

## Differential diagnosis in encephalic vascular malformation in a child: Case report

*Diagnóstico diferencial de malformação vascular encefálica em uma criança: relato de caso*

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### ABSTRACT

**Aims:** To report a case of encephalic venous malformation, which is not commonly described in the literature, despite its importance as one of the main differential diagnosis in intracranial hemorrhage in children.

**Case Description:** A five-year old girl presented chronic headache and had the first episode of seizure. Possible intracranial alterations were investigated as etiological factors, and an alteration in the Labbé vein was identified, possibly associated to a cavernoma in the left temporal region, which caused the hemorrhage.

**Conclusions:** Primary seizures and headaches are relatively common in children. However, secondary seizures such as those caused by hemorrhage, despite being less common, must be suspected and investigated, as they may lead to severe complications.

**KEY WORDS:** INTRACRANIAL ARTERIOVENOUS MALFORMATIONS; CEREBRAL VEINS; INTRACRANIAL HEMORRHAGES; DIAGNOSIS, DIFFERENTIAL; EPILEPSY; CHILD; CASE REPORTS.

### RESUMO

**Objetivos:** relatar um caso de malformação venosa encefálica, não comumente descrito na literatura, apesar de sua importância como um dos principais diagnósticos diferenciais em hemorragia intracraniana em crianças.

**Descrição do Caso:** uma menina de cinco anos de idade apresentava cefaleia crônica e teve o primeiro episódio de convulsão. Possíveis alterações intracranianas foram investigadas como fatores etiológicos, e foi identificada uma alteração na veia de Labbé, possivelmente associada a um cavernoma na região temporal esquerda, o que provocou a hemorragia.

**Conclusões:** convulsões primárias e cefaleia são relativamente comuns em crianças. No entanto, convulsões secundárias, tais como aquelas causadas por hemorragia, apesar de serem menos comuns, devem ser suspeitadas e investigadas, pois podem levar a complicações graves.

**DESCRIPTORIOS:** MALFORMAÇÕES ARTERIOVENOSAS INTRACRANIANAS; VEIAS CEREBRAIS; HEMORRAGIAS INTRACRANIANAS; DIAGNÓSTICO DIFERENCIAL; EPILEPSIA; CRIANÇA; RELATOS DE CASOS.

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## INTRODUCTION

Arteriovenous malformations correspond to 36% of the cases of encephalic vascular malformations. They have the shape of a worm tangle and are mainly supratentorial (67%).<sup>1-3</sup> Although 50% are asymptomatic, the most common initial presentation is hemorrhage (40-50%), being these malformations the leading cause of intracranial hemorrhage among those under 15 years of age.<sup>1,2,4,5</sup> The second most common presentation is seizure, in 30% of the cases; 20% of those as generalized tonic-clonic seizure.<sup>1,3</sup>

Venous malformations or abnormalities of venous development consist of a net of abnormal enlarged veins that centripetally drain to a dilated venous trunk without an arterial component and separated by normal neural tissue. Supratentorial hemorrhages rarely occur, though being more common in the malformations of posterior fossa.<sup>2,6</sup> Angiomas or cavernomas are responsible for 7 to 8% of these alterations. They are an agglomeration of thin veins without important nourishing arteries with little or none nervous tissue in between, commonly associated with abnormalities of venous development. The clinical presentation is similar to the AVM's, even though they are more commonly asymptomatic. They are generally less than 1 mm wide, yet they can become as wide as several centimeters, and 15 to 20% calcify. If they are incidentally found, they are generally conservatively treated, due to the low potential of bleeding or symptomatology.<sup>1,3</sup>

For children who present their first seizure, strong headache or a subtle change in the characteristics of a previous headache, as well as nuchal rigidity, it is essential that intracranial hemorrhage be ruled out as a cause. Brain computerized tomography (BCT) is the most adequate exam for screening; however, to define the etiology of the bleeding, brain angiography is the gold-standard procedure.

This article presents and discusses the possible differential diagnosis in a case of venous malformation suspicion in a 5-year-old girl. As the symptoms presented are very common, it is important to discuss their differential diagnosis, in order to avoid missing the diagnosis of such a severe disease.

The patient's legal guardian signed an informed consent form, in accordance with the resolution 196 of the Brazilian National Health Council.

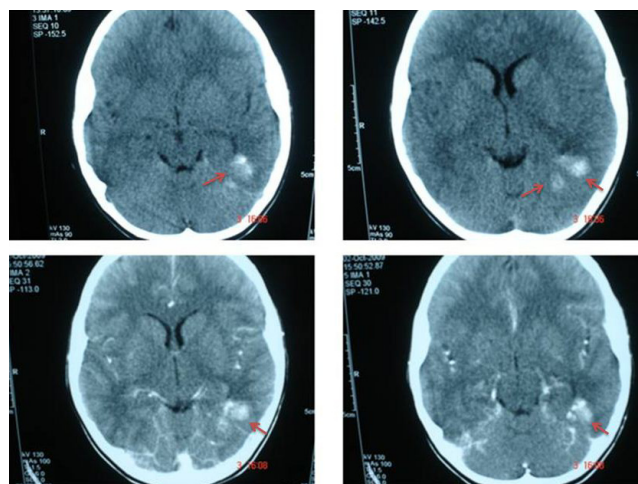
## CASE REPORT

A white five-year-old girl was first seen in the emergency room, complaining of having fainted the day before. According to her mother, the patient

was on the car's back seat when she vomited, lost consciousness and her limbs became rigid for twenty minutes. After that, she woke up, with no memory of what had happened. There were no involuntary movements or other alterations.

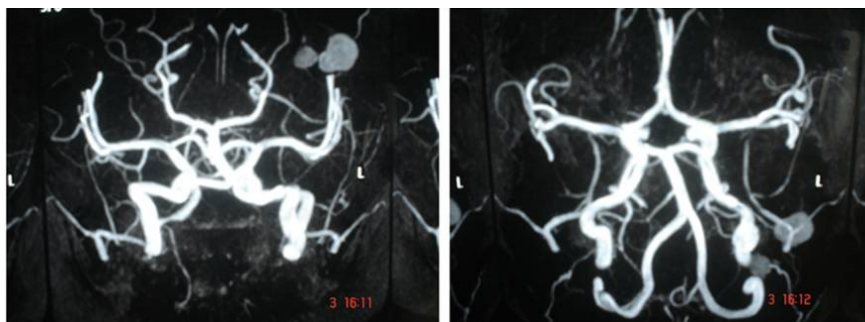
The patient presented a chronic headache, whose characteristics were hard to define; she had had no complications during her birth or neuro-psychomotor development. Her father had presented seizures during adolescence, which had been adequately treated.

Her neurological exam presented no alterations. A BCT was required, which revealed a hyperdense lesion in the left temporal region as well as contrast keeping in the area (Fig. 1). These findings were interpreted as a hemorrhagic stroke, due to an arteriovenous malformation. The patient was admitted and a magnetic resonance imaging (MRI), an angioresonance and an angiography were required. The cerebral spinal fluid presented no alterations and she was medicated with phenytoin 100 mg a day.

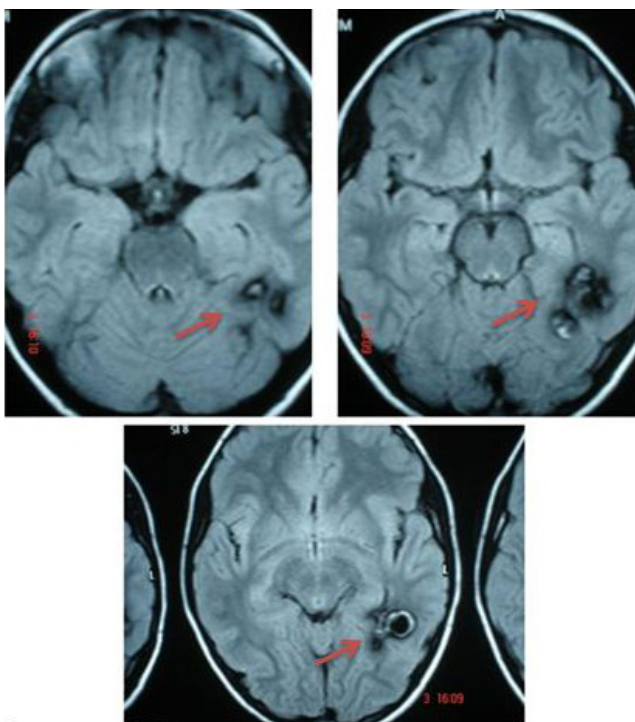


**Figure 1.** Brain computed tomography, above without contrast, below with contrast. Subcortical spontaneous hyperdense nodular heterogeneous lesions in left temporal region, keeping of contrast. Discrete perilesional hypodense blur suggestive of edema or re-absorption

The patient had the MRI performed (Figs. 2 and 3) and was discharged the following day, with a brain angiography still pending and a prescription of carbamazepine, 10 mg/kg/day. She presented no new episodes of seizures since then, and given the imaging findings the diagnosis was defined as an abnormality of the venous development (a looping in Labbé's vein) (Fig. 4), associated with a bleeding cavernoma.



**Figure 2.** Brain angioresonance showing normal arterial phase, circle of Willis without malformed or aneurismatic dilation.

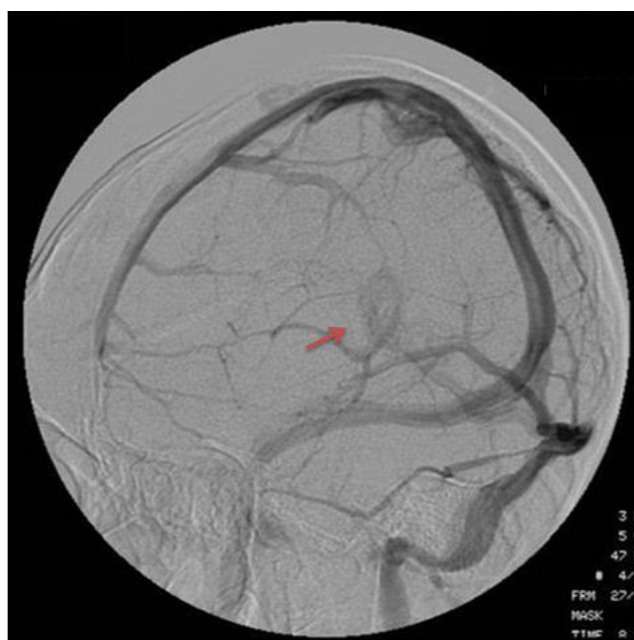


**Figure 3.** Brain magnetic resonance imaging showing small subcortical heterogeneous focal lesions on left temporal region, with hyper attenuation in flair sequence, suggestive of hemorrhagic focuses and degradation of hemoglobin with desoxyhemoglobin in the center and hemosiderin in the periphery.

## DISCUSSION

The first necessary step in a case like this is to identify the origin of the seizure. Metabolic disorders, as changes in blood sugar and electrolytes, should be first excluded. At the same time it is important to identify any intracranial alteration. In this case, a BCT is the most indicated exam. As a result, an alteration compatible with bleeding, calcification or an active lesion was found in this case. More imaging resources were required for the differential diagnosis.

When a child presents an acute headache or subtle change of its characteristics, or the first seizure, a vessel



**Figure 4.** Venous angiography with no aneurismatic or malformation lesion. No tumoral blush. A loop of the Labbe vein on the left.

malformation must be considered. Malformations make the vessel's wall thinner and more prone to bleeding, thus causing neurological symptoms through compression and edema of nearby areas, and may calcify. In this case, the symptoms were probably caused by a subtle bleeding and its edema. Even though displasias are more related to seizures when venous malformations are concerned, it is possible for a bleeding to trigger a crisis, and its edema to cause loss of conscience.<sup>7</sup>

The MRI identified the lesion on the posterior part of the left temporal lobe. Four cases in which there was a venous malformation on the left temporal lobe were presented by Pereira et al.<sup>7</sup> in 2008. Three of them had

initially a seizure, two were associated to headache, and one presented proptosis and pain in the left eye, associated with focal atrophy in the MRI. One of the cases showed focal edema; another presented a venous infarct, and the last had a hemorrhage (both associated with seizure and headache).<sup>7</sup>

Bergui<sup>8</sup> described a case of parenchymatous hemorrhage of the left temporoinular region, manifested as headache, aphasia and right hemiparesis. Koc et al.<sup>9</sup> presented a case of left temporal parenchymatous hemorrhage that showed aphasia and right hemiparesis. Seki<sup>10</sup> reported a case of congestive hemorrhage on the left temporoparietal region, with headache, right hemiparesis and coma.

Clinical presentation, therefore, may be variable depending on the extension of the affected area. In the case described herein, the lesion was predominantly temporal and, in its most inferior region, near the insular lobe. Of the seven cases found in the literature, three had a seizure as the initial presentation, and four a headache of acute onset.

Differential diagnosis of headache may be quite hard. The most common causes for primary headache in children are migraine and tension-type headache; most cases disappear during child's development. Defining the type of headache that this patient presented was complicated. We could not define whether she had a chronic primary headache, or whether she had recurrent acute headaches, due to repetitive intracranial bleedings.

Considering the location and the characteristics of the malformation, no surgical procedure could be performed. Thus, the most adequate treatment is preventing new seizures and being attentive to new bleedings, which can be presented as an intense headache of subtle onset, seizures or focal neurological deficits. New crisis were prevented initially by prescribing phenytoin, the most commonly used drug for preventing seizures. After the diagnosis and at the time of hospital discharge, carbamazepine was preferred, due to the characteristics of the disease.

If no new bleedings and/or seizures occur (or other secondary complications), it is possible that no surgical procedure will be necessary.

It is important for the general practitioner or the pediatrician to be aware of the secondary causes of seizures and the importance of performing a BCT for its differential diagnosis. It is also extremely important that the patient is treated at once, even before defining the diagnosis, in order to avoid complications and further lesions to the brain tissue.

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