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Stenosis of the Proximal External Carotid Artery in an Adult With Moyamoya Disease: Moyamoya or Atherosclerotic Change?

—Case Report—

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Abstract

A 55-year-old woman presented with moyamoya disease manifesting as recurrent transient ischemic attacks despite taking aspirin and antihypertensive agent. Angiography showed the characteristic angiographic appearance with bilateral internal carotid artery occlusion and abnormal collateral vessels. Left external carotid angiography demonstrated moderate stenosis of the proximal external carotid artery (ECA). A self-expandable stent was successfully placed in the left ECA to improve ipsilateral cerebral perfusion. The patient had an uneventful outcome after a 1-year follow up. Involvement of the proximal ECA is very unusual in moyamoya disease, and might result from hemodynamic stress or degenerative atherosclerosis. Revascularization procedures for stenoses of proximal ECA may improve cerebral perfusion in patients with moyamoya disease.

Key words: moyamoya disease, external carotid artery, revascularization, stent

Introduction

Moyamoya disease is a progressive occlusive arteriopathy, usually affecting the bilateral internal carotid arteries (ICAs).[6,9] The diagnosis depends on the angiographic features of stenosis of the bilateral ICAs and compensatory enlargement of the preferring vessels at the base of the brain.[7] The symptoms and signs in children usually result from chronic cerebral ischemia, whereas those in adults are commonly due to intracranial hemorrhage.[4] Moyamoya disease is uncommon outside the basal carotid regions and rarely affects the extracranial vasculature.[3,5]

The external carotid artery (ECA) is an important collateral pathway supplying the brain through anastomotic connections in the presence of ICA occlusion. Such reliance on the proximal ECA may increase the risk of transient ischemic attack and/or cerebral infarction in areas with insufficient perfusion.[10] The significance of the ECA in the surgical correction of cerebrovascular insufficiency was recognized decades ago when neurological symptoms were resolved in patients with occluded ICAs and stenotic ECA by surgical endarterectomy and patch angioplasty of the ECA stenoses.[11] Patients with moyamoya disease can be treated by extracranial-intracranial (EC-IC) bypass, anastomosing the superficial temporal artery with the middle cerebral artery (MCA). Carotid artery stenting has recently become an alternative to carotid endarterectomy for ICA stenosis.[8]

We describe a case of progressive stenosis of the proximal ECA in a patient with moyamoya disease who was treated with endovascular stent placement.

Case Report

A 55-year-old woman was admitted to our hospital due to right arm weakness on November 5, 2001. The patient had a 3-year history of periodic transient paresis of her right arm treated with aspirin (100 mg per day). Two weeks before admission she experienced progressive speech disturbance with facial palsy. She had been treated with perioral antihypertensive agent for 2 years.

Neurological examination revealed mild right-

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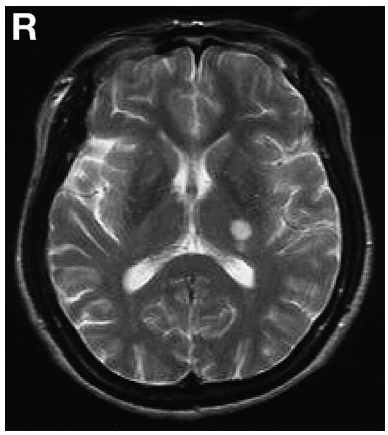

Fig. 1 Axial T_2-weighted magnetic resonance image revealing a hyperintense area in the left thalamus.

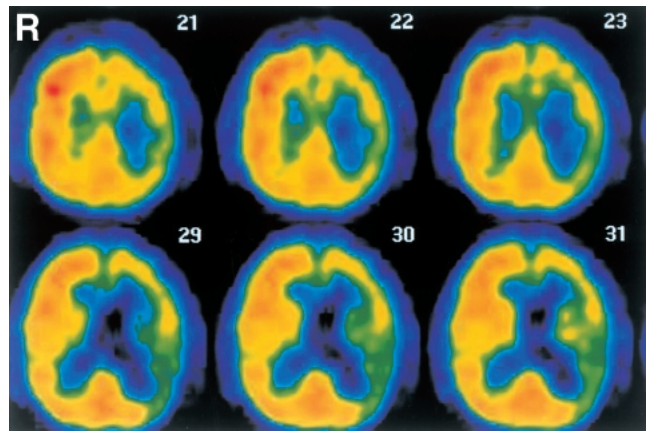

Fig. 3 Brain technetium-99m exametazime single photon emission computed tomography scans revealing a region of decreased perfusion in the left frontoparietal cortex.

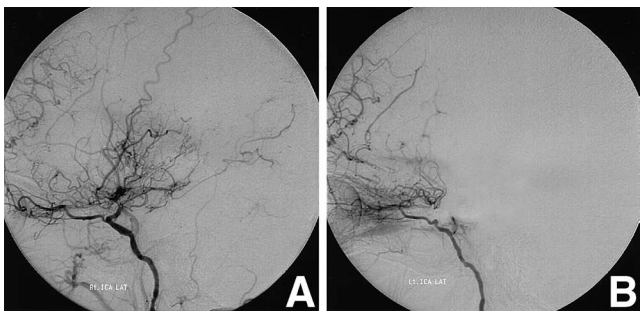

Fig. 2 (A) Right carotid angiogram demonstrating complete occlusion of the supraclinoid segment of the internal carotid artery (ICA) with multiple basal collateral vessels. (B) Left carotid angiogram showing complete occlusion at the level of cavernous segment of the ICA, just distal to the ophthalmic artery, and the left middle cerebral artery territory supplied by transdural collateral circulation from the left middle meningeal artery.

sided facial palsy but no definite limb weakness. T_2-weighted magnetic resonance (MR) imaging revealed a hyperintense area in the left thalamus (Fig. 1). T_1-weighted MR imaging with contrast medium showed multiple enhanced vascular structures in the bilateral basal ganglia and slow flow of cortical vessels. Moreover, axial T_2-weighted MR imaging failed to show the supraclinoid portions of the bilateral ICAs.

Right carotid angiography demonstrated complete occlusion of the supraclinoid segment of the ICA with multiple basal collateral vessels (Fig. 2A). The right frontal lobe was supplied by the anterior falcine artery from the ophthalmic artery as well as transdural collateral circulation from the right middle meningeal artery (MMA). Left carotid angiography showed complete occlusion at the level of cavernous segment of the ICA, just distal to the ophthalmic artery (Fig. 2B). The left MCA territory was supplied by transdural collateral circulation from the left MMA. Brain technetium-99m exametazime (Ceretec; Amersham, Buckinghamshire, U.K.) single photon emission computed tomography revealed a region of decreased perfusion in the left frontoparietal cortex (Fig. 3). Left external carotid angiography demonstrated moderate stenosis of the proximal portion of the left ECA (Fig. 4A). Vertebral angiography showed adequate leptomeningeal collateral circulation from the bilateral posterior cerebral arteries. The extracranial vertebral arteries had normal caliber.

The diagnosis was ''bilateral'' moyamoya disease with stenosis of the left ECA. The stenosis of the left ECA increased the risk of transient ischemic attack and/or cerebral infarction in the anterior circulation area supplied by the rich transdural circulation from the left ECA branches. Therefore, angioplasty with self-expanding stent placement and, if necessary, balloon angioplasty to directly restore the normal caliber were planned. If ECA angioplasty failed to restore adequate perfusion, superficial temporal artery-MCA anastomosis could be considered as the next treatment option. The patient's family gave informed written consent for stent-assisted or balloon angioplasty.

Systemic heparinization was achieved by continuous drip infusion of heparin (5,000 IU in 1 l normal saline). After puncture of the right femoral artery,

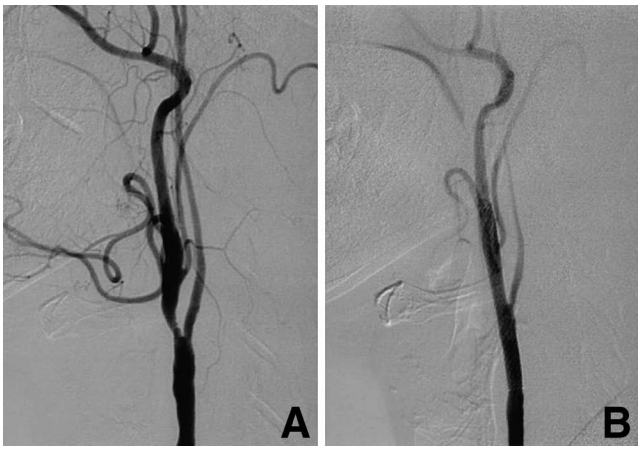

Fig. 4 **(A) Left common carotid angiogram demonstrating moderate stenosis of the proximal portion of the left external carotid artery. (B) Left common carotid angiogram after stent and balloon angioplasty revealing almost normal caliber of the external carotid artery.**

an 8-Fr femoral introducing sheath was inserted. The 8-Fr guiding catheter was introduced into the left common carotid artery. A 5 mm × 43 mm stent (Easy Wall; Boston Scientific Co., Natick, Mass., U.S.A.) was introduced from the ECA to the common carotid artery. After deployment of the stent, left carotid angiography showed persistent narrowing of the lumen. Therefore, balloon angioplasty was performed with a balloon catheter. After angioplasty, final carotid angiography showed almost normal caliber of the ECA (Fig. 4B). Heparin administration was discontinued after the procedure.

The postoperative course was uneventful and the patient was discharged at 4 days after the procedure. The patient received aspirin (100 mg per day) and clopidogrel (75 mg per day) for 3 months. Follow-up angiography at 1 year revealed patency of the stented artery with mild in-stent restenosis (Fig. 5A). Left carotid angiography showed abundant transdural collateral circulation from the MMA (Fig. 5B). The patient had developed no new neurological deficits including transient ischemic attacks. Her progress has been closely observed with continuing aspirin administration but no further treatment.

Discussion

This unique case demonstrates that the typical moyamoya changes associated with proximal ECA stenosis can be successfully treated with stent placement.

The etiology of moyamoya disease is unknown.

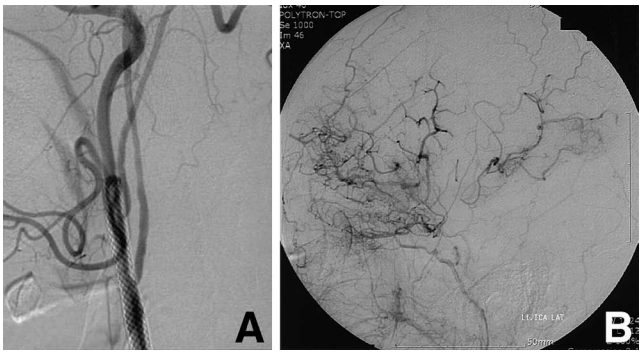

Fig. 5 **(A) Follow-up left common carotid angiogram at 1 year revealing patency of the stented artery with mild in-stent restenosis. (B) Left carotid angiogram showing abundant transdural collateral circulation from middle meningeal artery.**

Although most cases are sporadic, both genetic factors and environmental triggers are thought to be involved. The typical histological characteristics of moyamoya disease consist of lipid-poor fibrocellular intimal thickening.[10] These histological changes are not always restricted to the intracranial arteries, but may also occur in branches of the ECA and peripheral arteries.[1,11,12] Therefore, moyamoya disease may also affect the extracranial arteries, so constituting a systemic disorder. In our patient, the proximal ECA was progressively involved in moyamoya disease. This association may be infrequent but part of the natural history of moyamoya disease.

Atherosclerotic changes may also occur together with moyamoya disease.[3] An autopsy study found the essential arterial changes in the vertebrobasilar system in moyamoya disease were hypertensive in nature.[9] Such changes might be due to the increased pressure loading on the vertebrobasilar system to compensate for the occlusive state of the ICA system. Medial hypertrophy in the vertebrobasilar system is suggestive of the complexity of hemodynamic changes and consequent tissue responses in moyamoya disease. In our case, the involvement of the ECA might have resulted from hemodynamic stress or degenerative atherosclerosis.

The collateral circulations to the brain are very important and include large interarterial connections through the circle of Willis, small interarterial connections arising from leptomeningeal collaterals on the cerebral surfaces, and EC-IC connections. The most important of the EC-IC collaterals is through the ECA, collateralizing primarily through the periorbital plexus. The ipsilateral ECA is likely

to be essential in the presence of ICA occlusion associated with moyamoya disease. If these important pathways of collateral cerebral blood flow become obstructed, ischemic symptoms may occur. For example, the ipsilateral ECA contributes significantly to intracranial blood flow and oxygen saturation in patients with severe ICA stenosis.[2] Ipsilateral ECA revascularization can improve cerebral perfusion. However, we could not provide any firm biological evidence that the extracranial disease was indeed related to moyamoya disease in our case. We did not perform any postprocedural perfusion study and did not demonstrate that cerebral perfusion was improved after stenting to support the rationale for endovascular stenting in the presence of stable neurological conditions.

Stent placement is much less invasive than surgery and may prevent recurrent embolic or hemodynamic symptoms in moyamoya disease patients with the ECA stenoses, but remains controversial. In this case, the procedure carried the risk of placing of a stent across the origin of the left ICA which was supplying the ophthalmic artery. The ophthalmic artery is also important for collateral flow in moyamoya disease. Careful follow-up angiography is required to monitor for ICA flow compromise and restenosis of the stented ECA segment in this unusual case.

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