

Rare Diagnosis in Emergency Room: Moyamoya Disease

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Abstract : Moyamoya disease is a unique chronic progressive cerebrovascular disease characterized by bilateral stenosis or occlusion of the arteries around the circle of Willis with prominent arterial collateral circulation. The occurrence of Moyamoya disease is related to immune, genetic and other factors. There is no curative treatment for Moyamoya disease. Secondary prevention for patients with symptomatic Moyamoya disease is largely centered on surgical revascularization techniques. We present here a 62-year old male presented with headache and vision loss for 2 days. He was previously diagnosed with hypertension and glaucoma. On physical examination, left eye movements were restricted medially, both eyes were hyperemic and their movements were painful. Other neurological and physical examination were normal. His vital signs and laboratory results were within normal limits. Computed tomography (CT) showed dilated vascular structures around both lateral ventricles and atherosclerotic changes inside the walls of internal carotid artery (ICA). Magnetic resonance imaging (MRI) and angiography (MRA) revealed dilated venous vascular structures around lateral ventricles and hyper-intense gliosis in periventricular white matter. Ischemic gliosis around the lateral ventricles were present in the Digital Subtracted Angiography (DSA). After the neurology, ophthalmology and neurosurgery consultation, the patient was diagnosed with Moyamoya disease, pulse steroid therapy was started for vision loss, and super-selective DSA was planned for further investigation. Moyamoya disease is a rare condition, but it can be an important cause of stroke in both children and adults. It generally affects anterior circulation, but posterior cerebral circulation may also be affected, as well. In the differential diagnosis of acute vision loss, occipital stroke related to Moyamoya disease should be considered. Direct and indirect surgical revascularization surgeries may be used to effectively revascularize affected brain areas, and have been shown to reduce risk of stroke.

Keywords : headache, Moyamoya disease, stroke, visual loss

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