Intraventricular hemorrhage secondary to AVM of the septum pellucidum

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ABSTRACT

Arteriovenous malformations (AVMs) of the brain include congenital vascular lesions that represent about 2% of all hemorrhagic strokes. Despite the relative rarity of the disease (with a detection rate estimated at approximately 1/100.000 person-years), AVMs represent a significant neurological problem. Affected patients are mostly young and healthy. AVMs located in the medial region of the cerebral hemisphere and the limbic system comprise a special group with difficult access and consequent difficulties of resection. Furthermore, there is a high incidence of intracranial hemorrhage resulting from a complex venous drainage, sometimes directed to the superficial veins, but mostly to the deep venous system. The present case report illustrates a patient with an AVM centered on the septum pellucidum, whose initial clinical resulted from an intra-ventricular hemorrhage.

KEYWORDS

Arteriovenous malformations, vascular malformations, cerebral hemorrhage.

RESUMO

***Hemorragia intraventricular secundária a MAV de* septum pellucidum**

Malformações arteriovenosas (MAVs) encefálicas são lesões vasculares congênitas que representam cerca de 2% de todos os acidentes vasculares hemorrágicos. Embora se tratando de uma doença relativamente rara (com prevalência estimada em aproximadamente 1/100.000 pessoas-ano), MAVs representam um problema neurológico significativo. Os pacientes afetados são na maioria jovens e hígidos. MAVs localizadas na região medial do hemisfério cerebral e no sistema límbico correspondem a um grupo especial pela dificuldade de acesso e consequente dificuldade de ressecção. Além disso, possuem alta incidência de hemorragia intracraniana resultante de uma drenagem venosa complexa, por vezes direta às veias superficiais, mas na maioria para os sistemas venosos profundos. O caso relatado a seguir ilustra um paciente com uma MAV centrada no septum pellucidum*, cuja apresentação clínica inicial resultou de uma hemorragia intraventricular.*

PALAVRAS-CHAVE

Malformações arteriovenosas, malformações vasculares, hemorragia cerebral.

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Introduction

Cerebral arteriovenous malformations are benign lesions composed of a tangle of dilated blood vessels, which are characteristically cerebral arteries that drain directly into the veins.

These are sporadic and congenital abnormalities that usually manifest clinically by seizures (epilepsy) or intracranial hemorrhage, the latter with a risk of 4% per year, cumulative throughout life.[1,2]

Although the clinical presentation can occur at any period of life, the highest incidence is concentrated between the second and fourth decade of life.[1] There is a slight predominance of incidence in females, but without statistic significance.[2]

Recent advances in diagnostic techniques, microsurgery, endovascular therapy, and stereotactic radiosurgery have significantly improved the treatment outcome of vascular malformations of the central nervous system. Better information regarding the natural history of the various types of lesions has allowed to weigh the natural risk of the untreated disease, versus the morbidity and mortality of different treatment options.[2] The ultimate goal of treatment should be the prevention of future complications from the lesion, while minimizing the therapeutic risk to the patient. In most patients this is best achieved by total elimination of the lesion. In certain instances, however, only palliative treatment or expectant medical management is the least risky alternative for a given patient.

Below is a rare case of MAV *septum pellucidum*, addressing clinical and therapeutic aspects.

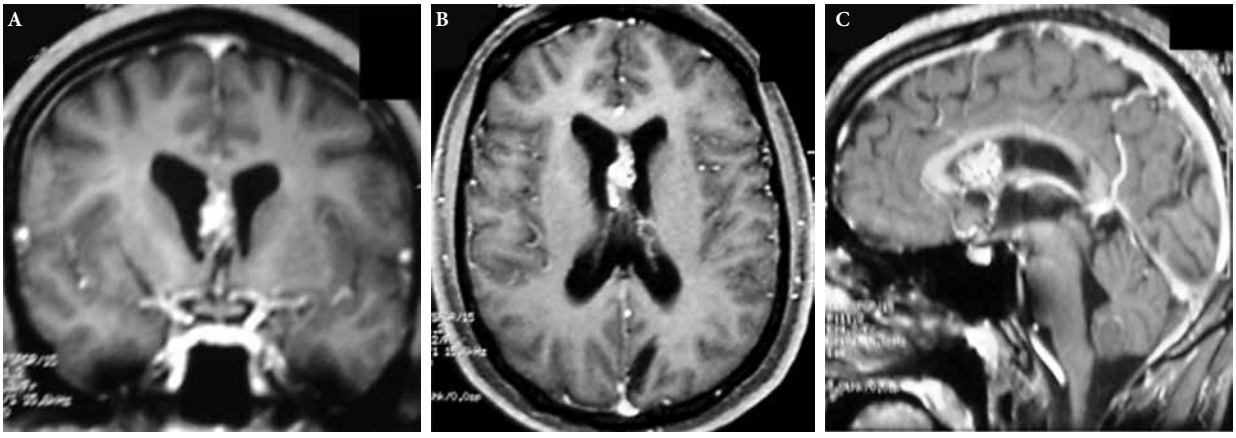
Case report

RP, male, white, 36 years old, presented with a history of three episodes of subarachnoid hemorrhage (SAH) without investigation in the city of origin. The first one occurred 12 years prior to admission, with sudden headache accompanied by neck stiffness and vomiting, a positive lumbar puncture and remaining asymptomatic since. One month prior to admission he developed a new episode of intense and sudden holocranial headache, leading to four days in coma, requiring external ventricular drainage (EVD) and cares in the intensive care unit (ICU) due to panventricular hemorrhage. The patient recovered well and was discharged without sequelae, but a week later he had a new episode of headache and a repeat computarized tomography (CT) scan revealed new bleeding. The patient developed paresis of the left lower limb with full recovery.

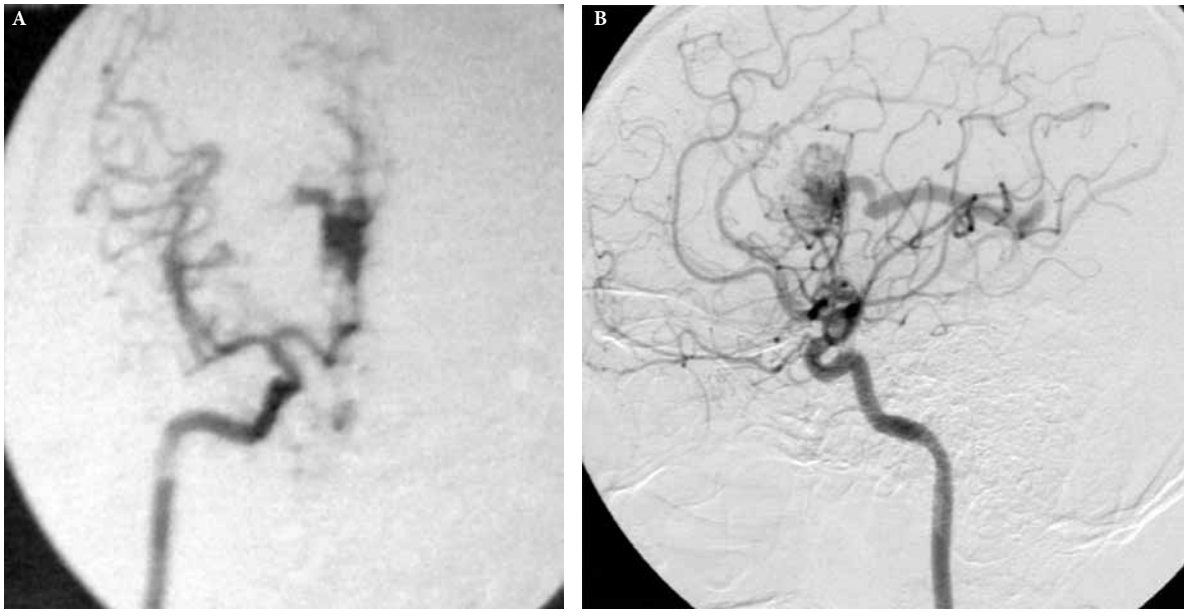
On admission, the patient was in good general condition, without neurological signs. Magnetic resonance imaging (MRI)-angiography showed a deep AVM along the midline in the *septum pellucidum* (Figures 1A, 1B, 1C). Angiography confirmed an AVM centered on the *septum pellucidum*, nourished by the septal branches of the anterior cerebral arteries and with little involvement of right posterior choroidal artery, with deep venous drainage into the internal cerebral vein (Figures 2A, 2B).

The first embolization resulted in the occlusion of the feeding pedicle through selective catheterization of the right internal carotid artery and the basal frontal midline AVM, with occlusion of 40% of the total volume of the nidus. A second embolization was completed a month later with the closure of approximately 60% of the total volume of the AVM.

The lesion was resected one week later through an anterior transcallosal approach with resection of the *septum pellucidum* AVM with microsurgical and endoscopic support and placement of EVD to control intracranial pressure. The patient recovered fully after surgery without neurological deficits.



Figures 1A, 1B and 1C – Angio resonance imaging brain: coronal, axial and sagittal sections, respectively, showing deep arteriovenous malformation, along the midline.



***Figures 2A and 2B – Selective angiography of the right internal carotid artery in anteroposterior and lateral projections, respectively, confirming the arteriovenous malformation centered on the* septum pellucidum.**

Post-operative cerebral angiography two months later showed resolution of the AVM, transverse and right sigmoid sinus occlusion (Figure 3). Follow-up MRI showed postoperative changes involving the anterior segment of the corpus callosum, with no evidence of residual AVM. On four year follow-up the patient remains without clinical or neurological sequelae.

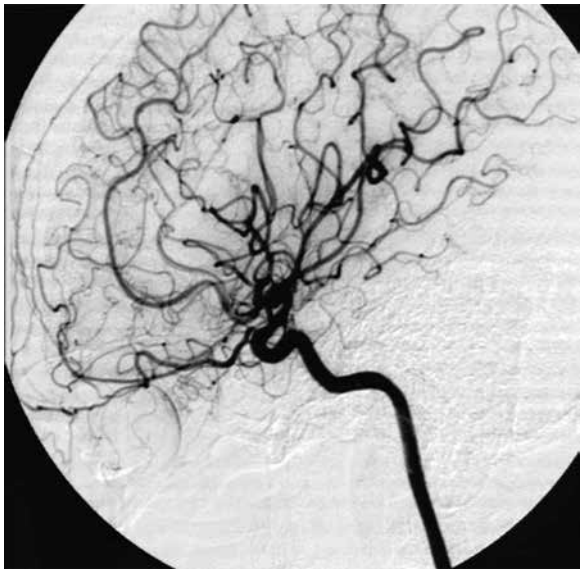


Figure 3 – The same angiographic projection in profile of the right internal carotid artery after two months of surgery and two sessions endovascular embolization shows resolution of the arteriovenous malformation, with thrombosis of the right transverse and sigmoid sinuses.

Discussion

Using the classification proposed by Stein,[3] this case can be classified as affecting the region F, namely an AVM located in the region the fronto-medial septum. In Stein's series with 164 cases, only 15% of all AVMs were located in the limbic or medial hemisphere and, of these, only two cases presented with lesions in the region F, confirming the rarity of this localization.

The clinical presentation of the patient reported conforms to the literature, wherein approximately half of all cases of cerebral AVMs presents with an intracranial hemorrhage.[1,4] The annual risk of bleeding is about 2% to 4% per year.[2,5] Hartmann *et al.*[6] in a series of 115 patients presenting with intracranial hemorrhage secondary to AVM, found the following distribution: 30% SAH, intraparenchymal hemorrhage 25%, 16% intraventricular and 31% combined hemorrhage. In the same series Hartmann *et al.*[6] found a higher association of focal neurological deficits in parenchymal hemorrhage (51.9%), followed by SAH (41.2%) and, finally, exclusively intraventricular bleeding (27.8%). In our case, despite recurrent episodes of bleeding, presenting over 20 years between them, the patient had no definitive neurological sequelae probably because the hemorrhage was predominantly intraventricular. Furthermore, the younger age of patients with AVMs may explain the better recovery when compared with aneurismal subarachnoid hemorrhage; as proposed by Hartmann *et al.*[6]

The angioarchitecture and the location of the AVM may have also influenced the type of hemorrhage

observed in the present case. Turjman *et al.*[7] correlated six parameters of the vascular architecture predicting the type of hemorrhage. Among these, two characteristics were demonstrated by angiographic studies of this patient: midline feeding perforating branches and the deep venous drainage.[8,9] Turjman *et al.*,[7] reported that the fact that the AVM is located in the *septum pellucidum* is considered a risk factor for intraventricular hemorrhage.

The main reason to treat AVMs is prophylaxis against bleeding. Multiple techniques exist currently to treat intracranial AVMs, and each modality has its own complications.[10-12] Surgical treatment is safe and a more efficient to protect the majority of patients with low grade AVMs from the disastrous effects of a rupture.[13-18] However, Stein[3] said AVMs located on the medial surface of the cerebral hemisphere represent a major challenge for the neurosurgeon to get a proper and complete resection exposure. Currently, the most accepted concept is to individualize therapy modality for AVMs based on patient age, lesion location, the angioarchitecture and current medical conditions. Thus, is formulated that the best treatment option applies to each patient to maximize clinical benefit, minimize neurological sequelae and optimize their quality of life.

A combination therapy was selected to treat this patient based on the concepts listed above. The rational employed directed the initial use of the endovascular approach with preoperative embolization to minimize the operative risk. Spetzler and Martin[18] classified AVMs based on three parameters: the size of the AVM, the venous drainage and involvement of eloquent areas. The risk to the patient increases with the size of the malformation, deep venous drainage, and location in an eloquent area. In the present case, the malformation was located deep in the midline of the cerebral hemispheres, had deep venous drainage into the internal cerebral vein, and was located adjacent to the midline hemispheric and septal areas. It was thus necessary to reduce the risk of a surgical approach, by the immediate reduction in blood flow to the AVM by endovascular intervention, decreasing the intracranial hypertension by external ventricular drainage, softening the brain, and limiting subsequent surgical catastrophic sequela.

Competing interests

The authors declare no conflict of interest.

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