Biological and Clinical Sciences Research Journal
eISSN: 2708-2261; pISSN: 2958-4728
www.bcsrj.com
DOI: https://doi.org/10.54112/bcsrj.v6i1.1523
*Biol. Clin. Sci. Res. J., Volume 6(1), **2025**: 1523*
Case Report

Moyamoya Disease: Acute presentation with fever and fits in a 5-Year-Old Female Patient: A Case Report

Muhammad Saad*[1], Aimen Yaseen[2], Faiqa Shafiq[3]

[1]khyber Medical University Peshawar, Pakistan
[2]department Neurology Sheikh Zayed Hospital Rahim Yar Khan, Pakistan
[3]Indus Hospital & Health Network – Al-Ghazi Campus, Bhong, Pakistan
**Corresponding author`s email address:* Muhmmadsaad460@gmail.com

(Received, 24th November 2024, Revised 9th January 2025, Published 31st January 2025)

Abstract: *Moyamoya disease is a rare cerebrovascular disorder characterised by the narrowing or blockage of the carotid artery, leading to reduced blood flow to the brain. I present the case of a 5-year-old female patient who experienced recurrent strokes and was diagnosed with Moyamoya disease. The patient exhibited a complex clinical course involving episodes of stroke, fever, and seizures. Detailed examinations, diagnostic tests, and medical interventions were conducted to manage her condition. This case underscores the importance of recognising Moyamoya disease in pediatric patients presenting with recurrent strokes and highlights the challenges associated with its diagnosis and management.*

Keywords: Moyamoya, recurrent stroke, fever, seizures

[***How to Cite:*** Saad M, Yaseen A, Shafiq F. Moyamoya disease: acute presentation with fever and fits in a 5-year-old female patient: a case report. *Biol. Clin. Sci. Res. J.*, **2025;** 6(1): 88-89. doi: https://doi.org/10.54112/bcsrj.v6i1.1523

Introduction

Moyamoya disease is a progressive obstructive condition affecting cerebral vessels, mainly characterised by the narrowing of distal internal carotid arteries on both sides and the anterior and middle cerebral arteries. (1). This results in the formation of collateral vessels to compensate for the blockage (2). The disease gets its name from Japanese, translating to 'puff of smoke (3). Although initially perceived as predominantly impacting individuals of Asian heritage, it is now observed globally across various ethnic backgrounds. The reported incidence stands at 0.086 per 100,000 population. While it is linked to conditions like Sickle cell disease, Down's syndrome, and Neurofibromatosis-1, it is important to note that there are almost twice as many female patients as male patients (2). It manifests two age peaks: around 5 years in children, often associated with strokes or transient ischemic attacks, and around 40 years in adults, typically marked by haemorrhages (4). The treatment strategies for MMD involve both pharmacological and surgical approaches for stroke prevention alongside symptomatic medications (5). "Moyamoya disease" (MMD) and "Moyamoya syndrome" (MMS) are cerebrovascular conditions affecting specific arteries. While MMD indicates a genetic predisposition with bilateral angiographic findings, MMS encompasses various conditions triggering vasculopathy, often with unilateral angiographic features and associated risk factors (6).

A 5-year-old female was brought to the hospital with a history of recurrent strokes and fever. She had experienced two episodes of stroke, initially presenting with left-sided weakness followed by right-sided weakness. Seizure-like episodes were also noted. She had been previously admitted to different hospitals for a total of 15 days due to her condition. The patient also had a history of fever with fits.

Birth History: She was born through expected vaginal delivery with an immediate cry. No complications were reported during the birth process.
Family History: She is the fifth child of her parents in a consanguineous marriage.
Developmental History: The patient exhibited developmental delays and speech problems.
Vaccination History: She was vaccinated according to the EPI schedule.
Dietary History: She had everyday dietary habits.
Allergic History: She had no known allergies.

Physical examination:
Vitals: Normal
Weight: 16.2 Kg
Cardiovascular examination: She had a systolic murmur.
Respiratory examination: She had clear bilateral lung fields.
Abdominal examination: Her abdomen was soft, non-distended and non-tender.
Central Nervous System examination: Her sensory and motor systems were intact.
Initial Evaluation:
Following the initial workup was done.
Anti DS DNA: Negative
Coagulation profile: This included:
Bleeding time: 2:00 minutes
Prothrombin time: 12.5 seconds
INR: 1.04
APTT: 32 seconds
Echocardiogram: Normal
Electroencephalogram: It showed electrographic seizure activity with focal epileptiform discharges.
Confirmatory Evaluation:
CT Cerebral Angiography: Showed sequelae of previous ischemic insult in left frontal and right parietal lobes. It also showed significant lamina narrowing of supra-clinoid portions of bilateral internal carotid arteries with collateral formation along the bilateral cerebral hemisphere, which suggests Moyamoya disease.
Diagnosis:
Based on CT findings, a diagnosis of recurrent stroke due to Moyamoya disease was made.
Management:
Strict vital monitoring was maintained throughout the patient's hospitalisation. She was commenced on the antibiotic Oxidil (Ceftriaxone Na). She was also given Syrup Epival (Sodium Valproate) 5 ml twice daily and Tablet Ascard (Aspirin) 25 mg once daily. With no active complaints, she was considered clinically and vitally stable. She was discharged with home medications, which included Syrup Epival and Tablet Ascard, which she had to use for a long time. She was also given

Syrup Cefspan DS (Cefexime), one teaspoon once daily for 5 days. She has called for a follow-up after one month.

Discussion

Moyamoya disease (MMD) is an unusual condition characterised by the slow and progressive narrowing of the supraclinoid part of the internal carotid artery, commonly affecting both the middle and anterior cerebral arteries (7). Originating in 1957 as "hypoplasia of the bilateral internal carotid arteries" and later defined as a "hemangiomatous malformation of bilateral internal carotid arteries at the base of the brain" in 1968, the term "moyamoya disease" was coined in 1969 to describe the hazy appearance of the vascular collateral network on angiography. Despite extensive reviews and guidelines spanning 50 years, the fundamental features persist: bilateral stenosis in intracranial arteries and the development of collateral vessels. Recent studies, especially in the last decade, have deepened our insights into moyamoya disease, yet the driving force behind ongoing research remains unclear (8).

Conclusion

In conclusion, the presented case highlights the importance of recognising Moyamoya disease in pediatric patients with recurrent strokes. A comprehensive, multidisciplinary approach involving detailed examinations and diagnostic assessments is essential for accurate and timely management. The discussion covers various aspects of Moyamoya disease, including its epidemiology, aetiology, pathophysiology, clinical presentation, diagnosis, treatment options, outcomes, prognosis, and differential diagnosis. Surgical interventions, particularly direct bypass, appear promising in preventing subsequent strokes, emphasising the need for ongoing research to enhance our understanding of this intricate cerebrovascular disorder.

Declarations

Data Availability statement
All data generated or analysed during the study are included in the manuscript.
Ethics approval and consent to participate
Approved by the department concerned.
Consent for publication
Approved
Funding
Not applicable

Conflict of interest

The authors declared the absence of a conflict of interest.

Author Contribution

All Authors contributed equally

All authors reviewed the results and approved the final version of the manuscript. They are also accountable for the integrity of the study.

References

1. Burke GM, Burke AM, Sharma AK, et al. Moyamoya disease: a summary. Neurosurg Focus 2009; 26: 1–10.
2. Gudepu RK, Qureshi MA, Qureshi IA, et al. Case Report: A case report of Moyamoya disease in a 36-year-old African American woman. F1000Res; 3. Epub ahead of print 8 December 2014. DOI: 10.12688/F1000RESEARCH.5859.1.
3. Tarasów E, Kułakowska A, Łukasiewicz A, et al. Moyamoya disease: Diagnostic imaging. Pol J Radiol 2011; 76: 73.
4. Azzam DB, Sharma AN, Tiourin E, et al. A Case Report of Moyamoya Disease Presenting as Headache in a 35-year-old Hispanic Man. Cureus; 11. Epub ahead of print 10 April 2019. DOI: 10.7759/CUREUS.4426.
5. Canavero I, Vetrano IG, Zedde M, et al. Clinical Management of Moyamoya Patients. J Clin Med 2021; 10: 3628.
6. Manorenj S, Shaik RS, Manorenj S, et al. Clinical Aspects of Moyamoya Disease. Moyamoya Disease - A Disease to Count On in Your Daily Practice. Epub ahead of print 26 February 2021. DOI: 10.5772/INTECHOPEN.96322.
7. Mamadaliev D, Feletti A, Mewada T, et al. Moyamoya Disease, Basic Concepts of Diagnostics, and Treatment. Neurovascular Surgery 2019; 223–232.
8. Kuribara T, Akiyama Y, Mikami T, et al. Macrohistory of Moyamoya Disease Analyzed Using Artificial Intelligence. Cerebrovascular Diseases 2022; 51: 413–426.
9. Roder C, Nayak NR, Khan N, et al. Genetics of Moyamoya disease. Journal of Human Genetics 2010 55:11 2010; 55: 711–716.
10. Zhang X, Xiao W, Zhang Q, et al. Progression in Moyamoya Disease: Clinical Features, Neuroimaging Evaluation, and Treatment. Curr Neuropharmacol 2022; 20: 292.
11. Rupareliya C, Lui F. Moyamoya Disease. StatPearls, https://www.ncbi.nlm.nih.gov/books/NBK535455/ (2023, accessed 31 December 2023).
12. Kuroda S, Houkin K. Moyamoya disease: current concepts and future perspectives. Lancet Neurol 2008; 7: 1056–1066.
13. Estmann A, Toftedal P, Schou A. Moyamoya disease: an important differential diagnosis for cerebral vasculitis. Pediatr Rheumatol Online J 2011; 9: P85.
14. Zhang H, Zheng L, Feng L. Epidemiology, diagnosis and treatment of moyamoya disease. Exp Ther Med 2019; 17: 1977.
15. Gonzalez NR, Amin-Hanjani S, Bang OY, et al. Adult Moyamoya Disease and Syndrome: Current Perspectives and Future Directions: A Scientific Statement From the American Heart Association/American Stroke Association. Stroke 2023; 54: E465–E479.
16. Bersano A, Guey S, Bedini G, et al. Research Progresses in Understanding the Pathophysiology of Moyamoya Disease. Cerebrovascular Diseases 2016; 41: 105–118.
17. Berry JA, Cortez V, Toor H, et al. Moyamoya: An Update and Review. Epub ahead of print 2020. DOI: 10.7759/cureus.10994.
18. Mertens R, Graupera M, Gerhardt H, et al. The Genetic Basis of Moyamoya Disease. Transl Stroke Res 2022; 13: 25.
19. Demartini Z, Teixeira BCA, Koppe GL, et al. Moyamoya disease and syndrome: a review. Radiol Bras 2022; 55: 31–37.
20. Kim JS. Moyamoya Disease: Epidemiology, Clinical Features, and Diagnosis. J Stroke 2016; 18: 2–11.
21. Mehndiratta MM, Goyal I, Aggarwal V, et al. Moyamoya Disease Worldwide-Global Burden East and West. Moyamoya Disease - A Disease to Count On in Your Daily Practice. Epub ahead of print 4 March 2021. DOI: 10.5772/INTECHOPEN.96137.
22. Guey S, Tournier-Lasserve E, Hervé D, et al. Moyamoya disease and syndromes: From genetics to clinical management. Application of Clinical Genetics 2015; 8: 49–68.
23. Kim T, Oh CW, Bang JS, et al. Moyamoya Disease: Treatment and Outcomes. J Stroke 2016; 18: 21.
24. Gupta A, Tyagi A, Romo M, et al. Moyamoya Disease: A Review of Current Literature. Cureus; 12. Epub ahead of print 30 August 2020. DOI: 10.7759/CUREUS.10141.