Case Report

Moyamoya disease presenting as ischemic stroke and interventricular hemorrhage in a young adult: A case report

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ABSTRACT

Moyamoya disease is a chronic, non-atherosclerotic cerebrovascular disorder characterized by progressive stenosis of the terminal internal carotid arteries along with the development of abnormal basal collateral networks. It typically presents with ischemic events in children, whereas hemorrhagic manifestations are more common in adults. We report the case of a 17-year-old female with a 10-day history of altered sensorium. Magnetic resonance imaging of the brain with contrast shows features suggestive of multiple acute infarcts with hemorrhagic transformation in the genu of the corpus callosum, along with interventricular hemorrhage. MR angiography shows occlusion of the supraclinoid right internal carotid artery, middle cerebral artery, posterior cerebral artery, bilateral anterior cerebral artery, and severe stenosis of the left supraclinoid internal carotid artery and left middle cerebral artery, along with extensive basal and cortical collateral vessels with ~95% distal basilar artery stenosis. This report highlights a rare presentation of combined ischemic and hemorrhagic stroke in a young adult female.

Key words: Interventricular hemorrhage, Ischemic stroke, Moyamoya disease, Magnetic resonance angiography

Moyamoya disease (MMD), derived from the Japanese term meaning "hazy puff of smoke," is a chronic, non-atherosclerotic cerebrovascular occlusive disorder characterized by bilateral stenosis of the terminal internal carotid arteries (ICA) and/or the proximal anterior and middle cerebral arteries [1]. The Japanese Ministry of Health and Welfare classifies MMD into four clinical types based on presentation: Ischemic (63.4%), hemorrhagic (21.6%), epileptic (7.6%), and other forms (7.5%) [2]. The identification of RNF213, encoding an unconventional E3 ubiquitin ligase, as a major susceptibility gene for MMD in East Asian populations has provided new insights into disease pathogenesis and potential therapeutic targets [3]. The characteristic histopathological changes in the steno-occlusive arteries include fibrocellular intimal thickening with proliferation of smooth muscle cells and a markedly tortuous, often duplicated internal elastic lamina, typically in the absence of atheromatous plaque formation [4].

CASE PRESENTATION

A 17-year-old young Indian female presented in our emergency department with a chief complaint of altered sensorium for 10 days. There was no history of fever, vomiting, or seizures.

Her vitals were blood pressure 110/78 mmHg, pulse rate 88/min, oxygen saturation 95%, random blood sugar 112 mg/dL, and temperature 98.8°F. Her Glasgow Coma Scale (GCS) was $E_3V_3M_5$, not oriented to time, place, or person. Pupil was mid-dilated and reactive. Plantar Response was bilateral extensor response. Neck rigidity and Kernig sign were absent.

Cerebrospinal fluid (CSF) analysis revealed a glucose level of 63.3 mg/dL, a protein level of 40.5 mg/dL, and a total cell count of 800 cells/mm^3 with neutrophilic predominance (65% neutrophils and 35% lymphocytes). Adenosine deaminase level was 3.42 IU/L. Non-contrast computed tomography of the brain demonstrated ill-defined hypodensities involving the left parieto-temporal lobe, left ganglio-capsular region, corona radiata, and corpus callosum, consistent with an acute infarct. Associated hyperdense foci within the infarcted areas and along the bilateral ventricles were suggestive of hemorrhagic transformation.

Magnetic resonance imaging (MRI) of the brain revealed areas of T2/Fluid-attenuated inversion recovery hyperintensity involving the left parieto-temporal region, anterior left corona radiata, left centrum semiovale, and the genu and splenium of the corpus callosum, with

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corresponding diffusion restriction on diffusion-weighted imaging and low apparent diffusion coefficient values. Similar signal changes were noted within the bilateral lateral, third, and fourth ventricles, with susceptibility-weighted imaging hypointensities in the bilateral lateral ventricles and genu of the corpus callosum, consistent with multiple acute infarcts with hemorrhagic transformation and associated intraventricular hemorrhage. MR angiography of the brain demonstrated non-visualization of the supraclinod segment of the right ICA with occlusion of the right M1 segment of the middle cerebral artery, right posterior cerebral artery, and bilateral anterior cerebral arteries. Severe stenosis was noted in the supraclinoid segment of the left internal carotid artery and the left M1 segment, with additional stenosis involving the lacerum and cavernous segments of the right ICA. Extensive collateral vessels (moyamoya vessels) were seen within the bilateral middle cerebral artery (MCA) cisterns, interpeduncular cistern, right Sylvian fissure, and along the bilateral frontal convexities, supplying the right basal ganglio-thalamic region. Severe (~95%) stenosis of the distal basilar artery was also observed. MR venography revealed mild stenosis involving the mid-portion of the superior sagittal sinus. The patient was diagnosed with MMD, having acute infarcts in the corpus callosum and temporo-parietal region, along with interventricular hemorrhage. However, digital subtraction angiography scan and genetic studies could not be done due to technical limitations.

The patient was managed conservatively with the help of antibiotics and osmotic diuretics (mannitol). After some days, her GCS started improving and was ultimately referred to a higher center for further evaluation and management.

DISCUSSION

Although MMD has been recognized as having genetic associations, the majority of cases occur sporadically. In addition, some individuals remain asymptomatic, with the disease being detected incidentally during angiographic evaluation [5]. Suzuki and Takaku described six progressive angiographic stages that characterize the natural evolution of MMD [6]. Studies have shown that patients with MMD exhibit markedly higher concentrations of basic fibroblast growth factor (bFGF) in the CSF, with average levels of 64.0 pg/mL compared to 6.5 pg/mL in individuals without MMD [7]. Yoshimoto *et al*. suggested that this elevation is specific to MMD rather than a non-specific consequence of cerebral ischemia, highlighting bFGF as a potential disease-specific biomarker. Intracerebral hemorrhage continues to be the primary cause of mortality in patients with MMD [4].

The case is remarkable for the concurrent presence of ischemic cerebral infarction with hemorrhagic transformation and intraventricular hemorrhage in a young adult, an infrequent and diagnostically challenging clinical scenario. While ischemic stroke represents the predominant manifestation of MMD in younger individuals, the simultaneous occurrence of intraventricular hemorrhage indicates rupture of fragile collateral vessels, underscoring the complex interplay between ischemic and hemorrhagic mechanisms characteristic of this disorder.

Another notable aspect of this case is the extensive multivessel involvement, including bilateral anterior and middle cerebral arteries, posterior cerebral artery, and severe stenosis of the distal basilar artery. Involvement of the posterior circulation is increasingly recognized in advanced stages of MMD and is associated with a higher risk of neurological deterioration. The angiographic features in this patient are consistent with advanced Suzuki stages, indicating widespread disease progression. This case emphasizes the need to consider MMD in young patients presenting with stroke, particularly when neuroimaging reveals both ischemic and hemorrhagic components along with multivessel involvement.

The majority of patients are treated surgically, as surgical intervention has demonstrated superior efficacy compared with non-surgical management. Medical therapy is largely adjunctive and consists of vasodilators, antiplatelet agents, and anticonvulsants. Surgical revascularization remains the treatment of choice and includes procedures such as superficial temporal artery-MCA anastomosis, encephaloduroarteriosynangiosis, and encephalomyosynangiosis [8].

CONCLUSION

With newer imaging modalities and diagnostic techniques, the rate of MMD diagnosis continues to rise. Continued genetic analysis of moyamoya vessels is expected to enhance understanding of the disease process and facilitate earlier diagnosis, allowing timely therapeutic intervention.

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