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Hemorrhage from moyamoya-like vessels associated with a cerebral arteriovenous malformation

Case report

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✓ The authors describe a case of subarachnoid hemorrhage from moyamoya-like vessels associated with an arteriovenous malformation (AVM) in a 44-year-old Hispanic man who presented with severe headache. The AVM was located in the left parietal lobe and the ipsilateral middle cerebral artery was occluded. Although the patient was initially neurologically intact, he began to experience neurological deficits from mild vasospasm, illustrating the sensitivity of the underperfused portion of brain surrounding an AVM. His neurological deficits improved with aggressive hydration and elevated blood pressure. After a 3-week period, the AVM was resected without complication and all of the patient's neurological deficits resolved. The authors review radiographic findings of this unique case.

Key Words • moyamoya disease • arteriovenous malformation • subarachnoid hemorrhage

Cerebral AVMs are known to present with cerebral hemorrhage including intraparenchymal, subarachnoid, and intraventricular hemorrhage.[1] Moyamoya vessels, most commonly seen in patients with moyamoya disease, are also known to result in cerebral hemorrhage. The authors present an unusual case of SAH from moyamoya-like vessels that were associated with an occluded MCA and an AVM.

Case Report

History. This 44-year-old Hispanic man presented to a local hospital with a 3-day history of severe headache. His initial examination included a CT scan of the head, which depicted a Fisher Grade 2 SAH centered in the left sylvian fissure (Fig. 1). He was markedly hypertensive with a systolic blood pressure greater than 200 mm Hg. He was given intravenous antihypertension therapy and transferred to our facility for further examination.

Examination. On arrival at our facility 3 days after the onset of symptoms, the patient continued to have headache and nausea. No neurological deficits were noted during the physical examination. The patient's vital signs stabilized, but he required nitroprusside for blood pressure control. Cerebral angiograms revealed a Spetzler–Martin Grade 3 AVM in the left parietal region with a very unusual filling pattern (Fig. 2). The M_1 segment of the left MCA was occluded with distal reconstitution of the MCA branches through pial collateral vessels from the left ACA. The superior portion of the AVM was directly supplied from the left ACA. The left MCA branches secondarily filled the lower bulk of the AVM in a delayed fashion by flow through the ACA collateral vessels. Superficial venous drainage was noted and no outlet stenosis or aneurysm was noted.

Moyamoya-like vessels were also noted in the parenchyma of the region normally supplied by the occluded M_1 segment and the proximal M_2 branches. Given the pattern of hemorrhage, which was an SAH with no evidence of intraparenchymal hemorrhage associated with the AVM, we suspected that the moyamoya-like vessels, and not the actual AVM, had been the source of the bleeding.

Preoperative Course. The patient was given hydration therapy and monitored closely in the intensive care unit. Six days after his initial hemorrhage he experienced a right hemiparesis and was unable to move his right arm on examination. He also displayed a moderate expressive aphasia. Another CT scan of the head was obtained; it demonstrated some hypodensity surrounding the area of the AVM. A cerebral angiogram demonstrated moderate vasospasm in the ACA vessels. We concluded that the patient's deficits were likely due to the combination of moderate vasospasm and tenuous blood supply around the AVM, which was caused by a vascular steal phenomenon, and occlusion of the M_1

Abbreviations used in this paper: ACA = anterior cerebral artery; AVM = arteriovenous malformation; CT = computerized tomography; MCA = middle cerebral artery; SAH = subarachnoid hemorrhage.

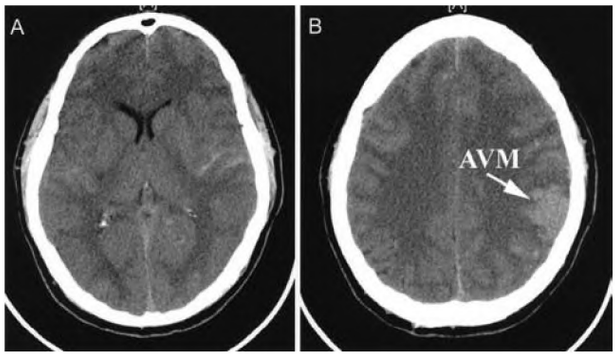

FIG. 1. Initial CT scans of the head demonstrating a hyperdense area in the left parietal region, which represents the AVM (A) and an SAH in the left sylvian fissure (B). The area of SAH is distinctly separate from the AVM.

segment. The patient was treated by cessation of blood pressure control and increased intravenous fluids to maximize cerebral perfusion. Antihypertensive medications were stopped and the patient's systolic blood pressure was maintained in a range of 160 to 180 mm Hg without vasopressors. His aphasia gradually resolved and high right upper-extremity weakness improved, but the fine motor control in his right hand remained diminished.

Surgery and Postoperative Course. After a 3-week period (beyond the time course for SAH-associated vasospasm), resection of the left parietal AVM was performed without complication. At surgery, some gliosis surrounding the AVM was noted, but there was no evidence of hemorrhage in the region of the AVM. The patient's postoperative course was uncomplicated, and he experienced improvement in neurological function with better fine motor control in his right hand. He was discharged home on postoperative Day 5 with no neurological deficits. A postoperative angiogram confirmed complete resection of the AVM.

Discussion

This case illustrates the potential for formation of moyamoya-like vessels in ischemic brain parenchyma and the potential for hemorrhage from these small friable vessels. The likely inciting event for the development of the moyamoya-like vessel pattern in this case was the combination of an occluded left M_1 segment and the vascular steal phenomenon associated with the AVM. Although a Medline lit-

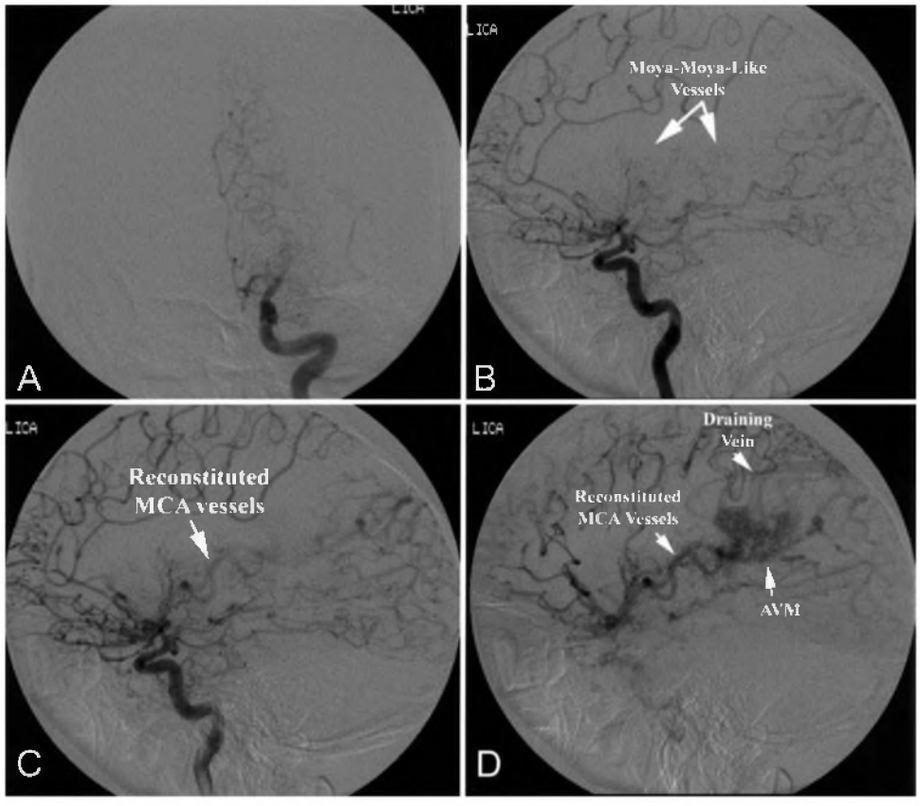

FIG. 2. Cerebral angiograms. A: Early left internal carotid artery (ICA) injection (anteroposterior view) demonstrating occlusion of the left M_1 segment. B: Early left ICA injection (lateral view) revealing moyamoya-like vessels and the occluded left M_1 segment. C: Middle arterial phase of the left ICA injection (lateral view) depicting the reconstitution of the MCA branches from the ACA collateral vessels. D: Late arterial phase of the left ICA injection (lateral view) demonstrating the left parietal AVM with a prominent cortical draining vein. Note that this particular AVM has delayed filling from left MCA vessels that have been reconstituted from the ACA collateral vessels.

erature review found no previously reported association of moyamoya-like vessels with an AVM, it seems reasonable that such an association may exist, given that the poorly perfused parenchyma near the M_1 occlusion, exacerbated by the sump effect of a high-flow AVM, provided the ischemic environment needed for moyamoya-like vessel formation.

This case also demonstrates the inadequate perfusion of brain tissue adjacent to an AVM with a low threshold for neurological deficits in response to additional ischemia. Neurological deficits have been reported to occur in approximately 7% of patients with cerebral AVMs because of the vascular steal phenomenon.[2] In this particular case, we speculate that the parenchyma surrounding the AVM was likely hypoperfused before the hemorrhagic event. With just moderate changes in local perfusion, caused by mild vasospasm from the SAH, neurological deficits developed. On removal of the AVM, the patient's neurological deficits improved. We speculate that the patient's improved neurological status was a result of increased blood flow to the previously ischemic parenchyma after elimination of the vascular steal phenomenon associated with the AVM.

Conclusions

This case demonstrates an association betweene MCA occlusion and moyamoya-like vessels associated with an AVM. These small friable vessels have the potential for hemorrhage. To our knowledge, this association has not been previously reported.

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