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REVIEW

A case report: Spetzler Martin grade III arteriovenous malformation

Reporte de un caso: malformación arteriovenosa grado III de Spetzler Martin

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ABSTRACT

Arteriovenous malformations are defined as an abnormal connection between the arteries that supply the brain tissue and the drainage veins forming a communication through a network of vessels in the brain tissue.It is presented by congenital errors in vascular morphogenesis caused by failures. Barrera (2018). This article describes a case of a 14-year-old patient with no apparent pathological history who presented with a generalized headache of a pulsatile nature without amelioration, for a period of 6 months of evolution.

Keywords: Arteriovenous malformations; Computed tomography; magnetic resonance imaging with contrast; Cerebral angiography; Spetzler-Martin grading scale.

RESUMEN

Las malformaciones arteriovenosas se definen como una conexión anormal entre las arterias que irrigan el tejido cerebral y las venas de drenaje, formando una comunicación a través de una red de vasos en el tejido cerebral. Se presentan por errores congénitos en la morfogénesis vascular causados por fallas. Barrera (2018). En este artículo se describe el caso de un paciente de 14 años, sin antecedentes patológicos aparentes, que presentó cefalea generalizada de carácter pulsátil, sin mejoría, con un tiempo de evolución de 6 meses.

Palabras clave: Malformaciones arteriovenosas; tomografía computarizada; resonancia magnética con contraste; angiografía cerebral; escala de clasificación de Spetzler-Martin.

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INTRODUCTION

Cerebral arteriovenous malformations (AVMs) are anomalies of cerebral vascular development located in the subpial region and can generally be parenchymal or dural. They are classified according to their location, size, and angioarchitecture.(1)

Arteriovenous malformations are relatively rare, with an incidence of 1-2 cases per 100 000 inhabitants. Most are diagnosed before the age of 30.

This type of malformation is believed to be due to abnormalities in the development of the cerebral vascular system during pregnancy and childbirth, but they can also occur sporadically or as manifestations of syndromic lesions. A combination of genetic factors, such as mutations in the MAP2K1 gene together with mutations in the RASA1, EPHB4, ENG, BMP9, ACVRL1, SMAD4, and BMPR2 genes, and environmental factors may contribute to their development.(1)

They progress slowly and can cause bleeding, epilepsy, and neurological deficits due to ischemia and hypoxia. Blood pressure and vascular resistance may increase the risk of bleeding.(2)

The diagnosis of arteriovenous malformations is based on clinical evaluation and images generated by imaging techniques such as computed tomography (CT) and magnetic resonance imaging (MRI).

Cerebral angiography is a more specific diagnostic technique that allows abnormal connections between arteries and veins to be visualized and is usually the option that distinguishes this condition from other diseases.

On the other hand, treatment will depend on the patient and the characteristics of the arteriovenous malformation, such as its size and location. Treatment options include surgery, angiographic embolization, radiosurgery, and conservative treatment.

The Spetzler-Martin grading system is the most widely used therapeutic system due to its simple application in analysis, based on size, location, and venous drainage.(3)

The score ranged from 1 to 5. The final score is calculated by adding the points for each category, and the degrees of cerebral arteriovenous malformations range from grade I to grade V, considering that the risk is progressively higher after grades I and II.(3)

Table 1. Spetzler Martin Grading Scale

Variables	Characteristics	Points
Size	Small (<3 cm)	1
	Medium (3-6 cm)	2
	Large (>6 cm)	3
Eloquent	Not eloquent	0
	Eloquent	1
Venous drainage	Superficial	0
	Deep	1

According to the author Guizado Infante (4), it is concluded that the risk is progressively higher from small, medium/deep, and medium/eloquent.

By adding up the points obtained, different degrees of risk are established; degrees one and two (1-2 points) present low morbidity and mortality, while degree three presents an intermediate risk (3 points) and degrees four and five a high risk (4-5 points), ranging between 30-50 % according to some prospective studies.(4)

Theoretical basis

- Alteration of cerebral blood flow: Arteriovenous malformations are characterized by abnormal direct connections between arteries and veins, without the normal intermediate capillary bed. This creates a vascular short circuit that diverts arterial blood flow, depriving portions of brain tissue of oxygen and nutrients, which is observable in contrast imaging studies.(5)
- Risk of hemorrhage: The walls of blood vessels in arteriovenous malformations are more weak and prone to rupture, which can cause intracranial hemorrhages. These hemorrhages can be massive and life-threatening.(6)
- Mass effect: Arteriovenous malformations (AVMs), especially large ones, can exert a mass effect on the surrounding brain tissue, causing compression and displacement of vital structures. (7,8,9,10)

- Impaired neurological function: Depending on the location of the arteriovenous malformation, areas of the brain responsible for functions such as language, motor skills, and sensitivity may be affected. This manifests clinically as focal neurological deficits.(10)
- Increased intracranial pressure: The presence of arteriovenous malformation and vascular shunting can increase pressure within the skull, which can cause headache, nausea, vomiting, and altered consciousness.(5)

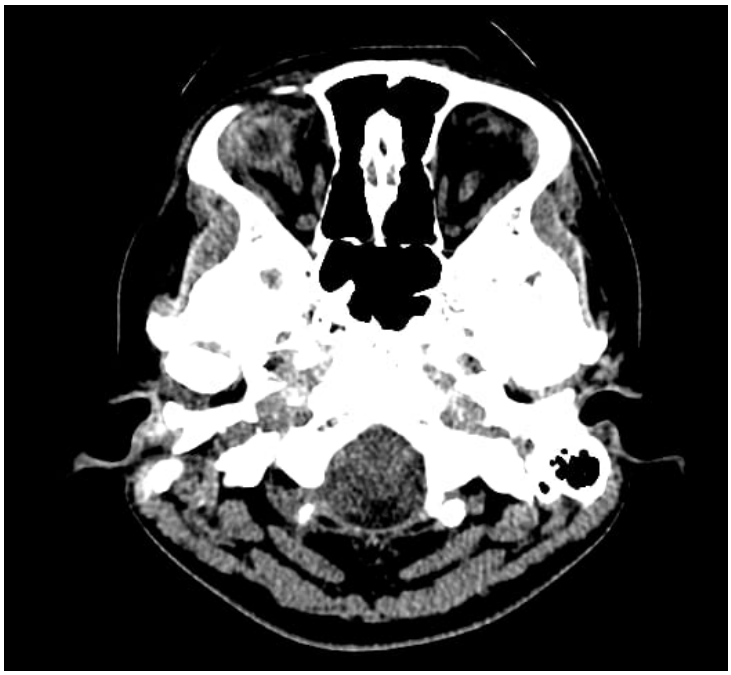
CASE REPORT

A 14-year-old female patient with no apparent medical history presented with global pulsating headache of six months' duration without acalmia, which began in mid-2017 and had been chronic since onset. The headache reached an intensity of 7/10 on the visual analog scale (VAS), forcing the patient to lie down to relieve the symptoms. There was no vomiting, photophobia, or ataxia, which is why she sought medical attention and it was decided to perform additional tests.

An evaluation of the autonomic nervous system was performed, including RR interval variation and sympathetic skin testing. The results showed a normal RR interval at rest, normal cardiovagal response, and normal plantar and palmar responses.

On July 21, 2018, a computed tomography scan was performed, showing a heterogeneous image located in the right semioval center, in which hyperdense areas are seen with others of lower density. It is not associated with edema or mass effect.

Figure 1. Computed tomography of the patient

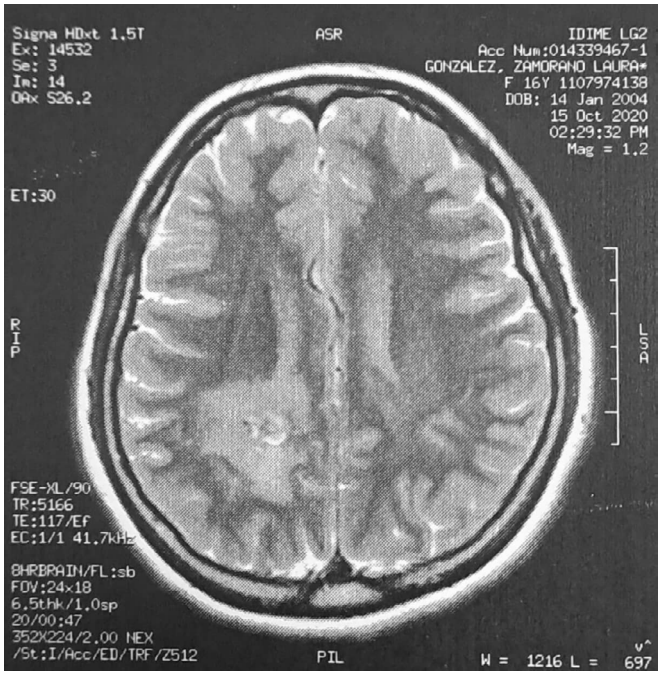


Source: Study conducted at Open Side, Panama

An electroencephalogram performed on July 20, 2018, showed low-amplitude beta rhythm, slow theta waves, and activity characteristic of stage 1 sleep, including attenuation of alpha rhythm. A rare epileptiform disturbance was identified in the right frontal region, associated with focal discharges. Valproic acid (500 mg) was initially prescribed but was discontinued due to poor tolerance; it was subsequently replaced by flunarizine. The patient used acetaminophen (2 g daily) to manage her headaches.

On August 1 of the same year, a gadolinium-enhanced brain MRI revealed dilated vascular structures in the right anterior parietal region, forming a nest measuring approximately 20 x 15 mm. The nest was supplied by a branch of the middle cerebral artery and the right pericallosal artery, with drainage to the Galen vein and the superior sagittal sinus. This finding led to a diagnosis of right parietal plexiform arteriovenous malformation (AVM).

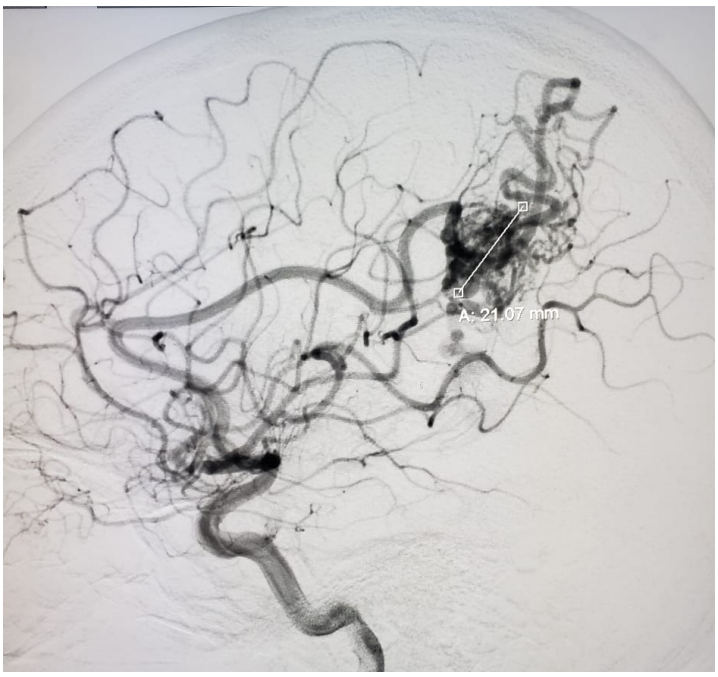
Figure 2. Brain MRI with gadolinium



Source: Study conducted at Open Side, Panama

Following this study, a cerebral angiography was performed to confirm the diagnosis, revealing a deep medial parietal arteriovenous malformation, a compact nest 18 mm high and 15 mm wide, afferent to the right pericallosal artery (tortuous), deep venous drainage through tortuous parietal vessels and tension that flow into the Galen vein. No internodal aneurysms.

Figure 3. Cerebral angiography



Source: Procedure performed at the Institute of Medical Diagnosis, Colombia

DISCUSSION

The investigations carried out corroborated a grade III arteriovenous malformation (AVM) in the brain, according to the Spetzler-Martin scale, which indicates a moderate risk of morbidity and mortality.

Computed tomography showed a varied image located in the right semioval center, without swelling or mass change, which facilitated the identification of initial features that could be compatible with arteriovenous malformations without serious problems.

Subsequently, gadolinium-enhanced magnetic resonance imaging revealed a dilated vascular nest measuring approximately 20 x 15 mm, fed by branches of the right middle and pericallosal arteries, with deep drainage into the Galen vein. This discovery was essential to confirm the diagnosis of plexiform arteriovenous malformation and to further investigate the angioarchitecture of the nest.

Finally, cerebral angiography corroborated these findings, describing a compact, h y nest with tortuous arterial afferents and deep drainage without the presence of internodal aneurysms, which reduced the risk of additional hemorrhagic complications.

These findings have significant clinical and therapeutic implications. The validation of grade III according to the Spetzler-Martin scale, based on size, location, and venous drainage, underscores the importance of multidisciplinary management.

Treatment options for arteriovenous anomalies include embolization, radiation, neurosurgery, and a combination of techniques.

Factors such as the following should be considered when choosing the correct intervention:

Table 2. Factors to consider for the intervention of an arteriovenous malformation

Natural progression of the disease	If there is bleeding and it is not treated immediately, it could recur bleed again in 6 to 17 % of cases during the first year. If there has been no bleeding, the risk after 2-4 years is 4 %.
Age	The risk of bleeding from a malformation that has not bled is 2-4 %, which is cumulative.
Symptoms	If symptoms are present, there is a higher chance of bleeding.
Radiological findings	If the malformations are small, if the nests are located in the periventricular or intraventricular area, if the venous drainage is deep or single, and if there are intranidal or pedicular aneurysms.

Source: (11)

CONCLUSION

The clinical case of our patient with cerebral arteriovenous malformation (AVM) was approached with a multimodal approach that included embolization and gamma knife treatment. These procedures, performed to reduce abnormal blood flow and minimize the risk of cerebral hemorrhage, are recognized interventions for their effectiveness in the management of AVMs.

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CONFLICT OF INTEREST

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