JACEP OPEN
JOURNAL OF THE AMERICAN COLLEGE OF EMERGENCY PHYSICIANS OPEN
Open Access
A GLOBAL JOURNAL OF EMERGENCY MEDICINE
American College of Emergency Physicians®
ADVANCING EMERGENCY CARE

CASE REPORT

Neurology

Check for updates

Moyamoya Disease- A Clinical Mimic for Psychiatric Disorders in the Emergency Setting: A Case Report

Daniel F. Leiva DO[1], Katie J. Arey PA[2], William E. Soares III MD, MS[2]

[1]*Department of Emergency Medicine, Cedars Sinai Medical Center, Los Angeles, California, USA*
[2]*Department of Emergency Medicine, University of Massachusetts Medical School –Baystate, Springfield, Massachusetts, USA*

Correspondence

Daniel F. Levia, DO, Department of Emergency Medicine, Cedars Sinai Medical Center, Los Angeles, CA 90048, USA. Email: dfleiva@gmail.com

Received: March 16, 2023
Revised: June 4, 2024
Accepted: June 17, 2024
https://doi.org/10.1016/j.acepjo.2025.100061

Abstract

Patient presentations to the emergency department for mental health evaluations are common, with an estimated 1 in 4 adult visits made for this reason. These presentations are often accompanied by vague symptomatology, which may make it difficult to determine if they are because of another medical condition. Previous evaluations may bias future presentations, leading to premature closure before correctly identifying a causative underlying medical condition. Accurate, timely diagnosis improves health care costs by decreasing inappropriate treatments and unnecessary admissions and lowering the risk of recidivism. A 32-year-old woman presented with a complaint of recurrent neuropsychological symptoms attributed incorrectly to a primary mental disorder. On representation, she was found on computed tomography angiogram imaging to have pathognomonic findings for moyamoya disease. A short review of neuropsychological presentations previously attributed to moyamoya disease is reviewed. Because of the frequency with which we encounter patients for a mental health evaluation and the multifaceted harms of misdiagnosis, emergency providers should be familiar with moyamoya disease as a cause of mental disorders due to another medical condition.

Keywords: *moyamoya, medical mimic, secondary psychosis, cerebrovascular accident, stroke*

1 INTRODUCTION

Mental health evaluations (MHEs) for a mental disorder, as defined in the Diagnostic and Statistical Manual of Mental Disorders, including psychiatric or behavioral health issues, constitute a large number of emergency department (ED) visits each year, with an estimated 1 in 4 adult visits made for an MHE.[1] Substance use or homelessness in these patients further increases ED utilization, recidivism, and admission rates.[2,3] These trends have been seen in adult and pediatric populations both domestically and internationally.[4,5] It is important to diagnose mental disorders correctly as misdiagnosis can lead to repeat ED visits, unnecessary or harmful treatments, increased length of stay, unnecessary hospitalizations, and increased health care expenditures. We present a case of an

Supervising Editor: Bernard Chang, MD, PhD

incorrectly diagnosed mental disorder who was later identified as possibly having a secondary mental disorder due to primary moyamoya disease. We will also review other neuropsychological mimics of this disease previously identified in the literature.

2 CASE REPORT

A 32-year-old Hispanic woman with a history of schizoaffective disorder with depression vs depression with psychotic symptoms presented to an urban academic ED with a chief complaint of altered mental status. The patient was confused and unable to offer any pertinent information during the evaluation. The patient's significant other (SO) stated that she had become increasingly confused over the past 3 days with difficulty recognizing familiar faces. The patient had multiple recent falls and complained frequently of headaches and dizziness. The morning of her presentation, the patient stated, "I'm not connected to my body," and refused to take her medications. The SO denied any recent fever, cough, abdominal pain, vomiting, diarrhea, or urinary complaints. The patient had no recent medication changes and no known alcohol or illicit drug use.

The patient had a history of depression and auditory hallucinations diagnosed at age 30 years, but before this was previously healthy and without mental disorders. At the onset of her psychiatric disease, a magnetic resonance imaging (MRI) of the brain without contrast was performed, which demonstrated minimal subcortical white matter abnormalities in the right frontal lobe that were felt to be of doubtful significance. More recently, her primary care physician noted that she had become less interactive, that her affect was often flat, and that she was an increasingly poor historian over the past year. She was admitted twice through the ED to inpatient psychiatric facilities for altered mental status and decompensated psychiatric disease due to medication noncompliance. Home visiting nurse services were also recently started to assist with activities of daily living and medication administration due to severe psychiatric illness.

A physical examination revealed a moderately obese female in no acute distress. Vital signs on arrival were an oral temperature of 36.8 °C (98.2 °F), heart rate of 87 beats/min, blood pressure of 140/63 mm Hg, respiration of 18 breaths/min, and oxygen saturation of 96% on room air. The patient was awake but confused, mumbling words and localizing pain, with a Glasgow Coma Scale of 13. There was no evidence of external head trauma. Pupils were equal, round, and reactive to light. The skin was warm and dry without evidence of rash or infection. Her cardiovascular, pulmonary, and abdominal examinations were unremarkable. The neurologic examination demonstrated an National Institute of Health Stroke Scale/Score of 9 with an incorrect question/answer pair, partial gaze palsy, right-leg drift, decreased sensation, and right-sided hemineglect, as evidenced by responding to a Spanish interpreter on the left while ignoring the emergency physician and her SO on her right. In addition, the patient held her right arm in spastic flexion with a full range of motion of the left upper and lower extremities. Ambulation testing was deferred, given the patient's presentation.

Blood testing demonstrated a minimal leukocytosis of 11.9 k/mm^3, hemoglobin of 12.5 g/dL, and platelet count of 411 k/mm^3. Her chemistry testing demonstrated sodium of 139 mmol/L, potassium of 4.7 mmol/L, chloride of 100 mmol/L, bicarbonate of 26 mmol/L with an anion gap of 13, and glucose of 100 mg/dL. Her blood ethanol level was 0, and a urinalysis with a toxicology screen was unremarkable. Her pregnancy testing was also negative. Her thyroid-stimulating hormone was normal at 0.55 mIU/mL.

A computed tomography (CT) of the head without contrast demonstrated a subacute infarct of the left occipital lobe with additional remote left frontal and left frontoparietal infarcts. A CT angiogram of the head and neck demonstrated multifocal stenosis involving the supraclinoid internal carotid arteries and proximal anterior and middle cerebral arteries (Fig 1). The patient was admitted and received consultations from neurology, rehabilitation medicine, and psychiatry. An MRI of the brain was performed 48 hours later, demonstrating a subacute left parasagittal watershed infarct (Fig 2). She was diagnosed with multiple ischemic cerebral infarcts secondary to moyamoya disease, which was thought to be the underlying etiology of her previously diagnosed mental disorder. She was discharged to a rehabilitation facility in good condition.

3 DISCUSSION

Moyamoya disease is a chronic cerebrovascular disease characterized by stenosis and occlusion of the carotid arteries and large vessels of the Circle of Willis, including the cerebral arteries. This leads to the development of collateral, small arteries, giving it a pathognomonic hazy appearance on angiography imaging, including CT and MRI studies. There is evidence to suggest that these vascular abnormalities can be

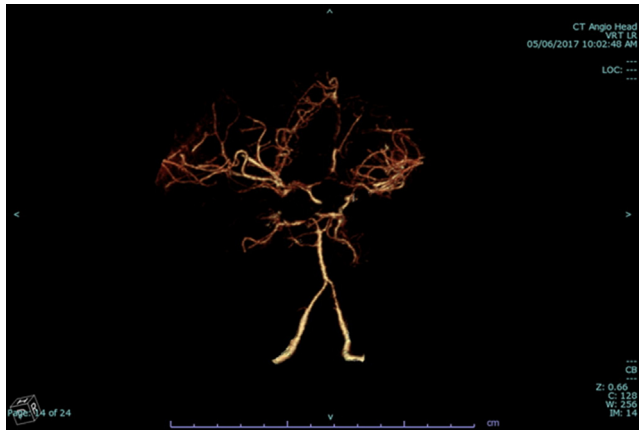

FIGURE 1. Multifocal stenoses involving the supraclinoid internal carotid arteries with a plexus of tiny collateral vessels in the region of the middle cerebral artery bilaterally. CT, computed tomography.

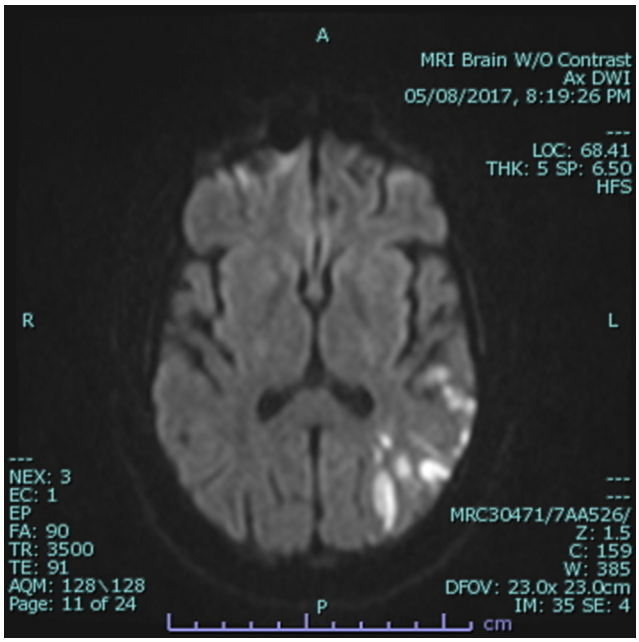

FIGURE 2. Subacute left parasagittal watershed infarct. MRI, magnetic resonance imaging.

congenital in origin but may also be acquired due to conditions including vasculitides.[6–8] Epidemiologic data have shown an increased incidence in the Asian population, with 15% of patients demonstrating familial occurrence (both autosomal dominant with incomplete penetrance and x-linked recessive patterns have been identified). There has also been found an increased incidence in females with a bimodal age distribution at 5 to 10 years old and 35 to 45 years old.[9] Neurocognitive impairments can be dramatic, particularly in the pediatric population.[10–12] Quality of life can also be severely affected by sequelae of the disease process in all age groups.[13]

Commonly identified complaints include headache, dizziness, and visual disturbances.[14–16] Examination findings may include difficulty speaking, extremity weakness or numbness, and abnormal limb movements (including unilateral or bilateral dyskinetic, choreiform, or dystonic movements).[17–19] Alterations in mental status from mild confusion to complete obtundation have also been seen.[20] Patients may also present with cerebrovascular syndromes or recurrent or refractive seizures, including febrile infection-related epilepsy syndrome due to the abnormal vasculature.[21–23] Attention should be paid to intractable or worsening headaches, especially if accompanied by or with a history of other transient or persistent neurologic symptoms.[16,24,25] Due to the sometimes transient nature of symptoms, misidentification due to other conditions like multiple sclerosis has also occurred.[26]

Patients who present for an MHE should be carefully examined to determine if their complaint and presentation is a primary or secondary process due to another medical condition, historically described as a functional vs organic psychosis. Differentiating between a primary and secondary process can be challenging due to the lack of specific testing for primary mental disorders. For this reason, patients may inadvertently be given a diagnosis of a treatment-resistant primary mental disorder when it may be secondary to an underlying condition like moyamoya. Due to the prevalence of mental disorders, not due to another medical condition, it is difficult to differentiate causation from correlation. Regardless, stroke syndromes have previously been found to be a causative factor in various mood disturbances not present prior to the vascular event.[27–29]

One of the earliest cases documented of moyamoya masquerading as a primary mental disorder was in 1981 in an 11-year-old male patient.[30] This patient had prodromal symptoms as described above, including the presence of a fever and abnormal movements of the extremities. This progressed to violent irritability accompanied by incoherent speech. The presence of other positive, negative, and cognitive schizophrenic features was also identified. Another case in 1991 describes a 19-year-old male with delusions and hallucinations in addition to a proceeding history of learning difficulties and transient hemiplegia, highlighting the importance of identifying transient neurologic complaints.[31] Other cases of psychosis with varying degrees of positive and negative symptoms have since been reported. A review of the literature has demonstrated other psychiatric manifestations in patients with moyamoya disease, including:

- Neurodevelopmental disorders, including intellectual disabilities, attention deficit hyperactivity disorder, and Tourette's;[32,33]
- Bipolar-related disorders characterized by mania and irritability with psychosis;[27]
- Depressive disorders;[34] and
- Anxiety disorders.[35]

Abnormal pathognomonic brain findings include areas of infarct, in particular patterns on CT and MRI, stenotic lesions with collateral formation on CT and MR angiography studies, and the "rebuild-up" phenomenon on electroencephalogram.[36–40] Treatment is primarily symptomatic in nature, including practices seen with other stroke syndromes (eg, resuscitation, optimization of cerebral perfusion pressure, and seizure control). Thrombolytic therapy has not been studied in patients with moyamoya disease. Consultation with our neurosurgical colleagues should be considered, as vascular bypass has previously been used in these patients with success. In addition to surgical revascularization, antithrombotic therapy may be utilized to decrease the risk of future ischemic events. Identification of patients with moyamoya can help providers be vigilant for significant sequelae, including stroke and acute coronary syndromes.[41,42]

Our patient was noted to have had headaches, dizziness, and left arm numbness before her first MRI. She also had progressive decompensated mental disorder presentations. Earlier evaluations may have biased future ED presentations, leading to premature closure and attribution of the current presentation to

a nonmedical cause of decompensation, including medication. Timely recognition can lead to early intervention, decreasing the risk of developing sequelae, including stroke syndromes, and may lead to improvement or resolution of secondary symptoms in addition to positive changes for long-term neurocognitive and psychosocial development.[33,43,44]

FUNDING AND SUPPORT

Dr Leiva receives funding from an NIH T32 grant (5T32HL120823-5). Dr Soares receives funding from an NIH K08 grant (5K08DA045933-02).

CONFLICT OF INTEREST

All authors have affirmed they have no conflicts of interest to declare.

ORCID

Daniel F. Leiva DO https://orcid.org/0000-0003-2745-5411

Daniel F. Leiva DO https://twitter.com/DanielFLeiva
William E. Soares III MD, MS https://twitter.com/BillSoaresIII

REFERENCES

1. Capp R, Hardy R, Lindrooth R, Wiler J. National trends in emergency department visits by adults with mental health disorders. *J Emerg Med*. 2016;51(2):131-135.e1. http://doi.org/10.1016/j.jemermed.2016.05.002
2. Lam CN, Arora S, Menchine M. Increased 30-day emergency department revisits among homeless patients with mental health conditions. *West J Emerg Med*. 2016;17(5):607-612. http://doi.org/10.5811/westjem.2016.6.30690
3. Azar KMJ, Petersen JP, Shen Z, et al. Serious mental illness and health-related factors associated with regional emergency department utilization. *Popul Health Manag*. 2020;23(6):430-437. http://doi.org/10.1089/pop.2019.0161
4. Shafiei T, Gaynor N, Farrell G. The characteristics, management and outcomes of people identified with mental health issues in an emergency department, Melbourne, Australia. *J Psychiatr Ment Health Nurs*. 2011;18(1):9-16. http://doi.org/10.1111/j.1365-2850.2010.01632.x
5. Hoffmann JA, Stack AM, Samnaliev M, Monuteaux MC, Lee LK. Trends in visits and costs for mental health emergencies in a pediatric emergency department, 2010–2016. *Acad Pediatr*. 2019;19(4):386-393. http://doi.org/10.1016/j.acap.2019.02.006
6. Shiari R, Tabatabaei Nodushan SMH, Mohebbi MM, Karimzadeh P, Javadzadeh M. Moyamoya syndrome associated with Henoch-Schönlein Purpura. *Iran J Child Neurol*. 2016;10(4):71-74.
7. Tanghetti B, Capra R, Giunta F, Marini G, Orlandini A. Moyamoya syndrome in only one of two identical twins. Case report. *J Neurosurg*. 1983;59(6):1092-1094. http://doi.org/10.3171/jns.1983.59.6.1092
8. Aoyama J, Nariai T, Mukawa M, Inaji M, Tanaka Y, Maehara T. Case of familial Moyamoya disease presenting 10 years after initial negative magnetic resonance screening in childhood. *World Neurosurg*. 2017;105:1035.e1-1035.e4. http://doi.org/10.1016/j.wneu.2017.06.036
9. Hertza J, Loughan A, Perna R, Davis AS, Segraves K, Tiberi NL. Moyamoya disease: a review of the literature. *Appl Neuropsychol Adult*. 2014;21(1):21-27. http://doi.org/10.1080/09084282.2012.721147
10. Karzmark P, Zeifert PD, Tan S, Dorfman LJ, Bell-Stephens TE, Steinberg GK. Effect of Moyamoya disease on neuropsychological functioning in adults. *Neurosurgery*. 2008;62(5):1048-1051; discussion 1051-2. http://doi.org/10.1227/01.neu.0000325866.29634.4c
11. Araki Y, Takagi Y, Ueda K, et al. Cognitive function of patients with adult Moyamoya disease. *J Stroke Cerebrovasc Dis*. 2014;23(7):1789-1794. http://doi.org/10.1016/j.jstrokecerebrovasdis.2014.04.032
12. Su SH, Hai J, Zhang L, Wu YF, Yu F. Quality of life and psychological impact in adult patients with hemorrhagic Moyamoya disease who received no surgical revascularization. *J Neurol Sci*. 2013;328(1-2):32-36. http://doi.org/10.1016/j.jns.2013.02.010
13. Kronenburg A, van den Berg E, van Schooneveld MM, et al. Cognitive functions in children and adults with moyamoya vasculopathy: a systematic review and meta-analysis. *J Stroke*. 2018;20(3):332-341. http://doi.org/10.5853/jos.2018.01550
14. Ma Y, Guo Q, Yan Y, et al. Moyamoya disease: a retrospective study of 198 cases. *Med Clin (Barc)*. 2019;153(12):441-445. http://doi.org/10.1016/j.medcli.2019.02.024
15. Wunderle KB, McCulley C. Smoke and mirrors: Moyamoya syndrome. *Am J Med*. 2017;130(9):1045-1048. http://doi.org/10.1016/j.amjmed.2017.05.008
16. Zach V, Bezov D, Lipton RB, Ashina S. Headache associated with Moyamoya disease: a case story and literature review. *J Headache Pain*. 2010;11(1):79-82. http://doi.org/10.1007/s10194-009-0181-8
17. Pandey P, Bell-Stephens T, Steinberg GK. Patients with Moyamoya disease presenting with movement disorder. *J Neurosurg Pediatr*. 2010;6(6):559-566. http://doi.org/10.3171/2010.9.PEDS10192
18. Baik JS, Lee MS. Movement disorders associated with Moyamoya disease: a report of 4 new cases and a review of literatures. *Mov Disord*. 2010;25(10):1482-1486. http://doi.org/10.1002/mds.23130
19. Jehangir W, Singh Z, Bhatt H, et al. A new early finding of Moyamoya-like phenomena. *J Clin Med Res*. 2015;7(8):643-645. http://doi.org/10.14740/jocmr2193w
20. Nakanishi M, Kaito M, Gondo Y, Takada H, Matsui M, Hirose G. Moyamoya disease presenting with an acute confusional state in an elderly patient. *J Stroke Cerebrovasc Dis*. 2010;19(3):247-250. http://doi.org/10.1016/j.jstrokecerebrovasdis.2009.04.003
21. Kleinloog R, Regli L, Rinkel GJE, Klijn CJM. Regional differences in incidence and patient characteristics of Moyamoya disease: a systematic review. *J Neurol Neurosurg Psychiatry*. 2012;83(5):531-536. http://doi.org/10.1136/jnnp-2011-301387
22. Maheshwari S, Anthony A, Kushwaha S, Singh S, Desai R, Madan D. Moyamoya disease presenting as alternating hemiparesis with relapsing remitting hemichorea: an unusual manifestation. *J Pediatr Neurosci*. 2018;13(4):514-516. http://doi.org/10.4103/JPN.JPN_85_18
23. Lee S, Rivkin MJ, Kirton A, DeVeber G, Elbers J; International Pediatric Stroke Study. Moyamoya disease in children: results from the international pediatric stroke study. *J Child Neurol*. 2017;32(11):924-929. http://doi.org/10.1177/0883073817718730
24. Siddiqui MR, Khan SU, Hoque MA, Rahman KM, Mondol MBA, Mohammad QD. Moyamoya disease presented as a case of hemiplegic migraine. *BMJ Case Rep*. 2010;2010:bcr0220102764. http://doi.org/10.1136/bcr.02.2010.2764
25. Diaz UJ, Cabán-Martinez AJ, Halder GE. Presentation with recurrent intractable headache: a patient with Moyamoya syndrome–case report. *Neurol Med Chir (Tokyo)*. 2014;54(2):133-135. http://doi.org/10.2176/nmc.cr2012-0134
26. Dorfman LJ, Fischbein NJ, Woodard JI, Choudhri O, Bell-Stephens TE, Steinberg GK. Moyamoya disease can masquerade as multiple sclerosis. *Neurologist*. 2012;18(6):398-403. http://doi.org/10.1097/NRL.0b013e31826a99a1

27. Behere RV, John M, Sureshkumar R, et al. Mood disorder in association with Moyamoya disease. *Psychiatry Clin Neurosci*. 2012;66(2):163-164. http://doi.org/10.1111/j.1440-1819.2011.02317.x
28. Richards M, Grzenda A, Nelson E, Gitlin M. Psychiatric comorbidity in Moyamoya disease and preliminary guidelines for treatment. *Am J Psychiatry*. 2019;176(4):269-274. http://doi.org/10.1176/appi.ajp.2018.18040404
29. Mitchell AJ, Sheth B, Gill J, et al. Prevalence and predictors of post-stroke mood disorders: a meta-analysis and meta-regression of depression, anxiety and adjustment disorder. *Gen Hosp Psychiatry*. 2017;47:48-60. http://doi.org/10.1016/J.GENHOSPPSYCH.2017.04.001
30. D 'netto TB, Sudarsanan S. Moyamoya disease (presenting as schizophrenia). *Indian J Psychiatry*. 1981;23(2):188-189.
31. McDade G. Symptomatic schizophrenia with moya moya disease. *Behav Neurol*. 1991;4(1):25-28. http://doi.org/10.3233/BEN-1991-4103
32. Patra S, Patnaik A. Pediatric moyamoya disease presenting as attention deficit hyperactivity disorder: time to pay attention. *Indian J Psychiatry*. 2019;61(5):544-545. http://doi.org/10.4103/psychiatry.IndianJPsychiatry_403_18
33. Bose R, Banerjee AD. Paediatric Moyamoya disease and Tourette's syndrome: an unusual association responding favourably to direct and indirect cerebral revascularisation surgery. *Pediatr Neurosurg*. 2019;54(6):436-440. http://doi.org/10.1159/000503114
34. Lai J, Patel A, Dandurand C, Gooderham P, Lu S. Depression and catatonia: a case of neuropsychiatric complications of moyamoya disease. *Cureus*. 2018;10(10):e3460. http://doi.org/10.7759/cureus.3460
35. Lin MZ, Tsai SJ, Hsu JW, Huang KL, Chen MH. A child with Moyamoya disease: mimicking the presentation of panic attack. *Psychiatry Res*. 2020;285:112720. http://doi.org/10.1016/j.psychres.2019.112720
36. Cho A, Chae JH, Kim HM, et al. Electroencephalography in pediatric Moyamoya disease: reappraisal of clinical value. *Childs Nerv Syst*. 2014;30(3):449-459. http://doi.org/10.1007/s00381-013-2215-4
37. Kodama N, Aoki Y, Hiraga H, Wada T, Suzuki J. Electroencephalographic findings in children with Moyamoya disease. *Arch Neurol*. 1979;36(1):16-19. http://doi.org/10.1001/archneur.1979.00500370046008
38. Yamada I, Nakagawa T, Matsushima Y, Shibuya H. High-resolution turbo magnetic resonance angiography for diagnosis of Moyamoya disease. *Stroke*. 2001;32(8):1825-1831. http://doi.org/10.1161/01.str.32.8.1825
39. Tsuchiya K, Makita K, Furui S. Moyamoya disease: diagnosis with three-dimensional CT angiography. *Neuroradiology*. 1994;36(6):432-434. http://doi.org/10.1007/BF00593677
40. Fujimura M, Tominaga T. Diagnosis of Moyamoya disease: international standard and regional differences. *Neurol Med Chir (Tokyo)*. 2015;55(3):189-193. http://doi.org/10.2176/nmc.ra.2014-0307
41. Komiyama M, Nishikawa M, Yasui T, Otsuka M, Haze K. Moyamoya disease and coronary artery disease–case report. *Neurol Med Chir (Tokyo)*. 2001;41(1):37-41. http://doi.org/10.2176/nmc.41.37
42. Livesay J, Johnson J. ST-elevation myocardial infarction (STEMI) in a patient with moyamoya disease. *Case Rep Cardiol*. 2019;2019:7124072. http://doi.org/10.1155/2019/7124072
43. Jefferson AL, Glosser G, Detre JA, Sinson G, Liebeskind DS. Neuropsychological and perfusion MR imaging correlates of revascularization in a case of Moyamoya syndrome. *AJNR Am J Neuroradiol*. 2006;27(1):98-100.
44. Nehra A, Kaur H. Moyamoya disease: a ray of hope from a psychosocial perspective. *Ann Neurosci*. 2015;22(2):121-124. http://doi.org/10.5214/ans.0972.7531.221212

How to cite this article: Leiva DF, Arey KJ, Soares WE. Moyamoya Disease- A Clinical Mimic for Psychiatric Disorders in the Emergency Setting: A Case Report. JACEP Open. 2025;6:100061.

https://doi.org/10.1016/j.acepjo.2025.100061