SAS Journal of Medicine
Abbreviated Key Title: SAS J Med
ISSN 2454-5112
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Moya-Moya Disease a Case Report and Literature Review

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DOI: 10.36347/sasjm.2024.v10i03.005 | **Received:** 05.01.2022 | **Accepted:** 08.02.2022 | **Published:** 04.03.2024

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Abstract **Case Report**

Moya-Moya disease is a rare chronic cerebral vascular disease (arteriopathy), which can affect both children and adults and gives rise to severe clinical manifestations of ischemic or hemorrhagic origin. This angiogenic disease is an intrinsic primary pathological process, characterized by a narrowing of the distal internal carotid artery that extends to the proximal segments of the middle and anterior cerebral arteries, inducing the formation of replacement vessels that give a characteristic appearance on angiography forming a smoke cloud: Moya Moya network. The involvement of the posterior circulation is very rare. Its etiology is still poorly understood and represents 10 to 15% of the causes of strokes, with 2 age peaks where it is more frequent: children around 5 years and adults around 40 years. Its evolution can be slow with intermittent symptoms or fulminant with a rapid neurological decline. Imaging is the reference examination in the diagnosis of Moya-Moya. MRI (Magnetic Resonance Imaging) has an important and growing role in the management of the disease. We report a case of a 48 year old patient with diabetes and hypertension, hospitalized for a deep left hematoma.

Keywords: Moya-Moya – MRI.

INTRODUCTION

Moya-Moya disease (MMD) is a chronic arteriopathy of unknown origin, characterized by progressive stenosis of the terminal segment of the internal carotid arteries (ICA) and the proximal segment of the middle (MCA) and anterior (ACA) cerebral arteries with the development of a network of basal arterial bypasses. The involvement of the posterior circulation is very rare [1]. It is to be differentiated from the Moya-Moya syndrome (MMS) when the involvement is associated with extra-cerebral signs realizing a syndromic framework or secondary to a chronic pathology. This angiogenic disease can be the cause of various manifestations such as transient or constitutive ischemic attacks, hemorrhagic strokes, headaches, seizures, ataxia, cognitive decline and choreiform movements [2]. Imaging plays a crucial role, especially CT and MRI (Magnetic Resonance Imaging), in the diagnosis, pre-treatment assessment and monitoring of the disease. The treatment of choice in Moya-Moya disease is surgical revascularization which can be direct or indirect, aiming to promote the development of neovascularization, or by multiple drill holes. Finally, it can be mixed by combining direct and indirect techniques [2, 3].

We report a case in a 48-year-old patient with diabetes and hypertension, under medical treatment, hospitalized for a deep left hematoma on the initial MRI.

CASE PRESENTATION

This is a 48-year-old woman, hypertensive and diabetic, under treatment, hospitalized in neurology for left deep hematoma on initial MRI.

Brain MRI showed a characteristic aspect of Moya Moya disease, by the presence of a temporal and Capsulo insular left range in T2 hypersignal (Fig1), T1 hyposignal and Flair (Fig2), seat of empty signals on the gradient echo sequence (Fig3), with moderate dilatation of the occipital horn of the left lateral ventricle. On the arterial agiographic sequence, there is a small aspect of the intra cavernous portion of the left internal carotid artery without visualization of the left sylvian artery, with multiple nodular vascular structures in front of it realizing a cloudy aspect (Fig 4 a and b).

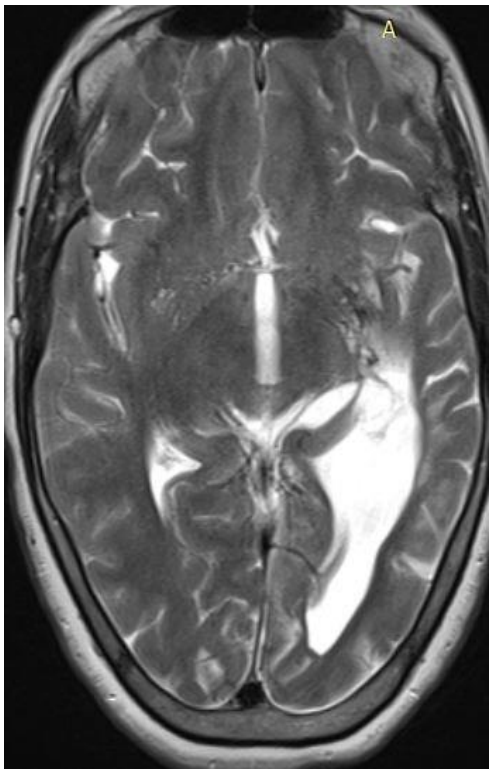

Fig-1: T2 axial sequence

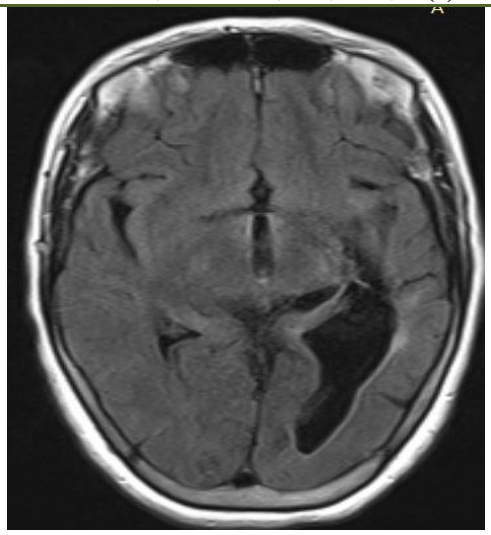

Fig-2: Axiale Flair sequence

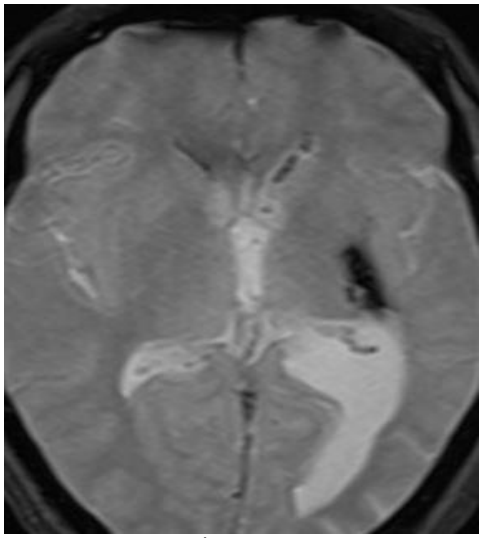

Fig-3: T2* axiale sequence

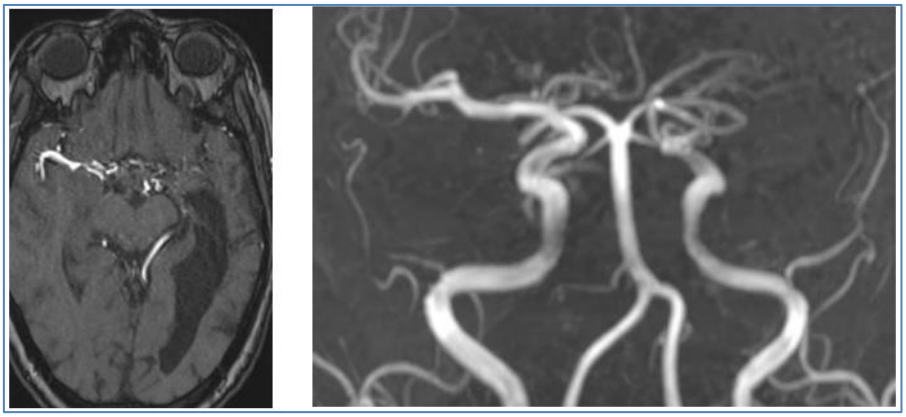

Fig-4 a et b: séquence axiale 3DTOF et agiographique artérielle

Tab-1

The Suzuki staging system

The Suzuki staging system for Moyamoya is based on the severity of the angiographic appearance

Suzuki Stages	Angiographic appearance
Stage I	Narrowing of the terminal internal carotid bifurcation
Stage II	Initial development of the first Moyamoya collateral vessels at the base of the brain with dilation of the intracerebral main arteries
Stage III	The collateral Moyamoya vessels intensify, becoming more prominent, and the major trunks of the anterior circulation become severely stenotic and start to occlude
Stage IV	Posterior cerebral arteries become occluded, moyamoya vessels start to diminish, and collaterals from the external carotid arteries begin to form
Stage V	Moyamoya collateral vessels begin to completely disappear, and the extracranial collaterals become more and more prominent
Stage VI	Disappearance of the moyamoya collaterals and major named cerebral arteries; the cerebral hemispheres receive blood almost exclusively from abnormal external carotid anastomosis

Tab-2

Suzuki Stage III sub-staging system

The majority of patients fall into Suzuki Stage III, and the table shows the critical subsets of Stage III scoring

Suzuki Stage III sub-stages	Angiographic appearance
Stage III A	Partial non-filling of the anterior cerebral arteries and the middle cerebral arteries
Stage III B	Partial preservation of the anterior cerebral arteries and the middle cerebral arteries
Stage III C	Complete lack of the anterior cerebral arteries and the middle cerebral arteries

DISCUSSION

Moya Moya disease is a chronic cerebral vascular disease characterized by progressive stenosis and occlusion of the terminations of the intracranial internal carotid arteries and the proximal part of the arteries of the polygon of Willis, associated with the development of an abnormal vascular network at the base of the skull. This network often has a "cloudy" or "smoke-like" appearance, which is translated into Japanese as "Moya Moya" [4]. It is common in East Asian countries, such as Japan and the Republic of Korea, where its annual incidence is estimated to be 0.35-0.94 per 100,000 population [5]. MMD is rare in the United States, with only 0.086 newly diagnosed cases per 100,000 people per year, or approximately one case per million new cases per year. Risk factors for MMD include East Asian ancestry and predisposing conditions such as neurofibromatosis and Down syndrome. It is more common in women than in men [6, 7], with a sex ratio of 2.18 [7]. There are two peaks in the frequency of the disease, 5 years and before 40 years [8]. In Morocco its incidence is not yet elucidated due to its rarity.

Ischemic manifestations are more frequent in children, while hemorrhagic manifestations predominate in adults [2,9]. These may be cerebral or meningeal hemorrhages related to rupture of constitutionally fragile neovessels (microaneurysm, thinned arterial wall) or leptomeningeal anastomoses subjected to unusually high pressures. Changes in the circulation within the polygon of Willis and the introduction of high-flow collateral circulation via the communicating arteries may also lead to the formation of intracranial aneurysms in "healthy" arteries.

Headache is reported in 20-60% of cases at the time of diagnosis. They are sometimes unusual and related to the occurrence of a stroke, but more often the link between the headache and the intracranial

angiopathy is difficult to establish. Seizures are also observed in 20% of cases, particularly after cerebral hemorrhage or infarction. Cognitive disorders may accompany the occurrence of a stroke or be observed in the absence of brain parenchyma lesions. In the latter case, the involvement of chronic cerebral hypoperfusion is suggested by the improvement in clinical performance after surgical restoration of efficient intracerebral hemodynamics [10, 11].

The diagnosis of Moya-Moya disease is certain in the presence of bilateral anomalies, probable [12]. Depending on the study, 10-39% of unilateral forms at initial diagnosis become bilateral during follow-up [13]. Imaging has a crucial role in the diagnosis, pre-treatment assessment and follow-up of the disease. Angiography remains the preferred examination for the precise anatomical assessment of the disease, while nuclear medicine techniques are the reference for the study of the hemodynamic impact. Nevertheless, other more accessible and less invasive imaging techniques, such as MRI, CT and ultrasound, now have a well-established place in management.

Recent and/or sequelae ischemic lesions are detected by diffusion imaging and FLAIR sequences. T2*-weighted gradient echo sequences allow the detection of hemorrhagic complications of the disease. Finally, some arteries on the surface of the brain (distal branches of the pial arteries or neovessels) can be visualized in FLAIR hypersignal in relation to slow flow.

The gold standard in diagnosis and grading based on the Moyamoya image is a catheter-based DCA. Suzuki and Takaku first classified the development of MMD into a six-stage system known as the Suzuki staging system (Table1) [14]. The majority of cases are in stage III, and an in-depth and more detailed staging system specifically for stage III has been developed (Table2) [15].

The use of transcranial Doppler (TCD) can be employed to assess mean arterial velocity and resistance index before and after surgery for a comparison of the effectiveness of revascularization [16].

Currently, there is no proven medical therapy to stop the natural progression of MMD. All medical therapies are aimed at preventing secondary complications of the disease process, such as the use of antiplatelet agents to reduce the incidence of thrombus formation as cerebral vessels become progressively obstructed.

Treatment of Moya-Moya disease requires multidisciplinary expertise involving neurologists, neurosurgeons and anesthesiologists. The treatment of choice in Moya-Moya disease is surgical revascularization. It can be direct by anastomosis between the superficial temporal artery (STA) and the ACM. It can be indirect, aiming to promote the development of neovascularization, by encephalo-myo-synangiosis, encephalo-duro-synangiosis, encephalo-arterio-synangiosis (these three techniques can be combined) or by multiple drill holes. The treatment can also be mixed by combining direct and indirect techniques [1, 2].

Direct or indirect surgical revascularization is the most effective treatment option for MMD. Surgical revascularization is designed to prevent strokes and restore adequate cerebral blood flow to subperfused regions. If a patient has bilateral disease, it is essential to treat the more symptomatic side first; however, it is not uncommon to perform bilateral operations in a single surgery. For adults with an initial presentation of an ischemic event, it appears that direct bypass surgery is more effective in preventing future ischemic strokes than indirect bypass surgery [17].

The prognosis of the disease is severe. Depending on the study, 50-90% of patients have neurological deficits after repeated ischemic strokes, and in 3-11% of cases the outcome will be fatal [18]. This prognosis is all the worse when the onset of symptoms is early (before the age of 7 years).

CONCLUSION

Moya-Moya disease is a chronic and progressive disease, constituting a non-negligible cause of ischemic or hemorrhagic stroke, with no effective medical or endovascular management options; however, surgical intervention can stop the disease.

Angioscan and angiogram are currently the gold standard imaging for initial diagnosis and monitoring, but arteriography remains important for the accurate diagnosis and lesion assessment of this condition. Surgical treatment should be strongly considered for symptomatic patients to improve hemodynamic flow to physiologically underperfused areas in the ICA, proximal MCA, and proximal ACA territory of the brain parenchyma. In pediatric patients, early diagnosis and surgical intervention are necessary to prevent irreversible cerebrovascular infarction.

Competing Interests

The authors declare no conflict of interest.

Contributions from authors

All the authors contributed to the conduct of this work. They also state that they have read and approved the final version of the manuscript.

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