Incidental Finding of Moyamoya Disease in a Second-Level Hospital in Honduras

Hallazgo Incidental de Enfermedad de Moyamoya en un Hospital de Segundo Nivel en Honduras

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ABSTRACT
Introduction: Highly rare in the Hispanic population, Moyamoya disease is an occlusive idiopathic arteriopathy that bilaterally affects the anterior cerebral circulation, primarily the internal carotid artery and its branches, progressing to the stenosis of its terminal portions. The aim of this study was to present the clinical history, medical management, and evolution of a patient with this disease to expand scientific knowledge in our Latin American region, thereby optimizing the likelihood of diagnostic suspicion among medical personnel.

Case presentation: A 51-year-old male patient visited the medical service due to fainting, headache, imbalance, and psychogenic vertigo. This is the first case of Moyamoya disease to be reported in Honduras. It was incidentally diagnosed, evaluated in neurosurgery for the possible need for external drainage, and ultimately conventionally treated with a favorable prognosis. The patient currently remains under outpatient neurological follow-up.

Discussion: The diagnosis of Moyamoya disease is a challenge, given its rarity and the scarcity of reports in Latin America. For this reason, most diagnoses are incidental. Conclusion: The proper management of Moyamoya disease is currently being debated, with no known curative treatment. Medical criteria are important for choosing the most convenient therapy according to each patient’s characteristics.

Keywords: Neurosurgery; Moyamoya disease; Honduras

RESUMEN
Introducción: La enfermedad Moyamoya es una arteriopatía idiopática oclusiva de muy escasa aparición en la población hispana, afecta la circulación cerebral anterior de forma bilateral, principalmente a la arteria carótida interna y sus ramas, progresivamente culminan estas en estenosis de sus porciones terminales. El objetivo del trabajo radica en presentar la historia clínica, manejo médico y evolución de un paciente con esta enfermedad a fin de aumentar el conocimiento científico en nuestra región latinoamericana y con ello optimizar las probabilidades de sospecha diagnóstica del personal médico. Presentación de caso: Paciente masculino de 51 años, que acudió al servicio médico por cuadro de lipotimia, cefalea, desequilibrio frente al entorno y vértigo psicógeno. Se presenta el primer caso reportado en Honduras de Enfermedad de Moyamoya, diagnosticado de manera incidental, evaluado en neurocirugía por posible necesidad de drenaje externo, tratado finalmente de manera médica convencional con evolución favorable, actualmente permanece bajo vigilancia neurológica ambulatoria. Discusión: El diagnóstico es un reto, dada la rareza de la enfermedad y la escasez de reportes
en latinoamericana, por ello la mayoría de los diagnósticos son incidentales. **Conclusión:** El manejo adecuado de la enfermedad de moyamoya es causa de debate actualmente, no hay tratamiento conocido que sea curativo. El criterio médico es importante en la elección de la terapia más conveniente según las características propias de cada paciente.

**Palabras-Clave:** Neurocirugía; Enfermedad de Moyamoya; Honduras

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**INTRODUCTION**

Moyamoya disease (MMD) is an idiopathic pathology that bilaterally affects the anterior cerebral circulation, primarily the internal carotid artery and its branches, subsequently progressing to stenosis in the terminal portion of these arteries\(^1\). As a result, the external carotid artery becomes the main blood supply\(^2\). This disease was named after the Japanese word moyamoya which means "puff of smoke", describing the appearance of the collateral blood vessels that form at the base of the skull\(^3-6\).

In 2018, estimates indicated that Asia, especially Japan, and South Korea had the highest recorded prevalence rates, with Japan reaching a prevalence of 3 cases per 100,000 individuals per year, predominantly in the pediatric population\(^7\). The total incidence among Japanese inhabitants is 0.94 per million inhabitants\(^8\). In Europe, the recorded incidence is one-tenth of that reported in Japan\(^7\). In turn, in the United States of America, approximately one new case is recorded per year per million inhabitants\(^1\). In 2018, Nathal et al.\(^1\) reported that in the United States, from 2005 to 2008, the incidence rate of this disease was 11.9%, among Hispanics aged 19 to 45 years, and 5.9% among Hispanics older than 45 years of age. This was in contrast to findings reported in 2005 by Uchino et al.\(^9\), who suggested an incidence rate of 0.03 cases per 100,000 Hispanic inhabitants in the state of California and a national estimate of 10 new cases of MMD and Moyamoya syndrome (MMS) among Hispanics per year.

MMD can start as early as 5 years of age, progressing quickly and with an unfavorable prognosis\(^7,10,11\). In adulthood, the mean age of onset is 40 year\(^7\). MMD can manifest as cerebral ischemia or hemorrhage, and its prevalence seems to be linked to the geographical area. In the United States, ischemia is reported as the main cause of MMD in both adults and children, but hemorrhage occurs much more frequently in the adult population than in the pediatric population, at 20% and 2.8%, respectively. In contrast, in the Asian population, 42% of adult patients with MMD are diagnosed with the hemorrhagic form of the disease\(^7\). In addition to ischemia and hemorrhage, the most common manifestations in both children and adults are seizures, headache, intellectual disability, and involuntary movements\(^6,10,11\). Involuntary movements are more common in women and have been reported as a risk factor in pregnant women. The mechanisms that cause the disease are ischemia of the basal ganglia-thalamocortical circuits, increased levels of sex hormones either during pregnancy or due to intake of oral contraceptives, and hyperthyroidism\(^5\).

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**CASE PRESENTATION**

This study presents the clinical case of a 51-year-old male mestizo patient from an urban area in Honduras. He had a history of type 2 diabetes mellitus, which was diagnosed in 2015 and was treated with 500 mg/day of metformin. Approximately 10 years prior, he suffered from dyspepsia and heartburn, which were not treated with drugs.

On October 25, 2020, he fainted twice while he was at work. Thus, his co-workers took him to a private practice in the locality, where a diagnosis or cause of the clinical symptoms could not be established. The patient also presented with gastric symptoms a decade ago, for which he was prescribed treatment with one sucrassyl tablet every 8 hours, simethicone every 8 hours, and 30 mg/day of lansoprazole. He was subsequently referred for evaluation and treatment to a secondary hospital in San Pedro Sula.

Four days later, on the 29th of the same month, the patient voluntarily visited the Emergency Department of the Honduran Institute of Social Security (Instituto Hondureño de Seguridad Social – IHSS) in the company of relatives. Upon admission, he reported mild-to-moderate, sudden-onset holocranial headaches, which started approximately 3 days prior, did not respond to mitigation, was constant throughout the day, and only disappeared...
when the patient fell asleep. He also mentioned a feeling of imbalance and psychogenic vertigo. He expressed fear that he was showing symptoms of coronavirus disease (COVID-19), which was subsequently ruled out after a nasopharyngeal swab and polymerase chain reaction test. He denied having symptoms such as vomiting, hyporexia, blurred vision, anosmia, and fever. He also denied excessive alcohol consumption and high blood pressure.

On physical examination, mesomorphic biotype, facial expression of pain, hydrated mucous membranes and skin with adequate coloration; mobile upper and lower extremities without edema; and thorax and abdomen without apparent abnormalities were noted. The patient’s vital signs were: heart rate, 65 beats per minute; P/A, 160/100 mmHg; respiratory rate, 12 breaths per minute; and oxygen saturation, 97%. There were no abnormalities on hematocrit, hemoglobin, leukocyte formula, platelet count, coagulation profile, blood glucose levels, liver and kidney function, lipid profile, electrolytes, and thyroid profile. Serological tests for syphilis and the human immunodeficiency virus were negative.

A hypertensive crisis was suspected as the cause of the clinical symptoms; therefore, intravenous administration of enalaprilat (one ampoule diluted in 250 mL of normal saline solution) was continued. The patient remained under strict surveillance. After four hours, he presented with a marked increase in blood pressure, reaching values up to 171/98 mmHg.

Results of neurological evaluation indicated that the patient was disoriented with regards to space, and time, scoring 14/15 on the Glasgow Coma Scale, with isochoric and light-reactive pupils and preserved external eye movements; eye fundus without abnormalities; facial symmetry without deviation of the labial commissure and intact nasolabial fold; present cough and gag reflex; 5/5 motor strength on the Daniels scale in all four extremities; superficial and deep sensitivity without abnormalities and intermittent episodes of lateralization due to psychomotor agitation; normal osteotendinous reflexes without signs of meningeal irritation; and coordination (symmetrical and unaltered gait).

Findings from imaging studies through brain multislice tomography showed areas of hyperdensity at the level of the ventricular system, with dilatation of the left lateral ventricle. For this reason, external drainage was considered to reduce intracranial hypertension (Figure 1).

Cerebral angiography was performed to rule out arteriovenous malformations and aneurysms, revealing a symmetrical occlusion of the internal carotid artery, which produces collateral circulation, faintly outlining the middle and anterior cerebral arteries. This was characteristic of the angiographic smoke cloud pattern of Moyamoya disease. A possible bleeding pattern due to arterial hypertension was also ruled out (Figures 2 and 3).

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Figure 1. A. Axial computed tomography of the brain at the level of the lateral ventricles, showing a hyperdense image highlighting dilatation of the left lateral ventricle; B. Midline sagittal computed tomography of the brain showing hyperdense content at the level of the ventricular system.
The patient remained disoriented, presenting with a moderately intense holocranial headache. Therefore, a second tomographic control was necessary, which showed a progressive decrease in intracranial hypertension.

Pharmacotherapy was initiated with candesartan, 40 mg of esomeprazole, 100 mg of phenytoin, 25 mg of dexketoprofen, and 1 g of acetaminophen. After this drug treatment, the patient evolved favorably with a gradual decrease in symptoms in approximately 5 hours. Thus, external drainage was not required.

Three days after hospitalization, the patient was discharged while conscious, with an improved health status, Glasgow score of 15, and readiness to return to work. He currently remains...
under medical surveillance through follow-up appointments and imaging studies.

**DISCUSSION**

Diagnosis of MMD is challenging, given the rarity of the disease incidence and the scarcity of reports in our Latin American region. It is usually prevalent among the Asian population, particularly the Japanese, but MMD is also reported among children aged 5-9 years who present with arterial ischemic stroke or transient ischemic attacks and among adults aged 45-49 years who present with intracranial hemorrhages (ICH), as in our patient; these demographics are in line with the current literature. Pediatric patients with ICH and adults with an ischemic pattern may also have MMD. In turn, suspicion increases if the patient is female, in view of a 2:1 relationship between the sexes.

While most cases are sporadic, reported that 10-15% of patients with MMD have a family history of MMD. Although the etiology remains unclear, research indicates the special involvement of chromosome 17, though other studies also have reported on disorders of chromosomes 3p, 8q23, 10q23, and 4q32, increasingly supporting the assumption of a multifactorial and polygenic disease. In our case, no genetic analysis were performed due to limited resources; however, the patient denied having neurological diseases or a family history of such.

Pathophysiologically, MMD is not yet well elucidated but is known as a primary disease with low morbidity, which causes bilateral progressive stenosis (affecting both cerebral hemispheres) possibly due to concentric and eccentric fibrocellular thickening of the intima at the distal level of the internal carotid artery, in the initial segment of the middle and anterior cerebral arteries. Posterior circulation involvement is rare and usually indicative of a poor prognosis. The most striking compensatory response is neovascularization and vessel thickening in regions near the base of the skull, accounting for the angiographic “puff of smoke”.

In contrast to MMD, MMS is a mostly unilateral secondary vasculopathy that occurs in response to other risk factors, such as neurofibromatosis, Down syndrome, thyroid diseases, cranial irradiation, and sickle cell anemia. Therefore, MMD and MMS form a highly heterogeneous group with diverse clinical presentations and variable hereditary penetrance. Clinical history taking and laboratory and imaging studies confirmed that our patient was not associated with any pathological condition consistent with MMS. Thus, he was diagnosed with MMD.

The typical clinical presentation of MMD varies between cerebral ischemia and ICH. In the former, the symptoms usually appear in children, and they are multiple and recurrent. The most frequent manifestations are mono- or hemiparesis, migraine-like headaches, cognitive impairment, sensory disorders, dysarthria, or aphasia. Atypical symptoms include syncope, vertigo, visual disturbances, seizures, or pseudopsychiatric symptoms. In pediatric patients, they are usually triggered after hyperventilation caused by physical or intellectual activity, dehydration, fever, and/or crying. Healthcare professionals must remain attentive to the “morning glory syndrome” (MGS); if MGS is present, MMD diagnosis should be considered. Therefore, when MMD is detected, magnetic resonance angiography is recommended to rule out this disease or any other vascular abnormality of the carotid system.

Unlike the symptoms of the ischemic pattern, according to a study published in 2015 by Guey et al., ICH occurs in more than 20% of adult patients with MMD. Furthermore, in 2019, Hui Zhang et al. reported that intracranial hemorrhage was the first symptom in half of the adult patients, whereas ischemia occurred first in the other half. The location of the bleeding can be intraventricular, intraparenchymal, or subarachnoid, and is attributed to the cessation of the blood vessels. In case of deep collaterals, a deep intraparenchymal hemorrhage occurs either at the basal ganglia, deep periventricular white matter, or even intraventricular level. A condition of the basal ganglia can lead to the onset of choreic movements in both adult and pediatric patients. Subarachnoid hemorrhage is rare and attributed to changes in circulatory patterns at the base of the brain, which can lead to the development of cerebral aneurysms located in the circle of Willis, whose rupture causes hemorrhage.

There are currently several imaging techniques which can help in the diagnosis of MMD, each of them with unique purposes. The choice is based on the availability and risk-benefit balance. Cerebral angiography is considered the gold standard because it reveals the location of the occlusion, although the Doppler study has provided good results in preoperative and postoperative follow-ups.

Currently, no known therapy can either stop or reverse changes generated at the vascular level in patients with MMD. Treatment

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is directed at alleviating symptoms, limiting cognitive decline, and preventing subsequent cerebrovascular events. The measures used to stabilize the patient can be medical or surgical\(^1\).

Medical treatment commonly consists of using vasodilators, steroids, and anticoagulants. However, appropriate MMD treatment is still being debated due to the lack of scientific evidence supporting its efficacy. The goal for these patients is to maintain an adequate cerebral vascular perfusion in the acute phase. In ischemic conditions, aspirin is administered to the patient, who is kept hydrated\(^2\). In our case, after a careful evaluation, the medical team decided not to intervene surgically and, instead, to treat the symptoms. Our patient improved favorably and avoided a prolonged hospital stay amid the crisis caused by the COVID-19 pandemic.

An effective diagnosis and treatment is essential because poor management may require a prolonged prescription of antiplatelet drugs, which can lead to the conversion from ischemic to hemorrhagic MMD\(^3\).

In the present case, the bilaterality of the occlusion with a smoke puff pattern was observed in the angiography, and the absence of risk factors demonstrated MMD. Moreover, the age at onset and the clinical presentation were also in line with the current literature.

CONCLUSION

Moyamoya cerebrovascular disease is a rare disease in Latin America. Few cases have been described thus far. Its incidence is reportedly highest among patients of Asian descent and lowest among Hispanics. The diagnosis of this disease is a challenge, with many reports referring to an incidental finding, and cerebral angiography remains the gold diagnostic standard. The proper management of MMD is currently being debated, with no known curative treatment.

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