

VEIN OF GALEN ANEURYSM IN AN ADULT

Case report

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ABSTRACT - Vein of Galen aneurysm is a rare pathology, representing less than 1% of intracranial vascular malformations. We report on a 65 years-old man who experienced a generalized tonic-clonic seizure. Brain imaging showed a large calcified expanding mass in the pineal region, confirming the diagnosis of a vein of Galen aneurysm. Because of the spontaneous thrombosis of the malformation, there was no need for microsurgical or endovascular treatment and he is being regularly followed since that.

KEY WORDS: vein of Galen aneurysm, vein of Galen malformation, cerebral vein, congenital vascular malformation.

Aneurisma da veia de Galeno em adulto: relato de caso

RESUMO - Aneurisma da veia de Galeno é patologia rara, representando menos de 1% das malformações vasculares intracranianas. Apresentamos o caso de um homem de 65 anos que teve episódio de crise convulsiva tônico-clônica generalizada. Exames de imagem evidenciaram grande processo expansivo calcificado na região pineal, confirmando o diagnóstico de aneurisma trombosado de veia de Galeno. Devido à trombose espontânea da malformação, foi excluída a possibilidade de tratamento endovascular bem como microcirúrgico, mantendo-se o acompanhamento clínico.

PALAVRAS-CHAVE: aneurisma da veia de Galeno, mal-formação da veia de Galeno, veias cerebrais, mal-formação vascular congênita.

Vein of Galen aneurysm (VGA) is a rare vascular malformation representing less than 1% of vascular intracranial abnormalities. It is a congenital process frequently detected between the 6th and 11th months of gestational age¹, during early childhood or neonatal period². This finding in adult age is very rare, presenting or not symptoms throughout childhood³. Clinical manifestations can present at any age in the form of heart failure, delayed neuropsychomotor development, hydrocephalus and seizures⁴.

We describe a case of VGA reporting clinical symptoms, radiological findings and management relating to current published literature.

CASE

A 65 year-old man was brought to the emergency room at the University Hospital of the Pontifical Catholic University of Paraná, Brazil because of generalized tonic-clonic seizures. On admission, he was treated with anticonvulsant therapy for status epilepticus. Past medical history was sig-

nificant for treated high blood pressure and heart failure diagnosed late in life. Also significant for an episode of ischemic cerebrovascular disease two years before, resulting in left hemiparesis. During childhood and adult life there was no neuropsychomotor development delay, no signs of cardiopathy, no neurological signs or symptoms as well as no history of epilepsy.

CT scan showed a calcified expanding process in the pineal region (Fig 1). MRI showed a heterogenic round-shaped mass localized in the supra-vermian cistern with dimensions of 35x32x30 mm showing high signal in T2-weight imaging (Fig 2). The MRI images were suggestive of a thrombosed VGA. Cerebral angiography showed a thrombosed VGA also (Fig 3).

Considering the fact that the aneurysm was already thrombosed and that there were no signs or symptoms of intracranial hypertension, nor hydrocephalus, the patient was not referred for surgery or endovascular treatment. Improvement in the level of consciousness and absence of seizures episodes were noted during the follow-up period while maintaining the clinical therapy for seizures.

The patient provided informed consent agreeing with this case publication.

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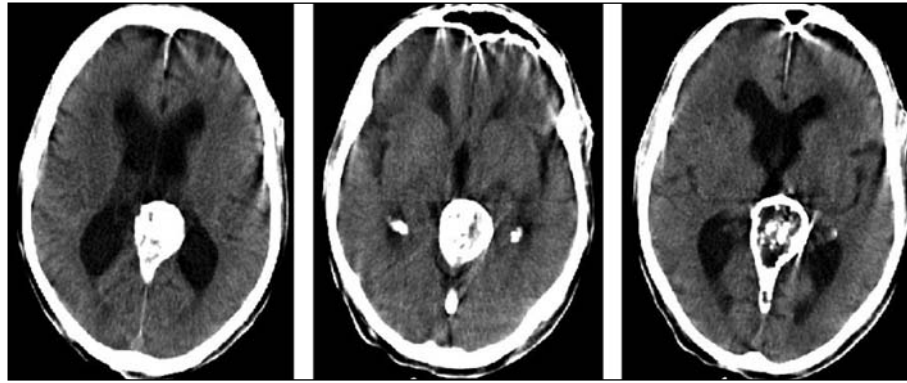


Fig 1. CT scan showing calcified expanding process at the pineal region.

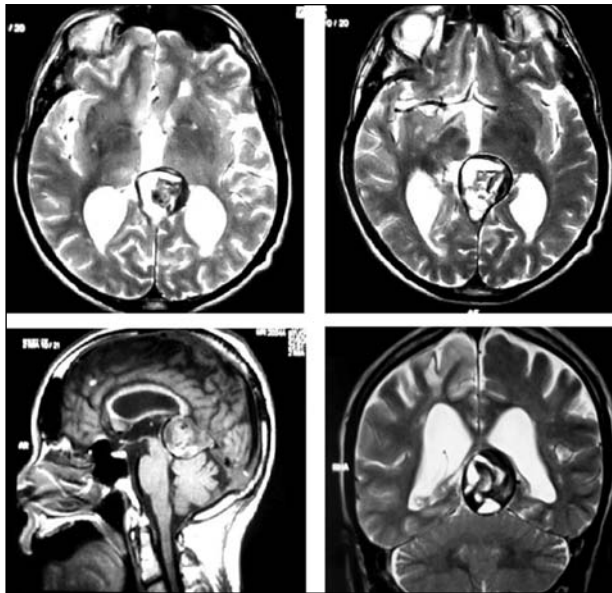


Fig 2. MRI showing thrombosed vein of Galen aneurysm.

DISCUSSION

VGA is a vascular abnormality typically found in children⁵. Raybaud et al.¹ believe it is a reminiscent of fetal anatomy produced by frequent occlusions of posterior fossa dural sinus, especially at the sigmoid sinus. Even though the VGA can be asymptomatic in adults^{6,7}, it is typically diagnosed during the neonatal period or in childhood with heart failure signs, macrocephaly and/or cranial murmurs⁸. Children with slow flow fistulas have a better extra uterine adaptation⁵. Adults usually present slow flow fistulas, headaches, seizures, hydrocephalus and rounded calcified masses in the pineal region⁸. Subarachnoid and intracerebral hemorrhages can occur because of blood flow reorganization to pial veins⁹.

There are many classifications for vein of Galen malformations. The two most used are those propos-

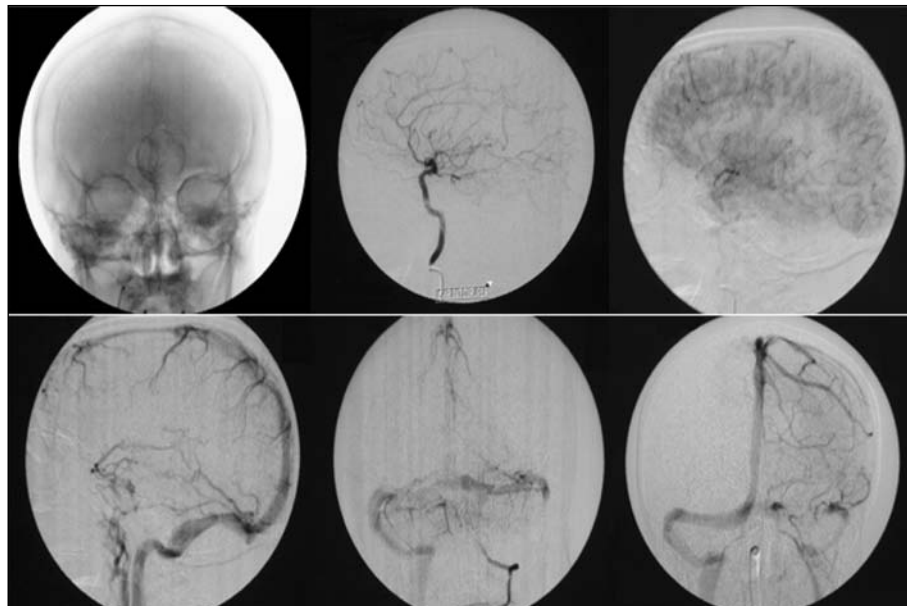


Fig 3. Angiography evidencing the thrombosed vein of Galen aneurysm.

ed by Yasargil¹⁰ and Lasjaunias et al.⁴. Yasargil's lesions types 1, 2 and 3 are direct fistulas between the malformations and the vein of Galen. Lesion type 4 are parenchymal arteriovenous malformations, which drain directly into the vein of Galen. According to Lasjaunias et al.⁴, these malformations can be divided into mural and coroidal types depending on the fistula localization. The coroidal type is characterized by multiple fistulas at the anterior and terminal segment of the median prosencephalic vein. This type usually presents at the neonatal period causing serious heart failure unfolding to multiple organ failure and death. The mural type has the fistula at the vascular wall usually at the lateral-inferior wall of the median prosencephalic vein. They are commonly slow flow and asymptomatic⁹. VGA differ from Galen dilatation, which results from an obstruction of the normal vein of Galen. This alteration is mentioned as a dilatation of the persistent median prosencephalic vein, also known as the Markowski vein¹¹.

Cerebral angiography is the gold standard for the diagnosis of VGA. The exam shows the dynamic aspect of the cerebral venous system and vascular relationships to the fistula^{12,13}.

In adults, this pathology has been described by some authors^{3,6,7,14-20}. Because of its rarity in adults, and despite what is known from these studies, there is still insufficient information about this disease during adult age¹³. Use of oral contraceptives²¹, postpartum status²², sickle cell anemia²³, and aseptic meningitis²⁴ were risk factors related to thrombosis of the vein of Galen.

Even considering recent micro-neurosurgery advances, the lesion resection usually is not possible. Therefore, advances in interventional neuroradiology have assured good results. The indications of these treatments are based on case-specific clinical manifestations⁹. Sasidharan et al.²⁰, suggested conservative conduct and patient monitoring for those with severe heart failure. The absence of blood flow at the deep cerebral venous system due to malformation thrombosis, absence of signs or symptoms of intracranial hypertension, and pharmacological control of seizures led us to conservative treatment of the patient presented here.

Treating a VGA must be analyzed case-by-case. There is no standard treatment that applies universally. Nowadays, endovascular techniques are the procedures of choice considering interventional treat-

ment. However, conservative treatment must be considered, especially for the elderly with co-morbidities, and neurological stable patients without severe neurological signs or symptoms.

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