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# CHOROID PLEXUS ARTERIOVENOUS MALFORMATIONS IN A 5-YEAR-OLD FEMALE PATIENT. CASE REPORT

**Keywords:** Intracranial Arteriovenous Malformations; Vascular Diseases; Pediatrics.
**Palabras clave:** Malformaciones Arteriovenosas Intracraneales; Enfermedades Vasculares; Pediatría.

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## RESUMEN

**Introducción.** Las malformaciones arteriovenosas (MAV) son anomalías infrecuentes que ocurren en aproximadamente 0.1% de la población general. Las MAV cerebrales suelen ser asintomáticas, pero las manifestaciones clínicas más comunes incluyen hemorragia intracerebral no traumática, siendo esta una de las complicaciones con mayor carga de morbimortalidad; estas MAV tienen una prevalencia del 0.02% en población pediátrica.
**Presentación del caso.** Niña de 5 años, previamente sana, quien fue llevada a un servicio de urgencias por dolor cervical asociado a síntomas neurológicos. La tomografía computarizada cerebral reportó hemorragia intraventricular, por lo que se realizó una ventriculostomía en la cual se colocó un catéter externo para iniciar drenaje. En la panangiografía realizada por parte del servicio de radiología intervencionista se observó un nido capilar arteriovenoso tipo II según la escala de Spetzler Martin en el plexo coroideo no candidato a manejo endovascular. Un año después del diagnóstico inicial se realizó la resección quirúrgica de la MAV de manera exitosa y la paciente evolucionó satisfactoriamente.
**Conclusión.** Las MAV cerebrales son raras y su diagnóstico y tratamiento dependen de la experiencia médica debido a la limitada literatura disponible sobre el tema. Por tanto, realizar estudios de imagen ante síntomas neurológicos inespecíficos es de gran importancia para lograr un diagnóstico oportuno e instaurar el tratamiento adecuado, evitando así desenlaces adversos.



## ABSTRACT

**Introduction:** An arteriovenous malformation (AVMs) is a rare defect that occurs in approximately 0.1% of the general population. Brain AVMs are usually asymptomatic, but the most common clinical manifestations include nontraumatic intracerebral hemorrhage, which is one of the complications with the highest morbidity and mortality burden; this type of AVM has a prevalence of 0.02% in the pediatric population.
**Case report:** A 5-year-old girl, previously healthy, was taken to the emergency department due to cervical pain associated with neurological symptoms. A brain computed tomography scan showed intraventricular hemorrhage, so a ventriculostomy was performed and an external catheter was placed to initiate drainage. A panangiography performed by the interventional radiology service showed a type 2 arteriovenous capillary nidus on the Spetzler Martin Grading Scale located in the choroid plexus, which was not eligible for endovascular treatment. One year after the initial diagnosis, the AVM was resected successfully, and the patient progressed satisfactorily.
**Conclusion:** Cerebral AVMs are rare, and their diagnosis and treatment depend on medical expertise due to the limited literature available on the subject. Therefore, performing imaging studies when encountering nonspecific neurological symptoms is particularly important to achieve a timely diagnosis and to establish the proper treatment, thus avoiding adverse outcomes.

## INTRODUCTION

Arteriovenous malformations (AVMs) are abnormal connections between arteries and veins without an intervening capillary bed. These malformations can appear anywhere in the body, but are most frequently found in the cerebral cortex, the brainstem (pons and midbrain), and the cerebellum (1). AVMs are rare defects, and their prevalence rate is estimated at 0.1% of the general population (2). In the pediatric population, brain AVMs are extremely rare, with a prevalence rate of 0.02% (3).

AVMs are lesions that occur since birth but are only perceived at that stage in 50-60% of cases (4), while they become evident in 20-30% of cases during adolescence and in 10-20% of cases during adulthood (5). Malformations differ among age groups and, consequently, symptoms and diagnostic tools vary with age; however, the diagnosis mainly involves diagnostic imaging such as computed tomography (CT), magnetic resonance imaging (MRI), and angiography (6).

AVMs are challenging for medical professionals because their pathophysiology is not very clear. The prevailing hypothesis of its etiopathogenesis suggests that they are caused by a defect in the development of the vascular plexus during embryogenesis, which generates communication errors between arteries and veins when they are in direct contact without intervening capillaries; however, it has been described that certain AVMs may be de novo and develop as acquired lesions (7).

Pediatric AVMs are difficult lesions to treat because of the devastating impact of bleeding and because the long lifespan of a child confers increased cumulative risk over time. Therefore, most care centers endorse an aggressive approach to the treatment of AVMs in the pediatric population (8). However, in children, as in adults, the therapeutic options have increased thanks to technological advances in microsurgical resection and radiosurgery, with surgical resection being the treatment of choice as it allows achieving a complete cure. Still, depending on the location of the AVM, conservative treatment may be preferred in some cases (8). It should be noted that a multidisciplinary approach is recommended to ensure the best outcomes.

The following is a case report of a pediatric patient with AVM who underwent surgery and had a satisfactory outcome.

## CASE PRESENTATION

A 5-year-old girl was taken to the resuscitation area of the emergency department of a quaternary care hospital in Floridablanca (Colombia) due to the sudden onset of cervical pain followed by 5 episodes of vomiting, with subsequent hypoactivity, loss of postural tone, mucocutaneous pallor, and poor response to stimuli. The patient, who was born at term and in good general condition, had no significant personal or family history.

On admission (half an hour after the onset of symptoms), the patient showed a moderate level of altered consciousness (11/15 on the Glasgow scale), high blood pressure (128/73 mmHg), low heart rate (48 bpm), excessive sweating, normal

oxygen saturation level (98%), slightly reactive 3mm pupils, and hemiparesis and muscle hypertonia on the right side of the body. Given the findings, supplemental oxygen at 2L/min and 0.9% saline solution (200mL slow bolus) was started, leading to an improvement in heart rate.

A blood glucose test was taken on admission yielding a normal result (183mg/dL), and an electrocardiogram was also performed which revealed sinus bradycardia and incomplete right bundle branch block. Following the electrocardiogram, a brain CT scan revealed intraventricular hemorrhage predominantly in the left lateral ventricle, with extension of the hemorrhage to the third and fourth ventricles, together with ventricular dilatation. Consequently, the patient was transferred to the pediatric intensive care unit (PICU) one hour after being admitted.

A multidisciplinary assessment including pediatric neurology and pediatric neurosurgery was performed in the PICU and, considering the rapid neurological deterioration and the very high risk of complications, elective intubation and protective ventilation were undertaken, while brain protection measures were initiated. A new evaluation by the pediatric neurosurgery department performed 3.5 hours after admission found signs of intracranial hypertension, because of intraventricular hemorrhage, as well as secondary hydrocephalus. Thus, a drainage of the hemorrhage through an external catheter placed by ventriculostomy was immediately requested. The procedure was performed without complications and allowed an improvement of hemodynamic parameters. The patient continued under sedation with open drainage 10cm above the ear canal.

A computed tomography angiogram was taken three hours after surgery and showed a possible aneurysm of the left posterior cerebral artery located in the occipital horn of the lateral ventricle, adjacent to the fluid collection, but without ruling out a possible AVM. The results of coagulation, renal function and electrolyte panel tests taken on admission were within normal parameters. However, anemia was observed but no transfusion criteria were met.

Two days after admission, the patient was successfully extubated, presenting good neurological evolution (no signs of focal neurologic signs or intracranial hypertension) and a functioning ventricular catheter. Four days after admission, the neurosurgery department decided to close the catheter to evaluate tolerance to the treatment, which was successful and had no complications.

Five days after admission, the interventional radiology service performed a panangiography in which a type 2 arteriovenous capillary nidus on the Spetzler Martin Grading Scale was observed in the choroid plexus (Figure 1), which was dependent on afferent capillaries passing from the angular artery and had a venous drainage to the deep temporal vein that drained into the great cerebral vein. Endovascular intervention was not indicated due to the size and characteristics of the malformation.

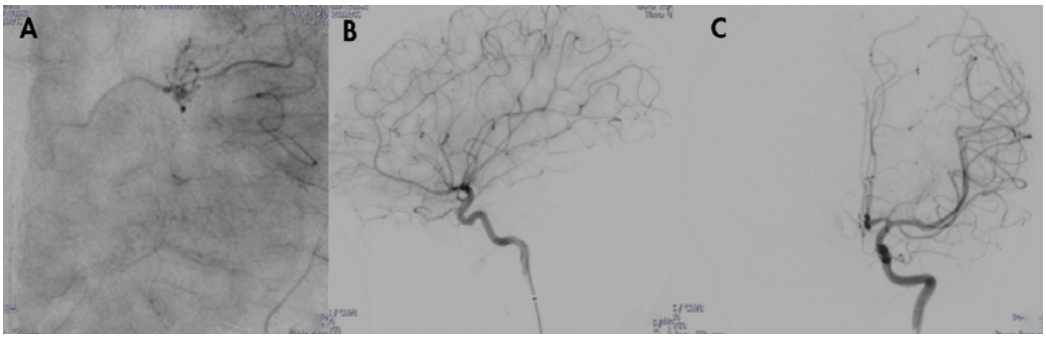


Figure 1. Cerebral panangiography showing left subcortical, parieto-occipital arteriovenous malformation dependent on afferent capillary arteries passing from the angular artery and with venous drainage to the deep temporal vein that flows into the great cerebral vein.

Source: Images obtained while conducting the study.

The catheter placed during the ventriculostomy was removed on the sixth day of PICU stay and a follow-up CT scan was performed, which showed a decrease in bleeding and resolution of dilatation of ventricles 3 and 4. That same day, the patient was transferred to the general hospital ward, where she had an adequate progression, with no new symptoms. She was discharged after 13 days of monitoring.

Two weeks after being discharged, the patient was taken to a follow-up appointment with the interventional radiology service, showing an adequate progress and no new symptoms or complications. It was decided to keep the follow-up with pediatric neurosurgery and radiosurgery to establish possible treatments considering the patient's age, since the arteries afferent to the AVM in children are difficult to catheterize due to their size. Moreover, regular follow-up imaging was considered, so a month after the first follow-up appointment with interventional radiology, a panangiography was performed, confirming the diagnosis established in the first study of this type.

Six months after discharge, a new pangiography was performed, which reported the persistence of the 6x7mm AVM located in the left subcortical, parieto-occipital region, without changes with respect to previous studies (Spetzler Martin type 2 classification), extending adjacent to the occipital horn of the lateral ventricle, and no compression or cerebral edema. Additionally, there were signs of bleeding with areas of gliosis in the right frontal lobe, which was the result of the ventricular shunt. In view of these findings, the pediatric neurosurgery service considered surgical treatment of the lesion due to the high risk of rebleeding.

Owing to the COVID-19 pandemic and the difficulties inherent to the Colombian Health System, the surgery had to be postponed on several occasions, and almost a year after being discharged from the hospital, neuronavigation-guided AVM resection was scheduled, which was performed successfully and without complications.

After the procedure, the patient was admitted to the hospital and had an adequate neurological progression. The follow-up CT scan, performed on the sixth postoperative day, ruled out major complications (Figure 2), so one month after the surgery she was discharged with recommendations on possible warning signs.

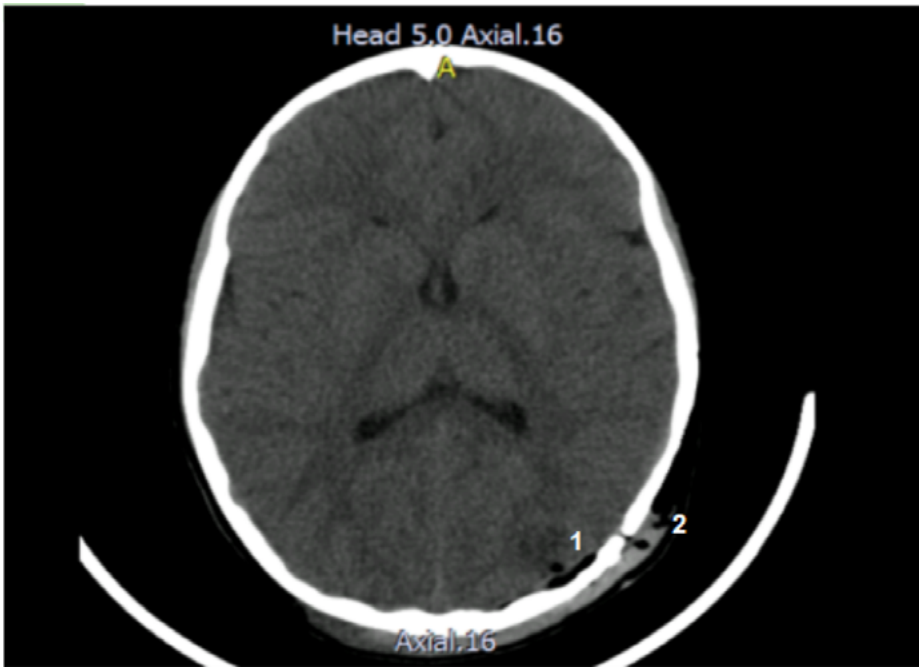


Figure 2. Postoperative simple computed tomography of the skull. 1) contusion area in the left parietal region with adjacent pneumoencephalon and post-surgical changes; 2) soft tissue swelling in the left parietal region and post-surgical changes.
Source: Image obtained while conducting the study.

The patient continued to be followed up and nine months after surgery the neuro-ophthalmology service ruled out structural damage to the optic disc. At the time of this case report, she was awaiting cycloplegia and continued to be followed up by the pediatric neurosurgery service and undergoing regular imaging studies.

Figure 3 presents the algorithm of the therapeutic approach to the patient in the present case.

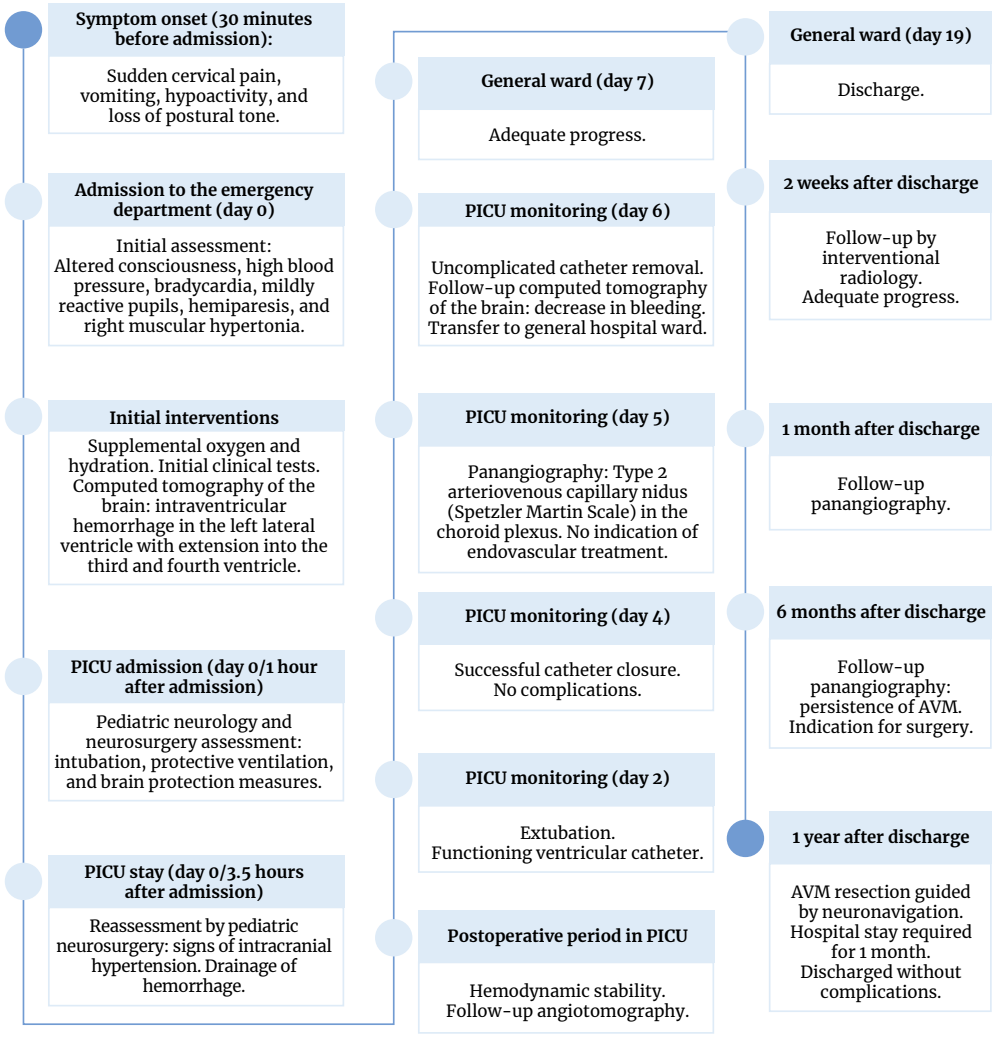


Figure 3. Algorithm of the therapeutic approach to the patient.
PICU: pediatric intensive care unit; AVM: arteriovenous malformation.
Source: Own elaboration.

## DISCUSSION

AVMs are lesions characterized by the presence of arteriovenous shunting through a nidus of coiled and tortuous vascular connections that connect afferent arteries to draining veins (9). Brain AVMs are rare and have an estimated prevalence of 0.05% among otherwise healthy individuals (10). The incidence of brain AVMs in pediatric patients is rare but leads to a higher rupture rate than their adult counterparts, which could be attributed to the detection of most pediatric AVMs only after they rupture (11). Thus, it has been established that the prevalence in the pediatric

population is 0.02 and the annual incidence of symptomatic AVMs is 1.1 per 100 000 cases (3). Furthermore, although AVMs can occur in any region of the brain, it is extremely rare for them to involve the choroid plexus of the ventricular system (12), as was the case presented here. Although AVMs have been consistently regarded as congenital malformations of the brain vasculature, case reports have described de novo AVMs in adults (13).

Brain AVMs are often clinically silent, but the most common clinical symptoms include hemorrhage, seizures, progressive neurological deficit, and headaches (14). In the reported case, the initial symptoms were nonspecific; however, imaging revealed intracranial hemorrhage and associated hydrocephalus. The most common initial symptomatic presentation of AVM is hemorrhage, which is usually intraparenchymal, subarachnoid, or intraventricular (as in the present case), but it can also be subdural with a lower frequency (15).

Regarding complications, intraventricular bleeding is associated with significant morbidity and mortality since there is a risk of hemorrhage due to continuous or recurrent bleeding (16). Some studies, such as Fukuda *et al.* (17), have demonstrated that, after the initial hemorrhage, 26% of patients have no neurological deficit, 29% have mild to moderate deficits, and 45% have severe deficits. The risk of rupture and re-rupture of an AVM persists until it is completely obliterated, so the goal of treatment is to eliminate the likelihood of further hemorrhage that could be fatal or cause severe neurological deficits (18).

The diagnostic imaging method of choice for diagnosing AVMs is conventional brain angiography, which allows evaluating the characteristics of the angioarchitecture of the malformation and, therefore, to better plan the treatment. In the case presented, CT angiography was the first diagnostic study performed, but considering that the institution had interventional radiology equipment, a panangiography was performed due to its diagnostic capacity.

In the present case, the AVM was classified as type 2 on the Spetzler-Martin scale due to its size, location, and deep venous drainage. This classification indicates a favorable probability of obtaining good outcomes with surgical treatment (19), as was the case with the treatment provided.

Invasive treatment is recommended for younger patients with one or more of the high-risk features for an AVM rupture, but in older individuals without high-risk features, the usual best treatment is medical management. Open microsurgical excision is the mainstay of treatment and offers the cure for patients considered at high risk of bleeding. Radiation therapy and endovascular embolization are useful alternatives to surgical treatment in patients at high risk for surgical therapy and may also be useful adjuncts to the main surgical treatment (20).

Although AVM is a rare condition, health personnel who care for the pediatric population must consider it in the differential diagnosis of patients with neurological symptoms, as some studies suggest that mortality is higher in the pediatric

population than in adults, possibly due to a higher probability of hemorrhage in children (21). The timely treatment provided to the patient in the case reported (despite the presentation of nonspecific symptoms) resulted in an adequate clinical progress of the patient, unlike other cases reported in the literature (22,23). This confirms the need to perform imaging studies to rule out AVM and/or intraventricular hemorrhages in the presence of neurological symptoms in children, to provide timely initial management while the medical team performs exhaustive studies that allow establishing a definitive diagnosis.

In our case, the patient was successfully treated and showed a satisfactory clinical course despite the delay in the definitive treatment due to the problems arising from the measures implemented to control COVID-19, which had repercussions in multiple procedures. In this sense, it is essential to encourage the promotion and prevention of this type of conditions to avoid underdiagnosis and reduce the neurological sequelae and associated morbidity and mortality in these patients.

## CONCLUSIONS

Due to the low prevalence of brain AVMs, the available literature on the subject is scarce, which means that suspicion and treatment of AVMs depend on the expertise of the treating physician. In this regard, in the presence of nonspecific neurological symptoms, imaging studies should be performed as a priority to obtain a timely diagnostic impression and establish proper medical care, thus avoiding a fatal outcome.

## PATIENT'S PERSPECTIVE

The patient's parents agreed with the actions taken during their daughter's hospital stay and consented to share the case with the authors to contribute to the scientific literature.

## ETHICAL CONSIDERATIONS

An informed consent form was signed by the patient's father, who authorized the publication of his daughter's medical records.

## CONFLICTS OF INTEREST

None stated by the authors.

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