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# The importance of Nuclear Magnetic Resonance (MRI) in the diagnosis of cerebral cavernomas. Case presentation

**Dan Bentia[1], Andrei Alexandru Marinescu[2], Florin Papagheorghe[3], Georgeta Popa[4], Alexandru Vlad Ciurea[5,6]**

[1] Department of Neurosurgery, Sanador Clinical Hospital, Bucharest, ROMANIA
[2] Department of Neurosurgery, National Institute of Neurology and Neurovascular Diseases, Bucharest, ROMANIA
[3] Department of Neurosurgery, Bucharest Emergency University Hospital, Bucharest, ROMANIA
[4] Department of Radiology and Medical Imaging, Sanador Clinical Hospital, Bucharest, ROMANIA
[5] Department of Neurosurgery Sanador Clinical Hospital, Bucharest, ROMANIA
[6] "Carol Davila" University of Medicine and Pharmacy, Bucharest, ROMANIA

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Corresponding author:
**Alexandru Vlad Ciurea**

Professor. "Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

prof.avciurea@gmail.com

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## ABSTRACT

**Introduction**: Cavernomas are one of the four types of vascular malformations which develop in the central nervous system. From an imaging point of view, they are low flow small vessel malformations that cannot be seen on conventional angiography and are rarely visible on CT. They can be seen on MRI because of the para-magnetic haemoglobin break-down products within them.

**Case presentation**: A 35 years old male presented at the emergency room following a tonic-clonic seizure, without urine emission. After a clinical exam, it was decided to perform a cerebral MRI with contrast substance and angiographic sequence. Clinical and imagistic results showed a superficial left temporal cavernoma, which showed signs of recent bleeding. After the surgery, the patient has no neurological deficits and is discharged afebrile and without any other symptoms. At 6 weeks after the surgery, the patients returned for neurosurgical reevaluation in perfect neurological condition, without motor deficit or intracranial hypertension.

**Conclusions**: Cavernomas are angiographic cryptic vascular lesions that need an MRI investigation in order to appreciate them correctly.

## INTRODUCTION

Cavernomas are one of the five types of vascular malformations which develop in the central nervous system, representing about 5-15% in

total. Cerebral cavernomas are cryptic angiographic lesions, classically defined as dilated vascular structures, with thin walls, located in the CNS (central nervous system), without any cerebral parenchyma interposed. [1,2]

In 1984, McCormick presented a study in which out of 5734 autopsies, only 19 cavernomas were discovered. This results in an incidence of 0.34%. A few years later, Otten et al report 131 cases of cavernomas out of a group of 24,535 autopsies. Thus, the incidence was 0.53%. [3]

According to McCormick's 1966 classification, neurovascular malformations can be divided into 5 large categories: [4]

- Telangiectasias
- Varicose veins
- Cavernous malformations
- Arteriovenous malformations (AVM)
- Venous angiomas

This classification was later modified: varicose veins were combined with venous malformations/venous angiomas and were put together in the category of venous developmental abnormalities (ADV). Several pathological classification criteria for each malformation have been suggested in the literature, but their nomenclature and structuring criteria are ambiguous and variable. In addition, cases of mixed forms and transitions from one form to another have been reported. Also, in the literature there are cases of coexistence of these malformations. The most common malformation associated with cavernomas is ADV. Another common combination is capillary telangiectasia. A few similarities such as pontin damage, the presence of a family form and multiplicity are reasons to consider telangiectasias a precursor to cavernomas. [3]

The term "cavernoma" was used for the first time by Rokitansky in 1846 and it was referring to a vascular lesion located in the brain. Later, in 1956 Rudolf Virchow made for the first time a histopathological description of these lesions. In 1956, Crawford and Russel introduced the term cryptic, to describe an arterio-venous pathology or venous hamartomas located in the encephalon and which often led to hemorrhage. Later, this term was used for the cerebrovascular lesions which couldn't be observed on angiography, along with the term „angiographically occult vascular malformation". The current nomenclature includes the following terms: cavernoma, cavernous angioma, cavernous hemangioma or cavernous malformations.[5]

The use of MRI leads to the diagnosis of cavernomas even before the histopathological diagnosis. The right treatment of these lesions requires a good knowledge of epidemiology, histopathological characteristics and of their natural evolution.

The clinical picture of cavernous angiomas varies significantly, by including asymptomatic forms which are discovered on CT or MRI examination (performed for a headache syndrome or after a traumatic event), but also forms discovered during autopsy after a fatal intracranial hemorrhage.

## Neuroimaging diagnosis

Today, the advances in neuroimaging allow the diagnosis of cavernomas even before the histopathological exam.

**The CT examination** shows a spontaneously hyperdense lesion, given the inhomogeneous hemosiderin impregnation, with a round well-defined contour or irregular shape. After administering the contrast substance, cavernomas have different behaviors, in some cases appearing filled with contrast substance, especially if the examination is performed tardy. It is worth noting that these lesions are characterized by a slow flow and that in some cases, calcifications can be seen at this level.

**The MRI examination** represents the „gold standard" for the diagnosis in most cases. The typical image for a cavernoma is a well-circumscribed lesion, with an inhomogeneous "popcorn" appearance and an hypointense ring on the outside which represents the hemosiderin impregnation. Traces of older or more recent microhemorrhages can be seen in the lesion or in the adjacent areas. Rarely, a draining vein can be seen. In the T2 sequence of MRI we can see a hypointense area at the periphery of the lesion due to the edema of the parenchyma. (Figure 1)

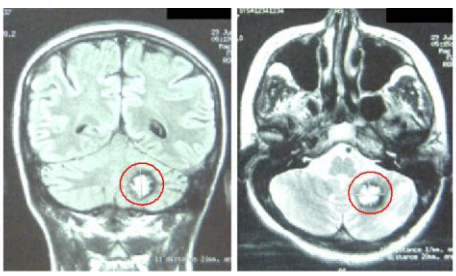

**Figure 1.** Cerebral MRI: left infratentorial cerebellar

cavernoma. (From the personal collection of Prof. Alexandru Vlad Ciurea, MD. PhD.)

**Cerebral angiography** is negative in most of the cases, cavernomas representing most of the occult vascular malformations. Rarely, it can show an avascular area or a capillary cluster. Usually, it is not indicated for cavernomas diagnosis unless it is necessary to obtain information regarding the vascular anatomy of the region or if an association with another vascular malformation is suspected.

## Therapeutic attitude

Cavernomas, being by their nature well-defined lesions, with feeding vessels with a reduced blood flow, offer the premises for a total surgical resect Currently, there is a general attitude of extending surgical indications even in young patients, minor symptoms, thus removing the risk hemorrhage and its consequences. therapeutical attitude is possible thanks to advances in neuroimaging and the introductio neuronavigation, it is worth noting complications may occur in lesions with critical locations: cortical or subcortical cavernomas of eloquent areas or deep regions, such as the basal nuclei, thalamus, corpus callosum, paraventricular or brainstem.

## Case presentation

Male patient, 35 years old, with no neurological pathology associated, came in the Emergency Room of Sanador Clinical Hospital following a tonic-clonic seizure, without urine emission. Following the neurological and neurosurgical consult, it was decided to perform a cerebral MRI with contrast substance and angiographic sequence. (Figure 2)

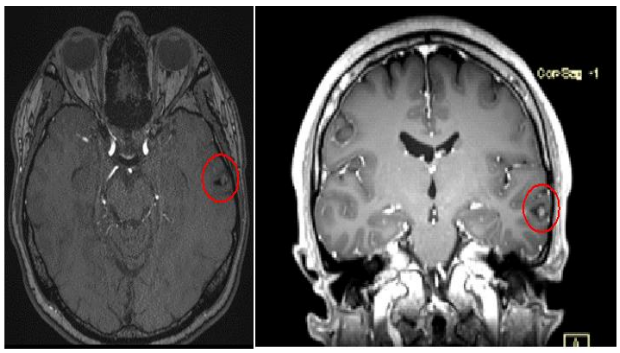

**Figure 2.** Cerebral MRI with contrast substance which shows a left temporal cavernoma, with recent haemorrhage.

Clinical and imagistic evaluation of the patient showed a superficial left temporal cavernoma, which showed signs of recent bleeding. Together with the neurosurgical team from Sanador Clinical Hospital, it is decided to perform the total ablation of the cavernoma using Kinevo 900 operating microscope and neuronavigation.

After the surgery, the patient has no neurological deficits and is discharged afebrile and without any other symptoms. At 6 weeks after the surgery, the patients returned for neurosurgical reevaluation in perfect neurological condition, without motor deficit or intracranial hypertension. This is confirmed by the postoperative MRI with contrast substance. (Figure 3)

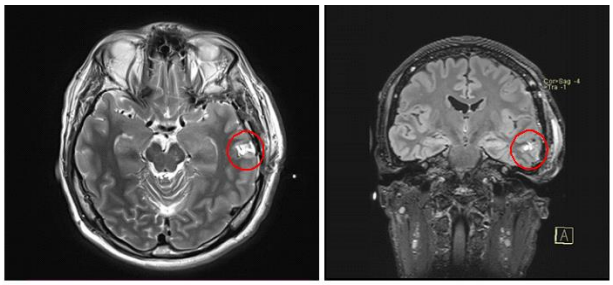

**Figure 3.** Postoperative cerebral MRI with contrast substance which shows total ablation of the left temporal cavernoma.

## Discussions

Cavernomas are low flow small vessel malformations that cannot be seen on conventional angiography and are rarely visible on CT, so if we want to be certain of the diagnosis, we must perform an MRI. This type of imaging can show us the cavernous malformations because of the para-magnetic hemoglobin break-down products within them.

Over time, it has been debated what is the best treatment for cavernomas, between surgical approach, radiotherapy (GKS) and conservative treatment. According to the literature, the best approach is the neurosurgical one in the case of symptomatic, superficial lesions that are not located in the eloquent area. For the other cavernomas (asymptomatic, multiple, located in eloquent or deep areas) the risk-benefit ratio must be balanced. So, it will be decided together with the patient, between the surgical approach and the conservative treatment.

H. Bertalanffy et al. (2001) presented an analysis that consist 72 patients operated in 5 years, of which 24 had the lesion located within the brainstem, 18

within the deep white matter of the hemispheres, 12 in the basal ganglia or thalamus, 11 in superficial areas of the hemispheres and seven within the cerebellum. They reinforce the fact that angiography was the first imaging study used to see cerebrovascular malformations, but it shows no pathological changes. In the mid-1970s, the computed tomography (CT) became available and the detection of symptomatic cavernomas increased significantly, but this kind of imaging study has poor specificity. The most important and with the highest accuracy imaging study is MRI.[6] Rigamonti et al., being one of the firsts to describe the MRI features of cavernomas, are recalled in this study for their article in which they presented the differences between CT scan and MRI. From 10 patients, CT scans detected 14 lesions, while MRI detected 27 distinct lesions, so it can be seen how specific is MRI besides CT scan for the cavernous malformations diagnosis.[7]

D. Cavalcanti et al. (2011) described that genetics have an important part in cavernous malformations. Cavernomas can be both sporadic and familial. According to them, "half of CCMs are familial, and they inherited in an autosomal dominant fashion with variable penetrance". The most of familial CCMs are identified on MRI as multiple lesions. This multiplicity of lesions is characteristic in up to 84% of familial cases, whereas it is reported in 10%-33% of supposed sporadic cases. Annually, up to 6.4% of familial cavernomas can bleed and become symptomatic. To date, 3 distinct loci have been mapped in different families, CCM1, CCM2 and CCM3. These 3 loci account for 70%-80% of all cases of familial CCMs. Multilocus linkage analysis showed 40% of inherited cases that CCM1 account, 20% for CCM2 and 40% for CCM3.[8]

The radiotherapeutic approach for cavernomas has been in constant debate since the invention of Gamma Knife Surgery. Still, this controversy reached a conclusion when one of the founders of GKS, Ladislau Steiner published his comprehensive comparative study on this subject. L. Steiner et al. (2010) came to the conclusion that GKS it is not an effective approach for cavernous malformations and the surgical approach remains the best choice in treating this pathology.[9]

J. K. Liu et al. (2010) found in their article about cavernous malformations that the CT scans showed the OPH CMs as suprasellar hyperdense masses. Cerebral angiography did not reveal any arterial feeding vessels. MRI was performed at 88% of cases and the cavernomas could be seen as heterogenous with mixed signal intensities suggestive of different ages of blood, this appearance often described as a "popcorn-like" lesion.[10]

In another case presentations similar to ours, P. Feizi et al. (2020) described that a 50 years old woman has presented to the emergency room following new onset seizures and express her symptoms as right arm jerking, drooling, and encephalopathy. CT scans without contrast were effectuated and the results were multiple supratentorial and infratentorial hyperdensities measuring up to 8 mm, which were concerning for possible hemorrhagic metastases in the setting of history of malignancy. [11]

An MRI was performed too, with and without contrast, and was demonstrated that some of the lesions had peripheral vasogenic edema, and internal hyperperfusion. Based on those imaging and clinical data and the patient`s known history of thyroid carcinoma, the first diagnosis was "possible multiple hemorrhagic brain metastases". Three-month follow-up MRI of the brain demonstrated vasogenic edema and left parietal lesion with a surrounding hemosiderin ring and heterogeneous internal signal now able to be visualized. So, the diagnosis of cavernous malformations became evident.[11]

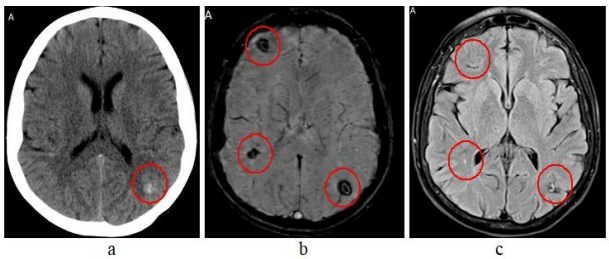


**Figure 4.** (a) CT scan; (b) MRI with SWI sequence; (c) T1 MRI
Image source: Feizi P, Lakhani DA, Kataria S, et al. Multiple cerebral cavernous haemangiomas masquerading as haemorrhagic brain metastases. Radiol Case Rep. 2020 Aug 20;15(10):1973-1977. Licensed under CC BY-NC-ND 4.0; © 2020 The Authors

## Conclusions

Cavernomas are angiographic cryptic vascular lesions which need an MRI investigation in order to appreciate them correctly. In the case of small cavernomas, such as the one presented, the CT examination does not reveal the lesion. Thus, MRI is a valuable investigation for neurosurgical

emergencies, especially if the patient has a new comitial seizure, is young and does not have other neurological pathologies associated.

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