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Moyamoya Disease in an Adult Female: A Case Report

K. El Mansouri[1*], I. Oussayeh[1], Y. El Ouardi[1], M. Khallouki[1]

[1]CHU Mohamed VI Marrakech, Morocco

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***Corresponding author:** Khalid El Mansouri
CHU Mohamed VI Marrakech, Morocco

Abstract **Case Report**

Moyamoya is a progressive vasoocclusive disease of large intracranial arteries with characteristic collaterals formation. It has a bimodal distribution and more frequent in females compared to males. We present a case of 20 years female who presented with a loss of consciousness following headache. She was investigated with a head CT scan. And digital subtraction angiography which led to a diagnosis of Moyamoya disease. She was treated conservatively and scheduled for follow-up. Although ischemic attacks are the common presenting symptoms in cases of Moyamoya disease, hemorrhagic forms are seen too especially among adults compared to children. Identifying Moyamoya disease can significantly affect the treatment options and give insight into managing the chronic nature of the disease to both the physicians and patients. This case highlights the importance of searching for underlying cause in a young female presenting with intracranial haemorrhage for the first time and keeping Moyamoya disease as a differential.

Keywords: Moyamoya Disease, Angiography, Intracranial Haemorrhage, Ischemic Attacks, Young age.

1. INTRODUCTION

Moyamoya disease is a rare cerebrovascular disease characterized by progressive stenosis of the terminal part of the internal carotid artery and its main branches. It is associated with the development of dilated and fragile collateral vessels at the base of the brain, called vessels of Moyamoya [1].

Moyamoya, which means “puff of smoke” in Japanese, the ministry of Health and Welfare of Japan has defined 4 types of Moyamoya disease (MMD): ischemic, hemorrhagic, epileptic and “other”. It has been shown that the ischemic type predominates in childhood, while the hemorrhagic type is more often seen in the adult population [2].

Patients may present with headaches, Seizures or cognitive impairment due to an ischemic cerebrovascular accident (CVA) and a large proportion of patients, the majority of patients, present with a hemorrhagic stroke [3].

In this article, we present the case of a young woman with a hemorrhagic stroke, who was diagnosed with Moyamoya disease by cerebral arteriography complicated by postoperative meningitis.

2. PRESENTATION OF CASE

A 20 years female, with a history of an ovarian cyst, operated 3 months ago before admission, presented, sudden onset, with a chief complaint of headache followed by a loss of consciousness. There was no associated fever, neck rigidity, abnormal body movements, dizziness, vision, auditory, speech, and gait abnormalities. She does not have a significant history of past medical illness, surgical intervention, drug allergies or family history of any similar illness. the clinical examination found the patient obsessed and agitated. The Glasgow scale at 7/15 (E1V1M5) associated with signs of intracranial hypertension with anisocoria. An orotracheal intubation and invasive ventilation are performed.

On CT scan: deep left parietal hematoma with intraventricular extension and diffuse cerebral oedema (Figure 1). An emergency EVD was placed and postoperatively she was admitted to the ICU.

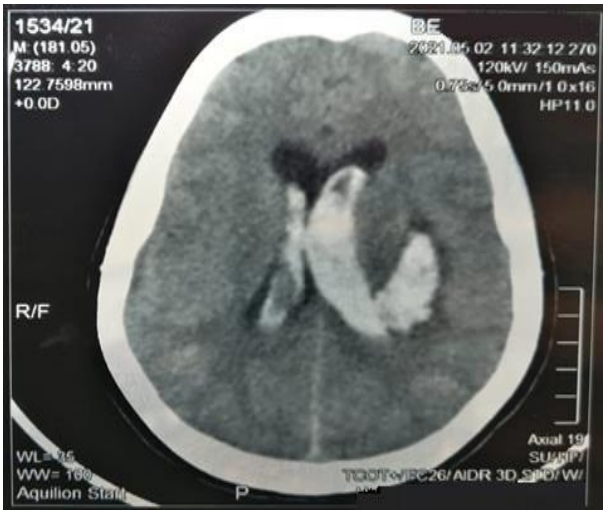

Fig-1: Deep left parietal hematoma with ventricular flooding

The subsequent etiological assessment by arteriography revealed grade III Moyamoya disease according to the Suziki and Takaku classification (Figure 2). In the ICU, the patient was quickly extubated after lifting the sedation. The evolution was marked by the installation of bacterial meningitis on day 18 of her hospitalization.

3. DISCUSSION

Moyamoya disease is common in Eastern Asia particularly in Japan, China, and Korea and there is a female predominance with a ratio of 1.9 [4]. It has a bimodal distribution with one of the peaks at 5–9 years and the other peak at 35–39 years of age [5]. Our patient was 20 years old, of African origin. . Although ischemic attacks are the common presenting symptoms in cases of Moyamoya disease, hemorrhagic forms are seen too especially among adults compared to children [6]. Moyamoya disease can present with features of stroke-like hemiparesis and speech impairment. They may also present with headaches or seizures. Seizures are less common usually. However, patients may be asymptomatic as well. Our patient presented with headaches, vomiting, and loss of consciousness. So, Moyamoya disease is one of the differentials that should be kept in mind in cases of young females with a severe headache. Studies have found haemorrhagic presentation in 10% of adults compared to a mere 2.5% in cases of children [8, 9]. Intraventricular haemorrhage was commonly found in Korea with or without intracerebral haemorrhage [10]. The presentation of our patient with deep left parietal hematoma with profuse tetra-ventricular hemorrhage and diffuse cerebral edema was similar to the findings of studies done in countries of China and Taiwan where intracerebral haemorrhage was found frequently [11]. Neuroimaging is important for the diagnosis of Moyamoya disease. The characteristic angiography finding is bilateral stenosis affecting the distal internal carotid arteries and proximal circle of Willis vessels, along with the involvement of prominent collateral vessels and it produces a characteristic hazy appearance like a puff of smoke in the air and hence, been named Moyamoya, a Japanese term meaning the same [12,13]. Although conventional cerebral angiogram is the gold standard for the diagnosis of Moyamoya Disease, non-invasive imaging like CTA and MRA has taken over as initial imaging modality in multiple centers around the world [14]. Assessment of the severity of vascular abnormality through angiographic staging is important for the determination of future risks of ischemia or haemorrhage [8]. Our patient underwent cerebral arteriography which revealed an aspect related to Moyamoya grade III disease according to the classification of Suzuki and Takaku.

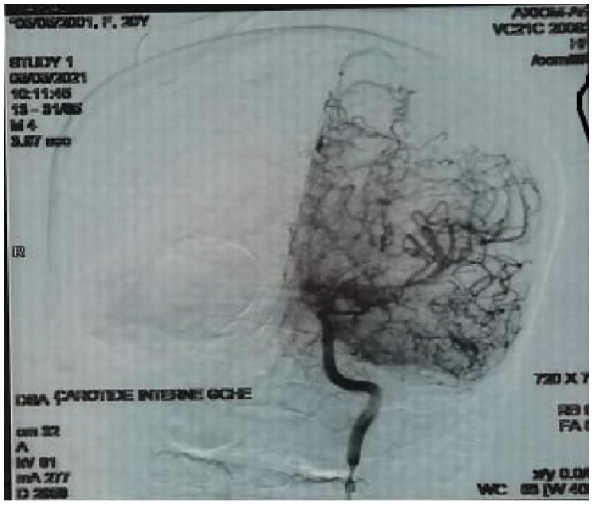

Fig-2: Cerebral arteriography with the aspect of Moyamoya disease

The treatment of acute episodes is mainly symptomatic with a focus on the reduction of elevated intracranial pressure, improvement of cerebral blood flow, and prevention of seizures [15]. An external ventricular drain was placed to reduce the intracranial pressure in our patient who had an intraventricular haemorrhage. It is important to manage pain and avoid hypotension, hyperthermia, and hypocarbia. Secondary prevention is mainly based on surgical revascularization which may be direct or indirect [6, 14]. Surgical revascularization is superior to conservative treatment with benefits with a great reduction in the risk of stroke even in a cluster of patients who had haemorrhage [16]. Our patient received conservative medical treatment with a placement of an external ventricular drain without surgical revascularization. The evolution was marked by a neurological degradation on multiresistant Enterobacter Cloacae meningitis, put under amikacin and colistin.

4. CONCLUSION

Moyamoya is a rare disease in the context of Africa. It should be considered a differential diagnosis in patients presenting with recurrent headaches or neurological symptoms in the bimodal age distribution it is seen. This case highlights the importance of

considering Moyamoya disease as a differential diagnosis in cases of young females presenting with intracranial bleed. Identifying Moyamoya disease can significantly affect the treatment options and give insight into managing the chronic nature of the disease to both the physicians and patients.

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