Perimesencephalic Cerebral Arteriovenous Malformation (AVM): case report

Malformação Arteriovenosa Cerebral Perimesencefálica (MAV): relato de caso

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ABSTRACT

Background: Cerebral arteriovenous malformation (AVM) evolves asymptptomatically and can cause serious injuries, most of the time irreversible, with difficult diagnosis, controversial treatment and restricted access to reference therapeutic centers. Case Presentation: a case of intraparenchymal hemorrhagic stroke with presence of a left perimesencephalic AVM in a 66-year-old male patient is reported. Regarding the therapeutic approach, the AVM correction procedure through micro neurosurgical resection is described here. Conclusion: With this in mind, the present study aimed to meet this expectation, as well as to describe the correction of silent AVM after intraparenchymal hemorrhagic accident.

Keywords: Arteriovenous Malformation; AVM; Intraparenchymal hemorrhage; Treatment

RESUMO

Introdução: A malformação arteriovenosa cerebral (MAV) evolui de forma assintomática e pode causar lesões graves, na maioria das vezes irreversíveis, com diagnóstico difícil, tratamento controverso e acesso restrito a centros terapêuticos de referência. Relato de caso: relato de um caso de acidente vascular cerebral hemorrágico intraparenquimatoso com presença de MAV perimesencefálica esquerda, em paciente do sexo masculino de 66 anos. Em relação à abordagem terapêutica, descreve-se aqui o procedimento de correção da MAV por meio de micro-ressecção neurocirúrgica.

Conclusão: Pensando nisso, o presente estudo teve como objetivo discorrer a história clínica, bem como descrever a correção da MAV silenciosa após acidente hemorrágico intraparenquimatoso.

Palavras-Chave: Malformação Arteriovenosa; MAV; Hemorragia intraparenquimatosa; Tratamento

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BACKGROUND

Vascular malformations are categorized as capillary, arterial, venous or lymphatic, being that the coexistence of different vessels in the same lesion may be common. According to the blood flow, can be divided in high or low. Those with high flow comprise arterial malformation (AM), arteriovenous fistula (AVF) or arteriovenous malformation (AVM). The low flow ones are venous malformation (VM), lymphatic malformation (ML) and capillary malformation (MC). In addition, there may be complex combined malformations, e.g. lymphatic capillary (MCL), venous capillary (MVC), lymphatic venous (MLV), arterial capillary (MAC), capillary venous arterial (MCAV) and lymphatic venous arterial capillary (MCAVL). They can also be classified as localized or diffuse. When ruptured, the subsequent risk of death increase, depends on association with aneurysms, deep locations, deep drainage, age and others risk factors.

Regarding the diagnosis, it can occur by means of angioimaging, revealing the delimitation of the malformation. These morphological changes consequently lead to functional changes directly and indirectly on the affected tissue. Regarding the prognosis, these may be non-modifiable or generate structural and/or functional sequels, and may even cause risk of death.

The diagnosis, treatment and follow-up of the patient is multidisciplinary and interprofessional, increasing resolution and effectiveness. Thereby, the actions to be carried should be clear and disclosing, for those better protocols diagnosis and therapeutical can be executed and better prognostics can be reached.

CASE PRESENTATION

This case report description was approved and accepted by the research Ethics Committee (CEP – Plataforma Brasil), under register 4,947,483. The participant and any identifiable individuals consented to publication of his image.

A 66-year-old male patient arrived at the service with right hemiplegia, dysarthria, lowered level of consciousness and Glasgow eleven, and in the following hours was seven, resulting in intubation. At the first cranial computed tomography (CT) imaging exam, left mesencephalic hemorrhage and presence of blood in the ventricle was revealed (Figure 1A). His previous morbid history indexed to the medical record did not present any comorbidity. Thus, in line with the previous findings, arteriography was performed in which perimesencephalic AVM could be seen on the left SM3 (Figures 1B and C).

With the diagnosis of intraparenchymal hemorrhagic stroke with the presence of a left perimesencephalic AVM, the chosen therapeutic approach was to perform microsurgery for resection of the AVM in the posterior fossa. The surgical procedure took place with the patient in a sitting position, head in flexion, without rotation or lateral flexion (Figure 2A). Straight incision of approximately 12 cm was performed in the sub-occipital region in the midline from IO to C1. The upper portion of the incision was curved to avoid a superficial pressure ulcer. The muscular plane was dissected through the nuchal ligament until the exposure of the external occipital protuberance, superior nuchal lines, external occipital crest, posterior border of foramen magnum (opistium), posterior atlanto-occipital membrane and posterior C1 arch (Figure 2B). Supra and infratentorial suboccipital craniotomy was performed using three burr holes, one of which located in the midline 2 cm above the external occipital protuberance and the other two holes located just below the superior nuchal line, every 5 cm apart of the midline. With the posterior arch of C1 preserved, V-shaped durotomy was performed, thus allowing the flaps to be folded upwards and the suboccipital surface of the two cerebellar hemispheres to be exposed (superior and inferior semilunar lobes, biventral lobe, pyramid and uvula) (Figure 2C).

To reach the AVM, the tentorium-pontine-cerebellar veins were cauterized, allowing the visualization and identification of the uvula, pyramid, lobes of the cerebellar hemispheres and arachnoid membrane. The cistern arachnoid was then dissected, thus revealing the AVM (Figure 3A). Nidus and afferent branches of the AVM were identified (Figure 3B) and cauterized, in the same way, the main drainage vein (precentral cerebellar vein). With the completion of the resection process, hemostasis of the quadrigeminal cistern region was verified and the cavity was revised and it was identified the maintenance of pulsation of vascular structures (Figure 3D).
After resection, cranioplasty was performed. The surgical procedure was uneventful. In the postoperative evaluation, the patient was bedridden, non-compacting, afebrile, in a persistent vegetative state, and had a sacral decubitus ulcer. A new postoperative arteriography was then performed, which revealed normal anatomy of the basilar artery (BA), the posterior cerebral artery (PCA), the superior cerebellar arteries (SCA), the antero-inferior cerebellar artery (AICA) and the postero-inferior cerebellar artery (PICA), and the absence of perimesencephalic AVM was also observed (Figures 4A-C).

Figure 1. A. Angiotomography showing intraventricular hemorrhage and cannula shunt on the left. B. beginning of contrast flow in the left posterior cerebral artery (LPCA). C. evidence of contrast accumulation in the center of the third branch of the LPCA, AVM of the SM3.

Figure 2. A. Positioning the patient in a seated position with head in flexion. B. dissected muscular plane with exposure of the occipital bone. C. Craniotomy and durotomy completed with exposure of the cerebellar hemispheres.
DISCUSSION

The AVM case reported was a silent condition, which, after an hemorrhagic accident, caused irreversible damage to the healthy patient and who had no previous comorbidities. Cases like this call for early diagnosis and resolute therapies, since due to the complicated screening of vascular irregularities, the diagnosis usually occurs only after vessel dehiscence. In this patient the intraparenchymal hemorrhagic episode have been caused by rupture of the AVM, generating the beginning of the signals and symptoms. With this scenario, the initial diagnostic hypothesis was stroke, so there was a delay between the differentiation between hemorrhagic stroke and confirmation of AVM (hours). The structure of the health system was very important for

Figure 3. A and B. AVM afferents can be visualized through the branches of the posteromedial and lateral choroidal arteries of the posterior cerebral artery. C. nidus coagulation; and D. extraction of the AVM nidus and draining vein with normal staining.

Figure 4. A. Preoperative arteriography, with the yellow dotted circle demarcating the perimesencephalic AVM area. B. arteriography showing the preservation of PCA, AICA, PICA, BA, and VA; and. C. Arteriography showing preservation of SCA.
achievement of results, the possibility of significant results for medical care.

Cerebral hemorrhage is a condition with low resolution that demonstrates high morbidity and mortality, in which patients who have injury may acquire focal motor deficits, functional sequelae, epilepsy, in addition to a permanent vegetative state and, in most severe cases, progress to death\(^8,9\). The present report goes according to the evolution to severe sequelae and death.

Early diagnosis is usually due to clinical investigation of other vascular comorbidities, as well as the presentation of nonspecific neurological manifestations (mainly due to bleeding episodes), seizures, headache and focal losses that can be motor, visual or somatosensory\(^10-13\). Therefore, the identification by neuroimaging and angiographic exams is essential, as well as diagnostic confirmation\(^14,15\). At the service where the patient was admitted, the diagnostic method used was initially computed tomography angiography, that indicated the lesion and facilitated the decision of the therapeutic approach. Microsurgery is the most widespread and used method, and the technique consists of anatomical exposure, occlusion of the feeding arteries, preservation of the vessels that pass through the region, circumferential dissection of the lesion, cessation of venous drainage and finally the removal of the nidus\(^16,17\).

The treatment and follow-up of the case was microsurgical, but other treatment options have the following modalities: conservative and interventional, which are divided into surgery, microsurgery, stereotaxic radiosurgery and endovascular embolization\(^18-20\).

Surgical resection presents high risk post-operatively, but this and the benefit colide when comparing with outcome of surgeries performed when total resection is achieved, in this context there is elimination of hemorrhagic danger and prolonged durability of hemostatic stabilization\(^19,21\). Complications of these methods consist mainly of hospital risks due to the long stay, stroke, intraoperative injury in the case of microsurgery, intraoperative or postoperative ischemic or hemorrhagic stroke in the case of embolization, and hemorrhagic risk in the latency period (time between radiation and tissue response with respect to radiosurgery)\(^11,12,22\). Therefore, in the present case no identification of new hemorrhagic focus or others complications were seen, however, the long stay was necessary for the other systemic issues.

The post-surgical vegetative state resulted in decompensatory systemic complications and culminated in death.

**CONCLUSION**

The correction of AVM by microneurosurgery is a therapeutic approach widely used in referral and high complexity centers. The description of resection procedures leads to the presentation of access and correction approaches that can contribute to clinical vision and therapeutic designs. With this in mind, the present study aimed to meet this expectation, as well as to describe the correction of silent AVM after intraparenchymal hemorrhagic accident.

**REFERENCES**

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