JNeViS
Journal of Neurointervention and Stroke

Journal of Neurointervention and Stroke 2025;1(1):45-48
doi: 10.63937/jnevis-2025.11.10

Case Report

Moyamoya Disease Involving Anterior and Posterior Circulation in Pediatric Ischemic Stroke: Rare Case Report

Subandi Subandi[1], Lothar Matheus Manson Vanende Silalahi[2]

[1] Department of Neurology, Faculty of Medicine, Universitas Sebelas Maret; Dr. Moewardi General Hospital, Surakarta, Indonesia
[2] Faculty of Medicine, Universitas Kristen Duta Wacana, Yogyakarta, Indonesia

Corresponding Author:
Lothar Matheus Manson Vanende Silalahi
Faculty of Medicine, Universitas Kristen Duta Wacana, Yogyakarta, Indonesia
Email: lothar@staff.ukdw.ac.id

Received: March 19, 2025
Revised: April 22, 2025
Accepted: April 24, 2025
Published: May 28, 2025

Introduction: Ischemic stroke is frequently observed in adults but may also occur in children. Moyamoya disease (MMD) is one of the causes of ischemic stroke in children. MMD is a progressive steno-occlusive large-vessel cerebral arteriopathy. MMD was initially defined as changes in the carotid artery with an abnormal vascular network, however, the changes were also noted in posterior circulation. **Case**: We reported the case of a 13-year-old male with right-sided weakness persisting for over 3 years. Magnetic resonance imaging revealed cortical infarction in the left parietal lobe. Digital subtraction angiography (DSA) confirmed moyamoya disease by revealing total occlusion of the bilateral terminal internal carotid artery and bilateral posterior cerebral artery, accompanied by a puff of smoke appearance. We treated this patient with antiplatelet therapy for secondary stroke prevention. **Conclusion:** MMD is one of the conditions that can cause ischemic stroke in children. The posterior circulation is another possible site of MMD manifestation, but it is mostly found in cerebral anterior circulation.

Keywords: Cerebrovascular disease, Digital Subtraction Angiography, Ischemic stroke, Moyamoya disease, Pediatric

Highlights
- Moyamoya disease is one of the causes of stroke in children
- Moyamoya disease typically involves anterior circulation
- Posterior circulation may also be involved in certain cases of Moyamoya disease

Introduction

Stroke is a neurological injury that causes significant morbidity and mortality. The underlying mechanism of stroke, occlusion of cerebral blood vessels, leads to ischemic strokes. Ischemic stroke is more frequent in adults, particularly those with risk factors such as hypertension, diabetes mellitus, and dyslipidemia, which contribute to atherosclerosis. However, ischemic stroke may also occur in pediatric populations due to underlying pathologies that differ from those seen in adults.[1]

Moyamoya disease (MMD) is one of the most common causes of ischemic stroke in children. It has a high prevalence and incidence in East Asia, particularly in Japan, Korea, and China.[2] In children, it mostly presents as progressive cerebral ischemia.[3]

MMD is primarily characterized by progressive narrowing of the internal carotid artery (ICA) bifurcation and the development of moyamoya vessels. However, posterior cerebral artery (PCA) involvement has also been reported.[4] A study found that 20-30% of MMD cases involve the PCA during the disease course.[5] In this case report, we presented a child with clinical presentations of ischemic stroke, which was confirmed as MMD involving both anterior and posterior circulation on digital subtraction angiography (DSA).

Case

A 13-year-old male was referred to our hospital with a three-year history of right-sided weakness, which had caused frequent falls. The weakness progressively worsened over time. There was no history of head trauma, speech or communication disorders, swallowing difficulties, seizures, or loss of consciousness. On physical examination, vital signs were within normal limits. A right hemiparesis with a positive Babinski reflex was found, along with a Medical Research Council (MRC) muscle strength score of 4. Cranial nerve examinations were normal, and no meningeal signs were present. Magnetic resonance imaging (MRI) of the head revealed cortical infarction in the left parietal lobe (Figure 1).

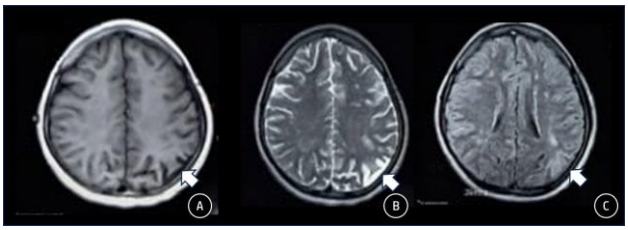

Figure 1. Head MRI image hypointense in T1 (a), hyperintense on T2 (b) and FLAIR (c) showing ischemic lesion in left parietal lobe (arrow).

DSA was then performed, and bilateral occlusion of the terminal branches of the ICA and PCA was found, along with a puff of smoke appearance that suggested moyamoya disease (Figure 2). We treated this patient with antiplatelet therapy for secondary stroke prevention.

Discussion

Pediatric stroke occurs at approximately 1.2 to 13 cases per 100,000 children under the age of 18. Attention to and investigation of the causes of stroke in pediatric patients are important for selecting management strategies and determining the prognosis.[1]

MMD is characterized by chronic occlusion of blood vessels in the circle of Willis. This chronic occlusion triggers the formation of abnormal collateral blood vessels around the affected vessels.[6] Studies have identified two main age groups for MMD onset, which are 5-10 years and 25-49 years[3]. The patient in this case aligns with these findings, presenting with weakness onset at 10 years of age.

The most common clinical manifestation of MMD is a cerebral infarction or hemorrhage. In children and adolescents, cerebral infarction is the most common (70-95% of cases), whereas in adults, cerebral hemorrhage is more frequent (19-40% of cases).[3] Consistent with this epidemiology, the patient in this case came with cerebral infarction, the most common manifestation of MMD in children and adolescents.

MMD can be diagnosed with MRI and MRA of the head, but DSA is the gold standard. According to the 2021 moyamoya diagnostic criteria, cerebral angiography

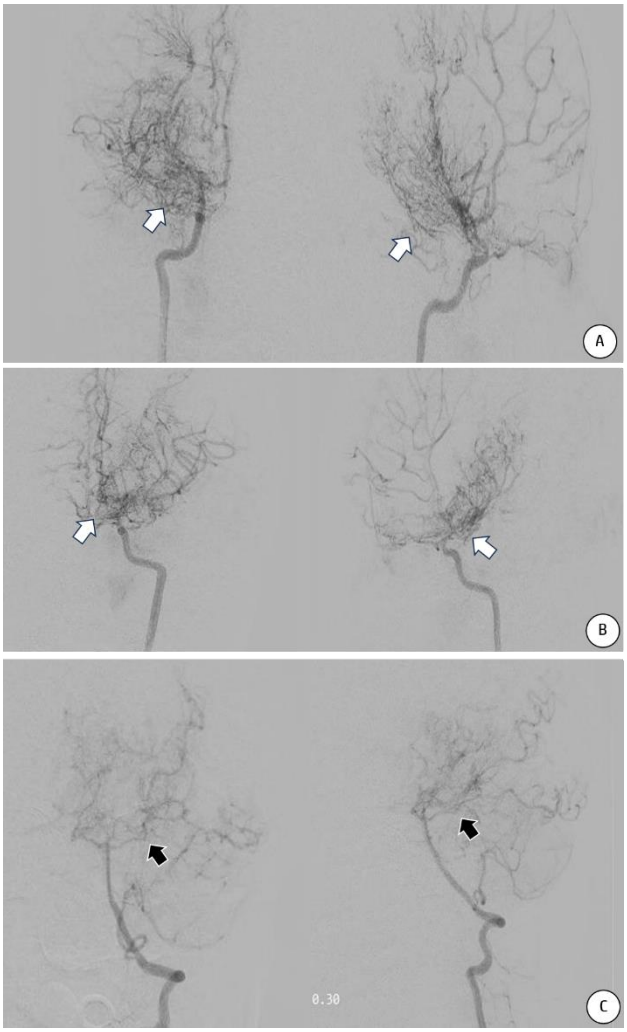

Figure 2. Injection of right ICA (a) and left ICA (b) showing bilateral total occlusion of terminal branches of ICA with puff of smoke appearance (white arrow). Injection of left vertebral artery (c) showing involvement of PCA with puff of smoke appearance (black arrow).

findings must include (i) stenosis or occlusion of arteries centered on the terminal segment of the internal carotid artery and (ii) the appearance of abnormal vascular networks around the occluded or stenotic vessels in the arterial phase.[7] DSA in our patient showed occlusion of the terminal internal carotid arteries and the presence of abnormal vascular networks around the occluded vessels, known as the “puff of smoke,” supporting MMD diagnosis.[8] Based on angiographic findings, MMD is classified into six stages (Table 1).[3] Based on the DSA findings, the patient in this study had stage 2 MMD due to the visible moyamoya collaterals around the narrowed vessels.

The diagnostic criteria for MMD primarily focused on changes in the ICA; however, it's important to note that MMD can also affect the posterior circulation, particularly the PCA.[9] Studies have shown that 20-30% of children diagnosed with MMD initially exhibit PCA stenosis.[5] Various classification systems for MMD have been developed, including one by Mugikura *et al.*, which categorizes angiographic findings related to PCA involvement (Table 2). Mugikura's classification system

indicates that the angiographic findings for the patient described in this case report align PCA stage 3.

Table 1. Angiographic stages of MMD by Suzuki et al[2]

Stage	Findings
1	Narrowing begins at the ICA bifurcation
2	Moyamoya collaterals seen around narrowed vessels
3	Worsening of collateral vessels
4	Exacerbation of narrowed vessels and initial weakening of the collaterals
5	Occlusion of large associated vessels and more pronounced reduction of surrounding moyamoya changes
6	Disappearance of the moyamoya collaterals and vessels of the ICA system, which have come to be supplied by the external carotid artery

Table 2. Angiographic PCA staging of MMD by Mugikura et al[9]

PCA Stage	Angiographic Findings
1	No occlusive changes in the PCA
2	Stenosis in the PCA with or without slightly developed PCA moyamoya
3	Severe stenosis or virtually complete occlusion of the PCA with well-developed PCA moyamoya
4	Occlusion of the PCA with decreased PCA moyamoya

According to the study, a significant factor associated with PCA stenosis in MMD is the presence of an initial infarction, as demonstrated by our patient's head MRI examination.[10] On the other hand, it has been postulated that involvement of the posterior circulation in MMD serve as a source of collateral channels for the anterior circulation.[11]

According to the study, a significant factor associated with PCA stenosis in MMD is the presence of an initial infarction, as demonstrated by our patient's head MRI examination.[10] On the other hand, it has been postulated that involvement of the posterior circulation in MMD serve as a source of collateral channels for the anterior circulation.[11]

Conservative and surgical approaches are currently the two main treatment options for moyamoya disease (MMD). However, there is no consensus on the optimal management strategy due to a lack of strong evidence comparing the long-term outcomes of these treatment options.[3] No pharmacotherapy can inhibit the progression of vascular disorders, even in asymptomatic conditions. Antiplatelet and anticoagulant medications can be used to prevent secondary stroke.[3,12]

Recent studies increasingly recommendsurgical intervention for the treatment of MMD, as conservative treatment has not been proven effective. Surgical intervention may be considered when cerebral hemodynamics are impaired, as indicated by angiographic findings in the Suzuki classification. The progressive nature of MMD and its potential to cause significant neurological disabilities, especially in children, have led to a growing preference for surgical approaches.[13]

However, surgery for MMD is still controversial, especially when it comes to perioperative complications.[2] A previous study found that rapid disease progression occurred in patients under three years of age; therefore, early surgical intervention is necessary without delay in this age group. In contrast, the urgency for surgical intervention may be lower in older children due to slower disease progression. Perioperative complication rates also appear to be lower when surgery is performed within three months of symptom onset.[14] In our case, however, the patient's symptoms began 3 years prior. The longer duration in this case may make conservative management more appropriate. The decision to treat this patient conservatively is also supported by the results of a study that found that reduction of the stroke risk by surgery is most striking in MMD presenting with hemorrhagic stroke but not in ischemic stroke.[13] MMD in children who are treated conservatively still had overall good neurological and functional outcomes.[15]

Conclusion

MMD causes ischemic strokes in children. DSA is the gold standard for diagnosing MMD. MMD primarily involves the anterior circulation but, in some cases, can also involve both the anterior and posterior circulation. The treatment of MMD can include medications and surgery. Surgery can be considered, but several factors must be taken into account, including age, MMD progression, and the possibility of perioperative complications. This case report provides clinicians worldwide with valuable insight into the diverse manifestations of MMD in children, as well as management options.

Acknowledgement
We sincerely appreciate the patients who volunteered to participate in our educational study

Conflict of Interest
All authors have no conflict of interest.

Patient consent for publication
Written informed consent for publication was obtained from the parent.

Funding
None

Author contribution
Subandi: Conceptualization, Methodology, Writing-Review and Editing. **Lothar Matheus Manson Vanende Silalahi:** Writing-Original Draft, Writing-Review and Editing.

References

1. Rawanduzy CA, Earl E, Mayer G, Lucke-Wold B. Pediatric stroke: a review of common etiologies and management strategies. Biomedicines. 2022; 11(1):2. DOI: 10.3390/biomedicines110100 02
2. Zhang H, Zheng L, Feng L. Epidemiology, diagnosis and treatment of moyamoya disease. Exp Ther Med. 2019;17(3):1977-84. DOI: 10.3892/etm.2019.7198
3. Demartini Jr Z, Teixeira BC, Koppe GL, Gatto LA, Roman A, Munhoz RP. Moyamoya disease and syndrome: a review. Radiologia brasileira. 2022; 55:31-7. DOI:10.1590/0100-3984.2021.00 10
4. Zhang Q, Zhang D, Wang R, Liu Y, Zhang Y, Wang S, et al. Clinical and Angiographic Features of Patients with Moyamoya Disease and the p.R4810K Heterozygous Variant. World Neurosurg. 2016;90:530-538. DOI: 10.1016/j.wneu.2015.12.093
5. Lee JY, Kim SK, Phi JH, Wang KC. Posterior cerebral artery insufficiency in pediatric moyamoya disease. Journal of Korean Neurosurgical Society. 2015;57(6):436-9. DOI: 10.3340/jkns.2015.57.6.436
6. Irhadi MuhF, Rachman ME, Kaelan C. Literature Review : Relationship Between Moyamoya Disease And Cerebral Iscemic Incidents. Jurnal Eduehelath. 2024;15:2024. Available at: https://ejournal.seaninstitute.or.id/index.php/healt/article/view/5405
7. Kuroda S, Fujimura M, Takahashi J, Kataoka H, Ogasawara K, Iwama T, et al. Diagnostic Criteria for Moyamoya Disease-2021 Revised Version. Neurol Med Chir (Tokyo). 2022;62(7):307–12. DOI: 10.2176/jns-nmc.2022-0072
8. Goswami M, Pandian R, Sharma S. Moyamoya disease—"A puff of smoke": A rare pediatric case report. Int J Clin Pediatr Dent. 2020;13(5):566–8. DOI: 10.5005/jp-journals-10005-1790
9. Bao XY, Tong HY, Wang QN, Wang XP, Gao G, Zhang Q, et al. A long-term study of posterior circulation changes after revascularization in patients with moyamoya disease. J Neurosurg. 2023;139(5):1281–6. DOI: 10.3171/2023.2.jns222649
10. Lee JY, Kim SK, Cheon JE, Choi JW, Phi JH, Kim IO, et al. Posterior cerebral artery involvement in moyamoya disease: Initial infarction and angle between PCA and basilar artery. Child's Nervous System. 2013;29(12):2263–9. DOI: 10.1007/s00381-013-2123-7
11. Hori S, Kashiwazaki D, Yamamoto S, Acker G, Czabanka M, Akioka N, et al. Impact of Interethnic Difference of Collateral Angioarchitectures on Prevalence of Hemorrhagic Stroke in Moyamoya Disease. Clin Neurosurg. 2019;85(1):134–45. DOI: 10.1093/neuros/nyy236
12. Dlamini N, Muthusami P, Amlie-Lefond C. Childhood Moyamoya: Looking Back to the Future. Vol. 91, Pediatric Neurology. Elsevier Inc.; 2019. p. 11–9. DOI: 10.1016/j.pediatrneurol.2018.10.006
13. Wouters A, Smets I, Van den Noortgate W, Steinberg GK, Lemmens R. Cerebrovascular events after surgery versus conservative therapy for moyamoya disease: a meta-analysis. Vol. 119, Acta Neurologica Belgica. Springer-Verlag Italia s.r.l.; 2019. p. 305–13. DOI: 10.1007/s13760-019-01165-9
14. Hayashi T, Kimiwada T, Karibe H, Shirane R, Sasaki T, Metoki H, et al. Preoperative Risks of Cerebral Infarction in Pediatric Moyamoya Disease. Stroke. 2021;52(7):2302–10. DOI: 10.1161/strokeaha.120.032699
15. Lim WK, Ong LC, Tan KA, Li L, Teh CM, Heng HS, et al. Clinical features and outcomes of paediatric moyamoya vasculopathy in Malaysia. Neurol Asia. 2022;27(3):617–27. DOI: 10.54029/2022pvt