Moyamoya Syndrome with Recurrent Intraparenchymal Hemorrhage from Hemoglobin C Disease

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Abstract

Moyamoya syndrome is an acquired idiopathic non-atherosclerotic vasculopathy that involves the terminal internal carotid artery and its branches. It is associated with numerous medical conditions such as hematologic abnormalities, autoimmune diseases, brain radiation, and tumors. These medical diseases cause chronic endothelial damage to cerebral vasculature resulting in both friability and stenosis of vessels, presenting as cerebral hemorrhage or infarction. There is an established link between moyamoya syndrome and hematologic diseases such as sickle cell and beta thalassemia. A few case reports of rare hemoglobinopathies have caused moyamoya syndrome; however, there are no reports of moyamoya due to hemoglobin C disease. This case report presents a patient with known hemoglobin C disease who presented with her second intracranial hemorrhage, found to have moyamoya syndrome.

Keywords: Moyamoya; Hemoglobin C disease; Intraparenchymal hemorrhage

Introduction

Moyamoya syndrome is an acquired idiopathic non-atherosclerotic vasculopathy that involves the terminal internal carotid artery and its branches. The syndrome is different from moyamoya disease as it is not a primary disease with associated genetic abnormality. Moyamoya syndrome has been associated with numerous medical diseases including but not limited to hematologic abnormalities, tumors, autoimmune diseases, or brain radiation. Both disease and syndrome usually have bilateral internal carotid and involvement; however can be unilateral. Angiography shows a characteristic “puff of smoke” appearance from collateralization of vasculature. Moyamoya syndrome presents more commonly as ischemic stroke in the pediatric population and hemorrhagic stroke in the adult population. Less common clinical presentations are epilepsy and movement disorders. Medical treatment consists of antiplatelet therapy to prevent thrombus formation. Surgical treatments include direct revascularization via anastomosis of the superficial temporal and middle cerebral artery which is more commonly used in children. Indirect revascularization consists of transposition of vasculature directly onto the cerebral cortex. It is unclear if surgical revascularization reduces the rate of hemorrhagic stroke.

Case Presentation

A 43-year-old right-handed female with type 1 von Willebrand’s disease, homozygous hemoglobin C disease and prior left thalamic hemorrhagic stroke in 2014 without residual deficits originally presented to an outside hospital with generalized shaking activity for 5-9 min at home. She was found to have a left basal ganglia hemorrhage with intraventricular extension and was transferred to our hospital for neurosurgical intervention. NIH stroke scale was 29. Intracranial hemorrhage score was 3 and was intubated. Patient was not taking aspirin or anticoagulation prior to admission. Upon transfer to our hospital, her exam showed a presence of oculocerebral, corneal and gag reflexes. She did not withdraw to noxious stimuli in the right upper extremity but did grimace, localized in the left upper extremity, and withdrew to noxious stimuli in the lower extremities bilaterally but more briskly on the left than the right. She did not have upper motor neuron signs, clonus, or up toe responses. Patient had an extra ventricular drain placed on admission.

Laboratory studies showed microcytic anemia with baseline hemoglobin ranging from 8-9 throughout admission. Computed tomography angiogram (CTA) showed left intracranial and extracranial stenosis with extensive collateralization around Circle of Willis. Given the CTA findings, patient underwent a diagnostic cerebral angiogram which showed late stage moyamoya of the left anterior circulation (Figures 1 and 2).

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Her hemoglobin C disease was associated with mild hemolytic anemia and mild splenomegaly, treated with daily folic acid and ferrous sulfate supplementation. On discharge, patient had right upper extremity and right hip flexor strength 4/5. 4 months after discharge, the patient underwent left encephalo-duro-arterio-synangiosis (EDAS). At her postoperative clinic visit, she had full strength in all muscle groups in her right upper and lower extremities. She is scheduled to have a 6 month postop diagnostic angiogram. 

Discussion

Moyamoya syndrome is a non-atherosclerotic vasculopathy involving the cerebral vasculature, most commonly involving the internal carotid artery and its branches. The term “moya moya” is Japanese for “puff of smoke,” referring to the characteristic angiographic appearance of vascular collateralization. Its pathogenesis is not well understood; however, basic fibroblast growth factor may play a role [1]. It is hypothesized that structurally abnormal red blood cells cause endothelial injury leading to endothelial proliferation resulting in vessel occlusion [1]. Histopathology shows intimal thickening from smooth muscle cell proliferation with medial fibrosis [1]. The medial fibrosis results in areas of discontinuity which then predisposes the vessel to hemodynamic stress [1]. In moyamoya vessels, it is noted that there is minimal or absent lipid deposition indicating a non-atherosclerotic process. Moyamoya has been associated with a variety of different hemoglobinopathies, genetic conditions that cause hemoglobin structural abnormalities resulting in abnormal red blood cells. The most widely known hemoglobinopathy is a mutation in hemoglobin S, or sickle cell disease. Sickle cell disease has been a well-established hematologic, and genetic factors. Am J Hematol 70: 206-15.

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