Giant Cell Glioblastoma Associated with Intrinsic Arteriovenous Malformation: a Case Report

Glioblastoma de Células Gigantes Associado a Malformação Arteriovenosa Intrínseca:

Relato de Caso

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ABSTRACT

Background: Primary brain tumors associated with cerebral arteriovenous malformations (AVM) although known is a rarely reported finding. There have been approximately 50 cases reported. Only two cases of a single lesion with coexistence of AVM and glioblastoma were described. Case Report. We report a case of a 46-year-old woman with headache and seizures for 2 months who performed a MR which displayed a large right frontal lesion suggesting a glioblastoma. Results. During surgical resection, two large veins could be seen draining tumoral vascularization in the surrounding cortex. After reaching the deepest portion of tumor, veins were coagulated and cut to allow tumor removal. However, unexpected bleeding started to occur with associated lesion growth and edema, resembling an AVM. Material sent to pathology revealed giant glioblastoma in association to an AVM with some thrombosed vessels and ischemic necrosis. Two previous studies discussed similar cases. Conclusion. The need of magnetic resonance in all AVM cases is highlighted in order to analyze brain parenchyma within AVM. We also remark the need of pathological analysis of whole surgical material, in order to characterize whole piece and to avoid not diagnosing an associated lesion.

Key words: Arteriovenous malformation; Glioblastoma; Differential diagnosis; Treatment

RESUMO

Introdução: Os tumores cerebrais primários associados a malformações arteriovenosas cerebrais (MAV) são um achado conhecido, mas raramente relatado. Cerca de 50 casos foram relatados. Apenas dois casos de uma única lesão com coexistência de MAV e glioblastoma foram descritos. Objetivo e Métodos. Relatamos o caso de uma mulher de 46 anos com cefaleias e convulsões por 2 meses que realizou RM que apresentou lesão frontal direita sugerindo glioblastoma. Resultados. Durante a ressecção cirúrgica, foi possível ver duas grandes veias drenando no córtex circundante. Quando a porção mais profunda do tumor foi atingida, as veias foram coaguladas e cortadas para permitir a remoção do tumor. Entretanto, um sangramento inesperado iniciou com crescimento de lesão associada e edema, semelhante a MAV. Material enviado para patologia revelou glioblastoma de células gigantes em associação a uma MAV com alguns vasos trombosados e necrose isquêmica. Dois estudos anteriores discutiram casos semelhantes. Conclusão. Destaca-se a necessidade de ressonância magnética em todos os casos de MAV, a fim de analisar o parênquima cerebral ao redor da MAV. Observamos também a necessidade de análise patológica de todo o material cirúrgico, a fim de caracterizar a peça inteira e permitir o diagnóstico de uma potencial lesão associada.

Palavras-chave: Malformação arteriovenosa; Glioblastoma; Diagnóstico diferencial; Tratamento

INTRODUCTION

Primary brain tumors associated with cerebral arteriovenous malformation (AVM) are a known but rarely reported finding1,2,3. There have been approximately 50 cases reported. In most cases, there is association of a low grade glioma or a meningioma in close proximity to an AVM1,2,3. Only two cases of a single lesion with coexistence of AVM and glioblastoma were reported2,3. We report a very similar case resembling glioblastoma which turned to be an association of both lesions.
CASE DESCRIPTION

A 46-year-old Angolan woman who was attended in Brazil one year ago to evaluate progressive headache for 2 months and 2 tonic-clonic seizures, and initially considered as a consequence of malaria, which was her previous known diagnosis. She presented with Karnofsky score (KS) of 100 and no neurological impairment. She underwent a MRI and an heterogeneous and contrast enhancing lesion in right frontal lobe was disclosed with rich vascularization and large drainage veins in cortical surface, highly suggesting glioblastoma (Figure 1).

Patient was submitted to surgical resection under microsurgical technique. Just after dural opening, it could be seen the lesion with a visible surgical plane and two large veins draining tumoral vascularization to surrounding cortex (Figure 2). Veins were crossing surgical plane and were left initially untouched to avoid bleeding. A delicate tumoral dissection was conducted along tumoral surface in the cortical and subcortical space.

After reaching the deepest portion of tumor, the veins were coagulated and cut to allow tumor removal. However, unexpected bleeding started to occur with associated lesion growth and edema. Some bleedings were without parenchyma manipulation and other with minor parenchymal “touches”, similarly to the behavior of AVMs with coagulated drainage veins. At this point, bleedings recurred and a faster resection was done to remove AVM nidus and avoid further bleedings. After complete resection, adequate hemostasis was carried out and the patient recovered well, leaving intensive care unit in the second postoperative day, being discharged in the fifth.

Material sent to pathology consisted of a 6x5x5 cm piece. Microscopical analysis revealed immature neuroepithelial neoplasm, with intense pleomorphism and giant multinucleated cells, some of them bizarre. Interface with normal brain could be seen in some surfaces. There were also intermediate size cells, with clear perinuclear circles and dystrophic calcifications, suggesting an oligo-like component. A high mitotic index, microvascular proliferation and microtumoral necrosis could also be observed. It could also be found an AVM within the tumor, with some thrombosed vessels and ischemic necrosis (Figure 3).

Postoperative image studies revealed complete resection of lesion. Patient was forwarded to oncological adjuvant therapy and performed radiotherapy and temozolomide for 1 year. Now, she returned and a recent MRI reveals no evidence of residual or recurrent tumor or AVM (Figure 1). Her KFS is 100 with normal life.
If there is a causal relationship for AVM and tumoral association or if it happens simply by chance remains unanswered. Cushing and Eisenhardt suggested that chronic irritation of the arachnoid cells caused by the increased blood flow may initiate a pathological process. Other authors have pointed factors such as endothelial cell turnover, elevated proangiogenic cytokine secretion (including angiopoietin-2 and vascular endothelial growth factor), and increased angiogenic receptor expression in genesis of tumor.

Bitoh et al. proposed a division of lesions into three types. Type I are lesions separated from each other and situated in different anatomical locations, type II are cerebral AVMs contiguous with the tumoral mass, with or without a clear cleavage plane from the latter, while type III are cerebral AVMs adjacent to the tumor within the same gyrus or lobe.

Literature review disclosed previous described association of AVM with several tumors, including meningiomas, hemangiomas, oligodendroglomas, astrocytomas and teratomas. Only two cases reported association with glioblastoma.

Aucourt et al. reported a glioblastoma coexisting with an arteriovenous malformation in a 65-year-old man. MRI revealed a necrotic and cystic lesion in the left hemisphere with dark vessel-like signals on T2 and susceptibility-weighted imaging (SWI). The AVM was confirmed by a preoperative cerebral angiography. Pathological report revealed atypical cells characteristic of glioblastoma and, in the same area, arteriovenous malformation.

Gmeiner et al. reported a 72-year old woman with MR imaging suggestive of a high-grade glioma. Intraoperatively, the lesion resembled a vascular malformation. Total extirpation of the lesion was performed and initial histopathological analysis revealed AVM. Two months later, multiple lesions were visible on MR and supported the diagnosis of malignant glioma. A review in histopathological sections could identify glioblastoma in only 5% of the section surrounded by AVM.

Similarly to Gmeiner’s case, we managed our patient initially as a glioblastoma with rich vascularization and did not perform preoperative angiographic study. Just during surgery we could notice venous anatomy which resembled an AVM. After vein coagulation, whole lesion became fragile and bled easily, behaving like AVM.

Notwithstanding either coincidental or causal relationship, the management of these lesions must address maximal surgical resection and further adjunctive therapies.

We highlight the need of performing a magnetic resonance in all AVM cases in order to analyze brain parenchyma within AVM. Finally, we also remark the need of pathological analysis of whole surgical material, in order to characterize whole piece and to avoid not diagnosing an associated lesion.

Figure 3. Histology image revealing immature neuroepithelial neoplasm with intense pleomorphism and giant multinucleated cells. It could also be found an AVM within the tumor, with some thrombosed vessels and ischemic necrosis.
REFERENCES


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