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Case Report

Massive Intracranial Calcifications Secondary to Vein of Galen Aneurysmal Malformation

Letícia Oliveira Faleiros[1], Joelma Karin Sagica Fernandes Paschoal[1], Eric Homero Albuquerque Paschoal[1], Eduardo Ferracioli Fusão[1], Ricardo Silva Pinho[1], Antonio José da Rocha[2], Luiz Celso Pereira Vilanova[1], Marcelo Rodrigues Masruha[1]*

[1]Department of Neurology and Neurosurgery, Federal University of São Paulo, Brazil

[2]Department of Radiology, Santa Casa de São Paulo, Brazil

***Corresponding author:** Marcelo Rodrigues Masruha, Department of Neurology and Neurosurgery, Division of Child Neurology, Federal University of São Paulo, Botucatu Street, São Paulo 720, Brazil, Zip Code: 04023-900, Email: mmasruha@ig.com.br

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Abstract

Massive intracranial calcifications are observed in several diseases, including arteriovenous malformations. The most frequent paediatric neurovascular malformations are vein of Galen arteriovenous malformations, followed by pial arteriovenous malformations and dural sinus malformations.

Most patients with arteriovenous malformations present initially with seizures, intracranial haemorrhage, or hydrocephalus. Patients may also present with progressive neurologic deficits including progressive hemiparesis, brainstem dysfunction or cognitive impairments. Eventually, an untreated arteriovenous malformation may progress to a chronic venous ischemia characterised by the development of dystrophic calcifications and subependymal atrophy with ventricular dilatation.

In this report, we present a case of massive intracranial calcifications in a two-year-old child with a vein of Galen aneurysmal malformation.

Keywords: Vein of Galen malformations; Arteriovenous malformations; Intracranial calcification; Magnetic resonance imaging; Tomography

Introduction

A vein of Galen aneurysmal malformation (VGAM) is a choroidal type of arteriovenous malformation. The lesion is supplied by the choroidal arteries. The choroidal shunt drains into a dilated vein, which is the median vein of the prosencephalon, the embryonic precursor of the vein of Galen. This embryonic vein drains only the choroidal system and does not connect with the deep venous system. It does not become the vein of Galen until communications with the thalamostriate and internal cerebral veins develop. In patients with VGAM, these latter communications do not form, and the thalamostriate veins drain into the posterior and inferior thalamic (diencephalic) veins and secondarily join the anterior confluence, a subtemporal vein, or (more often) the lateral mesencephalic vein to open into the superior petrosal sinuses, which demonstrate a typical epsilon shape ("epsilon vein") on the lateral angiogram [1].

In this report, we present a case of progressive neurologic impairment and early-onset multiple intracranial calcifications in a two year-old child with a VGAM. These findings are rare and have been poorly described previously in the literature.

Case Presentation

A two-year-old male patient was admitted to the emergency service with history of seizures and significant developmental delay. Birth, family, and medical history were unremarkable. Development was normal until seven months of age, after which the patient developed progressive loss of motor milestones. Approximately one year after the onset of the developmental decline, the patient developed excessive sleepiness, seizures, and loss of contact with the environment and his family.

On physical examination, he presented in an adequate nutritional status; head circumference was normal, without visceromegalies or evidence of cardiac failure, but with an irregular breathing pattern. The examination was remarkable for a bruit in the right mastoid. The neurological examination revealed a total absence of interaction

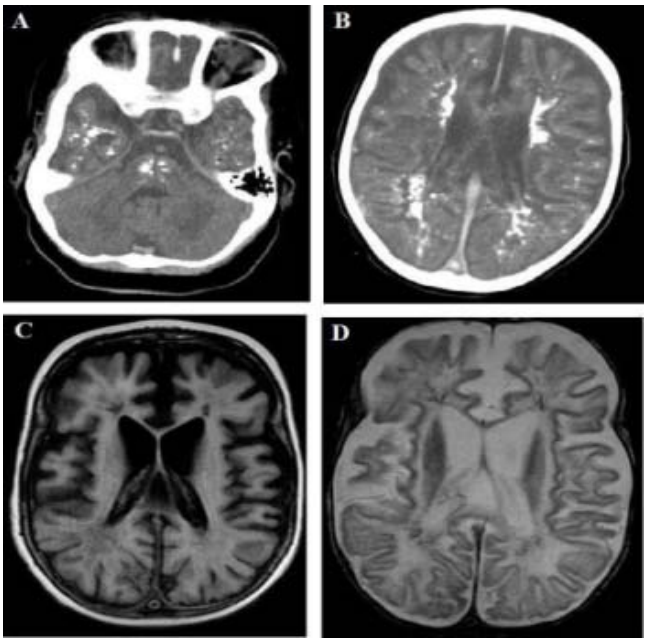

Figure 1: (A and B) Axial CT non-contrasted images showing multifocal supra- and infratentorial calcifications associated with a severe cortical and subcortical atrophy, indicating brain injury due to chronic venous hypertension. (C and D) T1 FSE (A-D) and T2 gradient-echo (T2* E-H) MR sequences on an axial plane, showing evident atrophy due to venous hypertension resulting in brain destruction. Note faint hypointensities on T2* corresponding to parenchymal calcifications.

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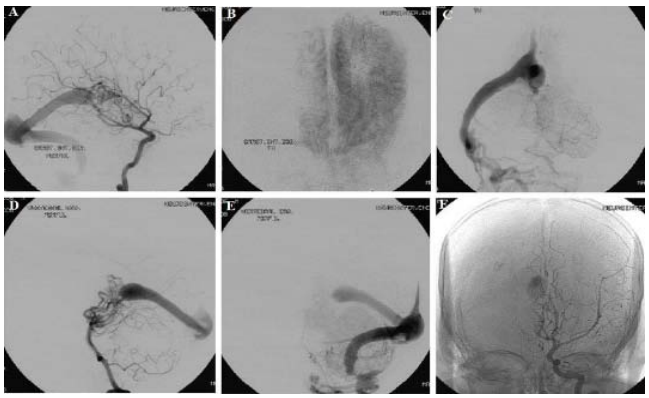

Figure 2: The digital subtraction cerebral angiogram showed (A) lateral right internal carotid findings of a shunt AV of the anterior choroidal artery with subependymal veins draining into the vein of Galen. (B) Capillary phase of the left internal carotid artery shows marked subcortical and cortical atrophy. (C) Occlusion of the left lateral sinus and bilateral stenoses of the jugular bulbs, after left vertebral artery injection. (D) A pial AVM at the quadrigeminal tectus nourished by the medial posterior choroidal and trans-mesencephalics arteries. (E) Vein of Galen dilated and signs of venous congestion caused by a right sigmoid sinus thrombosis. The cerebral venous outflow is performed with drainage by the cervical plexus. (F) Angiogram shows the arteriovenous shunt from the AVM and subcortical calcifications.

with either the environment or the examiner, failure to emit sounds, severe axial hypotonia, muscle weakness, signs of spasticity as well as pyramidal signs.

The imaging study showed extensive brain calcifications in the infra- and supratentorial structures, mild cortical atrophy and a markedly dilated venous sinus (Figure 1). Further angiography confirmed a vein of Galen aneurysmal malformation (VGAM) with chronic venous hypertension (Figure 2).

The patient's mother gave their informed consent prior to their inclusion in the study.

Comment

Clinical manifestations of patients who have VGAMs are mainly divided into those related to high output cardiac failure and those involving neurologic symptoms due to venous congestion and abnormal CSF flow. Intracranial venous hypertension resulting from the VGAM in combination with anatomic (the status of the cavernous sinuses and the degree of development of the base of the skull) and physiologic features (nonfunctioning pacchionian granulations) of the neonatal brain are the cause of neurologic deterioration. Their severity and tolerability are variable and are related to the angioarchitecture of the VGAM and the age of the child [1,2].

Symptoms in fetuses or neonates are generally the result of intracranial venous congestion or hemodynamic alterations owing to heart failure during intrauterine life and reflect early brain damage. In infants, macrocrania may be the first objective clinical sign of an abnormal central nervous system. An increasing head circumference is commonly associated with enlarged ventricles and generous perivascular spaces in patients with VGAMs. This fluid accumulation has little or no effect on the brain as long as the sutures enlarge, resulting in a balance between the intracranial pressure and the resistance of the cranial vault [3].

Untreated VGAMs evolve toward chronic venous ischemia manifested by the development of subcortical white matter calcifications and subependymal atrophy with ventricular dilatation. These calcifications reflect deep hydrovenous watershed failure, and they occur when the compliance of the medullary veins loses its normal ventricularcortical gradient. These calcifications are usually bilateral and symmetrical, but they may be asymmetrical and mostly unilateral in shunted children (often on the side opposite to the shunt). Subependymal atrophy is primarily seen in the occipital regions. It may be dramatic, and it seems to be at least partially related to an abnormal postnatal development of the corpus callosum. Spontaneous thromboses of isolated cortical veins are also possible in untreated patients with VGAM [1].

VGAMs should be included on the list of differential diagnosis for extensive intracranial calcifications, particularly with the peculiar distribution described herein.

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