

A RARE TOPOGRAPHY AND UNUSUAL REVELATION OF AN ARTERIOVENOUS MALFORMATION: A CASE REPORT

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ABSTRACT : An arteriovenous malformation is a congenital abnormality marked by a fistulous connection between arteries and veins without the interposition of a capillary bed. The supratentorial compartment of the brain is usually more affected, and the revelation is mainly made by the onset of hemorrhagic stroke. We report in this work, the uncommon presentation of a ventricular AVM (Arteriovenous malformation) and the management strategy planned for this adult patient.

Key words : Arteriovenous malformation (AVM), Hemorrhage, Ventricular.

INTRODUCTION

An AVM is a congenital vascular condition in which arteries and veins are connected through fistulous and abnormal channels, without the existence of a capillary bed that decreases the vascular flow in a progressively still the venous compartment in a healthy brain. The interposition of functional brain tissue within this tangle of vessels is not usual [1].

One of the major and familiar systems of classification was Spetzler - Martin grading system published more than 30 years ago [2]. It's an efficient scale for evaluating the possible therapeutic approach and the risk of surgery for a patient harboring an AVM.

In their natural history, the annual risk of hemorrhage of these malformations is around 2 to 4 % in most of the series 3-5, with a subsequent mortality of 1% per a year 4. During follow up in one study, the hemorrhagic stroke was intra parenchymatous in 28%, exclusively intraventricular in 38%, subarachnoidal in 10%, and combined intraventricular and intraparenchymal in 19% [6].

We aim to report a rare case of a ventricular AVM, revealed in an old patient which is also not usual. The management was in a hybrid way by the consideration of surgical and endovascular techniques which may be a perspective for other similar cases.

CASE REPORT

A male of 68 years old without previous history of cardiac morbidities or troubles of coagulation; was received in emergency. He presented brutal headaches and consciousness disorders. The GCS was at 13/15. All of the rest of metabolic parameters were almost normal for this patient.

Radiological evaluation by a brain CT showed a hyperdensity in the aqueductal region without an important associated hemorrhage in the rest of the ventricular system. This slight amount of blood caused an obstructive hydrocephalus.

Another tissular isodense formation was noticed in the right ventricular atrium.

The findings of the initial CT are shown on figure 1.

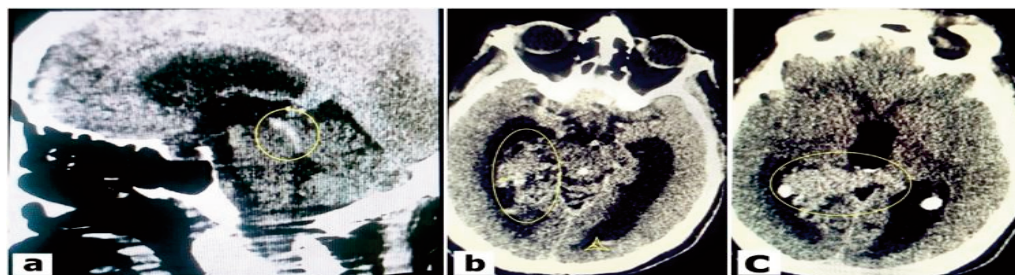


Figure 1 : initial Brain CT of the patient in two plans ; sagittal (a) and axial (b+c) without contrast administration. (a): sagittal section of CT that shows mainly the special location of the hemorrhage in the aqueduct (circle), which is completely occluded. (b): axial section demonstrating the nidus (yellow circle), and the minimal hemorrhage in the left temporal horn (yellow star). notice the important ventricular dilation. (c): the main part of the nidus is filling the right ventricular atrium (circle).

The initial management in emergency was to perform an external drainage of CSF to improve the clinical status of the patient and treat the hydrocephalus. The CSF was hemorrhagic at the moment of the surgical procedure.

The etiological exploration was completed by a brain MRI, and specified the nature of the ventricular mass to be an arteriovenous malformation in the right ventricular AVM

with a minimal posterior extension toward the right occipital horn.

The greater axe of this lesion was measuring 55 mm. The rest of sequences showed that feeding arteries was coming essentially from branches of the posterior cerebral artery. The drainage of the nidus was mainly to the superior sagittal sinus and the deep venous system via a dilated and abnormal network of veins (figure 2).

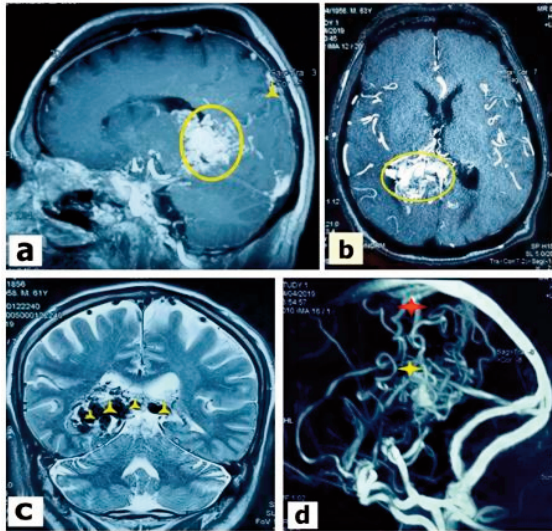


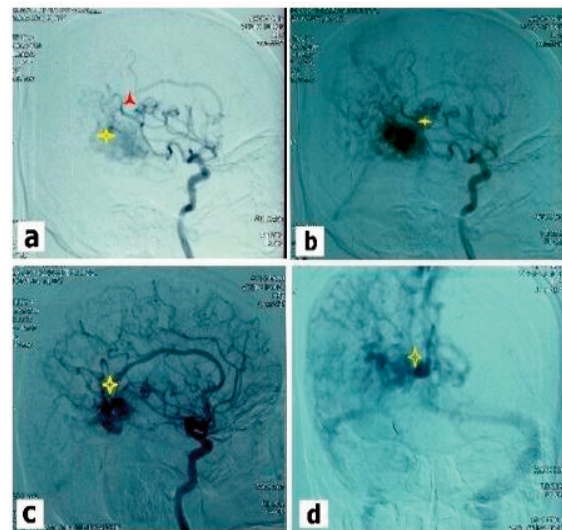
Figure 2 : brain MRI of the patient with contrast administration and ARM sequences. (a): sagittal T1 +contrast administration showing the nidus in atrial part of the ventricle yellow circle. notice the vein of drainage to the superior sagittal sinus (yellow star). (b): axial T1 sequence with contrast showing the important size of the nidus (yellow circle) ,and the tortuous and abnormal dilated vessels inside .

notice the normal size of ventricles after the placement of external drainage . (c): coronal T2 sequence showing a part of the nidus with this aspect of dilated or associated aneurysms (yellow stars). hard to define if they are of an arterial or venous nature. (d): 3D ARM sequence showing the nidus and the projection of superficial drainage (red star) and the deep veins (yellow star) of drainage to the galenic system.

Digital subtraction angiography (DSA) showed further that principal feeders are distal branches of; pericallosal artery, middle cerebral artery and posterolateral choroidal artery and the nidus was

measuring 56 mm. The pattern venous drainage was ; superficial toward the superior sagittal sinus and deep to the galenic vein.

Figure 3 : DSA (Digital Substruction Angiography) in different views (A-P)(d) , lateral (a,b,c) and phases ;arterial (a,b,c) and venous(d) . (a): lateral view of the right ICA injection in the early time showing the vascular blush concomitant with the nidus (yellow star), and a distal branche of the pericallosal artery feeding the AVM(red star). (b): intermediate time of the arterial phase showing an important tortuous vessel (probably another feeder) . (c): lateral view the late time the arterial phase in the injection of the right ICA showing the main feeders from pericallosal artery (yellow star) and other distal branches of the choroidal artery . (d): (A-P) view of the venous phase demonstrating the pattern of drainage. notice the drainage to the galenic vein which dilated . (yellow star).



The external drainage was maintained during 15 days and a significant improvement of the clinical status of the patient was noticed. After performing clamping test, the drain was removed.

Further management was a discussion made by a multidisciplinary team of neuroradiologist, cerebrovascular neurosurgeons and neuroanesthesiologists.

The main of final decision was to perform an embolization in a staged fashion, in order to occlude deep feeders ; choroidal arteries and branches of pericallosal artery and plan a resection surgery in a second stage. The procedure was performed in another institution, and the surgery is planned for a second time.

DISCUSSION

Hemorrhagic stroke is one of the most frequent modes of revelation of brain AVM. In Ondra reports 4, 40% of patients had at least one hemorrhage in the period of follow-up. This rate is higher in Yamane and Takakura series [7], affecting 57% of the enrolled patients .

The kind of hemorrhagic incident is different from one study to another ; it may have a parenchymal, ventricular or subarachnoidal topography. the ventricular space is affected in 28% of patients for Daniel H et al [8]. in the same work [8], ventricular AVMs were grouped within the deep form of AVM with other locations ; basal ganglia, thalamus and corpus callosum. Deep AVMs in this paper represented 15% of all cases.

More precise information about this concern were elucidated in Lawton's works. The ventricular and periventricular AVMs were classed together and their percentage was 7%. Among this group, 1.5% were located in the atrial part of lateral ventricle [9]. This percentage confirms more the scarcity of this form of AVM.

This risk of bleeding of AVMs was furthermore screened to predict factors that may promote or influence this serious event for patients. Bleeding was more frequent in AVMs smaller than 3 cm, compared with larger one (≥ 6 cm) (82% vs 21%) [10].

They found significantly higher feeding artery pressures in small AVMs, and suggested that differences in arterial feeding pressures may be responsible for the observed relationship between the size of the AVMs, the frequency and severity of hemorrhage [10].

Unlike that , In one study, the size of the nidus was not found to be predictive of hemorrhagic events or their severity [11]. The size of the AVM in our case was 55 mm in both MRI and DSA.

The pattern of venous drainage is an important parameter to consider in an AVM.

A strong correlation was existing between deep drainage and frequency of bleeding [12]. Single or abnormal (kinked or stenotic) venous drainage was an

additional factor that was correlated with bleeding [13].

In our case, the pattern of venous drainage was hybrid, to the superior sagittal sinus and to the the galenic vein. This drainage was through two large and dilated veins.

Another parameter that remains special in this patient was the severity of bleeding. Only a small volume of blood was present in the aqueduct. This fact remains also unusual and special for an AVM in this location. Authors believe that ventricular AVMs are more vulnerable and carry a higher risk of rupture because of the reduction of supportive parenchyma [14] .

In our analysis of the AVM's angioarchitecture, we observed that most of its characters are correlated with a significant risk of rupture. The pattern of drainage, where the flow is partially to the galenic system, and the absence of surrounding tissue against the nidus. Despite this favorable condition that promotes a serious rupture, the hemorrhagic stroke was minimal in the aqueduct .

The little volume of blood in the aqueduct and the presence of factors that could promote bleeding as the deep drainage and associated aneurysms, are conflicting findings in our case. New thoughts must be conceived and further research should be undertaken about the vascular dynamic of these lesions to understand more their mechanism of rupture.

The proximity of bleeding to nuclei of the reticular formation around the aqueduct, and hydrocephalus are two important etiologies of the initial clinical presentation marked by an impaired consciousness.

CONCLUSION

Ventricular AVM is a rare entity that could face neurosurgeons in their daily activity. Several kind of hemorrhagic stroke could be the mode of revelation and all probabilities and etiologic investigations should be performed adequately.

Staged embolization for large ventricular AVM s is an efficient therapeutic option to prepare the patient for a further microsurgical procedure in a second stage.

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