Investigation of thunderclap headache in cavernous angioma: when magnetic resonance makes the difference. Case report*

*Investigação de cefaleia em trovoada em angioma cavernoso: quando a ressonância magnética faz a diferença. Relato de caso **

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ABSTRACT

BACKGROUND AND OBJECTIVES: Brain magnetic resonance is a major exam to evaluate thunderclap headache, after excluding subarachnoid hemorrhage. This study aimed at reporting a case of brainstem cavernous angioma (cavernoma) where clinical presentation and computerized tomography have suggested intraventricular hemorrhage.

CASE REPORT: Female patient, 55 years, was referred to the hospital with a history of new headache 10 days ago. Pain onset was sudden, pressure-type, severe, located in the occipital region with irradiation throughout the head, followed by severe photophobia, nauseas, diplopia and blurred vision. Neurological evaluation has revealed anisocoria, complete ophthalmoplegia and right eyelid ptosis. Cranial CT has shown blood in the third ventricle. Conventional brain arteriography has not shown aneurysm, arteriovenous malformation or venous sinus thrombosis. At lumbar puncture, an opening water pressure of 45cm was found and liquor analysis was normal. Brain resonance has shown oval lesion (1.0x1.0x0.6cm) of exophytic aspect in the interpeduncular cistern and third ventricle, compatible with brainstem cavernoma.

CONCLUSION: In this case, magnetic resonance was essential for the diagnosis, since routine exams (brain tomography, liquor puncture and arteriography) could not define it. Further studies are needed to explain how magnetic resonance impacts investigation.

Keywords: Central nervous system cavernous angioma, Disorders secondary to headache, Magnetic resonance.

RESUMO

JUSTIFICATIVA E OBJETIVOS: A ressonância magnética cerebral é um exame importante na investigação da cefaleia em trovoada, após a exclusão de hemorragia subaracnoidea. O objetivo deste estudo foi relatar um caso de angioma cavernoso (cavernoma) no tronco cerebral, em que a apresentação clínica e tomografia computadorizada sugeriram uma hemorragia intraventricular.

RELATO DO CASO: Paciente do gênero feminino, 55 anos, foi encaminhada ao hospital com uma história de cefaleia nova há 10 dias. A dor teve início súbito, do tipo pressão, de forte intensidade, localizada na região occipital com irradiação para toda a cabeça, acompanhada de fotofobia intensa, náuseas, diplopia e visão turva. O exame neurológico revelou anisocoria, oftalmoplegia completa e ptose palpebral à direita. TC de crânio mostrou sangue no terceiro ventrículo. Arteriografia cerebral convencional não apresentou aneurisma, malformação arteriovenosa ou trombose de seios venosos. Na punção lombar, uma pressão de 45cm de água de abertura foi encontrada e a análise do líquido cefalorraquidiano foi normal. A ressonância de crânio revelou lesão oval (1,0x1,0x0,6cm) de aspecto exofítica na cisterna interpeduncular e terceiro ventrículo compatível com cavernoma de tronco cerebral.

CONCLUSÃO: No caso descrito, a ressonância magnética foi essencial para o diagnóstico, uma vez que os exames de rotina (tomografia de crânio, punção de líquor e arteriografia) não conseguiram defini-lo. Maiores estudos são necessários para esclarecer como a realização de ressonância magnética impacta a investigação.

Descritores: Hemangioma cavernoso do sistema nervoso central, Ressonância magnética, Transtornos secundários da cefaleia.

INTRODUCTION

Thunderclap Headache (TC) is a type of headache with sudden or hyper-acute onset. Most common phenotype of this headache, followed or not by neurological deficits, is associated to the rupture of intracranial aneurysm, or subarachnoid hemorrhage (SAH), but there might be other causes such as brain venous thrombosis, other intracranial hemor-

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rhages, pituitary apoplexy or reversible brain vasoconstriction syndrome[1]. In general, intracranial hemorrhages have high morbidity and mortality rates. Among them, intraventricular hemorrhage (IVH) is associated to poorer prognosis and higher need for care and assistance[2].

With regard to investigation, the first exam should be brain computerized tomography (CT) to detect possible intracranial bleeding. If this exam is normal, CSF puncture is indicated to rule out any bleeding which might have gone undetected by CT. If bleeding is detected, next step is arteriography to locate a possible brain aneurysm. If there is no brain aneurysm, brain magnetic resonance (MRI) becomes an important exam for the etiological investigation of thunderclap headache. It may detect, for example, some brain tumors such as cavernous angiomas, which may not be visible by conventional angiography and have unique findings on resonance. So, starting from the evidence of primary IVH, MRI would be a critical exam[3].

This study aimed at reporting a case of thunderclap headache caused by minor third ventricle hemorrhage, secondary to midbrain cavernous angioma (dorsal tegument), not previously diagnosed, as well as at reviewing major etiologies and exams for investigation.

CASE REPORT

Female patient, Caucasian, 55 years old, presented in April 2014 with a new explosive, occipital headache with whole brain irradiation, with severe photophobia, nauseas, followed by diplopia and blurred vision. Brain CT has shown small hyperdense image in third ventricle, compatible with bleeding. Patient was admitted to the Emergency Unit 10 days after ictus, and was scheduled for brain angiography.

Patient was in good general status, conscious, oriented, complaining of severe headache with the same initial characteristics, causing irritability and discomfort. Associated morbidities were systemic hypertension, smoking, chronic obstructive pulmonary disease (COPD) and major depressive disorder. Patient denied family history of neurologic diseases. Physical evaluation has shown anisocoric (D>E) photoreactive and brady-reactant pupils, in addition to left eye inferomedial deviation. Extrinsic eye movement and fundus evaluation was impaired by severe photophobia. Remaining neurological evaluation was normal.

At this moment, brain CT had no evidence of hemorrhage. Patient was submitted to arteriography, which has not shown evidence of aneurysm, vascular malformation or venous thrombosis. The day after admission, lumbar puncture was performed in L_3-L_4 with opening pressure of 45cm of water. CSF was clear, colorless, with 2 cells, 1.6 red cells and normal biochemistry.

Brain MRI was requested and has shown oval exophytic lesion (1.0x1.0x0.6cm) in interpenducular cistern and third ventricle, with hyperintense signal at T_1-T_2 sequence and hypointense signal halo at T_2 sequence (Figures 1 to 4), compatible with brainstem cavernous angioma. Patient evolved with partial headache remission with topiramate (treatment of intracranial hypertension, since acetazolamide was contraindicated – COPD hypercapnia). Patient maintains mild converging strabismus and is under Neurosurgery clinical-imaging follow-up.

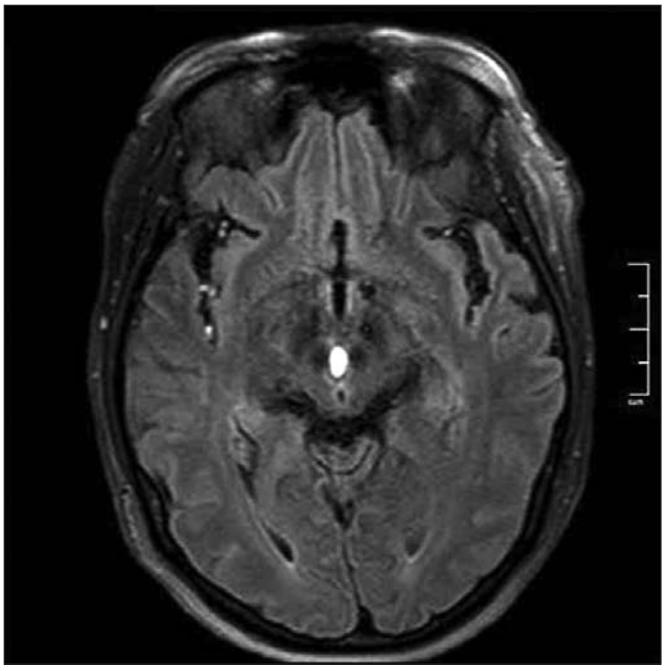

Figure 1. Resonance magnetic in axial cut, flair sequence

Oval formation with hyperintense signal at T_1 and T_2 sequences, located in central and anterior region of midbrain tegument, with exophytic aspect for the interpeduncular cistern and third ventricle, measuring approximately 1.0x1.0x0.6 cm, with hypointense signal halo at T_2 sequence.

There has been no contrast enhancement. There has been no opacification of altered circulation at angio-MRI sequences.

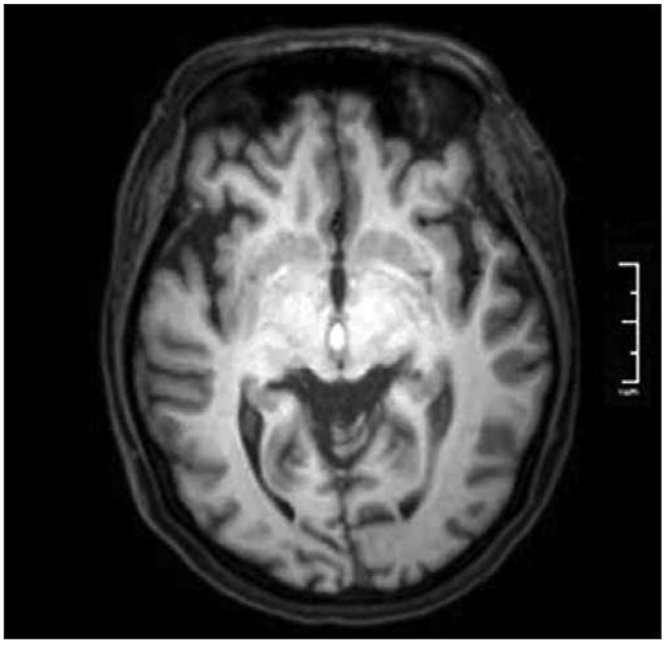

Figure 2. Weighted axial resonance magnetic in T_1

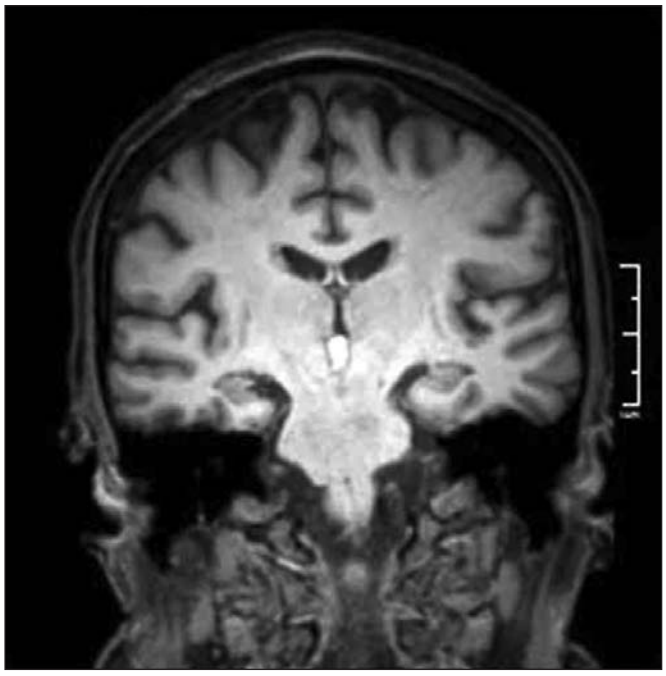

Figure 3. Weighted coronal resonance magnetic in T_1

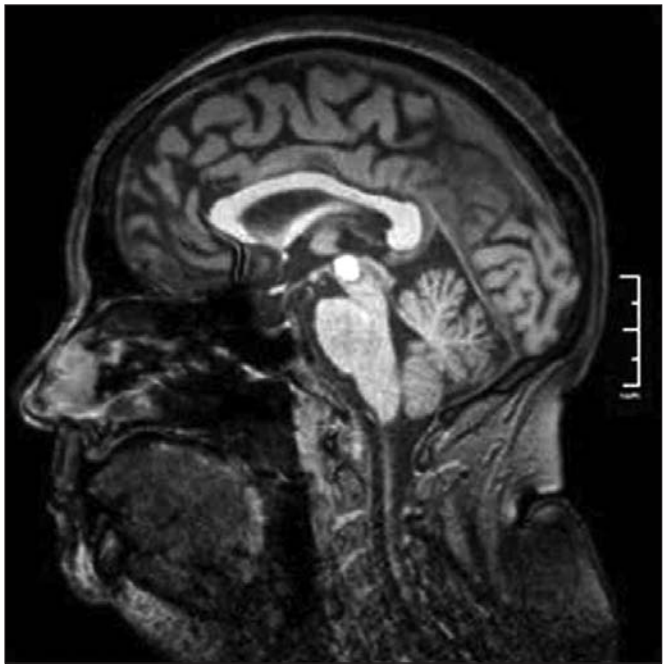

Figure 4. Sagital weighted magnetic resonance in T_1

DISCUSSION

Approximately 30% of IVH are primary, that is, originated from the ventricular system itself. They may come from intraventricular structure of from lesion close to the ventricle. They appear in situations of intraventricular trauma, aneurysm rupture or vascular malformation and tumor complication, for example. The other 70% of IVH are secondary, corresponding to the extension of intraparenchimal hemorrhage or SAH[2].

Cavernomas (synonym to cavernous angiomas) are slow flow capillary malformations, made up of roughly dilated vascular channels lined with a single layer of endothelial cells, without elastic tissue and smooth muscle. Blood breakdown and reactive gliosis products may be found in adjacent brain parenchyma and are described as looking like raspberries[3].

In our case, initial suspicion was SAH, of probable vascular origin by the evidence of primary IVH. However, brain arteriography was normal. In the meantime, patient remained symptomatic, with signs of intracranial hypertension (ICH) – diplopia and blurred vision – possibly associated to hemoventricle products breakdown and intervention on CSF flow[4,5]. Hypertensive CSF has confirmed such hypothesis, and topiramate was started. Considering previous COPD and associated hypercapnia, we decided to rule out acetazolamide to manage ICH. Brain MRI was essential to clarify the diagnosis.

Cavernomas are reported in 0.1-0.5% of general population, with bimodal presentation from 3 to 11 years of age (30%) and from 30 to 40 years of age (60%). Above 40 years of age, only 10% of diagnoses are made[5,10]. There is a well-known predominance of females (1.8:1), in addition to higher prevalence in Caucasian patients[6].

Solitary cavernous angioma occurs in 80% of cases and multiple cavernous angiomas in 20%. With regard to genetic predisposition, 80% are sporadic and 20% are familial[7]. Our patient was a 55-year old female with isolated and sporadic lesion.

With regard to location, 70% are supratentorial (predominance of frontal lobe), 25% are infratentorial and 5% are located in spinal cord. In brainstem, 57% of cavernomas are located in the pons, 29% in midbrain and 14% in the bulbus[3,10]. Bleeding location, size and presence lead to a broad variation of neurological repercussions, from asymptomatic to sudden death by acute hemorrhage, identified in 20% of cases[7].

Clinical manifestations may be nausea, vomiting, dizziness and epileptic crises[8]. Focal motor deficits are seen in approximately 46% at presentation. A range of 10% to 90% with reports of headache is due to different situations, from incidental diagnoses, where there is association with common migraine, to headache induced by ICH[9]. In our case, it was thunderclap headache with focal deficits, caused by expansive midbrain lesion.

Cavernomas are part of a group of vascular malformations angiographically occult or cryptic, like venous angiomas, capillary telangiectasis and some arteriovenous malformation. So, brain angiography is an exclusion exam.

Brain CT may evidence isodense lesion or with nonspecific focal hyperdensity due to recent hemorrhages or to microcalcifications and with poor contrast enhancement. Such findings are nonspecific and the diagnosis of cavernoma is often ignored[3,10].

Classic imaging exam is described as a "popcorn" lesion at MRI. In the T_2 sequence it shows an image with reticulated core, surrounded by a radiolucent halo (corresponding to repeated hemorrhages and hemosiderin deposition). There is no perilesional edema. In addition, punctuate lesions may be seen in gradient-eco, which is more sensitive[3,10].

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Risk factors predisposing to bleeding are gestation, cavernomas above 1.0cm, age below 35 years and, especially, previous bleeding. In the general mean, based on natural history evidences, bleeding rate per year in cases of supratentorial cavernomas is 3%, increasing to 5% for rebleeding. For infratentorial cavernomas, the same rates are 5% increasing to 15%, respectively[3,10].
The option for expectant follow-up is well established in cases of asymptomatic cavernomas, incidental findings or for patients without surgical conditions (due to inaccessibility of the lesion or to patient's clinical conditions). In this therapeutic modality, control is regularly performed with MRI to evaluate growth or new hemorrhages. In addition, gestation, intensive physical exercises and anticoagulants should be discouraged[3,7].

CONCLUSION

This case shows the importance of brain MRI in patient with thunderclap headache, however further studies are needed in this area to explain how this impacts the investigation.

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