Spontaneous regression of intracranial arteriovenous malformations

Report of three cases

MATTHEW F. OMOJOLA, M.D., ALLAN J. FOX, M.D., FERNANDO V. VIÑUELA, M.D., AND CHARLES G. DRAKE, M.D.

Departments of Diagnostic Radiology and Clinical Neuroscience, University of Western Ontario and University Hospital, London, Ontario, Canada

This is a report of spontaneous regression of intracranial arteriovenous malformations (AVM's) in three female patients; two of these patients had complete angiographic disappearance of the AVM, including an instance of intimate association of the AVM with an astrocytoma. The AVM's in these two patients were unicompartmental medium- to large-sized lesions supplied by a single feeder and draining principally through one large vein; spontaneous thrombosis is suggested as a cause of the AVM regression. Partial regression in the third patient might have been partially due to embolism from a clot-filled aneurysm on the feeding vessel. The significance of such disappearance of AVM's in relation to persistence or otherwise of the neurological status of these patients is discussed.

KEY WORDS
- intracranial arteriovenous malformation
- natural history
- spontaneous thrombosis
- embolism
- brain tumor
- astrocytoma

One of the less well understood aspects of the natural history of intracranial arteriovenous malformations (AVM's) is their potential for spontaneous regression, mostly due to the rarity of this event. The few reports of such instances in the literature are mostly of single cases.2,5,6,8,9,11,14,15,17 It is even rarer to encounter an intracranial AVM coexisting with a glioma,3,4,7,18 and the disappearance of an AVM in such combination has not to our knowledge been reported previously.

The purpose of this communication is to document three instances of spontaneous regression in the size of intracranial AVM's, two of which completely disappeared, as seen on angiography; these include a case of intimate coexistence of the AVM and an astrocytoma. We believe this case to be the first of its type. We also speculate on factors responsible for the spontaneous occlusion of intracranial AVM's.

Case Reports

Case 1

This 35-year-old right-handed woman suffered intracranial hemorrhage causing coma and left hemiplegia in 1976. Cerebral angiography revealed an AVM in the medial aspect of the right parietal lobe (Fig. 1 left). She was not treated surgically and gradually improved on a comprehensive rehabilitation program. Function in her left hand and arm recovered completely. Her left leg improved for several years to the point where she could walk with a cane. When she was seen in 1981, she complained that her leg had become worse, with increasing weakness of the left foot during the previous year. She described an episodic involuntary movement of the left leg, thought to be focal seizures, and these were controlled by Dilantin (phenytoin sodium). She also complained of daily headaches.

Examination was normal except for the left lower extremity which showed moderate paresis, hyperreflexia, and ankle clonus. Left plantar reflex was equivocally extensor. There was diminished sensation of pinprick and light touch over the entire left lower extremity.

Cerebral angiography did not show the previously known AVM (Fig. 1 right), but a computerized tomography (CT) scan showed a small residual low-
Spontaneous regression of AVM's

Fic. 1. Right carotid angiograms, lateral views.  Left: Initial appearance showing an arteriovenous malformation (AVM) supplied by a distal branch of the callosomarginal artery. Drainage is through a large vein into the superior sagittal sinus.  Right: Angiogram 5 years later showing no trace of the AVM. The left carotid and left vertebral angiograms were normal.

density lesion in the same area. The AVM was presumed to have spontaneously thrombosed.

Case 2

This 15-year-old right-handed girl presented in 1971 when she first became aware of some decreased power and clumsiness of her right hand. For the previous year she had had some difficulty in seeing objects on the right. She also complained of non-localized occasional severe headaches.

Examination revealed a slight right lower facial weakness and a right homonymous hemianopsia. Cerebral angiography demonstrated a left temporal AVM (Fig. 2 upper). Because the AVM was deep in the patient’s dominant hemisphere, she was treated conservatively. She subsequently regained a great deal of strength in her right hand and arm.

She returned in 1974 with a 1-week history of a feeling of paresthesia and excessive sensitivity in her right foot and hand, without any change in power or coordination. Examination showed no change in the right homonymous hemianopsia. There was increased tone in the right extremities, and slightly decreased power. Tendon reflexes were hyperactive on the right, with a flexor plantar response. There was hypalgesia to pinprick over the right foot to just above the ankle. No sensory changes were obvious in the hand.

Cerebral angiography did not show the previously demonstrated AVM (Fig. 2 lower). There was an obvious mass effect in the left temporal area near where the AVM had been shown previously. A CT scan showed a large low-density structure deep in the left hemisphere, with an enhancing mural nodule. This lesion was subsequently shown by surgical exploration and biopsy to be a low-grade cystic astrocytoma. The cyst was shunted into the ventricle as a palliative measure. When the patient was last seen in 1981, she was being maintained on Dilantin. Her neurological status otherwise remained unchanged and she was doing well in college. The CT scan (Fig. 3) had remained essentially unchanged since 1974.

Case 3

In 1979, this 49-year-old right-handed woman was discovered by her optometrist to have a left visual field defect, shown as an incongruous left homonymous quadrantanopia. She had suffered intermittent headaches for at least 10 years. The headaches would begin with flashing lights in the left visual field, followed by blurred vision. This disappeared after 10 minutes and left a moderately severe steady bifrontal headache which was relieved by aspirin. Both her husband and her son have a history of classical migraine.

Examination confirmed the left homonymous quadrantanopia. No other abnormality was found. Skull x-ray films showed some calcification in the right parieto-occipital area. ACT scan revealed a large contrast-enhancing mass lesion surrounded by a zone of low-density in the right occipital region. On cerebral angiography, a right occipital AVM with an aneurysm of the feeding artery was visualized (Fig. 4 upper). It was elected to treat the patient conservatively. She was prescribed high doses of dexamethasone for its possible effect on the low-density area surrounding the lesion (presumed to be edema), and discharged.

In 1980 (17 months later), there was no significant change in her neurological status, and the ophthalmological assessment remained the same. Repeat vertebral angiography at that time (Fig. 4 center) showed a decrease in the caliber and segmental stenosis of the right posterior cerebral artery and its branches, which fed the lesion. The flow was considerably slowed and the core of the AVM filled late; it was preserved but smaller. The big draining vein seen previously was no
longer apparent. The aneurysm at the origin of the right posterior cerebral artery was smaller and more irregular in shape than before.

Computerized tomography showed an appearance similar to that of the initial scan (Fig. 4 lower), although the part enhanced by contrast material was slightly smaller. It was again elected to treat the patient conservatively.

Discussion

The common findings in our three cases were that all the patients were female; all the lesions were medium to large in size (using criteria followed by Parkinson and Bachers12); and each was supplied by a single feeder and drained principally through a single vein. Spontaneous regression has been ascribed to intracranial hemorrhage or hematoma,8,10,11,13,16,17 and spontaneous thrombosis.5,15 The patient in our Case 1 did not experience intracranial hemorrhage between the two angiographic studies, but she did suffer a coma-producing hemorrhage before the initial angiogram. The part played by the initial hemorrhage in the subsequent thrombosis is not clear. Her initial improvement in neurological status, followed years later by increasing weakness of the left foot and focal seizures affecting the same extremity, suggests a separate event. This presumably was not caused by hemorrhage but was ischemic, perhaps due to spontaneous thrombosis extending into the surrounding normal vessels. There is nothing in the history of the patients in Cases 2 and 3 to suggest intracranial hemorrhage at any time.

The patient in Case 2 probably also had spontaneous thrombosis of her AVM. It is interesting to note that the coexisting tumor was overlooked on the initial angiogram, but became evident on the repeat study.
Spontaneous regression of AVM's

Welcker and Seidel\textsuperscript{19} reported a case in which an astrocytoma was found within an AVM, but the tumor had not been suspected clinically or radiologically and was only discovered at autopsy. There was, however, no thrombosis of the AVM. Crowell, \textit{et al.},\textsuperscript{3} reported a patient with a cystic tumor in the right temporal lobe which communicated with the ventricular system as demonstrated by ventriculography. No angiography was performed, but at surgery and histological examination it was discovered that the tumor had existed within the AVM. In another case reported by Heffner, \textit{et al.},\textsuperscript{7} a right frontal mass with large tortuous vessels in the subfrontal region was demonstrated by angiography, with early venous opacification by contrast material. This had been thought to represent a glioma which may have undergone malignant degeneration. Surgery and histological examination, however, confirmed this lesion to be a combination of a benign astrocytoma and an AVM that was confined to the nearby meninges. The histology in our present case did not show the thrombosed AVM, presumably because the biopsy was taken some centimeters away from the area where the AVM existed.

Gliomas are well known to be richly vascular neoplasms, but their vascularity is capillary and does not involve large tortuous vessels.\textsuperscript{3,7} We are convinced from the angiographic pictures that the tortuous vessels demonstrated in our Case 2 represented the AVM and not tumor vascularity. On angiography, the tumor was subsequently shown to be avascular, not hyper-vascular.

Our third patient showed a partial regression and obvious hemodynamic alteration in the flow pattern of the AVM. The apparent decrease in the size of the aneurysm at the origin of the feeding posterior cerebral artery is attributed to clot formation within the aneurysm. The multiple stenoses of the feeding vessels...
could possibly have been caused by remnants of re-
canalized emboli from the clot within the aneurysm. 
However, large irregular nodules of hyalinized intima 
and smooth muscle have been demonstrated histologi-
cally to project into the lumen of vessels in AVM's.1 
Unfortunately, there has been no histological verifi-
cation of those stenoses to indicate what stenotic 
angiopathy is present in the feeding vessels of AVM's. 
This could even be an accelerated process of athero-
sclerosis stimulated by the excess flows.

Causes of spontaneous regression in intracranial 
AVM's remain poorly understood, but as more of 
these cases are documented, common denominators 
may surface. Destruction and subsequent disappear-
ance of AVM's by intracerebral hematoma seems to 
be the cause most often encountered and most readily 
understood. However, in two of our patients, throm-
bosis was probably responsible for the spontaneous 
disappearance, while clot embolism from the aneu-
rystm is suggested as a possible additional cause for 
the partial regression in the third patient.

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Present address for Dr. Omojola: Department of Radiol-
yogy, Institute of Health Sciences, University of Port Har-
court, P.M.B. 5323, Port Harcourt Rivers State, Nigeria.
Address reprint requests to: Allan J. Fox, M.D., Depart-
ment of Diagnostic Radiology, University Hospital, 339 
Windermere Road, London, Ontario, Canada, N6A 5A5.