Infarctions and non-invasive diagnosis in Moyamoya Disease: Two case reports

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Abstract
An 31-year-old female and a 32-year-old male had clinical signs and angiographical confirmation of adult Moyamoya disease (MMD). Bilateral carotid siphon (C1), middle cerebral artery (M1), and anterior cerebral artery (A1) stenoses were diagnosed by means of transcranial Doppler sonography (TCD) and visualized during angio Magnetic Resonance Imaging (angio-MRI). In the woman, a ‘rete mirabile’ of dilated and tortuous lenticulostriate arteries could be visualized during proton weighted and T1-weighted MRI sequences. CO₂-dependent vasomotor reactivity was bilaterally reduced and completely exhausted in the territory of right middle cerebral artery in both patients. Cerebral perfusion reserve, defined as the ratio of cerebral blood flow (CBF) to cerebral blood volume (CBV) was assessed by SPECT, and was found to be dramatically reduced in the anterior region of the male patient. Both patients had bilateral small subcortical infarctions in the corona radiata. TCD and MRI are important noninvasive techniques for a preliminary diagnosis of Moyamoya disease. Infarctions in Moyamoya disease may be hemodynamically produced low-flow infarctions.

Keywords: Hemodynamic infarctions, Magnetic Resonance Imaging, Moyamoya disease, transcranial Doppler sonography.

1 Introduction
Moyamoya disease is a bilateral occluding process of the basal cerebral arteries of unknown etiology. Its diagnosis is based on angiographical criteria which show bilateral stenoses of the terminal portion of the internal carotid arteries and the proximal portion of the anterior and middle cerebral arteries with simultaneous occurrence of abnormal vascular networks, the so-called “rete mirabile” [1, 7].

We report transcranial Doppler sonographical findings and Magnetic resonance imaging in two cases with angiographically confirmed Moyamoya disease.

2 Case reports
A 31-year-old female presented with dysarthria and clumsy hand syndrome. Within a few days, all symptoms ceased. A transcranial Doppler sonography (TCD) study revealed a high frequency, “stenotic” flow signal within the left anterior cerebral artery (ACA). A low-flow signal was found at an insonation depth of 50 mm corresponding to severe downstream flow changes within the left middle cerebral artery (MCA). A high-grade stenosis with “musical murmur” was also found in the right MCA at a depth of 50 mm. The posterior cerebral arteries (PCA) were hyperperfused. CO₂-dependent vasomotor reactivity [5] in both MCA distributions showed complete loss of vasomotor response to hyper- and hypocapnia on the right and severely reduced vasomotor reactivity on the left. Selective arterial angiography confirmed the ACA and ICA siphon stenoses. The left MCA was not found, the right MCA showed a high degree stenosis of the M1 segment at the origin of the lenticulostriate arteries. The lenticulostriates were maximally dilated and tortuous. Magnet Resonance Imaging (MRI) showed the maximally dilated and tortuous lenticulostriate arteries arising from the stenosed MCA.

A 32-year-old male of Turkish origin suffered from repeated sensory motor right-sided hemiparesis and aphasia. Transcranial Doppler sonography showed low-flow signal in both MCA’s. A stenosis
of the C1 segment of the internal carotid artery and stenoses of both ACA's were also diagnosed. CO$_2$-dependent vasomotor reactivity was completely exhausted in the right, and severely reduced in the left MCA distribution. Angio Magnetic Resonance Imaging showed high degree stenoses of both C1-segments of the internal carotid arteries with no orthograde flow in either MCA. A large irregular arterial network was seen arising from the posterior cerebral arteries and was interpreted as "rete mirabile". Selective arterial angiography confirmed bilateral high grade stenoses of the C1, M1, and A1 segments. The M2 segments of the MCA's were either filled retrogradely by leptomeningeal anastomoses or anterogradely by an abnormal vascular network originating from the posterior circulation. The lenticulostriate arteries were maximally dilated on the left.

All laboratory investigations were normal in both cases. The regional cerebral blood flow (rCBV), regional cerebral blood volume (rCBV), and the rCBF/rCBV ratio, i.e., the regional cerebral perfusion reserve (rCPR) [3] were assessed as described previously [2]. A decreased cerebral perfusion reserve was calculated in both MCA territories as compared to the posterior circulation (cerebellum) in the male patient. rCPR was normal in the woman.

In both patients, MRI showed several bilateral hyperintense 2–8 mm lesions rostral to the lateral ventricles in the corona radiata on T2-weighted images.

3 Discussion

TCD and angio-MRI showed bilateral C1, A1 and M1 stenoses and major basal artery collateral networks in 2 young individuals with a stroke history and angiographically demonstrated Moyamoya disease. These findings confirm diagnosis of Moyamoya disease, when symptomatic Moyamoya syndromes can be ruled out (e.g. tuberculous angiitis, angioma) [1]. Results of non-invasive tests such as TCD and MRI can suggest the further direction for the diagnostic procedures, but until now only selective angiography can confirm the diagnosis. TCD and angio MRI are, for instance, not able to register very slow flow, so that small anastomoses can not convincingly be detected [6]. The dilated and tortuous arteries of the abnormal vascular network were demonstrated by T1-weighted MRI and angio-MRI imaging. The origin of these vessels, however, could only be elucidated by angiography. In the male patient, the vessels arose from the posterior circulation and filled the M2-segment of the occluded MCA. In the female patient maximally dilated and tortuous lenticulostriate arteries arising from a subtotal MCA stenosis formed the 'rete mirabile'. In this latter patient, the infarctions were located in the rostral terminal supply area of these arteries. This was not unexpected since these Moyamoya vessels, despite being compensatorily dilated, arose from a nearly occluded parent vessel, i.e., the MCA.

In evaluating cerebral hemodynamic reserve [3], pathophysiological tests such as assessment of CO$_2$-dependent vasomotor reactivity by TCD or rCPR as rCBF to rCBV ratio are superior to conventional purely morphological techniques [2, 3, 5]. The morphological pattern of brain infarctions in these two patients and the exhausted cerebral hemodynamic reserve suggest that these infarctions were hemodynamically produced, i.e., were low-flow infarctions [4]. Due to the high degree stenoses or occlusions of nearly all basal arteries, the compensatory power of the circle of Willis was severely reduced and the cerebral hemodynamic reserve was exhausted. Low-floe infarctions in the terminal supply area of the lenticulostriate arteries, which are end arteries in the true sense, resulted. An EC/IC-bypass operation may be considered in such patients. However the generally good prognosis of conservatively treated Moya-Moya disease makes one hesitate to recommended bypass surgery.