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External Carotid Artery Aneurysm Developing After Embolization of a Ruptured Posterior Inferior Cerebellar Artery Aneurysm in a Patient With Cervicocephalic Fibromuscular Dysplasia

—Case Report—

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Abstract

A 30-year-old man presented with an aneurysm of the left posterior inferior cerebellar artery manifesting as subarachnoid hemorrhage and cerebellar infarction. Angiography demonstrated string-of-beads sign typical of fibromuscular dysplasia (FMD) in the extracranial carotid and vertebral arteries. The aneurysm and the parent artery were successfully embolized with Guglielmi detachable coils. Severe vasospasm developed 1 week after admission, and was treated several times by selective injection of vasodilator. A new aneurysm of the left external carotid artery became evident 1 month later, whereas only slight dilation had previously been apparent. This angiographic sequence demonstrated a new arterial dissection. Despite the possibility of damage to the artery during multiple catheterizations, arterial wall changes caused by FMD appear to have been primarily responsible. This case emphasizes the need for particular care in performing vascular interventional procedures in the presence of FMD.

Key words: dissecting aneurysm, posterior inferior cerebellar artery, external carotid artery, Guglielmi detachable coil, fibromuscular dysplasia

Introduction

Fibromuscular dysplasia (FMD) is a multifocal vascular disease of unknown etiology. Cervicocephalic FMD most frequently involves the extracranial internal carotid artery (ICA) at the level of the second cervical vertebra, and occasionally affects the intracranial portion. Intracranial aneurysms occur in 25% to 50% of patients with cervicocephalic FMD, suggesting that FMD is associated with predisposition to aneurysm formation.[2,5] These patients have a high incidence of multiple aneurysms and a poor prognosis related to spasm after subarachnoid hemorrhage (SAH).[2] Angiographic findings indicating FMD have been found in 15% of patients with spontaneous craniocervical artery dissection, implicating FMD as a risk factor for arterial dissection.[6] Dissecting aneurysms of the intracranial posterior circulation are relatively uncommon, and dissecting aneurysms involving the posterior inferior cerebellar artery (PICA) but not the vertebral artery (VA) are extremely rare.[11]

We treated a 30-year-old man with cervicocephalic FMD who presented with SAH following rupture of a PICA aneurysm by intravascular embolization with Guglielmi detachable coils (GDCs). Vasospasm following the SAH required multiple intravascular interventions. One month later, a newly formed external carotid artery (ECA) aneurysm was detected and was treated with GDCs.

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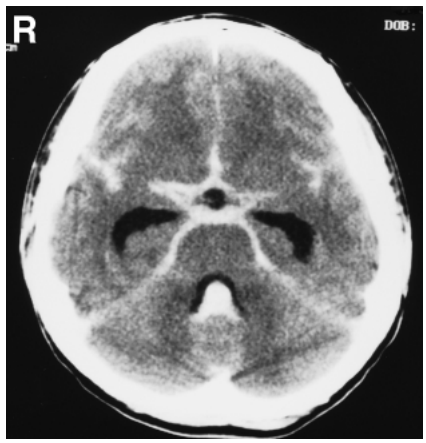

Fig. 1 Computed tomography scan on admission showing subarachnoid hemorrhage.

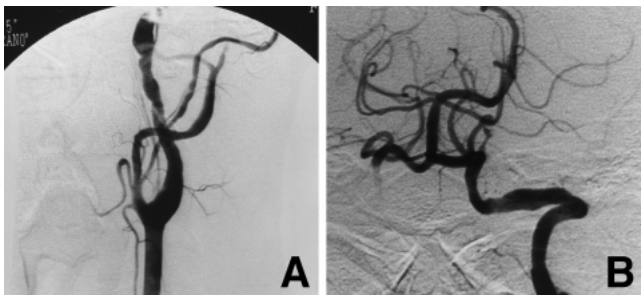

Fig. 2 Initial digital subtraction angiograms of (A) the left carotid artery, oblique view, showing string-of-beads sign in the left external carotid artery, and (B) the left vertebral artery, anteroposterior view, showing no aneurysm of the posterior inferior cerebellar artery.

Case Report

A 30-year-old man presented with sudden severe headache followed by loss of consciousness on September 22. Initial neurological examination found his level of consciousness was mildly decreased. Computed tomography (CT) demonstrated blood in the fourth ventricle suggesting an aneurysm involving the PICA (Fig. 1) and hydrocephalus. After admission the patient gradually became lethargic. Digital subtraction angiography showed stenosis of the proximal portion of the left PICA (Fig. 2A). Other angiographic findings included marked irregularity of the left ICA, ECA, and extracranial left VA (Fig. 2B). Ventricular drainage was performed to decrease cerebrospinal fluid pressure, and his level of consciousness gradually improved.

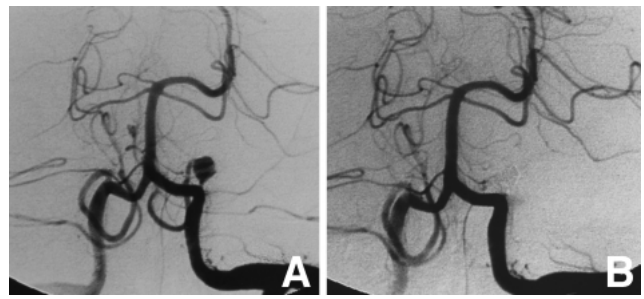

Fig. 3 Left vertebral angiograms, anteroposterior view, (A) 3 days after admission showing a fusiform aneurysm of the left posterior inferior cerebellar artery, and (B) successful embolization.

On the next day, CT demonstrated an area of low density in the cerebellar territory of the left PICA in the absence of a corresponding clinical neurological deficit. On hospital day 3, conventional angiography displayed a fusiform aneurysm of the PICA (Fig. 3). The diagnosis was SAH from a dissecting aneurysm of the PICA that had most likely recently enlarged. The aneurysm and PICA were successfully embolized with GDCs on hospital day 4. Tolerance to test occlusion was not assessed given his poor condition, but the immediate postintervention course was uneventful with no ischemic neurological deficits.

On hospital day 5, the patient became drowsy because of severe arterial spasm in the vertebrobasilar circulation. Fasudil hydrochloride (30 mg) was injected selectively into the left VA, achieving dilation of the left VA and the basilar artery with angiographically evident improvement of blood flow. Two more injections were given over the next 2 days, and his level of consciousness improved.

A mass measuring approximately 3×3 cm became evident below the left side of the jaw on October 20. The mass was rubbery, firm, and pulsatile. Selective left external carotid angiography demonstrated an aneurysm of the left ECA, arising where only a slight dilation had been demonstrated previously (Fig. 4). This sequence suggested a new arterial dissection. The aneurysm and the ECA were embolized with GDCs. The postoperative course was favorable. Blood test showed no evidence of coagulation disorder, α-1 antitrypsin deficiency, or collagen disease. No recurrence has developed over 1 year of postoperative follow up.

Discussion

The diagnosis of FMD is based on angiographic

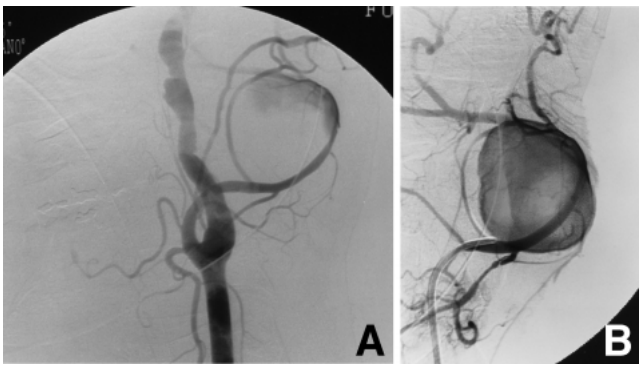

Fig. 4 A: Left carotid angiogram, oblique view, after 1 month showing an external carotid artery (ECA) aneurysm arising where only a slight dilation had been demonstrated previously. B: Selective left external carotid angiogram showing an aneurysm of the left ECA.

and/or histological findings. The characteristic angiographic string-of-beads sign is easily distinguished from signs of other vascular diseases.[10] Disruption of the mural layers of the involved vessel may become evident as areas of marked thinning and aneurysmal dilation. Our case demonstrated angiographic irregular segmental alternation of dilation and stenosis in the extracranial left ICA, ECA, and VA, indicating FMD with the typical string-of-beads appearance.

Dissecting aneurysms of the intracranial posterior circulation are generally treated by proximal clipping together with occipital artery-PICA anastomosis, considering the risk of injury to the brainstem. Fourteen of 16 well-characterized patients were treated surgically.[13] The alternative intravascular interventional treatment, PICA embolization after test occlusion of the PICA, is also well tolerated. However, only two cases including the present one have described treatment of isolated dissecting PICA aneurysm with GDC.[11]

Rebleeding occurs in 24% to 30% of patients with SAH caused by rupture of a dissecting aneurysm including the intracranial VA, resulting in a high mortality.[3] In our patient, the first conventional angiogram, obtained 3 days after SAH, suggested considerable enlargement of a fusiform aneurysm not seen in previous digital subtraction views. These observations suggested a dissecting aneurysm with a propensity to rebleed. CT demonstrated cerebellar infarction affecting the PICA territory, and angiography on hospital day 3 demonstrated filling of the proximal PICA, raising the concern that PICA embolization could cause infarction of the brainstem. However, test occlusion prior to embolization was deferred because of the patient's condition.

Interestingly, our patient developed a new dissecting aneurysm in the ECA after intravascular intervention to treat the PICA aneurysm. Documented de novo aneurysm formation associated with FMD is rare, but the possible mechanism has given rise to considerable speculation.[4,7,8] Hemodynamic stress might contribute to de novo formation of an aneurysm, in combination with the intrinsic weakness of the arterial wall induced by FMD.[4] However, ECA aneurysms are considerably less common than intracranial aneurysms, which most often occur as a traumatic pseudoaneurysm of the anatomically vulnerable superior temporal artery.[9] In our case, the ECA aneurysm might have resulted from iatrogenic injury in the course of multiple interventional procedures for treatment of severe vasospasm.

Iatrogenic dissection is an uncommon complication of cerebral angiography.[12] The prevalence of dissection was only 0.3% in diagnostic cerebral angiography, and 0.7% in interventional neuroradiology procedures.[1] The slightly elevated risk of iatrogenic dissection associated with interventional procedures may reflect the need for more vascular manipulation and more distal advancement of catheters than during diagnostic angiography. Nonetheless, pseudoaneurysms in series of patients with dissection as a complication of angiography are unusual, whereas angiographic narrowing of the true arterial lumen is common.[1]

Vasospasm is particularly likely to follow SAH in patients with FMD, suggesting many potential indications for the interventional treatment of vasospasm. In our case, angiography including selective intraarterial injection of vasodilator was performed several times. The ECA site where the new aneurysm later developed had appeared as a slight dilation on previous angiograms, supporting the diagnosis of dissecting aneurysm. Arterial wall fragility resulting from FMD most likely caused predisposition to aneurysm formation, although arterial wall damage from interventional treatments may also have been a contributing factor. Accordingly, particular care is needed when performing intravascular interventions in the presence of FMD.

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