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Case Report

Occipital Cavernous Malformation. Case Report and Literature Review

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Abstract

We present a 47-year-old female patient with a personal pathological history of squamous cell carcinoma of the cervix treated with multimodal therapy. She is rushed to our hospital because of a generalized tonic-clonic seizure. After performing computed axial tomography and magnetic resonance imaging studies of the skull, an occipital lesion was evident that was located above the tentorium cerebellum in relation to her right transverse sinus. An occipital approach is performed where said lesion is completely resected. The anatomical-pathological result shows a Cavernoma, an unusual vascular lesion of the nervous system.

***Keywords:** Brain vascular malformation, Cerebral cavernoma, cervix*

Introduction

Cerebral vascular malformations constitute congenital alterations of vascular morphogenesis caused by failures or dysfunction of the embryonic process of capillary maturation that result from the abnormal formation of arterial, venous or capillary channels with or without the presence of direct arteriovenous communications. (1)

Cerebral cavernous malformation is a cerebromicrovascular disease that affects up to 0.5% of the population. The decrease in endothelial cell-cell contact and the loss of junctional complexes lead to the loss of the integrity of the brain endothelial barrier and the formation of hemorrhagic lesions. Cavernomas, the third most common vascular malformation in some series, are composed of malformed and multilobulated capillaries that slowly increase in size and undergo thrombosis and progressive fibrosis with calcification; consisting of a group of sinusoidal vascular walls composed of a single layer of endothelium, absence of smooth muscle fibers and collagen. The capillaries are immediately adjacent to each other; and lack intermediate neural parenchyma or identifiable mature vessel wall elements. They are typically not associated with dilated afferent arteries or draining veins, and blood flow is low or stagnant. They are low-flow, low-resistance vascular lesions. (2)

The genes responsible for cavernous malformation in its familial form of presentation have been studied; these are mainly attributed to the genes: KRIT1, CCM2, PDCD10; on the other hand, cases of sporadic cavernous malformation have been associated with mutations in the RNF213 gene. (3)

It was decided to present the following case with the objective of describing the unusual vascular malformation and its treatment alternatives.

Case Presentation

Female patient, 47 years old, right-handed, with a personal pathological history of squamous cell carcinoma of the cervix treated with multimodal therapy (Surgery/Radiotherapy/Chemotherapy). She was rushed to our hospital due to a generalized tonic-clonic seizure, an episode that she repeated on several occasions despite treatment with anticonvulsants.

Physical Examination (Positive)

Glasgow Coma Scale: 15/15 points

Isochoric and isoreactive pupils.

No focal motor defect

Upper left homonymous quadranthopnosia

Fundus of the eye: without alterations

Complementary Exams

Simple Brain CT Scan. (Figure 1)

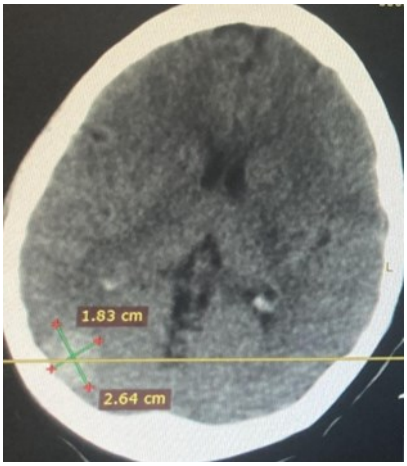

Figure 1: *A hyperdense lesion with well-defined edges measuring 1.83X 2.64X 1.67 cm for a volume of 4.03 cm 3 is observed in axial sections, in the right occipital region, which does not exert a significant mass effect on neighboring structures nor does it present perilesional edema.*

A CT scan of the chest, abdomen and pelvis was performed under the medical belief of a secondary injury to the nervous system. These studies were negative, so possible injuries to other organ systems were ruled out. A nuclear magnetic resonance imaging was performed (MRI) (0.35T) of Simple Skull to acquire more imaging information of said lesion. (Figure 2)

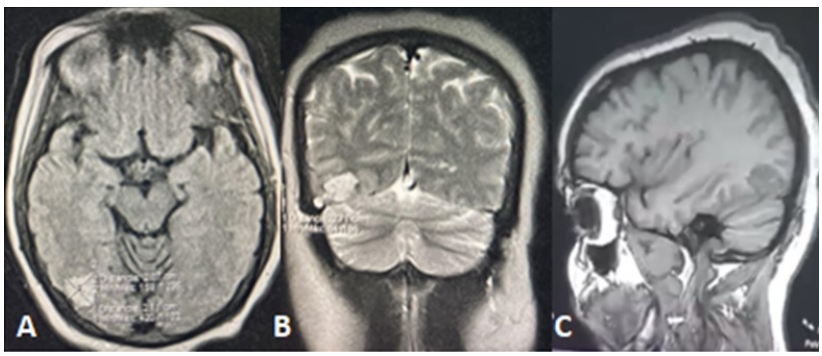

Figure 2. A: *Simple skull MRI 0.35 T. Axial sections. FLAIR weighting. A hyperintense lesion of probable tumor etiology with irregular edges without mass effect or perilesional edema is observed in the right occipital region.* ***B:*** *Coronal cuts. T2 weighting. The hyperintense lesion described above is observed that rides over the tentorium cerebellum and is located in relation to the right transverse sinus.* ***C:*** *Sagittal sections. T1-weighted: The lesion described above is observed, but with a significant decrease in its intensity (hypointense) in supratentorial subcortical T1.*

Based on the data obtained, it was decided to offer surgical treatment under the imaging suspicion of a brain metastasis, in which an occipital approach was applied with prior planning of the supposed location of the lesion in which it was possible to completely excise it. without transoperative or postoperative complications. (Figure 3)

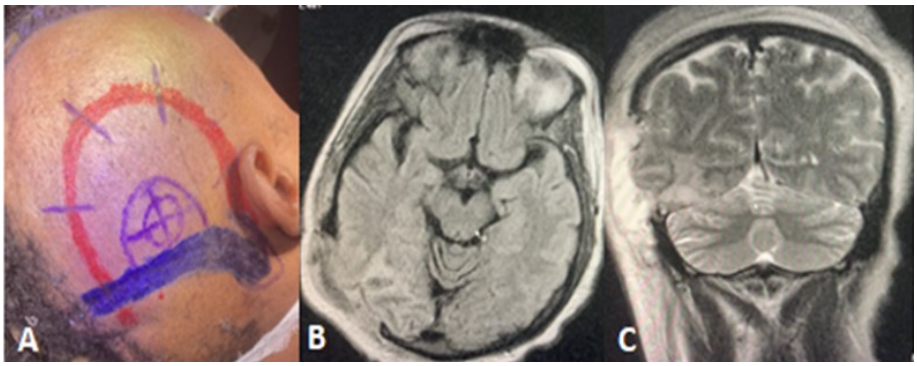

Figure 3: A: *Topographical planning of the lesion located above the tentorium in direct relation to the transverse sinus.* ***B, C:*** *Magnetic resonance imaging of the skull, axial and coronal section, FLAIR and T2 weighting respectively, 5 days after surgery, showing total resection of the lesion.*

The patient evolved favorably with a total absence of seizures after surgery; she had a hospital stay of 7 days.

After an exhaustive review by the neuropathology team, it was determined that the resected lesion presented the characteristics of a Cavernoma (Figure 4), which corresponds to an unusual brain vascular malformation whose main complications include intra-extralesional bleeding and seizures refractory to medical therapy, the latter present in the operated patient.

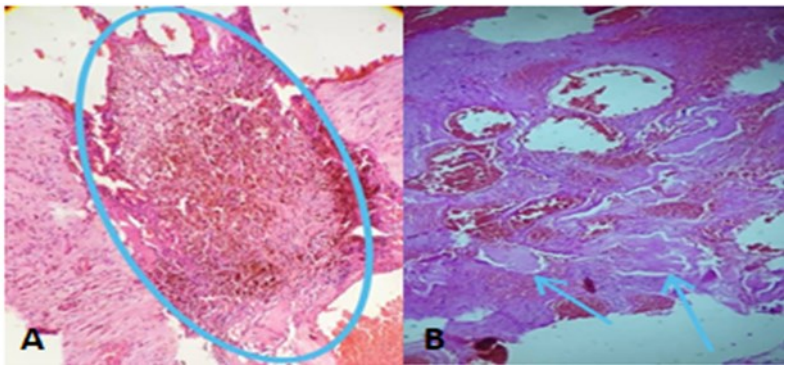

Figure 4: A: *Numerous cavernous vessels and foamy macrophages with hemosiderin deposits are observed.* ***B:*** *Brain tissue with numerous thrombosed cavernous vessels suggestive of cavernous cerebral vascular malformation.*

Discussion

For several years now, the therapeutic options to treat cavernous lesions of the nervous system have been surgery, radiosurgery and conservative therapy with serial imaging follow-up.

In 1994, Zabramski et al., under the supervision of Professor Spetzler, published a four-grade imaging classification with subtypes that didactically offered a prognosis of the possibility of bleeding per year in relation to the imaging and anatomo-pathological characteristics that the lesion presented, extremely important to define surgical conduct or radiosurgery, where they defined a greater risk for type I and II injuries.[4]

Last year, Fontanella et al.[5] validated an international system to define surgical treatment in patients with cerebral and cerebellar cavernomas, in which, through the evaluation of neuroimaging characteristics such as: size and serial growth of the lesion, bleeding and location , clinical features: focal motor defect and presence of seizures, and age of the patient, offer a score to determine when a patient with this type of injury is eligible for surgery.

Regarding the most effective treatment alternative, there are still controversies; a meta-analysis showed that the effectiveness in ensuring the prevention of hemorrhage was 97% in surgical treatment, 86% in radiosurgical treatment, and 77% in conservative, the lowest mortality (1%) was after radiosurgery and the highest persistent morbidity (22%) was in natural history series. It is concluded that surgical resection for cavernomas is effective in ensuring the prevention of hemorrhage with acceptable morbidity and mortality, but conservative and radiosurgical management is a justified treatment alternative. [6] The same position is demonstrated in similar meta-analyses. [7, 8, 9]

Other authors [10] justify surgery in lesions with a greater propensity for hemorrhagic and convulsive events, identified as those associated with the CCM3 gene, Zabramski type I and II lesions and location in the brain stem, with respect to the latter Lawton [11] proposed a scale specifically for brainstem cavernomas that suggests the type of lesion that should be operated on, as well as the most appropriate approach.

Another meta-analysis also shows the benefit of surgery in refractory epilepsy secondary to cerebral cavernomas, where it was shown that the epilepsy control rate was 88%, with a low risk of postoperative complications and mortality. [12]

Conclusion

We can conclude that surgery remains in most cases the most effective option in symptomatic cavernomas.

Conflict of Interest

The authors declare no conflict of interest.

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