

Furthermore high vascular lesions may present high perfusion as well as brain cancer.

The symmetrical calcifications and the diffuse enhancing nodular lesions are other important findings that suggest a diffuse cerebral condition. Actually the nodular enhancing lesions constitute pre-cystic lesions as same cases show in the literature. Actually the leucodystrophy described in this entity is rather related to vasogenic edema and gliosis surrounding the cysts and the nodules<sup>1-5</sup>.

LCC is a very rare disease and less than 30 cases reported since the original description by Labrune and colleagues, all of them with characteristically infantile or juvenile onset<sup>1-3</sup>. Sener et al. were the first to suggest the existence of an adult form of this disease, based on the observation of a slow clinical progression in a patient with onset in late adolescence<sup>4</sup>. Wargon et al. report a 30-year-old woman with a lacunar infarct as the first manifestation of LCC<sup>5</sup>. In our patient, the diagnosis of LCC was based on the highly suggestive neuroradiological findings which were almost identical to previous report. Our biopsy showed no neoplastic cells, but clotted blood within the lesion.

LCC is an entity can present with cystic expansive lesions with mass effect, hemorrhagic content and high vascular density that can be misunderstood as neoplastic lesions. Symmetrical brain calcifications and dif-

fuse enhancing hemorrhagic nodules are the other crucial findings. These abnormalities points out that this entity should rather be described as a vascular entity in the books than as a primary leucodystrophy.

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## LEUCOENFALOPATIA, CALCIFICAÇÕES CEREBRAIS E CISTOS: ENTIDADE QUE PODE SIMULAR UMA NEOPLASIA

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# Pial arteriovenous fistula in the posterior fossa

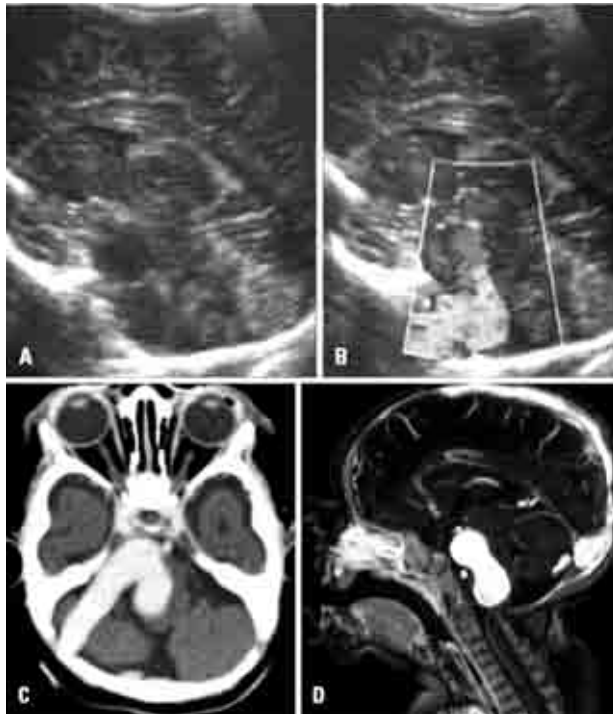
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Intracranial pial arteriovenous fistulas (AVFs) are rare congenital abnormality that can cause severe morbidity and mortality, particularly in neonates<sup>1</sup>. AVFs are rare cerebrovascular lesions of the brain that were considered to be distinct from other arteriovenous malformations (AVMs) by Lasjaunias, in 1986<sup>2</sup>. Intracranial pial AVFs have a single or multiple arterial connections to a single venous channel. They differ from brain AVMs in that they lack a true nidus. They differ from dural AVFs in that they derive their arterial supply from pial or cortical arteries and are not located within the dura mater<sup>3</sup>. They may be localized anywhere in the brain but display a preference for the supratentorial regions<sup>3</sup>.

Pial AVFs can be acquired traumatically or iatrogenically or may be congenital. Their natural evolution is unfavorable, conservative management of pial AVFs has been associated with mortality in 63% of patients<sup>4</sup>.

## CASE

A 6-month old boy present macrocephaly, asymptomatic, without apparent motor injury, did not show any skin lesion and no signs of heart failure. A transfontanele ultrasound Doppler showed evidence of large arteriovenous shunt (AVS) at posterior fossa with patent flow (Fig 1A and 1B). Additional investigation with computed tomography (Fig 1C) and magnetic resonance



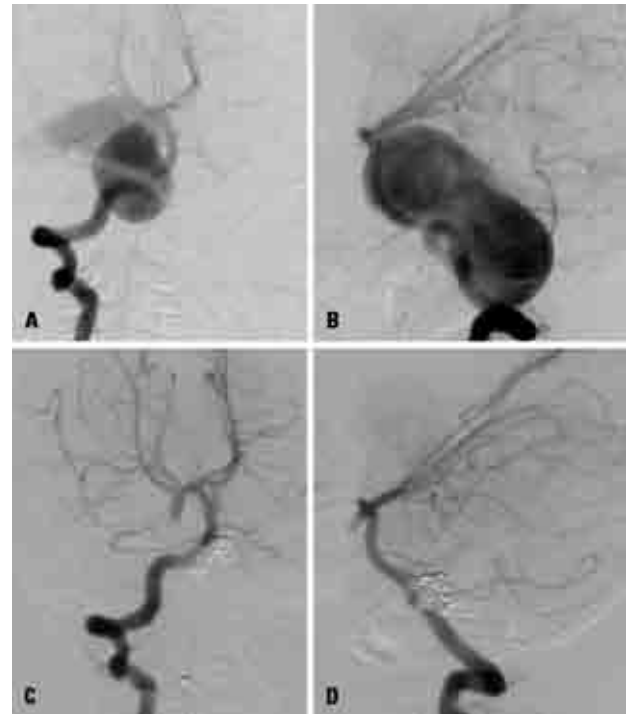
**Fig 1.** [A and B] Color Doppler transfontanelle ultrasound showing a tubular image with patent flow in posterior fossa. [C] CT scan demonstrated large vascular structure occupying part of posterior fossa. [D] MRI T1 weighted in sagittal views, demonstrated the extent of the injury and the compressive effect on the brain stem.

showed extensive vascular mass partially occupying posterior fossa, leading to displacement and compression of the brainstem and no signs of hydrocephalus. The magnetic resonance (Fig 1D) and digital subtraction arteriography (Fig 2A and 2B) confirmed AVF with single feeder originated from the vertebrobasilar junction and single draining in cerebellar vein toward torcule.

The AVF was treated via an endovascular route with detachable coils. Elective embolizations of the AVFs were done with coils at 6 months of age, resulting in complete exclusion of the lesions, as shown on the postembolization internal carotid angiogram on anterior posterior and lateral views (Fig 2C and 2D).

## DISCUSSION

Intracranial pial AVFs are rare cerebrovascular lesions. They are a direct communication between a pial artery and cerebellar vein, without an intervening nidus and located in the subpial meningeal space. This newly distinguished subtype of intradural AVS is necessary because it corresponds to an architecture that engenders specific symptoms, occurs in a special age group, constitutes the phenotype of a well-defined disease, and requires a specific therapeutic technical approach<sup>5</sup>.



**Fig 2.** [A] Angiography of right internal carotid artery anterior posterior views and in lateral views [B] confirmed AVF with single feeder originated from the vertebrobasilar junction and single draining in cerebellar vein toward torcule. [C] and [D] Postembolization internal carotid angiogram on anterior posterior and lateral views.

This pathology has different clinical presentations. Neonates can present with intractable cardiac failure and intracranial bruit. In infancy, it can present with hydrocephalus and seizures. Older children or young adults can present with intracranial hemorrhage<sup>1</sup>.

Owing to the absence of a nidus, closure of the shunt by either endovascular or surgical technique represents a satisfactory therapeutic procedure<sup>2</sup>. But the indications, the time, and the method of treatment chosen for management of AVFs depend on a variety of factors such as the age at presentation, presenting symptoms, the status of the brain, and the status of the other organs<sup>5</sup>.

In conclusion, emergent diagnosis and treatment of this pathology is crucial due to its associated with high morbidity and mortality. Endovascular techniques can provide safe and successful treatment for these infants.

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#### FISTULA ARTERIOVENOSA PIAL NA FOSSA POSTERIOR

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## Extradural cavernous hemangioma of thoracic spine

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Cavernous hemangioma of the central nervous system is a vascular malformation which is a developmental hamartoma, also known as cavernoma or cavernous malformation or venous angioma<sup>1</sup>.

Cavernomas may affect any segment of the neuraxis. Most of these malformations are intracranial: supratentorial compartment is a site usually affected<sup>2,3</sup>. Pure spinal epidural cavernomas represent approximately 12% of spinal cavernous anomalies<sup>4</sup> and the thoracic segment is the most frequently affected<sup>5</sup>. There are approximately 80 cases of epidural cavernous hemangioma published in the literature<sup>4,5</sup>.

We report on a case of thoracic extradural cavernous hemangioma, with emphasis on the clinical aspects.

#### CASE

A 63-years-old female caucasian patient with a previous history of cryptogenic epilepsy was referred to our emergency department with complaints of a one year history of gait ataxia, numbness and dysesthesia in both lower limbs. In the last 2 months, she also noticed reduced strength in the lower limbs.

Neurological examination showed reduced strength in the lower limbs, grade 4/5 in the right lower limb and grade 3/5 in the left one. There was reduced sensation for all sensory modalities below the T10-11 dermatomes. There was also lower limb hyperreflexia and hypertonia, with bilateral extensor plantar response.

An MRI showed an epidural space-occupying lesion involving the T9-T10 spinal canal levels and extending to



**Figure.** Preoperative MRI. Sagittal T2-weighted image showing isodense lesion in the posterior extradural space at T9-T10 with ventral displacement of the spinal cord.

the T9 left foramen, with an intense and homogeneous contrast enhancement (Figure).

The patient was submitted to a T9-T10 laminectomy, including a T9 left foraminectomy. A posterior soft and reddish epidural mass with a good cleavage plane was completely removed. The patient evolved with partial improvement of sensibility in both lower limbs and strength.

Histological examination revealed thin-walled blood vessels lined with a single layer of endothelial cells, surrounded by connective tissue. The microscopic appearance was compatible with cavernous hemangioma.