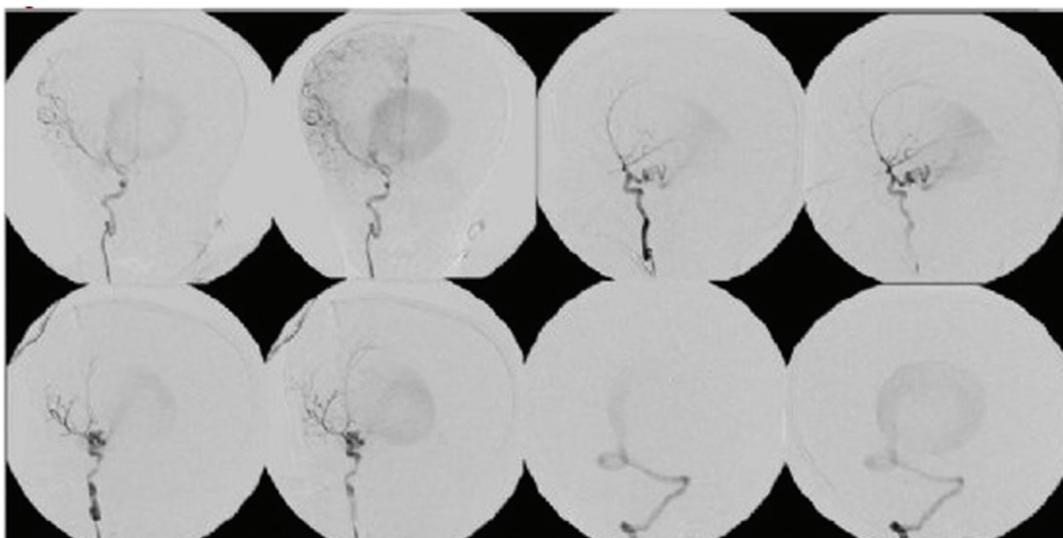
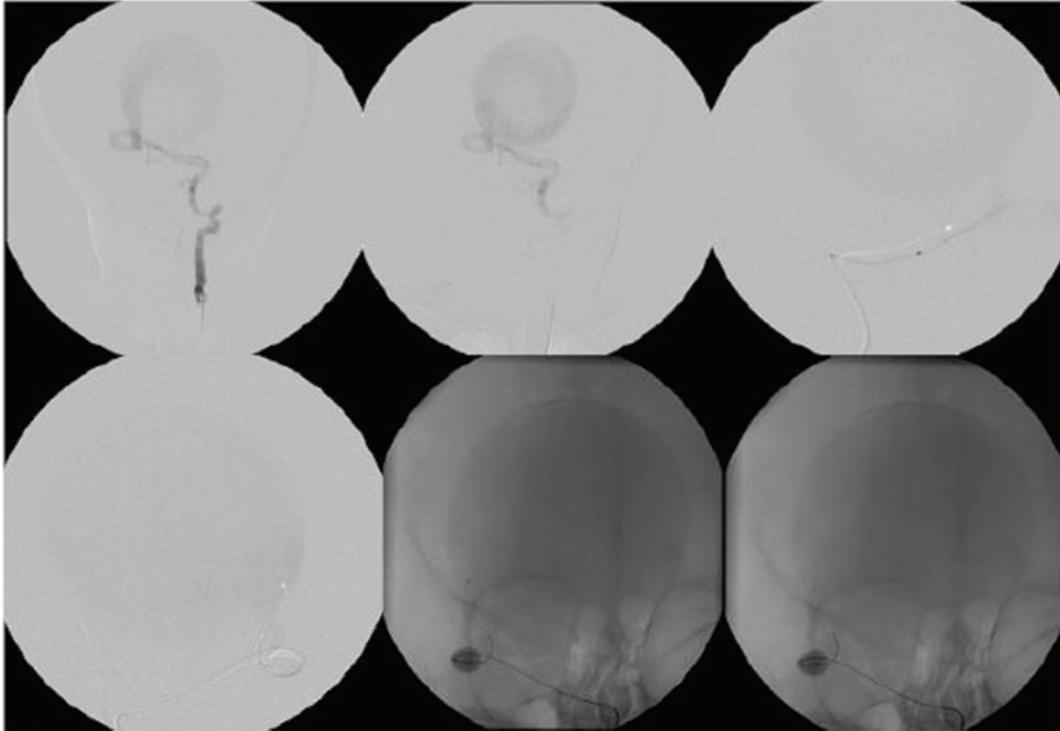


Figure 3. Right Internal Carotid Artery Injection Digital Subtraction Angiography Showing Intracranial Pial High Flow AV Fistula with Venous Varix



**Figure 4. Vertebral Artery Injection Digital Subtraction Angiography, Showing Intracranial Pial High Flow AV Fistula with Venous Varix**

malformations [11]. High-flow fistula causes hyperdynamic circulation leading to increased circulatory volume with intracranial venous hypertension, and increased cardiac load. Intracranial venous hypertension can cause congestive cardiac failure in the neonates, and some children may develop seizures, and rupture of intracranial arterialized cortical veins can lead to intracranial intracerebral, intraventricular, and subarachnoid hemorrhage. During infancy, intracranial venous hypertension may lead to hydrocephalus development. The current case also had head circumference enlargement which was noticed by the parents in the second year [6].

In 2012, Hetts et al. retrospectively reviewed the data of 25 patients with non-galenic arteriovenous fistula. They found 7.3% of pediatric intracranial vascular lesions with a nondual arteriovenous shunt. Seven cases that presented in the first month had congestive cardiac failure, with large, complex fistulas, but those presented later in childhood had comparatively simple angioarchitecture with predominantly single arterial feeder and usually presented with seizure. The researchers concluded that

patients presenting in the first two years of life were more likely to have a multi-hole fistula and had poorer clinical outcome [13].

Initial imaging is CT scan as it is easily available, less costly as well as less time consuming. Although MRI of the brain delineates the lesion better, the gold standard is still digital subtraction angiography and confirms the diagnosis of IPAVF and aids in planning on the therapeutic program, either surgical or endovascular, while endovascular treatment can be carried out in the same setting.

Management of IPAVF poses a challenge to both neurosurgical and neurointerventional teams as it is associated with high mortality and poor neurological outcome irrespective of the treatment used. The occlusion of the arteriovenous shunt is necessary [5,13,14].

The treatment should be decided according to both neurosurgical as well as neurointerventional team available at the treatment center. The endovascular approach allows radiological delineation of venous and arterial part of the pial fistula, even if the vein is enlarged to form varicose veins, which is a quite common association. However, an engorged, tortuous, dilated,

arterialized and varicose vein makes surgical exploration tedious.

Under certain circumstances, surgical method may be the only option, for instance, in cases where embolization is dangerous or catheterization of smaller feeder vessels is not possible or unfits due to angioarchitecture of IPAVF [14]. Cases with IPAVF require regular follow-up to check for new dural IPAVF development and recanalization [12,14].

Every child with focal neurological deficit along with enlarging head should be investigated for suspected IPAVF, although its occurrence is extremely rare. The authors of present case report advocate cranial screening with computed tomography for suspected early diagnosis and providing an opportunity to preserve good neurological outcomes after digital angiography as well as providing a tailor-made treatment.

### Funding

None

### Conflicts of Interest

The authors declare that they have no conflicts of interest.

### Authors' Contribution

**Conception and Design:** Luis Rafael Moscote-Salazar

**Data Collection:** Jorge Valderrama, Ricardo Garcia, Guru Satyarthee, Willem Guillermo Calderon-Miranda, Luis Rafael Moscote-Salazar

**Drafting the Article:** Willem Guillermo Calderon-Miranda, Samer Hoz, Alexis Narvaez Rojas, Luis Rafael Moscote-Salazar

**Critically Revising the Article:** Samer Hoz, Alexis Narvaez Rojas, Luis Rafael Moscote-Salazar

**Reviewed Submitted Version of the Manuscript:** All authors

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