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Recurrent Stroke in Young Adult as a Manifestation of Moyamoya Disease: Case Report

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ABSTRACT

Background: Moyamoya disease is a chronic cerebrovascular disorder characterized by progressive narrowing of the intracranial portions of the distal internal carotid artery, the initial proximal components of the middle cerebral artery, anterior cerebral artery, and posterior cerebral artery. Moyamoya disease can manifest as a stroke in young adult.
Methods: This report presents a young female patient with Moyamoya disease in Siloam Hospital Denpasar, Bali, Indonesia, diagnosed using MRI and Digital Subtraction Angiography.
Case Report: A 29-year-old woman with recurrent stroke, presented with right-side hemiparesis, urinary incontinence, disinhibition syndrome, hypertension, and a history of left hemiparesis from a previous stroke. The MRI brain showed hyperintensity throughout the sulci (ivy sign) in the bilateral frontoparietal with acute infarct in the left frontomedial region, chronic infarct in right frontomedial, occlusion of bilateral carotid arteries (supraclinoid segment), bilateral MCA and ACA. The cerebral DSA study showed stenosis in the right ICA, stenosis in the M1-M2 junction with poor collateral to ACA territory with a puff of smoke appearance, occlusion in the left ICA (supraclinoid segment), and severe stenosis in M1. The patient had undergone direct bypass in the right MCA and right superficial temporal artery. Unfortunately, the bypass did not succeed because of infection of the bypass site and the patient got oral medication with a single antiplatelet (asetosal) and angiotensin receptor blocker (valsartan).
Discussion: Moyamoya disease is a rare chronic cerebrovascular disorder that can manifest as an ischaemic stroke in young adult. This patient had a recurrent ischaemic stroke due to Moyamoya disease. According to a global multicenter stroke database, 90% of Moyamoya disease patients initially presented with an ischemic stroke, 7.5% with a transient ischemic attack, and 2.5% with a hemorrhagic stroke. DSA showed that the intracranial collateral system failed to fulfill the demand for brain circulation when the stroke occurred. The patient had failure to grow collateral after direct bypass surgery from the right superficial temporal artery to MCA and single oral antiplatelet as a secondary prevention for ischaemic stroke.
Conclusion: Moyamoya disease is a rare disease that needs further studies for its treatment to halt the progression and prevent secondary complications. Young patients with ischaemic stroke needed to be asses with brain MRI, MRA, and DSA to evaluate the etiology of ischaemic stroke.

Keywords: Moyamoya disease, Digital Substraction Angiography, ischaemic stroke

INTRODUCTION

In 1957, Japanese literature published the first description of the Moyamoya disease. In 1969, Suzuki and Takaku gave it the name "moyamoya illness." The same moyamoya phenomenon is represented by moyamoya syndrome, but it occurs in the context of either extra-neurological or neurological, inherited or acquired diseases.

As a result of persistent brain ischemia, Moyamoya vessels, which are described in Japanese as "something hazy like a puff of smoke drifting in the air," form mostly at the base of the brain. (Hertza et al., 2014)

A chronic cerebrovascular illness called Moyamoya disease is characterized by a progressive narrowing of the intracranial parts of the middle cerebral artery, anterior cerebral artery, and the distal internal carotid artery. It is extremely uncommon for the posterior vertebrobasilar circulation to be affected. The etiology of Moyamoya disease, a fundamental intrinsic pathological disease process, is not fully known. Reduced cerebral blood supply to the brain parenchyma due to the increasing stenosis results in ischemic stroke and a compensatory vascular response. Smaller blood vessels start to grow as the cerebral vasculature tries to make up for the loss of blood flow by establishing collateral circulation, and hypertrophy becomes increasingly apparent in diagnostic cerebral angiography. The angiographic appearance of a "puff of smoke," or "Moyamoya" in Japanese, is caused by the development of collateral blood arteries. This is the angiographic trait that distinguishes the condition. Recent investigations reveal that these collateral channels are just enlarged versions of preexisting micro-perforator arteries such as the lenticulostriate and thalamoperforating arteries, contrary to what was previously thought. (Berry et al., 2020)

CASE REPORT

A 29-year-old woman presented with right hemiparesis that occurs suddenly after waking up. She also complains about urinary incontinence. Her family said that she became easily angry. Physical examination of the vital signs is normal, and neurological examination shows grade 4 flaccid right hemiparesis. Neurobehaviour examination found disinhibition syndrome and frontal lobe syndrome. The patient has an ischemic stroke history from one year ago presented with left hemiparesis and got fully resolved, she got secondary stroke prevention with oral antiplatelet (acetylsalicylic acid). She also has a hypertension history and got medication with an angiotensin receptor blocker (candesartan).

Imaging examination from brain MRI and MRA show linear hyperintensity throughout the sulci (ivy sign) in bilateral frontomedial with acute infarct in the left frontal region, chronic infarct in right frontomedial, occlusion of bilateral carotid arteries (supraclinoid segment), bilateral MCA and ACA. Digital subtraction angiography showed the stenosis in the right internal carotid artery until T bifurcation with minimal flow to A1 with puff and stenosis in the left M1-M2 junction with a puff of smoke appearance.

Figure 1. Brain MRI with ADC sequence shows acute infarction in the left frontomedial region (blue arrow) and chronic infarction in the right frontomedial region (red arrow)

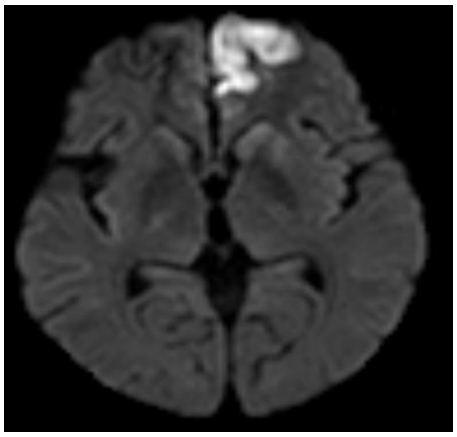

Figure 2. Brain MRI with DWI sequence shows acute infarction in left frontomedial region (blue arrow)

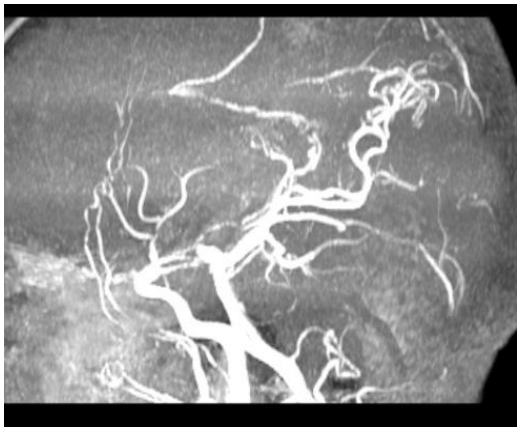

Figure 3. Brain MRA shows occluded ACA (blue arrow) and MCA (red arrow), collateral from posterior pericallosal artery to partial ACA territories (yellow arrow)

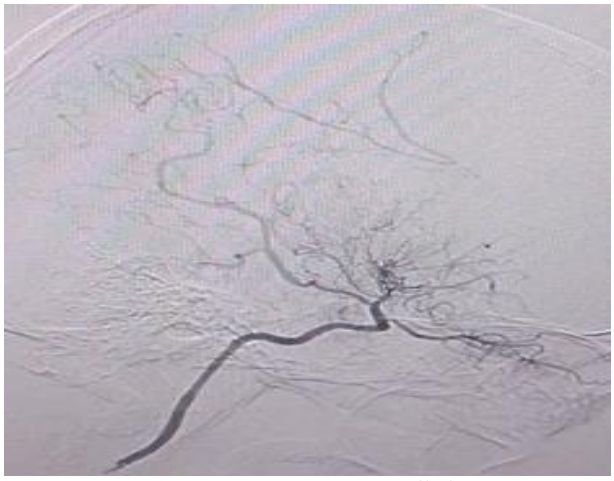

Figure 4. Right lateral brain DSA shows a puff of smoke appearance (blue arrow), occluded right ACA and MCA, and poor collateral from posterior pericallosal artery to partial ACA territories (red arrow)

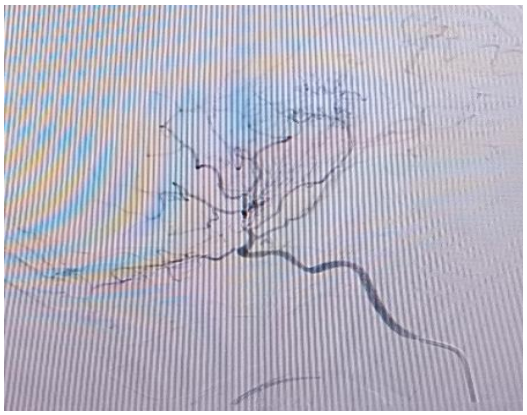

Figure 5. Left lateral brain DSA shows a puff of smoke appearance (blue arrow), occluded left ACA and MCA

The patient underwent surgical treatment for direct bypass from the right MCA to the right superficial temporal artery combined with oral medical management single oral antiplatelet. Brain MRI and MRA one month after bypass showed stenosis of supraclinoid segment of bilateral internal carotid arteries, stenosis and irregularity of bilateral middle cerebral arteries, stenosis of bilateral anterior cerebral arteries with multiple collaterals net-like vessels giving puff of smoke appearance. DSA post bypass showed distal occlusion at the supraclinoid segment of bilateral internal carotid arteries, both Pcom gives circulation to the right PCA then through the posterior pericallosal artery to anterior pericallosal artery to the ACA and MCA territories, showed puff of smoke appearance in both bilateral MCA and ACA arteries, there is no anastomosis from the right external carotid artery to the right internal carotid artery.

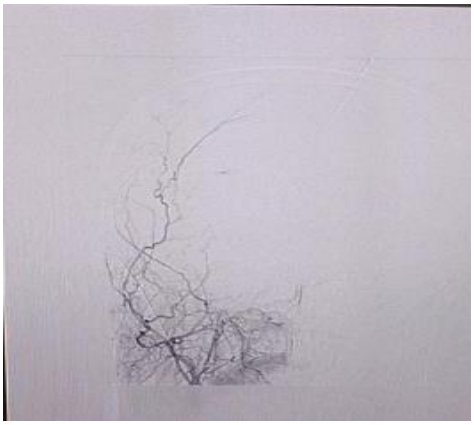

Figure 6. Right AP DSA post bypass shows no anastomosis from a right superior temporal artery (blue arrow) to the right middle cerebral artery.

Unfortunately, the bypass did not succeed because of an infection of the bypass site and the patient got oral medication with a single oral antiplatelet (acetylsalicylic acid) and angiotensin receptor blocker (valsartan).

DISCUSSION

A 29-year-old woman with right hemiparesis, urinary incontinence, disinhibition syndrome, and frontal lobe syndrome, neurological examination shows flaccid right hemiparesis grade 4 with ischemic stroke history from one year ago presented with left hemiparesis and get fully resolved. Moyamoya disease is a cerebrovascular disorder characterized by abundant, hazy, proximal collateral arteries that are known as "Moyamoya vessels,"

internal carotid artery blockage, and increasing vascular stenosis. For both children and adults with Moyamoya disease, these delicate, smoke-like arteries don't supply enough cerebral perfusion, which results in a variety of symptoms like ischemia or hemorrhagic stroke, seizures, cognitive decline, and disability or death. (Fang et al., 2021). The most frequent initial presentation is an ischemic episode, which is more common in children. Patients rarely experience hemorrhagic events; however, if they do, they are more likely to do so later in the disease's progression, around the time they approach adolescence. These rarely represent the patient's first presentation. 90% of Moyamoya disease patients originally presented with an ischemic stroke, 7.5% with a transient ischemic attack, and 2.5% with a hemorrhagic stroke, according to a worldwide multicenter stroke database. When Moyamoya disease is first identified, there is a bimodal distribution of age groups; in the pediatric population, the age of initial diagnosis peaks at age five, but in the adult population, the fourth decade of life is when it is most commonly originally diagnosed. In Moyamoya disease, the ratio of male to female adults is about 2:1. Patients with neurofibromatosis type I, sickle cell disease, and Down's syndrome have a greater prevalence of Moyamoya disease. Many of these individuals receive their diagnoses in their late teens, just before they reach maturity. Adults are more likely to have their Moyamoya syndrome diagnosed. (Lee et al., 2017; Berry et al., 2020).

Most cases of Moyamoya disease have been found in East Asia, and Japan, China, Taiwan, and Korea have particularly high yearly incidence rates. It was shown that genetic variables are closely connected with this regional and ethnic trait. The greatest Moyamoya disease vulnerable gene was hypothesized to be the polymorphism of R4810K in the RING (encoded by the Really Interesting New Gene 1) finger protein RNF213, which mediates protein-protein interactions and has ubiquitin-protein ligase activity. The pathogenesis of Moyamoya disease is a complicated route that combines genetic variables, environmental factors, and an intrinsic angiogenetic ability, however, not all Moyamoya disease patients have the RNF213 variation. A widely mentioned susceptibility gene for Moyamoya disease is the RING finger protein RNF213, which has been discovered in family instances and exhibits a significant ethnicity effect. There are several subtypes of Moyamoya disease caused by the RNF213 p.4810K mutation, including the ischemic type and the hemorrhagic type. (Fang et al., 2021).

Brain MRI and MRA show linear hyperintensity throughout the sulci (ivy sign) in bilateral frontomedial with acute infarct in the left frontal region, chronic infarct in right frontomedial, occlusion of bilateral carotid arteries (supraclinoid segment), bilateral MCA and ACA. Digital subtraction angiography showed the stenosis in the right internal carotid artery until T bifurcation with minimal flow to A1 with puff and stenosis in the left M1-M2 junction with puff of smoke appearance. A worsening clinical progression frequently correlates with increasing angiographic stages. While adult populations have a more steady trajectory and stay in a single angiographic stage, pediatric populations more frequently pass through the phases. The best way to see areas of the brain that have experienced acute ischemia or infarction is via an MRI. The "ivy sign" of Moyamoya disease is the emergence of a hyper-intense linear high signal sulcal pattern on the Fluid Attenuation Inversion Recovery (FLAIR) sequence. Significant flow gaps, often known as "termite nests," are seen on MRI images of Moyamoya disease patients' bilateral collateral arteries to the basal ganglia and thalami. This is regarded as almost a Moyamoya disease diagnosis. The DSA is essential for determining which ECA branch will be best for bypassing. There are several circumstances in which a strong occipital artery (OA) might be preferable over a

hypoplastic superficial temporal artery (STA). (Seo et al., 2015).

Smaller blood vessels start to grow as the cerebral vasculature tries to make up for the loss of blood flow by establishing collateral circulation, and hypertrophy becomes increasingly apparent in digital subtraction angiography (DSA). The angiographic appearance of a "puff of smoke," or "Moyamoya" in Japanese, is caused by the development of collateral blood arteries. This is the angiographic trait that distinguishes the condition. Recent investigations reveal that these collateral vessels are really the expansion of preexisting micro-perforator arteries such as the lenticulostriate and thalamoperforating arteries, contrary to what was previously thought. Moyamoya disease almost exclusively affects the anterior circulation, beginning in the distal portions of the intracranial ICA and the proximal portions of the MCA and ACA. As the disease progresses, hypo-perfused brain parenchyma begins to recruit vessels from the leptomeninges and eventually form anastomotic branches of the external carotid artery (ECA).

Moyamoya disease is characterized by bilateral artery angiographic indications of increasing stenosis in the anterior circulation, whereas Moyamoya syndrome is characterized by unilateral abnormalities. A secondary pathological process called Moyamoya syndrome happens as a result of a primary pathological disease. Patients who undergo radiation to the head and neck for cancer may later get substantial radiation-induced ICA stenosis as an illustration of this. These individuals eventually develop Moyamoya syndrome, a condition in which the body tries to make up for ICA stenosis by hypertrophying existing smaller blood channels such as the lenticulostriate perforating arteries. Moyamoya syndrome often involves only one artery, setting it apart from Moyamoya disease. (Berry et al., 2020).

The patient was treated with surgical treatment for direct bypass from the right MCA to the right superficial temporal artery combined with oral medical management single oral antiplatelet. Brain MRI and MRA one month after bypass showed stenosis of supraclinoid segment of bilateral internal carotid arteries, stenosis and irregularity of bilateral middle cerebral arteries, stenosis of bilateral anterior cerebral arteries with multiple collaterals net-like vessels giving puff of smoke appearance. DSA post bypass showed distal occlusion at the supraclinoid segment of bilateral internal carotid arteries, both Pcom gives circulation to the right PCA then through the posterior pericalosal artery to anterior pericalosal artery to the ACA and MCA territories, showed puff of smoke appearance in both bilateral MCA and ACA arteries, there is no anastomosis from the right external carotid artery to the right internal carotid artery. Surgical management for direct bypass failed due to infection in the post-operation site. In a cohort study from Jang et all in South Korea with adult symptomatic Moyamoya disease, bypass surgery reduced stroke recurrence in hemorrhagic Moyamoya disease, but its effectiveness on the prevention of new stroke in ischemic Moyamoya disease is unknown. Regarding mortality reduction, this study failed to show the benefit of bypass surgery compared with medical treatment in adults with Moyamoya disease. (Jang et al., 2017). A meta-analysis study from Yan et all suggesting that revascularization surgery significantly reduces recurrent stroke rates compared with conservative therapy in hemorrhagic-type adult patients with moyamoya. Despite the better angiographic outcomes found in direct bypass compared with that in indirect bypass, no significant differences in stroke prevention and perioperative complications were detected between surgical modalities. (Yan et al., 2019)

Declaration by Authors

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