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(CASE REPORT)

Moya –Moya disease revealed by a subarachnoid hemorrhage: A case report

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Abstract

Background: Hemorrhage is a major category of the clinical symptoms of moyamoya disease (MMD). Intracerebral and intraventricular hemorrhages are the most common hemorrhages in MMD, and subarachnoid hemorrhages (SAHs) are notably rare.

Case report: A 43 year old female patient, with no previous pathological history, admitted with a frank meningeal syndrome followed by a disorder of consciousness.She was admitted with a GCS at 15, which was subsequently reduced to 10. She was hemodynamically and respiratory stable with a BP of 12/8 cm/Hg, a respiratory rate of 16 cycles/min and a heart rate of 92 beats/min. Initial imaging showed subarachnoid hemorrhage Fisher IV. A cerebral angiography-CT showed vascular thrombosis of the internal carotid artery whitout aneurysm and arteriovenous malformation and dural fistulas. An EVD (external ventricular drain) was placed. After three days of hospitalization in the intensive care unit, a cerebral arteriogram was performed showing vascular arrest in the right internal carotid artery in its pre-cavernous portion with absence of visualization of the right sylvian artery and the presence of several perforating cerebral artery and external carotid artery bypass systems suggestive of Moya Moya.

Conclusion: Moya Moya disease is a rare entity of unknown origin. Subarachnoid hemorrhage is a rare manifestation. It's mainly occurs in a young adult. Cerebral angiography remains the gold standard for diagnosis.

Keywords: Moyamoya disease; Diagnosis; Sub-arachnoid hemorrage; Cerebral angiography

1. Introduction

Moya Moya disease is a rare chronic intracranial arteriopathy, which affects both internal carotid arteries. It is characterized by progressive occlusion of the terminal portions of the internal carotid arteries with development of a bypass circulation Although it primarily affects people of Asian descent, it is found in all populations and ethnicities [2,3]. The disease was first described in Japan in 1955 by "Takeushi and Shimizu" after reporting a case of bilateral hypoplasia of the internal carotid arteries. The disease was first named Moya Moya by "Suzuki and Takaku" in 1969 [1]. We report the clinical observation of a 43-year-old girl, without any notable pathological history, a frank meningeal syndrome followed by a disorder of consciousness of abrupt onset associated with an HTIC syndrome, admitted with a GCS 14 - 15 subsequently deteriorated to 10. The diagnosis of MMM was made on the basis of cerebral angiography data. Through reviews of the literature, we will recall the main epidemiological, diagnostic characteristics of this condition.

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2. Case presentation

2.1. Patient information

A 43-year-old female with no previously medical history presented severe headache,vomiting with consciousness disorder, without any neurological motor deficit.

2.2. Clinical findings

At the admission, the patient was conscious with a Glasgow Coma Scale of 15 . Tere was no motor-sensory defcit. He denied any changes in speech and facial, visual, swallow, bladder, or bowel disturbance at the onset. He denied any trauma or other inciting event. Subesequently, She suddenly showed an altered mental status on hospital day 3 (GCS score, 11).

2.3. Diagnostic assessment

The cerebral brain CT- scan showed a subarachnoid hemorrhage Fisher IV. A cerebral angiography-CT showed vascular thrombosis of the internal carotid artery whitout aneurysm and arteriovenous malformation and dural fistula.

2.4. Therapeutic intervention

The patient underwent surgery a external ventricular drain was placed.

2.5. Follow-up and outcome

During the hospitalisation , he benefited a cerebral arteriography showing a bilateral occlusion of the supraclinoid segments of both ACIs as well as the two AI segments suggestive of a Moya Moya disease. The patient did not well in the postoperative period, despite the combined surgical and medical treatment, the patient continued to deteriorate and subsequently died.

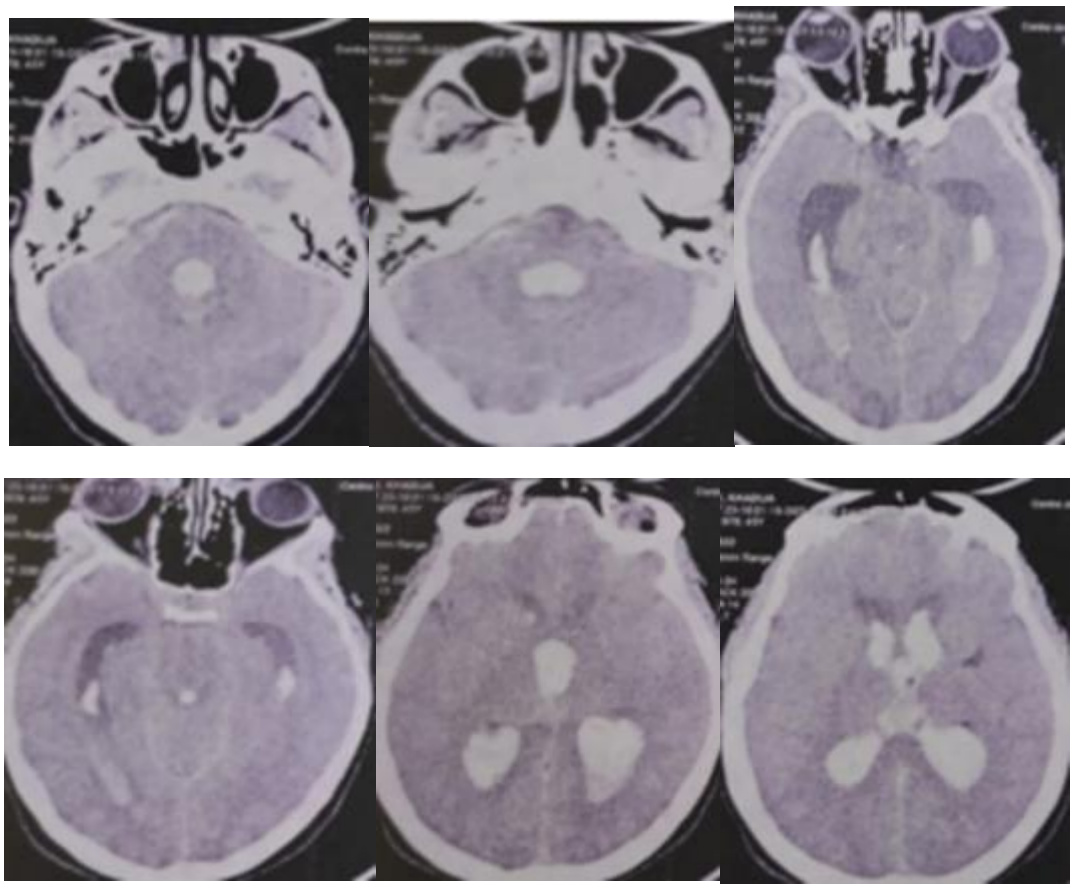

Figure 1 A large intraventricular hemorrhage

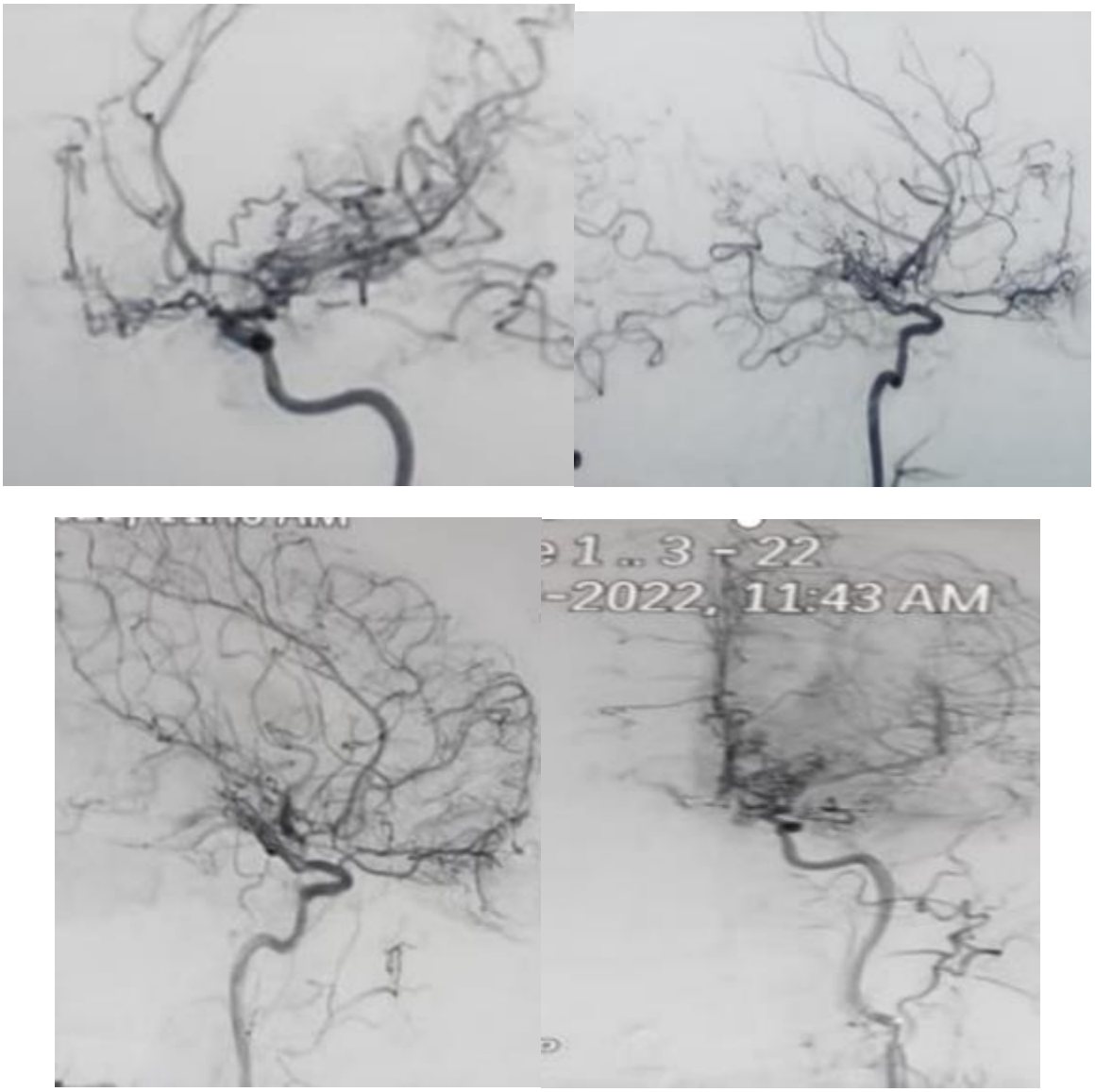

Figure 2 Cerebral Angiography showed bilateral occlusion of the supraclinoid segments of both ACIs as well as the two AI segments. Presence of a large anastomotic network at the base of the skull allowing iso-current opacification of the grafted segments of the A2 and ACA, as well as of the M1 segments and the branches of the sylvian divisions bilaterally

3. Discussion

Moyamoya disease is characterized by a progressive occlusion of the intracerebral internal carotid arteries and their main branches compensated by the appearance of a very thin "cigarette smoke" supply network. It was first reported in 1957 by Takeuchi and Shimizu, named "Moyamoya disease" by Suzuki and Takaku in 1969.

The epidemiological characteristics of Moyamoya disease are now better established with a more frequent incidence in non-Asian populations as shown by a North American cohort published in January 2013 [2]. It affects children more frequently in Asia, whereas in non-Asian populations its frequency is higher in adults. Our se' rie confirms this as the only patient of Asian origin was a 12-year-old child while the other cases were non-Asian adults. It is a progressively progressive condition responsible for occlusion of cerebral vessels including mainly distal stenosis of bilateral internal carotid, anterior, middle cerebral arteries [1]. Posterior cerebral arteries are also involved in some patients [3]. The familial form of Moya Moya is autosomal dominant with incomplete penetrance [4]. The cause of this disease remains unclear. Histologically, affected internal carotid arteries show intimal thickening, media attenuation [5].

The physiopathology of this syndrome is still poorly understood. However, several hypotheses seem to have been evoked: first of all, histopathology with a progressive vascular occlusion resulting from a hyperplasia of the smooth muscle cells, a fragmentation of the internal elastic boundary, a thinning of the media altering the intracranial arterial structure [6]. On the other hand, genetic factors playing a role in cerebral angiogenesis have been suggested in recent studies [7]. The role of the expression of several angiogenic factors promoting the development of the collateral arterial network also seems to be under exploration [8]. The etiology of the disease is unknown; however, the RNF213 gene located' at 17q25-ter has recently been identified as underlying a genetic susceptibility for Moya-Moya disease in the East Asian population [8-9]

The clinical features of Moyamoya syndrome or disease are highly polymorphic. According to the literature, the most common présentation in children,non-Asian adults is cerebral infarction [10-11]. Moreover, the presence of TIA during exercise in the pediatric population, as a consequence of the precarious cerebral arterial vasoconstriction during hyperpnea, is very suggestive, as it was the case in our child. In contrast to these data, epidemiological studies report a predominance of intracranial hemorrhages revealing Moyamoya in Asian adult populations [12,13] explained by a rupture of fragile arterial neovessels developing since childhood [14] The main manifestations of the disease are strokes, especially ischemic, which can cause speech, language or vision disorders, as well as balance, movement coordination problems, usually resulting in academic delay due to learning or memorization difficulties [15]. Although MRI angiography is widely used to confirm the diagnosis, to specify the anatomy of the vessels involved, Angiography is also useful for the visualization of intracranial vascular stenoses. It was' the key examination in our observation. Thus, in individuals with Moya-Moya disease, the most revealing images resemble the "smoke puffs" constituted by the collateral circulation [16] Arteriography is a reference examination for the diagnosis. It allows preoperative visualization of the intra- m extra-cranial vascularization. The diagnosis is based on the presence of stenosis/or bilateral occlusion of the carotid siphons a/or proximal portions of the ACM, ACA. The opacification at arterial time of a bilateral supplementary vascular network visible around the obstructive lesions, which is the basis of the classification of Suzuki, Takaku, who classified this pathology into 6 grades of increasing severity: Grade I: stenosis of the distal portions of the internal carotid arteries; Grade II: appearance of Moya Moya vessels; Grade III: increased number of vessels; Grade IV: decreased number of vessels; Grade V: significant decrease in the number of vessels; Grade VI: disappearance of Moya Moya vessels. Transcranial Doppler provides a non-invasive means of monitoring changes in blood flow. The natural history of Moyamoya syndrome has previously been described in several studies that show a risk in the five years following the first episode [17,18]. There seems to be a notable difference between Asian adults,the Caucasian population, which has a lower tendency to recurrence [19].

Therapeutically, there is no treatment that prevents cerebral artery narrowing, but there are ways to limit symptoms. Platelet anti-aggregants prescribed at an early stage can decrease or even disappear the deficient attacks. An alpha-blocker, vasodilator, antie'pileptic treatment (in case of epileptic seizures) can be proposed. Functional rehabilitation is essential. As for surgical treatment, not all patients are good "candidates" for surgery, which can sometimes be more dangerous than beneficial. Indeed, several surgical techniques of revascularization (direct or better indirect by encephalo-duro-arterio-synangiosis (EDAS) which is based on the principle of the appearance of a neovascularization [20], have been described to increase the cerebral blood flow by promoting collateral circulation. Finally, the various proposed treatments are still very controversial. They are mainly of two types: medical or surgical. The data in the literature concerning revascularization surgery in Moyamoya remain uncertain in the absence of a randomized study on the subject in particular concerning the selection of patients eligible for such surgery, the ideal time of intervention, which must be decided on a case by case basis. Nevertheless, there seems to be a benefit to revascularization surgery in the various retrospective studies performed in both Asian children [21,22] adults [23,24] with a significant reduction in the rate of ischemic recurrence, the aim of the surgery being to increase blood flow in hypo-perfused cortical regions. The prognosis of the disease is severe. Depending on the study, 50-90% of patients have a neurological deficit following repeated ischemic strokes, in 3-11% of cases the outcome will be fatal [25]

Therapeutics current data show the importance of surgical treatment as the reference method for the management of Moya syndrome, especially in patients with progressive recurrent symptoms. Several operative techniques using most often the external carotid artery circulation. Long-term studies have shown good results of surgical treatment The surgical indication should be discussed early enough within 3 months after the onset of symptoms, as early intervention can rapidly improve the patient's condition prevent ischemic recurrence [26, 27,28].

Abbreviations

- MMD (Moya Moya disease)
- GCS (Glasgow Coma Scale)
- EVG (encephalo-duro-arterio-synangiosis)
- MRI (Magnetic Resonance Imaging)
- ACM (Middle Cerebral Artery)
- ICA (Internal Carotid Artery)
- ACA (Anterior Cerebral Artery)
- EDAS (encéphale-duro-arterio-synangiosisc)
- HTIC (intracranial hyper tension)

4. Conclusion

Moya Moya is a rare chronic cerebral vascular disorder, idiopathic or secondary, constituting a significant cause of stroke. Diagnostic delay is frequent, due to its polymorphic symptomatology. Angio CT-scan, MRI are currently the reference imaging for initial diagnosis, monitoring. Arteriography remains a gold standard for the diagnosis,precise lesion assessment of this pathology. Vascular recurrence is frequent, particularly in the case of bilateral involvement, making it a serious disease that should be discussed on a case-by-case basis for surgical management of revascularization.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare that they have no competing interests.

Statement of informed consent

Consent for publication from the patient's guardians was taken.

Author contributions

DA MES collected the data wrote the manuscript YH,MH,NEL REM did the proofreading advised on corrections.

Reference

[1] Yasargil MG. Diagnosis indications for operations in cerebrovascular occlusive disease. In: Microsurgery applied to neurosurgery. Stuttgart: Georg Thieme Verlag, Academic Press; 1969;95–118

[2] Caldarelli M, Di Rocco C, Gaglini P. Surgical treatment of Moyamoya disease

[3] Suzuki J, Kodama N. Moyamoya disease – a review. Stroke 1983;14:104–9

[4] Suzuki J, Takaku A. Cerebrovascular ''moyamoya'' disease: Disease showing abnormal net-like vessels in base of brain. Arch Neurol 1969;20:288–99

[5] Bradley A, Du R. The natural history of Moyamoya in a North American adult cohort. J Cli Neurosci 2013;20:44–8

[6] Ikezaki K, Kono S, Fukui M. Etiology of Moya Moya disease: pathology, pathophysiology, genetics. In: Moyamoya disease. Rolling Meadows: American Association of Neurological Surgeons; 2001. 20–30

[7] Scott RM, Smith ER. Moyamoya disease and moyamoya syndrome. N Engl J Med 2009;360:1226–37

[8] Han H, Pyo CW. Associations of Moyamoya patients with HLA class I class II alleles in the Korean population. J Korean Med Sci 2003;18:876–80

[9] Fujimura M, Sonobe S. Genetics and biomarkers of Moyamoya disease: significance of RNF213 as a susceptibility gene. J Stroke 2014;16:65–72

[10] Liu W, Morito D, Takashima S, et al. Identification of RNF213 as a susceptibility gene for moyamoya disease and its possible role in vascular development. Plos One 2011;6:e22542.

[11] Calviere L, Catalaa I. Aspects cliniques et e´volutifs de la maladie de Moyamoya chez des adultes franc¸ais. Rev Neurol 2009;165:709–17.] Starke RM, Crowley RW. Moyamoya disorder in the United States. Neurosurgery 2012;71:93–9

[12] Kumar Garg A, Suri A. Ten year experience of 44 patients with moyamoya disease from a single institution. J Cli Neurosci 2010;17:460–3

[13] Fukui M. Guidelines for the diagnosis treatment of spontaneous occlusion of the circle of Willis ('moyamoya' disease). Research committee on spontaneous occlusion of the circle of Willis (Moyamoya disease) of the ministry of Health Welfare, Japan.)

[14] Hallemeier CL, Rich KM, Grubb Jr RL, Chicoine MR, Moran CJ, Cross 3rd DT, et al. Clinical features outcome in not American adults with moyamoya phenomenom. Stroke 2006;37:1490–6

[15] Arias EJ, Derdeyn CP. Advances surgical considerations in the treatment of moyamoya disease. Neurosurgery 2014;74(Suppl. 1):S116–25

[16] Hayashi K, Horie N, Nagata I. A case of unilateral moyamoya disease suffered from intracerebral hemorrhage due to the rupture of cerebral aneurysm, which appeared seven years later. Surg Neurol Int 2013;4:17.

[17] Scott RM, Smith ER. Moyamoya disease moyamoya syndrome. N Engl J Med 2009;360:1226–37

[18] Gudepu RK, Mohtashim A, Qureshi A, et al. Case report: a case report of Moyamoya disease in a 36-year-old African American woman. F1000Res 2014;3:297.] Gudepu RK, Mohtashim A, Qureshi A, et al. Case report: a case report of Moyamoya disease in a 36-year-old African American woman. F1000Res 2014;3:297

[19] Arias EJ, Derdeyn CP. Advances surgical considerations in the treatment of moyamoya disease. Neurosurgery 2014;74(Suppl. 1):S116–25

[20] Morel C, Rousselle C, Pelissou-Guyotat I, et al. Maladie de Moyamoya : intérêt d'un diagnostic et d'un traitement chirurgical pre´coces. A` propos de trois observations. Arch Pediatr 1999;6:1186–90

[21] Matsumoto H, Kohno K. Indications for one-stage extensive indirect vascular reconstructive surgery for pediatric moyamoya disease. Surg Neurol 2009;72:538–44

[22] Hallemeier CL, Rich KM, Grubb Jr RL, Chicoine MR, Moran CJ, Cross 3rd DT, et al. Clinical features outcome in north american adults with moyamoya phenomenom. Stroke 2006;37:1490–6.

[23] Arias EJ, Derdeyn CP, Dacey Jr RG, Zipfel GJ. Advances surgical considerations in the treatment of moyamoya disease. Neurosurgery 2014;74(Suppl. 1):116–25.