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LETTERS TO THE EDITOR

Accessory middle cerebral artery and moyamoya disease

A rare association of moyamoya disease with the accessory middle cerebral artery was seen in two patients. The terminal portions of bilateral internal carotid arteries and their vicinities were markedly stenotic and so-called moyamoya vessels developed at the base of the brain. The left accessory middle cerebral artery originating near the anterior communicating artery supplied the left anterior frontal lobe in both patients. Although the accessory middle cerebral artery coursed in the vicinity of the markedly stenotic terminal portion of the left internal carotid artery, the artery was not stenotic. This finding implies that the steno-occlusive changes in the cerebral vasculature in moyamoya disease have topological predilection to the distal internal carotid arteries.

Moyamoya disease is characterised by angiographic features of steno-occlusive changes of the terminal portions of bilateral intracranial internal carotid arteries as well as dilated perforating arteries at the base of the brain known as "moyamoya" vessels. The clinical manifestation of moyamoya disease is typically brain ischaemia in the paediatric population and brain haemorrhage in adults.[1] The accessory middle cerebral artery is a variation of middle cerebral artery branching and its incidence has been reported to be 0.3–4.0%.[2–4] The accessory middle cerebral artery originates from either the proximal or distal horizontal portion of the anterior cerebral artery coursing parallel to the horizontal portion of the middle cerebral artery and reaches the anterior frontal lobe.[5]

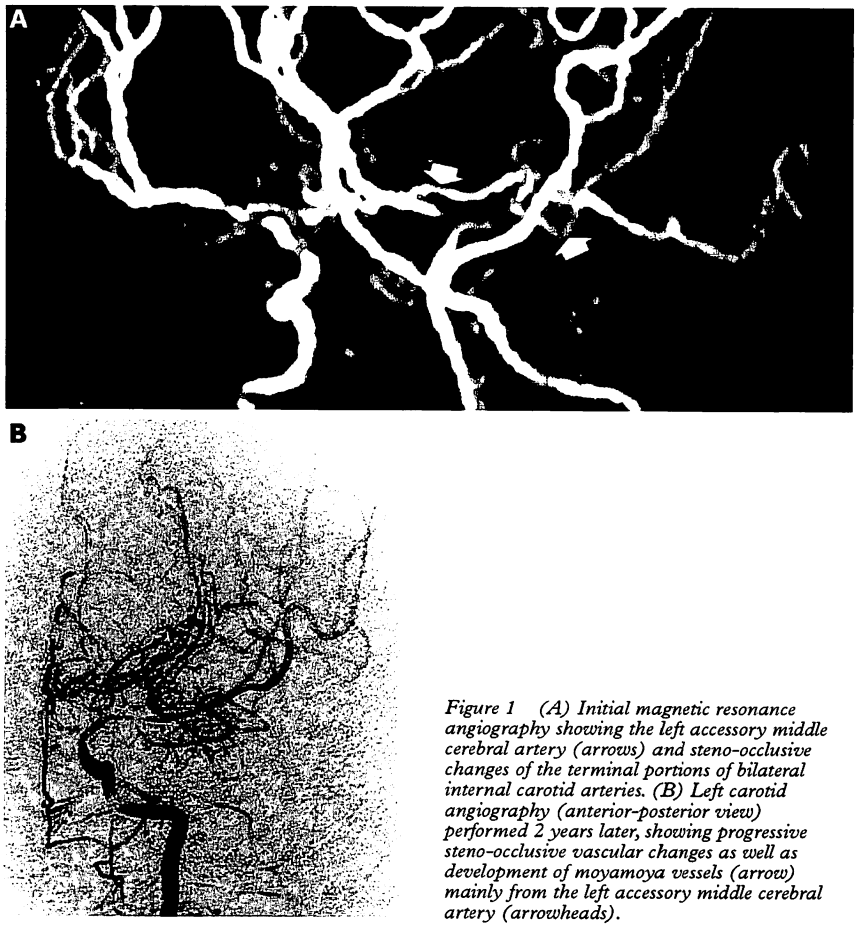

Figure 1 (A) Initial magnetic resonance angiography showing the left accessory middle cerebral artery (arrows) and steno-occlusive changes of the terminal portions of bilateral internal carotid arteries. (B) Left carotid angiography (anterior-posterior view) performed 2 years later, showing progressive steno-occlusive vascular changes as well as development of moyamoya vessels (arrow) mainly from the left accessory middle cerebral artery (arrowheads).

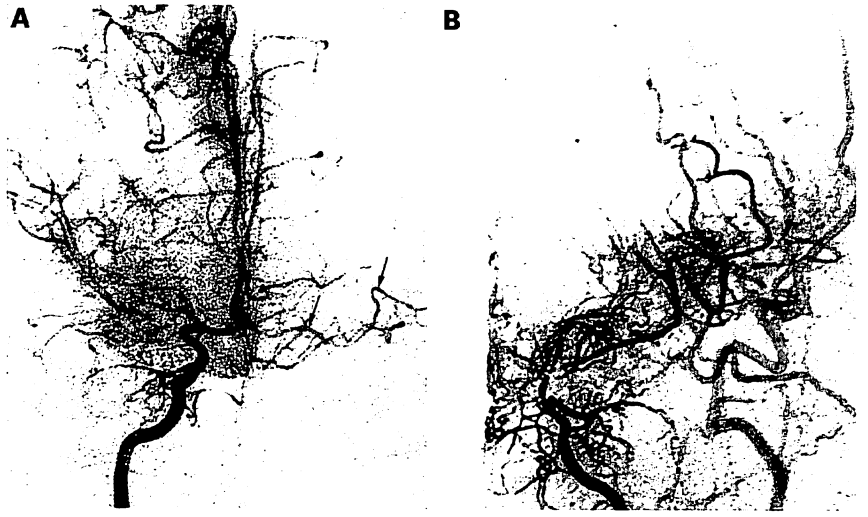

Figure 2 (A) Right carotid angiography (anterior-posterior view) shows stenotic change at the origin of the right middle cerebral artery, but the right anterior cerebral artery is normal. The left accessory middle cerebral artery (arrows) originates near the anterior communicating artery coursing parallel to the left middle cerebral artery. (B) Left carotid angiography (anterior-posterior view) shows severe steno-occlusive changes at the terminal portion of the internal carotid artery with development of moyamoya vessels.

Patient 1, a 32 year old woman, had presented with a transient ischaemic attack of right hemiparesis 2 years before the current episode. Results of initial MRI imaging performed at another hospital were interpreted as normal, but MR angiography showed steno-occlusive changes of the terminal portions of bilateral intracranial internal carotid arteries. Stenotic changes were more severe on the left than on the right side. Although the proximal portion of the left middle cerebral artery was markedly stenotic on MR angiography, the left accessory middle cerebral artery was clearly shown to be without stenosis (fig 1 A). The patient was treated conservatively.

No recurrent ischaemic attack had occurred over a period of 2 years until the patient began to experience transient ischaemic attacks of right hemiparesis several times a month. She was referred to us for further evaluation. She was neurologically normal and history was not contributory except for mild hypertension for 2 years. Digital subtraction angiography showed progressive steno-occlusive changes of the terminal portions of the internal carotid arteries as well as development of moyamoya vessels, which were consistent with the diagnosis of moyamoya disease (fig 1 B). Most moyamoya vessels on the left side originated from the accessory middle cerebral artery. This patient subsequently underwent bypass surgery bilaterally. Frequency of transient ischaemic attacks reduced markedly during the follow up period of 3 months postoperatively.

Patient 2 was a 30 year old man admitted for re-evaluation of moyamoya disease. This patient experienced occasional headache and vomiting at the age of 5 years. At the age of 11 years, transient ischaemic attacks of right hemiparesis developed to a rate of one a week. The diagnosis of moyamoya disease was established by cerebral angiography. He underwent bilateral bypass surgery subsequently at the age of 13 years in another hospital. He had experienced no ischaemic episodes for 17 years thereafter and he thought that moyamoya disease was cured. He came to our hospital when he had minor head trauma at the age of 30 years and was advised to re-evaluate the disease.

At admission, the patient was neurologically normal. Brain MRI showed no parenchymal abnormality. Single photon emission computed tomography was normal. Right carotid angiography showed severe stenotic

change of the proximal right middle cerebral artery, but the right anterior cerebral artery was normal. The left accessory middle cerebral artery originated near the anterior communicating artery (fig 2 A). Left carotid angiography showed severe stenosis at the terminal portion of the internal carotid artery with moderate development of moyamoya vessels (fig 2 B). The left accessory middle cerebral artery was not stenotic despite the vicinity of the markedly stenotic distal internal carotid artery and middle cerebral artery. Moyamoya vessels were not supplied by the left accessory middle cerebral artery. The patient was conservatively followed up for 6 months without any ischaemic episodes.

An association of the accessory middle cerebral artery and cerebral aneurysms has been well documented.[2 6] Moyamoya disease is highly associated with primitive carotid-basilar anastomosis, such as with the primitive trigeminal arteries and their variants.[7] To our knowledge, however, an association of the accessory middle cerebral artery with moyamoya disease has not been reported. The accessory middle cerebral artery was first regarded as a hypertrophied recurrent artery of Heubner.[8] However, it is now thought to be a cortical branch of the middle cerebral artery supplying the anterior frontal lobe, which is annexed to the embryological early artery, the anterior cerebral artery.[5] The accessory middle cerebral artery can serve as a collateral blood supply when the internal carotid artery or middle cerebral artery, or both are stenotic or occluded,[9] as was the case in our patients.

Our patients are interesting in that (*a*) the accessory middle cerebral artery was associated with moyamoya disease and (*b*) the accessory middle cerebral artery was not stenotic even though the distal internal carotid artery and the proximal middle cerebral artery showed steno-occlusive changes. Stenotic changes were not seen in the accessory middle cerebral artery although it coursed in the vicinity of the stenotic horizontal portion of the middle cerebral artery. This implies that susceptibility to arterial stenotic change is limited to the distal portion of the internal carotid artery and the proximal middle cerebral artery but not to the accessory artery even though all of these vessels are in close proximity. The cause of moyamoya disease is still unknown, but we think that there is a topographic difference in the predilection to stenotic changes in the cerebral vasculature in the disease.

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