Occlusion of Internal Carotid Artery and Formation of Anterior Communicating Artery Aneurysm in Cervicocephalic Fibromuscular Dysplasia

—Follow-up Case Report—

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Abstract

A 44-year-old female with cervicocephalic fibromuscular dysplasia (FMD) suffered a subarachnoid hemorrhage 10 years after presenting with left impaired extraocular movement. Carotid and vertebral angiography had shown string-of-beads sign in the left external carotid artery and fusiform aneurysms in the left carotid cavernous portion and in the left vertebral artery. On admission, the left carotid angiogram showed complete occlusion of the left internal carotid artery and the right carotid angiogram showed a new anterior communicating artery aneurysm. The territory of the left middle cerebral artery was supplied from the right internal carotid artery *via* the anterior communicating artery and from the basilar artery *via* the posterior communicating artery. Occlusion of the main arteries is rare in the clinical course of FMD. We suggest that hemodynamic stress due to occlusion of the internal carotid artery contributed to formation of the anterior communicating artery aneurysm, possibly associated with intrinsic changes of the arterial wall induced by FMD.

Key words: fibromuscular dysplasia, aneurysm formation, internal carotid artery occlusion, subarachnoid hemorrhage

Introduction

Cervicocephalic fibromuscular dysplasia (FMD) is more common in Western countries where 300 cases have occurred,¹²⁾ compared to about 20 in Japan.¹⁰⁾ This epidemiological disparity, and its predominance among middle-aged females, are well known. Cervicocephalic FMD is accompanied by an intracranial aneurysm in 21–51.4% of cases, suggesting that FMD may cause predisposition to the formation of intracranial aneurysm.^{7,12,13,18,19)} George *et al.*⁶⁾ found that cervicocephalic FMD carries a poor prognosis due to the high rate of associated vasospasm following rupture of aneurysms.

We describe a 44-year-old Japanese female, who had been diagnosed as having cervicocephalic FMD and intracranial fusiform aneurysms 10 years ear-

lier,¹⁰⁾ with subarachnoid hemorrhage (SAH) due to rupture of a newly formed anterior communicating artery aneurysm and complete occlusion of the left internal carotid artery.

Case Report

This 44-year-old female had previously developed impairment of left extraocular movement and double vision in January, 1983. Angiography then revealed string-of-beads sign in the left external carotid artery, a giant fusiform aneurysm in the left carotid cavernous portion, and another fusiform aneurysm in the left vertebral artery. The left internal carotid artery was patent, and no aneurysm was present in the anterior communicating artery (Fig. 1). She was treated conservatively.

On July 27, 1992, she experienced a sudden severe headache, and was admitted to our hospital. Neurological examination found she was alert

Fig. 1 *left*: Left carotid angiogram, lateral view, at first examination showing string-of-beads sign in the left external carotid artery and a giant fusiform aneurysm in the left cavernous sinus. The left internal carotid artery is patent. *right*: Right carotid angiogram, anteroposterior view, at first examination showing no aneurysm in the anterior communicating artery.

Fig. 2 CT scan on admission showing SAH.

without paresis, but her headache and neck stiffness were severe (Hunt and Kosnik grade 2). Left abducens nerve paresis with the absence of light reflex, resulting from the giant fusiform aneurysm in the left cavernous sinus, were also present. These symptoms had persisted for the last 10 years. Com-

puted tomography (CT) on this admission showed SAH, Fisher group 2 (Fig. 2). A left carotid angiogram revealed string-of-beads sign in the left external carotid artery and complete occlusion of the left internal carotid artery at a point near the bifurcation of the common carotid artery (Fig. 3 *left*). A right carotid angiogram showed a saccular aneurysm arising from the anterior communicating artery which had not been present 10 years earlier (Fig. 3 *center*). This aneurysm was thought to be the origin of the SAH. A left vertebral angiogram revealed the fusiform aneurysm in the left vertebral artery which had been noted 10 years earlier (Fig. 3

Fig. 3 *left*: Left carotid angiogram, lateral view, showing string-of-beads sign in the external carotid artery and complete occlusion of the internal carotid artery at a point near the bifurcation of the common carotid artery. *center*: Right carotid angiogram, right oblique anteroposterior view, showing an anterior communicating artery aneurysm (arrow). Blood is supplied to the contralateral side *via* the anterior communicating artery. *right*: Left vertebral angiogram, anteroposterior view, showing a fusiform aneurysm in the left vertebral artery and blood supply to the left middle cerebral artery region *via* the posterior communicating artery.

right). The size of the aneurysm had not changed. The territory of the left middle cerebral artery was supplied *via* the anterior communicating artery from the right internal carotid artery and *via* the posterior communicating artery from the basilar artery.

Two days after admission, the neck of the anterior communicating artery aneurysm was clipped using a No. 3 Sugita clip through a right trans-Sylvian approach. The surface of the A₁ portions of the bilateral anterior cerebral arteries and the dome of aneurysm demonstrated no macroscopic abnormalities. However, the intracranial portion of the left internal carotid artery had become hard and white.

Postoperatively, she developed meningitis, which responded well to the intravenous administration of antibiotics. Otherwise, her postoperative course was uneventful, and she was discharged without new neurological deficits on August 31. Follow-up single photon emission computed tomography showed no hypoperfusion in any area of the brain.

Discussion

Cervicocephalic FMD cases rarely demonstrate changes in the main arteries, with only four such cases reported.^{1,5,14,24} Only one patient presented with *de novo* formation of an aneurysm in the pericallosal artery based on angiographic evidence obtained at an interval of 9 years.¹ Our patient had developed two new lesions in the 10 years since the first admission, complete occlusion of the left internal carotid artery and a new saccular aneurysm of the anterior communicating artery.

The pathogenesis of intracranial saccular aneurysms involves two particularly important factors. The first is the presence of congenital developmental anomalies of the arterial wall.^{2,16} Intrinsic abnormalities in the wall of cerebral arteries may favor the formation and rupture of cerebral aneurysms.³ Congenital factor(s) may be more important in aneurysms arising in childhood, because these present different clinical aspects to aneurysms in adults.¹⁵ Our patient demonstrated macroscopic disorders of the arterial wall due to the underlying FMD, although in another location to the aneurysms. The second factor is the progressive alteration of the cerebral arteries due to the mechanical action of blood flow or to arteriosclerosis due to acquired factors.¹⁷ In moyamoya disease, cerebral aneurysms frequently occur in the vertebrobasilar system.²³ Saccular aneurysms are sometimes seen in the main feeders of arteriovenous

malformations.²¹ Matsuda *et al.*¹¹ reported seven cases of ruptured cerebral aneurysms arising in the collateral arteries in patients with occluded main cerebral arteries. The *de novo* formation of aneurysms following carotid ligation indicates the importance of hemodynamic stress in cerebral aneurysm formation.^{4,17} Cerebral aneurysms could be induced experimentally in hypertensive, β -aminopropionitrile-treated rats and monkeys with the carotid artery ligated unilaterally or bilaterally.^{8,9} In our patient, hemodynamic stress due to the internal carotid artery occlusion was probably involved in the *de novo* formation of the aneurysm in the anterior communicating artery.

The natural course of cervicocephalic FMD is considered to be relatively benign, and some authors warn against surgical intervention.^{20,22} However, in our patient the SAH necessitated clipping of the anterior communicating artery aneurysm associated with cervicocephalic FMD.

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