Case Report

Role of magnetic resonance perfusion studies in moyamoya disease

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Moyamoya disease, Japanese for ‘puff of smoke’, is a rare disease that presents most commonly with recurrent TIsA (transient ischemic attacks) / stroke in childhood. Ischemic symptoms in patients with moyamoya disease are usually due to hemodynamically-mediated perfusion failure. Identification of abnormal tissue perfusion is an important aspect of the evaluation of these patients. We present the radiological features including the Magnetic Resonance (MR) Perfusion findings illustrating the hemodynamic changes of cerebral ischemia in a case of moyamoya disease.

Key Words: Moyamoya disease, cerebral ischemia, magnetic resonance perfusion

Introduction

Moyamoya disease, a chronically progressive cerebrovascular occlusive disease affects the supraclinoid Internal Carotid Arteries (ICA) with prominent collateral formation, either of unknown etiology or secondary to neurofibromatosis and sickle cell anemia. Moyamoya affects the Japanese and Koreans and other ethnic groups. MR Perfusion allows non-invasive investigation of tissue perfusion. Simultaneously acquired structural and diffusion images identify tissue with a normal diffusion but abnormal perfusion. In an acute stroke, such areas represent tissue-at-risk; in chronic cerebral ischemia, hypoperfused areas, not irreversibly damaged, are evident. Conventional angiography, the ‘gold-standard’ for depicting collaterals, nevertheless requires MRI to arrive at a diagnosis. This case demonstrates the utility of MRI and MR Perfusion as a stand-alone modality for moyamoya.

Case Report

An 8-year-old male presented with seizures and choreiform movements since 3½ years of age with slowing of mentation. Family history was negative. Examination revealed memory impairment, disorientation, hypotonia and chorea. Routine hematology, biochemistry and serology were negative. Computed Tomography revealed non-enhancing hypodense areas in the occipital and frontal regions bilaterally, suggesting infarcts. Basal ganglia collaterals were noted on post-contrast scans and moyamoya was considered as one of the differential diagnoses.

Routine MRI, followed by Magnetic Resonance Angiography and MR Perfusion were obtained using a 1.5 Tesla System (Echospeed, GE Medical systems, Milwaukee, USA). MRI using T1 and T2 weighted (T1W, T2W) Spin Echo and FLAIR (Fluid-Attenuated Inversion Recovery) sequences, revealed well-defined wedge-shaped hyperintensities in bilateral frontal, parietal, parieto-occipital and left temporal cortices and subcortical white matter, suggestive of encephalomalacic areas (Figure 1).

MR Angiographic images through the circle of Willis revealed near-complete occlusion of both supraclinoid ICAs. Both middle cerebral arteries (MCA) were not visualized. Both anterior cerebral arteries seemed to fill from the collaterals. Prominent vascular channels in the skull-base region (leptomeningeal collaterals) and basal ganglia resembled a ‘puff of smoke’ (Figure 2). External carotid arteries (ECA) were prominent. Common carotid ar-

Figure 1: T2 weighted image depicting encephalomalacic areas in the bifrontal and left parieto-occipital cortices (arrows)

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Discussion

Children present typically with recurrent episodes of sudden hemiplegia that might alternate sides. Fine involuntary movements of the extremities, seizures, slowly progressive mental impairment and TIs.

Children display a progressive course with ample collaterals while adults often have an arrested course with altered circulatory dynamics.

Imaging focuses on this collateral circulation, which forms owing to the occlusion of major proximal intracranial arteries. Typically, laboratory data are unremarkable. Screening EEG can be distinctive. 20 to 60 seconds following the termination of hyperventilation, high-voltage, slow waves return. The most consistent CT finding is a low-density area in the temporal white matter, a feature absent in our patient. However, angiography depicts the characteristic ‘puff of smoke’, providing a definitive diagnosis. Also, Single Photon Emission Computed Tomography (SPECT) or Positron Emission Tomography (PET) are newer reference standards for assessing cerebral hemodynamics.

MR Angiography (MRA) also depicts collaterals. Limitations of CT and MRA can be overcome by a combination of diffusion-and perfusion-weighted MRI in the same imaging session. Ultrafast Echo Planar MR Imaging (EPI) sequences evaluate the passage of contrast through the intracranial vasculature. MR Perfusion is performed before and after the bolus of a paramagnetic contrast such as gadolinium or dysprosium. Voxel signal intensity is then plotted versus time (signal-intensity curves), allowing conversion of the information to cerebral blood volume (CBV). With ischemia, perfusion-imaging shows less magnetic susceptibility-induced signal loss and / or delay in peak signal loss in the affected vascular distribution, compared with normally perfused tissue. Perfusion imaging can also be used for follow-up after

Figure 2: MR Angiographic images (frontal view-above, lateral view-below) through the circle of Willis showing a profusion of collateral vessels resembling a ‘puff of smoke’ (arrows)

Figure 3: MR Perfusion through the territory supplied by the anterior circulation. Perfusion tracing (signal-intensity curve [signal intensity = y-axis; time = x-axis]) corresponding to the regions of interest (ROIs) on the T2 weighted image shows an increase in the time to peak (TTP, arrow) with a drop in the maximum signal intensity, in comparison to the posterior circulation (see Figure 4)

Figure 4: MR Perfusion through the territory supplied by the posterior circulation. In comparison with Figure 3, signal-intensity curve corresponding to the ROIs on the T2 weighted image shows a TTP (arrow) reduction with an increase in the MPC
duraencephalosynangiosis, an ECA-to-ICA bypass surgery. Diffusion MRI has demonstrated ischemic penumbral at the periphery of acute infarcts. In our case, diffusion studies were normal, thus highlighting the chronic nature of the infarcts.

In moyamoya, reduced regional cerebral blood flow (CBF) reflects the course of the disease, which starts from ICAs, MCAs and extends to the posterior cerebral arteries. The posterior circulation usually supplies the ischemic fronto-temporal lobes via the leptomeningeal collaterals, whereas the deep nuclei derive supply from the multiple basal collaterals. Such findings can be detected with perfusion-weighted EPI. The posterior circulation on perfusion scan is slightly delayed compared to the anterior in normals. This phenomenon was slightly reversed in our patient, indicating that perfusion of the posterior circulation was more brisk than that of the anterior, a feature consistent with moyamoya. The perfusion tracing through the anterior circulation, on the T2W image depicted an increase in the time to peak (TTP, seen as a dip on the tracing) with a drop in the maximum signal intensity, as compared to the posterior circulation. Perfusion through infarcted areas on routine MRI showed a flat curve indicative of an ischemic core. At the periphery of these lesions, a smaller ‘dip’ indicated an ischemic penumbra.

In conclusion, once the diagnosis of moyamoya has been reached using conventional angiography, MR Perfusion reliably depicts hemodynamic abnormalities and can also assess the patient postoperatively.

References


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