



# Moyamoya Disease Suzuki Stage III in Adult: Diagnosis and Conservative Management - A Case Report

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## ABSTRACT

**Introduction:** Moyamoya disease (MMD) is a rare, progressive cerebrovascular condition, predisposing patients to ischemic or hemorrhagic strokes—particularly among Asian populations. **Case:** A 38-year-old male with a prior ischemic stroke in 2023 presented with acute left lower limb weakness lasting two days. MRI showed an acute and chronic ischemic lesion, and an acute lacunar infarct in the right superior frontal gyrus. MRA and cerebral angiography confirmed Suzuki stage III MMD, with A1 right anterior cerebral artery (ACA) occlusion and collateral flow from the left posterior cerebral artery (PCA). Cerebral angiography revealed occlusion of the proximal A1 segment of the right ACA and a characteristic “puff of smoke” appearance from abnormal collateral vessels in the distal branches of the right middle cerebral artery (MCA). The patient was managed conservatively with cilostazol, rosuvastatin, and antiplatelet therapy, alongside physical rehabilitation, resulting in functional improvement. **Discussion:** MMD is a progressive vasculopathy that may cause recurrent cerebrovascular events due to impaired cerebral perfusion. Cerebral angiography remains essential for diagnosis and Suzuki staging. Although revascularization surgery is the standard treatment to improve cerebral blood flow and reduce stroke risk, conservative management may be considered in selected patients based on clinical stability, collateral circulation, and treatment indications. In this case, conservative therapy combined with rehabilitation was associated with clinical improvement, although continuous monitoring is required due to the progressive nature of the disease. **Conclusion:** Early diagnosis of MMD through advanced neuroimaging is crucial for preventing recurrent stroke and guiding treatment decisions. This case highlights that individualized conservative management may provide clinical benefits in adult Suzuki stage III MMD when surgical intervention is not immediately performed, while long-term follow-up remains essential.

**Keywords:** Case report, cerebrovascular, Moyamoya disease, Suzuki stage III, ischemic stroke.

## ABSTRAK

**Pendahuluan:** *Moyamoya disease* (MMD) merupakan kondisi serebrovaskular progresif yang jarang terjadi, yang dapat meningkatkan risiko terjadinya stroke iskemik ataupun hemoragik, terutama pada populasi Asia. **Kasus:** Seorang laki-laki berusia 38 tahun dengan riwayat stroke iskemik pada tahun 2023 datang dengan keluhan kelemahan akut pada ekstremitas bawah kiri yang berlangsung selama dua hari. Pemeriksaan *magnetic resonance imaging* (MRI) menunjukkan adanya lesi iskemik akut dan kronis, serta infark lakunar akut pada girus superior frontal kanan. Pemeriksaan *magnetic resonance angiography* (MRA) dan angiografi serebral mengonfirmasi diagnosis MMD Suzuki stadium III, dengan temuan oklusi A1 pada arteri serebri anterior (ACA) kanan dan adanya aliran kolateral dari arteri serebri posterior (PCA) kiri. Angiografi serebral menunjukkan oklusi pada segmen A1 proksimal ACA kanan serta gambaran khas “*puff of smoke*” yang berasal dari pembuluh darah kolateral abnormal pada cabang distal arteri serebri media (MCA) kanan. Pasien mendapatkan tata laksana konservatif berupa *cilostazol*, *rosuvastatin*, terapi *antiplatelet*, serta rehabilitasi fisik, yang menghasilkan perbaikan fungsi klinis. **Pembahasan:** MMD merupakan vaskulopati progresif yang dapat menyebabkan kejadian serebrovaskular berulang akibat gangguan perfusi serebral. Angiografi serebral tetap menjadi pemeriksaan penting dalam menegakkan diagnosis dan menentukan stadium Suzuki. Meskipun tindakan revaskularisasi merupakan terapi standar untuk meningkatkan aliran darah serebral dan menurunkan risiko stroke, tata laksana konservatif dapat dipertimbangkan pada pasien tertentu berdasarkan stabilitas klinis, sirkulasi kolateral, dan indikasi terapi. Pada kasus ini, terapi konservatif yang dikombinasikan dengan rehabilitasi memberikan perbaikan klinis, meskipun pemantauan berkelanjutan tetap diperlukan karena sifat penyakit yang progresif. **Simpulan:** Diagnosis dini MMD melalui pemeriksaan pencitraan lanjutan sangat penting untuk mencegah stroke berulang dan membantu menentukan keputusan terapi. Kasus ini menunjukkan bahwa pendekatan konservatif yang disesuaikan secara individual dapat memberikan manfaat klinis pada pasien dewasa dengan MMD Suzuki stadium III ketika tindakan pembedahan belum dilakukan, namun tindak lanjut jangka panjang tetap diperlukan. **Ni Made Ayu Candrayuni, Marsya Julia Riyadi, I Made Edwin Alberty Wardhana. Penyakit Moyamoya Suzuki Stage III pada Dewasa: Diagnosis dan Tata Laksana Konservatif - Laporan Kasus.**

**Kata Kunci:** Laporan kasus, serebrovaskular, penyakit Moyamoya, stadium Suzuki III, stroke iskemik

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## INTRODUCTION

Moyamoya disease (MMD) is a rare, chronic, and progressive cerebrovascular disorder characterized by stenosis or occlusion of the terminal internal carotid arteries (ICA) and their proximal branches, leading to the formation of fragile collateral vessels around the basal ganglia. These abnormal vessels produce the characteristic “puff of smoke” appearance on cerebral angiography, which is the hallmark of MMD.<sup>1</sup>

MMD is most common in East Asian countries such as Japan, Korea, and China, but cases have increasingly been reported worldwide, including in Indonesia.<sup>2</sup> In Indonesia, awareness of the disease is limited, and diagnosis is often missed because the symptoms resemble those of more common neurological conditions.<sup>2,3</sup> Furthermore, advanced imaging techniques required for diagnosis, such as magnetic resonance angiography (MRA) and digital subtraction angiography (DSA), are not widely available. MMD can affect both children and adults.<sup>4</sup>

The clinical presentation varies but frequently includes ischemic strokes, transient ischemic attacks (TIAs), headaches, seizures, and cognitive impairments. Because these symptoms are often non-specific, delays in diagnosis and treatment are common.<sup>5</sup> Treatment options are also limited by the availability of trained neurosurgeons and surgical facilities, particularly for revascularization procedures, which are considered the primary treatment in progressive cases. Consequently, conservative medical management remains the initial—and sometimes the only—available treatment. This report describes a case of Moyamoya disease in an adult Indonesian patient, highlighting the clinical presentation, imaging findings, and challenges in providing optimal management in a resource-limited setting.

This report presents a case of adult-onset Suzuki stage III MMD in an Indonesian patient with recurrent ischemic stroke who demonstrated clinical improvement following conservative management with medical therapy and rehabilitation. This case highlights the potential role of individualized conservative strategies in selected patients

when surgical intervention is not immediately performed, while emphasizing the importance of early diagnosis and long-term monitoring in progressive cerebrovascular disease.

## CASE

A 38-year-old male presented with a sudden onset of weakness on the left side of his body, which started two days before admission. He reported a sensation of heaviness and dragging of the left leg while walking. His medical history included an ischemic stroke in 2023, with residual weakness and numbness that improved with six months of treatment. The patient also had uncontrolled hyperlipidemia and a notable family history of stroke, with both his father and grandfather of Chinese descent having suffered similar cerebrovascular events.

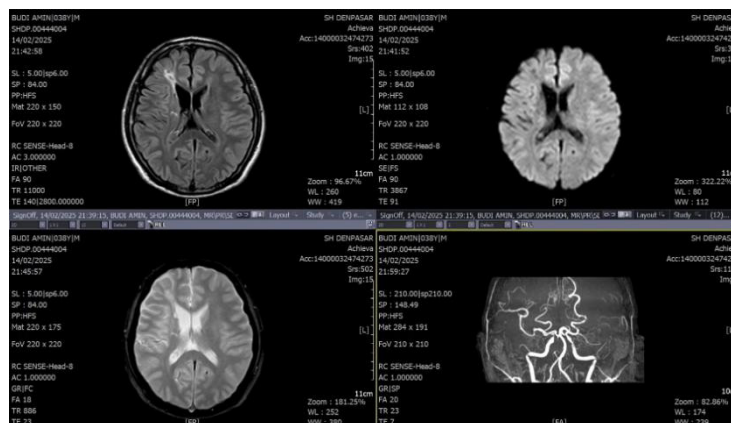
Neurological examination revealed mild weakness (grade 4/5) and paresthesia in the left leg, with preserved reflexes and no cranial nerve deficits. Brain MRI without contrast demonstrated an acute lacunar infarct in the right superior frontal gyrus, along with chronic infarcts in the basal ganglia, corona radiata, and right frontal lobe, consistent with prior ischemic insults. MR angiography showed stenosis of the anterior cerebral artery (ACA), raising suspicion for Moyamoya disease. This was confirmed by cerebral angiography, which revealed occlusion of the proximal A1 segment of the right ACA and a characteristic “puff of smoke” appearance due to abnormal collateral vessels in the distal branches of the right middle cerebral artery (MCA) (**Figure 1 and 2**). Additionally, occlusion was observed

in the distal left internal carotid artery (LICA), with collateral circulation from the left posterior cerebral artery (PCA) supplying the territories of the left ACA and MCA.

During hospitalization, the patient experienced transient worsening of left lower limb weakness, with MRC strength decreasing to 3/5. Conservative management with cilostazol, rosuvastatin, antiplatelet therapy, and physical rehabilitation was initiated. Following rehabilitation, muscle strength improved to MRC 4/5, allowing discharge with planned outpatient follow-up for neurological monitoring and prevention of recurrent cerebrovascular events.

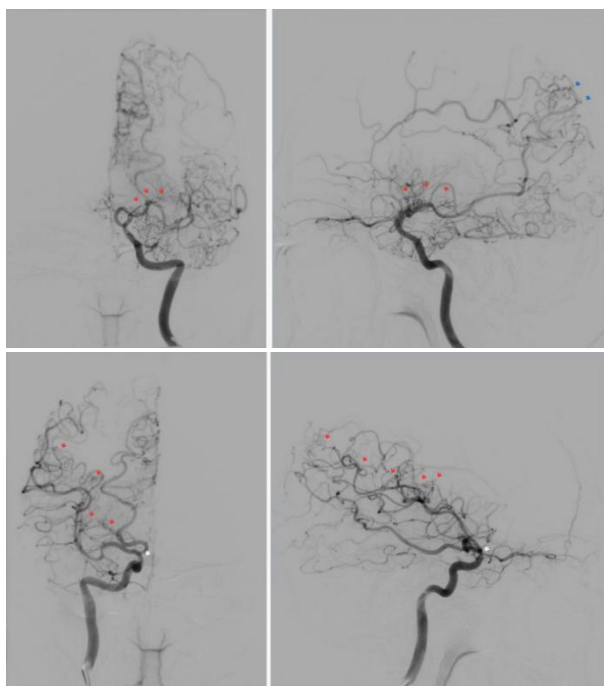
## DISCUSSION

Moyamoya disease (MMD) is a rare, progressive cerebrovascular disorder characterized by stenosis or occlusion of the terminal internal carotid arteries (ICAs) and their proximal branches, followed by the formation of abnormal collateral vessels.<sup>5,6</sup> Although MMD is more frequently reported in East Asian populations, particularly Japan, Korea, and China, increasing recognition of cases in other regions highlights the importance of clinical awareness worldwide.<sup>6</sup> In Indonesia, MMD remains an uncommon but clinically relevant cause of cerebrovascular disease. A recent multicenter Indonesian case series demonstrated that ischemic stroke was the most frequent presentation among patients with MMD, emphasizing the need to consider this diagnosis in patients with atypical or recurrent stroke patterns.<sup>3</sup>



\*Photo documentation by Ni Made Ayu Candrayuni.

**Figure 1.** MRI/MRA head noncontrast showed an acute lacunar infarct in the superior frontal gyrus, with chronic infarcts in the basal ganglia, corona radiata, and right frontal lobe (arrows).



\*Photo documentation by Ni Made Ayu Candrayuni.

**Figure 2.** Cerebral angiography showing occlusion of the proximal A1 segment of the right ACA, with a "puff of smoke" appearance in the distal branches of the right middle cerebral artery (MCA). Occluded LICA with a "puff of smoke" sign in the left deep basal ganglia area. PCA with collateral vessels that supplied the left ACA and MCA (arrows).

The pathogenesis of MMD involves a complex interaction between genetic susceptibility and progressive vascular remodeling. Genetic studies have identified RNF213 as the strongest susceptibility gene for MMD, particularly in East Asian populations.<sup>6,7</sup> The RNF213 gene is thought to contribute to abnormal angiogenesis, endothelial dysfunction, and vascular remodeling, although the exact molecular mechanism remains incompletely understood.<sup>7,8</sup> Familial clustering has also been reported, supporting the contribution of genetic factors despite the absence of a simple Mendelian inheritance pattern.<sup>9</sup> In this case, the patient's family history of stroke involving his father and grandfather of Chinese descent raises concern for a possible hereditary predisposition. Therefore, clinicians should consider genetic risk assessment and screening of first-degree relatives, particularly in families with multiple cerebrovascular events, when appropriate imaging facilities are available.<sup>6,9</sup>

Pathologically, MMD is characterized by progressive narrowing of intracranial arteries due to intimal thickening, smooth muscle cell proliferation, and vascular remodeling,

resulting in chronic cerebral hypoperfusion.<sup>10</sup> In response to reduced cerebral blood flow, fragile collateral networks develop at the basal ganglia region, producing the characteristic "puff of smoke" appearance on cerebral angiography.<sup>11</sup> In this patient, angiography demonstrated occlusion of the proximal A1 segment of the right anterior cerebral artery (ACA), distal left internal carotid artery involvement, and prominent abnormal collateral vessels, confirming the diagnosis of MMD.

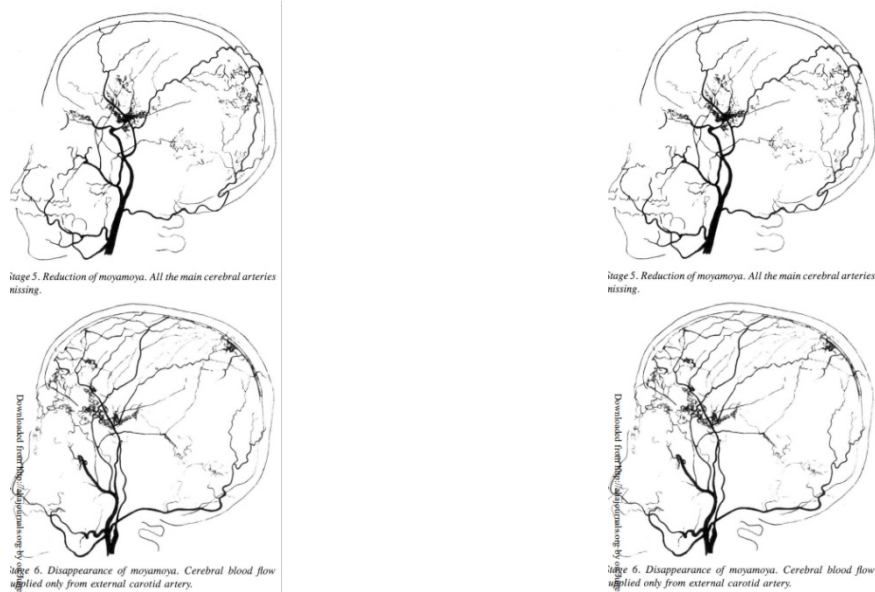
Disease severity in MMD is commonly assessed using the Suzuki and Takaku angiographic classification, which describes progression from initial ICA narrowing to advanced collateral changes and dependence on external carotid artery circulation (**Figure 3**).<sup>12</sup> In the Suzuki stage III, abnormal Moyamoya vessels become prominent due to progressive arterial occlusion and represent the peak phase of collateral formation.<sup>13</sup> In the present case, angiographic findings were consistent with Suzuki stage III, characterized by significant arterial occlusion with well-developed collateral vessels. However, despite collateral

development, these vessels are fragile and inefficient in maintaining adequate cerebral perfusion, contributing to the risk of recurrent ischemic events.<sup>14</sup> This mechanism correlates with the patient's recurrent stroke history and MRI findings showing acute infarction in the right superior frontal gyrus with chronic ischemic changes in the basal ganglia and frontal regions.

The treatment strategy for MMD requires individualized consideration based on clinical presentation, disease severity, and available resources. Surgical revascularization, including direct and indirect bypass procedures, remains the standard treatment for symptomatic MMD because it aims to improve cerebral perfusion and reduce future stroke risk.<sup>12,15</sup> Previous studies have demonstrated favorable outcomes following surgical intervention in reducing recurrent cerebrovascular events among adult patients.<sup>16,17</sup> Given that this patient had Suzuki stage III disease and a history of recurrent ischemic stroke, referral for neurosurgical evaluation and consideration of revascularization should be included in long-term management planning.

However, treatment decisions should consider individual clinical conditions, surgical accessibility, and healthcare resources. In many regions, including parts of Indonesia, limited specialized neurosurgical facilities and limited access to advanced treatments may delay surgical intervention. Therefore, conservative management may be considered as an initial or supportive approach in selected patients while awaiting further evaluation or when surgery is not immediately feasible. In this case, conservative treatment was chosen, consisting of antiplatelet therapy, statin therapy, and rehabilitation, with close clinical monitoring.

Cilostazol was selected as part of the medical management in this patient due to its dual pharmacological effects as an antiplatelet agent and a vasodilator through phosphodiesterase III inhibition. This mechanism may provide potential benefits in patients with cerebral hypoperfusion by reducing platelet aggregation and improving vascular function. Although medical therapy does not reverse the underlying



**Figure 3.** Classification of basal Moyamoya into six stages.<sup>14</sup>

arterial stenosis in MMD, it may help reduce thrombotic risk and support symptom stabilization.<sup>18</sup> In this case, conservative therapy combined with structured rehabilitation resulted in neurological improvement, with muscle strength improving from MRC grade 3/5 during hospitalization to 4/5 after rehabilitation, allowing discharge with outpatient follow-up.

Rehabilitation plays an important role in improving functional outcomes following ischemic stroke associated with MMD. Physical therapy focusing on strength training, gait rehabilitation, and functional mobility may enhance recovery and reduce disability.<sup>18,19</sup> In this patient, rehabilitation contributed to improvement in motor function and supported independence in daily activities. These findings highlight that multidisciplinary

management remains important, particularly when surgical intervention cannot be immediately performed.

Despite clinical improvement, this case has several limitations. First, long-term clinical and radiological follow-up data were limited; therefore, the progression of vascular changes and long-term stroke risk could not be fully assessed. Second, genetic testing, including RNF213 analysis, was not performed despite a significant family history. Future evaluation may provide additional insight into hereditary risk. Third, although conservative management resulted in short-term improvement, it should not replace consideration of revascularization in symptomatic progressive disease.

Overall, this case emphasizes the importance

of recognizing MMD as a potential cause of recurrent stroke in young adults, particularly those with family history or atypical vascular findings. Early vascular imaging, appropriate referral for surgical evaluation, control of vascular risk factors, family counseling, and long-term monitoring are essential components of comprehensive MMD management, especially in resource-limited settings.

## CONCLUSION

Moyamoya disease is a rare, progressive cerebrovascular disorder requiring individualized management. Clinicians in Indonesia should consider MMD in adult stroke patients with atypical features, including young-onset stroke, recurrent ischemic events, family history of stroke, or unexplained/bilateral intracranial arterial stenosis. Advanced vascular imaging with magnetic resonance angiography (MRA) should be considered in these patients to evaluate intracranial vascular abnormalities, while digital subtraction angiography (DSA) remains essential for diagnostic confirmation, Suzuki staging, and surgical revascularization planning. Conservative management with antiplatelet therapy, statins, and rehabilitation may provide clinical benefits in selected patients, as demonstrated in this case of Suzuki stage III MMD. Long-term follow-up, vascular risk factor control, and appropriate family screening are important to reduce future cerebrovascular complications.

## Informed Consent

The patient or a family member has provided written or verbal consent for the publication of the manuscript and all identifiable data.

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