Association between cavernous angioma and cerebral glioma. Report of two cases and literature review of so-called angiogliomas

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Summary

The association between vascular malformations and cerebral gliomas is unusual. While the association between cavernous angioma with gliomatous lesions is even more rare, it is considered by certain authors to be a particular pathological entity termed angioglioma. The authors report on two cases of association of a cavernous angioma with a ganglioglioma and an oligodendroglioma respectively. Subsequent review of the literature on the so-called angiogliomas was conducted. In the author's opinion, the entity of angiogliomas represents a general spectrum of angiomatous neoplasms that include gliomatous tumors, in the majority low-grade gliomas, associated with a major vascular component.


Introduction

Councilmann in 1930 was the first to report the name of “angiogliomas” describing a cerebellar tumor with a huge vascular component. From Councilmann until nowadays, only a few authors have reported this entity along with an attempt of a better definition of such pathology.

Indeed, Roussoy and Oberling in 1930 classified CNS neoplasms and described as well the angioglioma that it was defined as a tumor with a glial part, generally of low-grade, along with a notable vascular component. Although, Rubinstein accepted the term angioglioma; in a more recent publication he suggested the restriction of the term angiogliomas to denote a mixed tumor composed of an hemangioblastoma and astrocitoma. Sugita et al., after reporting a case of a xanthoastrocitoma highly vascularized, proposed the use of the term angiomatous glioma intending to describe a malignant glial neoplasms highly vascularized, while leaving the term angiogliomas just for mixed neoplasms composed of a low-grade glioma and a vascular malformation of any kind.

The presence of two angiogliomas in our own series of low-grade gliomas leaded us to review the literature on this rare pathological condition.

Case 1

A 16-year-old girl was admitted in our department for progressive bilateral facial palsy. Neurological examination evidenced a central bilateral VII cranial nerve dysfunction. A brain MRI with and without contrast demonstrated a hypothalamic chiasmatic tumor with disomogenous contrast enhancement, irregular borders and slight perilesional edema (Fig1). A bilateral subfrontal approach was carried out with subtotal resection of the tumor. During surgery, an intracerebral mass with thrombosed vessels and considerable vascular lakes was found. Histological diagnosis was ganglioglioma with glial component of pyloid type and associated vascular component of angiomatous type (angio-
glioma). The vascular component was observed in some regions of the tumoral mass. It presented itself with hyalinized ectasic vascular channels, perivascular hemosiderin deposits, gliosis and vascular calcifications. The gliomatous nature of the tumor was pathologically obvious, although in some points of the tumoral mass it was nearly darkened from this particular architecture of the vascular component (Fig 2a, b, c). Electron microscopy was not performed.

Postoperative course was complicated with a panhypopituitarism, which improved with pharmacological replacement therapy. Clinical and radiological follow-up of 76 months showed a non growing residual tumor and stable medical condition.

**Case 2**

A 38-year-old men was admitted in our department for bifrontal headaches for six months. Neurological exam was normal. A brain MRI with and without contrast demonstrated a right frontal lobe tumor with disomoge-
nous contrast enhancement and areas of hyperintensity surrounded by a hypointense ring due to the paramagnetic effects of hemosiderin (Fig. 3). A right frontal craniotomy was performed with a total removal of the tumor. Histological diagnosis was angiolglioma with a glial component of oligodendroglioma with polymorphous aspects; a prominent vascular component was observed in some regions of the tumoral mass. Hyalinized ecstatic vascular channels, perivascular hemosiderin deposits, gliosis and vascular calcification were noted (Fig. 4a, b). Postoperative evolution was uneventful. Clinical and radiological follow-up of 64 months showed a neurologically intact patient without tumoral recurrence on imaging studies.

Discussion

From a consecutive series of 168 low-grade gliomas surgically treated in our department from 1992 to 2003, we found 2 cases with histological diagnosis of “Cerebral Angioglioma”. After revision of these two cases it was evidenced that the vascular part assumed a cavernous-angioma like component, while the gliomatous part were a pylloid type ganglioglioma in one case and an oligodendroglioma in other case. In 1991 Lombardi et al.\textsuperscript{10} reviewed the histological diagnosis of a series of 1034 surgically treated MAV, and concluded that the entity of the angiolglioma belonged to a distinct and rare pathological category composed of a low-grade glial neoplasm with a rich vascular part, with same clinical and angiographic characteristics, along with prognosis, of those gliomas without an angioma-like vascular component. Palma et al.\textsuperscript{13} described other two cases with verified association between glioma and cavernous angioma, that were defined as examples of angiolgliomas.

Some authors\textsuperscript{1,5,6,11,20,21} expressed different etiological hypothesis including: genetical predisposition\textsuperscript{20}; reactive or malformative nature\textsuperscript{1,21}; viral origin\textsuperscript{5}; and exceptional
coincidence\textsuperscript{2,9}. White et al.\textsuperscript{20}, sustained the hypothesis of a genetical predisposition of these mixed tumors after reporting the autopsy of a 45 year-old male patient with cerebral, lung and hepatic cavernomas deceased after surgery for a septum pellucidum astrocytoma.

Crowell\textsuperscript{4}, Nazek\textsuperscript{12} and Lombardi\textsuperscript{10}, suggested that an exaggerated glial proliferation could be a reactive response to any kind of AVM, with subsequent chronic ischemia and white matter changes.

Fisher in 1982\textsuperscript{5}, reporting two cases of cavernous angioma associated with an oligodendroglioma and a pylocitic astrocytoma respectively, considered a viral cause as a possible pathogenesis. Indeed, cavernomas can be induced experimentally in rats after inoculation of Polyoma virus, while the neoplasm can be induced directly after inoculation of SV40 virus of the Papova family.

Nazek et al.\textsuperscript{12} reported 3 cases of AVM association with abnormal oligodendrogial proliferation, and Spetzler\textsuperscript{18} cited a personal case of association between an AVM and an oligodendroglialoma, affirming the existence of a rare coincidence between AVM and gliomas. In our first case the histological diagnosis was ganglioglioma with glial component of pyloid type and associated vascular component of angiomatous type, while in our second case histological examination showed a glial component of oligodendroglioma with polymorphous aspects; in both cases the vascular component was observed in some regions of the tumoral mass.

We can include cases of angiogliomas either as an exclusive pathological entity, or as a unique category of angiogliomatous tumors composed of a low-grade glioma associated with an important vascular component. However, after reviewing the literature and our cases, the following subcategories can be described:

- Hypervascularized low-grade gliomas.
- Mixed glioma, with an angiomatous and a glial component, both components secondary to oncogenic factors.
- Serendipity of an AVM with a glioma in two adjacent contiguous regions.
- Coincidence of an AVM with a glioma in two distant regions cannot be considered within angiogliomas and this entity should be considered as coexistence of two different pathologies.
- The exuberant glial proliferation that is seen in cases of AVM regions cannot be considered within angiogliomas, because it seems to be secondary to tissue changes after chronic ischemia and gliosis. These cases did not demonstrated progression to glial neoplasm and have a natural history similar to other AVMs.
- Prognosis of angiogliomatous tumors depends on different factors: a) intrinsic biological behavior, malignancy and tumoral grading; b) tumoral location; c) amenability and extent of resection; d) adjuvant therapies. In our first case follow-up of 76 months showed a non-growing residual tumor and stable medical condition; in the second case, follow-up of 64 months showed a neurologically intact patient without tumoral recurrence on MR images.

**Conclusions**

Based on our experience and on the literature review, we imply that the presence of rare cases of any kind of AVM associated with a glioma can be interpreted as a truly compound tumor, less likely to be an exceptional coincidence. We suggest that the entity of angiogliomas represents a general spectrum of angiomatous neoplasms that include gliomatous tumors, in the majority low-grade gliomas, associated with a major vascular component.

**References**


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