Speech Reorganization after an AVM Bleed Cured by Embolization

A Case Report and Review of the Literature

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Summary

Intracerebral arteriovenous malformations (AVMs) are defined as the direct communication of arteries to abnormal veins without interposing capillaries. Although AVMs can have various clinical presentations due to their dynamic nature, the most common presenting sign is intracerebral hemorrhage. Whenever an AVM is discovered, the therapeutic choice is often not obvious and it is influenced not only by the hemodynamic features of the AVM, but also by considerations of the extent of intervention-related morbidity and mortality.

A patient with a left frontal AVM is described. He bled three years after gamma knife radiosurgery and developed aphasia. The complete obliteration of the AVM was later achieved by embolization.

Functional compensatory brain reorganization and plasticity is discussed, since our patient presented with a fast recovery from aphasia and unexpected contralateral redistribution of the speech function and with preference for his second spoken language.

Introduction

Intracerebral arteriovenous malformations (AVMs) are defined as the direct communication of arteries to abnormally tortuous and dilated veins without interposing capillaries¹. Even though AVMs have long been considered

to be congenital in nature, the evidence suggests that they are not static lesions but dynamic, showing the ability to grow, regress and even reappear over time². This behavior can explain the extremely variable clinical presentations of AVMs, which can be related to mass effect, hemodynamic modifications or even focal area dysfunction.

Intracerebral hemorrhage associated with AVMs is the most common clinical presentation^{3,4}, and its causes and prognosis have been the subject of many reports⁵⁻⁷. Other presentations include seizures, mass effect, ischemic steal and also headache³. However, many AVMs are asymptomatic and are discovered incidentally^{3,4}. The natural history of this type of lesion is controversial: the annual risk of intracranial hemorrhage among people with AVMs who present with symptoms other than hemorrhage is 2% to 3% per year⁸.

As a consequence of AVMs' heterogeneous natural history, the treatment choice is not obvious. Location, size and hemodynamic features (e.g. venous drainage), together with the intervention-related morbidity and mortality, are the main characteristics that affect the selection of the best therapeutic approach⁹. Functional magnetic resonance imaging (fMRI) and positron emission tomography (PET) studies provide important information about adjacent eloquent cortex.

We describe a case with a left frontal AVM, who bled three years after being treated by

gamma knife radiosurgery, in order to discuss the possible implications of different therapeutic approaches in the evolution of this condition. We also describe the functional compensatory brain re-organization observed in this case, and we discuss it in relation to a long-standing lesion with an acute injury.

Case Report

A 21-year-old right-handed male presented in March 2004 with a first partial, secondary generalized motor seizure. The patient did not complain of a previous headache, vomiting or loss of consciousness. A CT scan with and without contrast disclosed a left frontal superficial arteriovenous malformation. At that time the patient was taking sodium valproate three times per day and no other seizures were reported except for the initial one.

The patient's medical history was unremarkable. The family history was negative for seizures or other neurological conditions. He was an occasional smoker. His psychomotor development was completely normal. He was bilingual: French was his mother tongue and he had learnt English in school.

The brain MRI confirmed the arteriovenous malformation located in the pars frontalis and triangularis of the left inferior frontal gyrus (Figure 1A). Subsequently, a cerebral angiogram demonstrated a 2 cm, superficial arteriovenous malformation, located in the region of the inferior frontal gyrus (Figure 2A). The AVM was supplied by two prefrontal branches of the left middle cerebral artery. Venous drainage was exclusively superficial via the vein of Labbé. Due to the location, probably in close contiguity to the speech area, the AVM was attributed as Grade II of the Spetzler-Martin Classification⁹.

A PET scan with brain MRI revealed a predominance of left-sided speech activation, located just posterior and anterior to the AVM (Figure 1B), while motor function was distant from the lesion. In consideration of this, the treatment options available (surgery, embolization, stereotactic radiosurgery) and their associated risks were discussed with the patient and his family. Although embolization followed by surgery was recommended by our treating team, the family chose a stereotactic radiosurgical option and was referred to another institution. The young-man was treated using gam-

ma-knife in April 2005 and he underwent periodic MRI follow-up examinations at that center; these scans did not demonstrate any obliteration of the AVM.

In December 2008, three years after the gamma-knife radiosurgery treatment, he was admitted to another hospital for sudden onset of headache, severe aphasia, fine motor impairment of the right hand, and right facial involvement. A CT scan demonstrated a left fronto-insular hemorrhage. He was then transferred to our Institute. The cerebral angiogram showed a reduction in size and a change in shape of the left frontal insular AVM. A new pseudoaneurysm at the origin of the venous drainage, and a focal stenosis of the draining vein of Labbé at its confluence with the left transverse sinus were noted. Mass effect on the left middle cerebral artery branches was evident caused by the intracerebral hemorrhage.

Clinically, the global aphasia evolved within three weeks into an expressive aphasia and a specific rehabilitation program of speech therapy in his mother tongue (French) was started during the hospitalization and was then continued on an out-patient basis. The patient's fine motor impairment and facial deficit recovered completely after just a few weeks.

The MRI of the brain performed later revealed the sequelae of the intracranial hemorrhage in the left inferior frontal gyrus. The venous pseudoaneurysm was located in the superior, medial and posterior aspect of the nidus and protruding into the cavity left by the hematoma (Figure 3). The size of the pseudoaneurysm progressed from December 2008 to April 2009, with a final diameter of 7 mm (Figure 2B).

In the meantime the patient recovered from aphasia, displaying no notable language difficulties and he returned to his university studies in the field of computer science. A PET scan study was repeated in 2009 and was compared to the 2004 scan (Figure 1C). Right hemisphere activation with a less lateralized localization was noted during performance on the same tasks of word repetition and synonym generation that were presented in the auditory modality. Also, on an object-action naming task presented visually, there was a predominance of right hemisphere CBF increases, with activation of the superior temporal gyrus, frontal areas and the thalamus. No activations were observed in the corresponding regions of the left

Figure 1 A) (top left) Axial MR PD WI demonstrating the left inferior frontal AVM. B) (top right) MRI-PET fusion examination obtained in 2004 showing the increase in CBF during synonym generation minus silent baseline and demonstrating the activity just adjacent to the AVM, in particular in the left frontal and temporal areas, and in the left thalamus. C) (bottom) MRI-PET fusion examination obtained in 2009 demonstrating the predominance of the right hemisphere CBF increases, with activation of the right superior temporal gyrus, right frontal areas and the right thalamus. The range of t-value for the PET data is coded by the color scale.

superior temporal gyrus and thalamus. Compared to the same examinations performed in 2004, the neurolinguistic tests assessed during the PET scan and also during an fMRI scan showed an increase of intrusions in response from a non-target language (English).

At that time embolization was suggested for nidus reduction and the patient underwent the procedure at the end of April 2009. The post embolization angiogram showed complete occlusion of the nidus and normal opacification of the vein of Labbé. Three months later, a small recanalization of the nidus occurred, requiring a second embolization which was completely successful (Figure 2C).

At the present time the patient is well and shows continued improvement in speech as measured by speech therapy. He complains of a slight reduction in concentration and in attention span, but this has not affected his work and daily activities.

Discussion

Several therapeutic options could be proposed for a symptomatic AVM. In each case, intervention should be undertaken with the goal of complete AVM obliteration, because subtotal therapy does not confer protection from hemorrhage⁸. Complete angiographic obliteration of the lesion is the gold standard of cure.

Stereotactic radiosurgery is now an established treatment for AVMs, together with neurosurgery and embolization^{3,10}. It is often recommended if an AVM is less than 3 cm in diameter and is located in an eloquent area³. Usually radiosurgery needs a period of approximately one to three years from the time of treatment to potentially complete obliteration of the AVM³. During this period rearrangements in angioarchitecture and hemodynamics of the AVM obviously can occur, secondary to the slow process leading to obliteration.

Figure 2 A) (top left) A lateral view of the left internal carotid angiogram showing the superficial AVM, supplied by two prefrontal branches of the left middle cerebral artery and the venous drainage via the vein of Labbé. B) (top right) Repeated angiogram after bleed showing the venous pseudoaneurysm and the stenosis at the confluence of the vein of Labbé with the transverse sinus. C) (bottom) Angiogram after second embolization demonstrating complete obliteration of the AVM.

In our case, we observed the modification in shape and size of the AVM occurred in a lag period of five years: an important change was the formation of a venous pseudoaneurysm along the drainage of the AVM.

As far as we know, the formation of venous pseudoaneurysms has never been described among possible complications of radiosurgery, which includes cyst formation, de novo formation of cavernomas, intracranial artery stenosis, dural arteriovenous fistula, and radiation necrosis^{3,11-13}.

The formation of the pseudoaneurysm could be the result of the modifications that occurred in the AVM over the years. AVMs are actually plastic lesions, which can change in shape and size, with consequent hemodynamic adjustments. Such mod-

ifications can occur in the natural course of the disease, but in our case it should also be asked if radiosurgery could have played a role in the formation of the pseudoaneurysm and, finally, lead to the hemorrhage. In fact, the venous pseudoaneurysm was probably the source of the hemorrhage, due to its location in the site of the hemorrhage.

The endothelial hyperplasia leading to the stenosis present at the junction of the vein of Labbé with the left transverse sinus could potentially have been responsible for the pseudoaneurysm formation.

Specifically, AVM-related venous pseudoaneurysms are reported in the literature^{3,14} and some authors^{8,15-17} consider their presence, especially if perinidal, a risk factor for rupture and hemorrhage.

Figure 3 Axial MR T2-weighted image after gadolinium injection, showing the venous pseudoaneurysm located in the medial and posterior aspect of the nidus of the AVM and protruding into the cavity left by the hematoma.

Embolization achieved complete obliteration of the AVM. Until now this treatment procedure, as a sole therapeutic modality, has usually been described for small lesions (less than 1 cm in diameter³) fed by no more than four arterial pedicles^{8,18-20}. In many series, permanent occlusion of brain AVMs by embolization was achieved in 10% to 30% of cases¹⁸. Our report provides an example of complete obliteration obtained with embolization, even in a case in which an AVM, fed by two distinct arteries, was complicated by venous pseudoaneurysm.

In patients with AVMs, the plasticity of the lesion is often accompanied by plasticity of the brain.

The nature and the extent of cerebral functions and recovery following neurologic disease are extremely variable. In particular, speech function in patients with chronic mass lesions, such as some AVMs, has been explored to clarify the characteristics of a compensatory network²¹ and a certain heterogeneity of results has been found. For example, a bilateral temporal lobe activation of language was shown in patients presenting with similar left temporal lobe AVMs located in regions that should be involved in language function^{22,23}. Another study suggested a redistribution of expres-

sive language function to the contralateral hemisphere in cases of AVM located in the left frontal language region, thus confirming the hypothesis of a contralateral re-organization of motor language behavior in this type of lesion²¹.

Nevertheless, our case showed a strict left hemispheric organization of language function at the first PET study, even though the AVM was located in the left inferior frontal gyrus. Surprisingly, after the AVM angioarchitectural re-organization and hemorrhage occurred, the language activations were now observed in the contralateral hemisphere even at a time when the patient's language functions completely recovered.

Follow-up investigations between functional studies help to understand if the language re-organization occurred secondary to the acute disease (hemorrhage) or, on the contrary, if it is the final result of a slow compensatory restructuring related to a long-standing lesion. The fast recovery from aphasia observed in this patient could be interpreted as supporting the second hypothesis, of a slow process of re-organization. On the other hand, before hemorrhage occurred, the AVM did not seem to influence the organization of language function, which was completely lateralized to the left hemisphere. Besides, a contralateral redistribution of speech function took place just after the hemorrhagic event.

Another interesting aspect of our case is the patients' bilingual recovery from aphasia. Our patient, a French-English bilingual, showed a peculiar preference for English words and definitions in the functional study performed five months after the hemorrhage. This could be unexpected given that the language testing and the speech therapy were carried out in the patient's first language (French). The Literature has been pointed out that a variety of recovery patterns are possible in patients who are bilingual with aphasia, depending on many factors such as age of second language acquisition, and relative level of proficiency and exposure²⁴⁻²⁶. In our case, the second language (English) was acquired at school age, after the classic critical period of plasticity of the developing brain. As a consequence, the preference for the second language noted in the functional study raises questions about the effective plasticity of the brain even into adulthood.

Conclusions

Functional imaging using PET and MRI are mandatory in the pre-operative assessment of this type of lesion, particularly when their loca-

tion is eloquent. Functional images give strategic information on brain plasticity and its evolution over time mainly when a dynamic lesion is present.

Reorganization of language may occur in situations of rapid anatomical change, such as in the case of our patient, where the bleed occurred in the speech area. The role of radiosurgery in modifying the angioarchitecture of AVM which may lead to hemorrhage is report-

ed. Finally, embolization of small AVM's is emphasized as the major treatment modality.

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