Amaurosis Fugax as the Presenting Manifestation of Dural Arteriovenous Malformation

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SUMMARY A 46-year-old woman under investigation for three episodes of amaurosis fugax in the left eye proved to have a left anterior-middle fossa dural arteriovenous malformation with pial venous drainage. The malformation received its main supply from the left middle meningeal artery, but its anterior part was fed by the recurrent meningeal branch of the left ophthalmic artery. Transient episodic lowering of retinal arterial pressure due to shunting of blood from the ophthalmic artery to the malformation is the most likely explanation for the occurrence of amaurosis fugax, indicating that this symptom occurs in some patients on a hemodynamic basis.

TRANSIENT ISCHEMIC ATTACKS rarely occur in relation to intracranial arteriovenous malformations (AVM's), and among them transient monocular blindness (TMB) seems to be exceptional. We report a patient with recurrent episodes of isolated TMB as the presenting manifestation of a dural AVM.

Patient Report

The patient, a 46-year-old white housewife, was admitted to University Hospital on July 17, 1984.

In 1979, she noted a continuous left frontal noise, without headache, which spontaneously resolved in a few minutes. One year before admission, she experienced transient monocular blindness in the left eye just after standing up from a chair, with no other associated symptoms. This loss of vision was characterized by a quick (2 seconds) centripetal darkening of vision to complete blindness; the eye maintained completely blind for 1 minute, followed by a progressive reappearance of the vision in the entire visual field within 30 seconds. Eight months later, while bending forward, the patient suffered a similar episode of transient monocular blindness in the left eye of slightly longer duration (5 minutes). Two months later, a similar event recurred while she was sitting in a boat, and lasted at least 15 minutes, but the patient experienced the loss of vision only in the lower half-field of the left eye, in an ascending curtain-like fashion. A few weeks later, she became aware of transient recurrence of the noise in the left forehead. A CT scan and a carotid angiogram were performed revealing two dural AVM's of the anterior-middle fossa: a larger one in the region of the pterion on the left and a smaller one in the area of the insertion of the falx anteriorly. She was referred to this institution for treatment.

On physical examination, the patient was alert and cooperative. Blood pressure was 120/70 mm.Hg. and cardiac auscultation was normal. No bruits were heard on the supraclavicular fossa, neck, eyes or head. Optic fundi were normal, without papilledema or vascular abnormality; visual acuity was 20/30 on the right and 20/25 on the left. The pupils were in mid-position and reacted normally. The iris and conjunctiva were normal. The ocular movements were full and the remainder of cranial nerves examination was normal, as was the rest of the neurological examination.

ECG, chest x-ray, and standard blood tests were normal. On July 18, bilateral internal and external carotid angiograms and a left vertebral angiogram were performed. A large AVM was demonstrated in the left middle fossa (fig. 1 and 2), receiving its blood supply from the left middle meningeal artery, branches of the internal maxillary artery and the recurrent meningeal branch of the left ophthalmic artery. The venous drainage was by way of enlarged cortical veins, which emptied into the basal vein of Rosenthal. A much smaller AVM was demonstrated on the right side, near the insertion of the falx anteriorly, mainly supplied by ethmoidal branches of the right ophthalmic artery. The venous drainage was by way of enlarged cortical veins, which emptied into the basal vein of Rosenthal. A much smaller AVM was demonstrated on the right side, near the insertion of the falx anteriorly, mainly supplied by ethmoidal branches of the right ophthalmic artery. On July 19, an ICBA embolization of the left middle fossa dural AVM was performed. After embolization, no change in the physical examination was observed, and the optic fundi remained normal. A post-embolization internal maxillary angiogram showed only small residual feeders from this artery, with very slow filling of the venous outlet of the malformation. A left internal carotid angiogram showed that the most medial part of the malformation nidus was still supplied by the left ophthalmic artery, with drainage into cortical veins. Control internal and external carotid angiograms performed on July 23 showed a slower flow in the residual arteriovenous malformation in the region of the left
FIGURE 1. *Left external carotid artery angiogram. A large dural AVM is present in the left middle fossa. It is mainly fed by the middle meningeal artery (black arrow), and also by the internal maxillary artery (open arrow) with cortical venous drainage.*

FIGURE 2. *Left internal carotid artery angiogram. The anterior part of the dural AVM, in the left anterior fossa, is fed by the recurrent meningeal branch of the left ophthalmic artery.*

FIGURE 1. Left external carotid artery angiogram. A large dural AVM is present in the left middle fossa. It is mainly fed by the middle meningeal artery (black arrow), and also by the internal maxillary artery (open arrow) with cortical venous drainage.

FIGURE 2. Left internal carotid artery angiogram. The anterior part of the dural AVM, in the left anterior fossa, is fed by the recurrent meningeal branch of the left ophthalmic artery.

superior orbital fissure, the main supply still being from the recurrent meningeal branch of the ophthalmic artery. During the following two months, the patient experienced two more episodes of TMB; further therapy was declined.

Discussion

A large dural AVM was discovered in the left anterior-middle fossa in a patient investigated for recurrent episodes of isolated left transient monocular blindness (TMB). She had experienced a transient frontal bruit four years before, but the episodes of transient monocular blindness were the major presenting neurological manifestation.

Patients with amaurosis fugax investigated by angiography were recently reviewed by Adams et al., who did not find any example of AVM among 59 patients from a personal series and a total of 751 from the literature, suggesting that TMB is very rarely associated with AVM's. Although Walsh and Hoyt mention this association, we were able to find only one documented case of TMB in the literature, related to an intracerebral AVM. However, in this case, the episodes of TMB were rather atypical, lasting only a few seconds, and the patient had papilledema. In our case, the episodes of TMB had the usual duration of typical amaurosis fugax and the optic fundi were normal. Two types of TMB were present: firstly, loss of the whole monocular visual field starting from the periphery; secondly, vertical progression of an altitudinal visual field loss.

In dural AVM’s, the occurrence of visual loss, usually with associated papilledema, has been reported, most often with posterior fossa dural AVM’s draining into the transverse-sigmoid sinus, but also in anterior fossa dural AVM’s. However, these cases of visual loss were permanent and never corresponded to TMB. Other ophthalmic symptoms as proptosis, red eye, elevated intraocular pressure, cranial nerve palsy, visual field defects, ptosis and bruit have been reported in dural AVM’s, but with no mention of TMB. Dural AVM’s with pial venous drainage are much more uncommon than those with sinus drainage, and their main neurological manifestations include epilepsy (50%), subarachnoid hemorrhage (42%), permanent neurological deficits without hemorrhage (50%) and dementia (25%). Transient ischemic attacks occur in as much as 17% and are more frequent than in dural AVM’s with direct sinus drainage (3%), but they have always involved the hemispheres and no case of TMB has been reported previously.

Typically, amaurosis fugax is related to microemboli of atherosclerotic material from a stenosed ipsilateral internal carotid artery. When this artery becomes occluded, similar episodes may occur due to migration of the microemboli through the reversed ophthalmic artery flow via the external carotid artery.
collaterals. Cases with presumed hemodynamic amaurosis fugax have less often been reported. Severe occlusive disease of the extracranial arteries or increased viscosity from polycythemia are the usual causes of these episodes of TMB, which are often triggered by orthostatic changes in blood pressure. In AVM’s, transient or permanent neurological deficits may occur on a hemodynamic basis, from a critical lowering of cerebral blood flow in discrete brain areas, a phenomenon which has been called intracranial “steal.” In the case of left TMB reported by Kosary et al., a brain AVM was fed by the right pericallosal artery receiving its supply from the left internal carotid artery. The authors hypothesized that blood was diverted from the left ophthalmic artery towards the AVM, producing the episodes of TMB. However, without direct involvement of the ophthalmic artery in the arterial feeding of the AVM, it is difficult to conceive of a significant selective lowering of ophthalmic and retinal arterial pressure that would produce the symptoms. In that case, funduscopy showed papilledema, a common cause of atypical short episodes of visual loss, and TMB in that patient was more probably related to papilledema than to a hypothetical remote “steal”. On the other hand, in our patient, the ophthalmic artery directly participated in the AVM supply, as the anterior part of the larger AVM was fed by the recurrent meningeal branch of the left ophthalmic artery, although the larger posterior part of the AVM was supplied by the branches of the left external carotid artery. This type of blood supply, with shunting of blood from the ophthalmic artery towards the AVM, may have produced episodic lowering of the ophthalmic artery/retinal arterial blood pressure (steal phenomenon), at the origin of the episodes of ipsilateral TMB on changing position. The recurrence of TMB after the embolization procedure may be explained by the persistence of residual flow in the malformation, with supply from the recurrent meningeval branch of the ophthalmic artery on control angiograms. Increased venous pressure may also contribute to flow problems in dural AVM’s, and the angiographic finding of markedly dilated veins is consistent with venous hypertension. However, in the absence of enlarged or abnormal retinal veins on funduscopy, it is difficult to ascribe the episodes of TMB to a venous flow disturbance.

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