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Case Report

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A psychiatric clinical picture revealing moyamoya disease

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ABSTRACT

Moyamoya disease (MMD) is a rare entity. It is a chronic cerebrovascular pathology characterized by stenosis and progressive occlusion of the termination of the carotid arteries. It is of reserved prognosis. Its diagnosis can be evoked on computerized tomography and magnetic resonance imaging (MRI) but is essentially based on arteriography. Its treatment must be early and essentially consists of revascularization surgery. We report the case of a patient admitted to the psychiatric emergency room for management of a behavioral disorder, mutism and auditory and visual hallucination, a cerebral MRI was performed urgently and the diagnosis of MMD was laid. Through this observation and a review of the literature, we discuss the characteristics of this rare condition, in particular the psychiatric comorbidity.

Keywords: moyamoya disease, neuropsychiatric disorders, patient

INTRODUCTION

Moyamoya disease (MMD) is a rare chronic intracranial arteriopathy of undetermined origin, characterized by the presence of steno-occlusive lesions of the bifurcation of the terminal internal carotid arteries, with the presence of a nearby neovascular network producing a (swirls of smoke) hence the Japanese name Moya-Moya. Symptoms of the disease are very variable. Some affected patients remain asymptomatic, others develop transient accidents, and others still severe neurological deficits as a result of ischemic or hemorrhagic accidents. The disease was described in Japan in 1955 by "Takeushi and Shimizu" and was named Moya "Suzuki and Takaku" in 1969 [1]. A systematic review revealed that the incidence per 100,000 patient years ranged in Japan from 0.35 to 0.94 (95% CI 0.69 to 1.19), in the USA from 0.05 (-0.04 to 0.12) in Iowa to 0.17 (-0.06 to 0.40) in Hawaii and were 0.41 (0.28 to 0.54) in Nanjing, China and 0.02 (0.003 to 0.040) in Taiwan [2]. Imaging has a fundamental role in the positive diagnosis. Angiography remains the gold standard for the anatomical assessment of the disease. The prognosis of MMD is poor, currently the treatment of choice remains surgical revascularization [3].

The clinical expression of this disease is highly variable, particularly in adults. It can be noisy and characterized by the occurrence of infarction and/or cerebral hemorrhage or paucisymptomatic or even asymptomatic [4]. A typical or atypical psychiatric clinical picture may be an expression of this rare disease [5].

Neuropsychiatric disorders have been linked to several other brain diseases [6], in the case of MMD, the most frequent manifestations are depression, anxiety and other cognitive disorders [5]. However, only a few case reports of onset of a psychotic disorder following a stroke have been found.

We report a clinical observation of a patient initially presenting with psychiatric symptoms before being diagnosed with MMD.

THE OBSERVATION OF THE PATIENT

A 47-year-old patient, married, without children, without profession, she presents as a history of obesity, primary sterility and a dysmorphic syndrome (height: 137 cm, low hairline) admitted to the psychiatric emergency room for treatment (ArRazi Psychiatric Hospital in Salé) charged with mutism, odd behavior, and auditory and visual hallucinations.

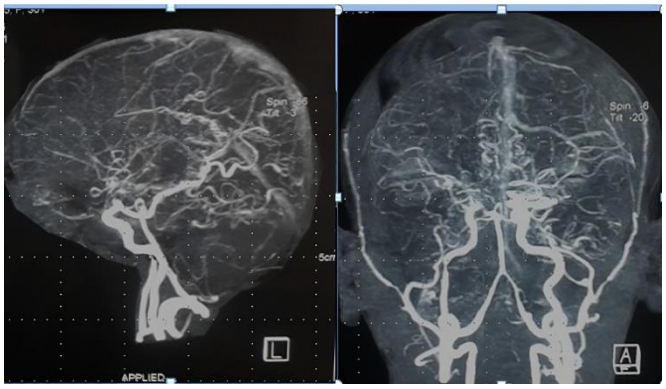

Figure 1.MRI angio: MR angio sequence: Absence of visualization of the 2 ACAs 'anterior cerebral artery' & occlusion of the two internal carotids with important arteriolar network on the level of the polygon of Willis (reprinted with permission of the patient)

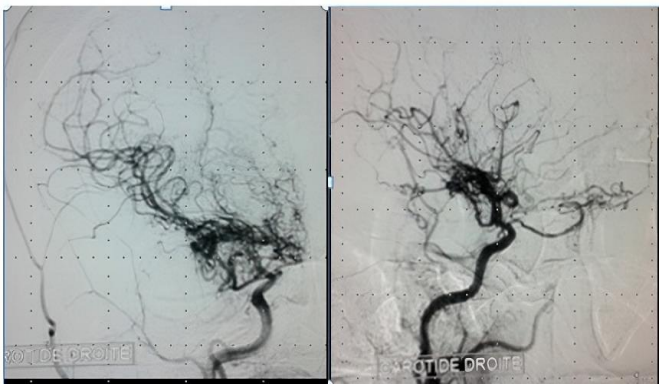

Figure 2. Images of the right carotid: Absence of opacification of the internal carotids' termination; fine anastomotic network creating a "smoke cloud" aspect at the level of the polygon of Willis; & moderate support of right M2 and anterior cerebral artery segments by this network (reprinted with permission of the patient)

The psychiatric interview with the patient and her entourage shows a change in behavior that goes back to one month by the installation of crises of agitation without triggering factor with notion of pseudo-epileptic seizures, incomprehensible vocalizations (cries) to answer his visual and auditory hallucinations and insomnia, the patient initially consulted at the health center and was referred to the psychiatric emergency room for treatment.

A paraclinical assessment was carried out, the biology returned without particularity, namely the blood count, the blood ionogram, the C-reactive protein, the glycemia, as well as the haemostasis and thrombophilia assessments. The fundus was also unremarkable. A radiological assessment was also requested showing calcified psammomas at the level of computerized tomography (CT) cerebral scan in spontaneous contrast, a cerebral magnetic resonance imaging (MRI) under sedation was requested in addition showing some small cerebral sequelae areas anterior deep and superficial bilateral junctions compatible with cerebrovascular accidents elders; an injection of gadolinium at the end of the examination showed occlusions of the terminal portions of the internal carotids with fine anastomotic networks against the current opposite the base of the skull, supplied mainly by the posterior cerebral (**Figure 1**).

An arteriogram was performed showing a lack of opacification of the internal carotid endings with fine anastomotic networks creating a "smoke cloud" appearance compatible with MMD (**Figure 2**), the patient was referred to the neurology department for complete supported.

The neurological examination finds as an anomaly a mute patient, the contact is difficult, an opposition at the level of the muscular tone, the coordination is difficult to evaluate, the examination of the sensitivity is difficult to evaluate, the cranial pairs and the superior functions are difficult to be evaluated and the patient is negative.

The patient received treatment with lorazepam, boluses of Solumedrol, and sodium valproate, enoxaparin with good clinical improvement of her negativism, the patient did not benefit from revascularization.

The evolution would be marked by a slight improvement on the clinical level, but the patient would have remained isolated in withdrawal, no longer doing her daily tasks and no longer taking care of herself, and she would also have presented a delirium of persecution centered on the entourage, which motivated the husband to consult in the psychiatric emergency room and the patient would be put on antipsychotic treatment based on risperidone four mg per day with very good clinical improvement

DISCUSSION

The pathogenesis of psychiatric symptoms in MMD appears to be multifactorial [7]. Cellular destruction of distribution territories of the internal carotid artery due to ischemic stroke, repeated transient ischemic attacks, or hemorrhage from aneurysms and collateral vascular networks may manifest immediately as psychiatric clinical pictures or progress with time with new vascular damage. However, patients without ischemic or hemorrhagic stroke also suffer from depression, anxiety and other symptoms, indicating a possible link with the pathogenesis of the disease, probably related to chronic hemodynamic insufficiency of small cortical vessels [7]. Finally, psychological precipitants, related to prognosis, poor quality of life and frustration due to cognitive impairment, probably contribute [8]. Early engagement in psychotherapy is strongly recommended.

Moyamoya, Depression, and Anxiety

A Korean report describes the case of a 45-year-old woman with depression and transient episodes of storytelling and inappropriate behavior. The authors initially thought her symptoms were secondary to depression until she developed right-sided hemiparesis and muscle weakness and MRI showed bilateral MMD [9].

In two studies [10, 11] of American adults with MMD almost one-third of patients reported suffering from clinically significant depression and clinically significant anxiety. Another Chinese study compared quality of life with MMD to

matched healthy control subjects, people with MMD reported higher rates of stress intolerance, poor interpersonal relationships and pessimism and 73% reported depression. Patients with MMD also had higher scores for obsessive-compulsive, depressive, and phobic anxiety symptoms [12]. An association was also observed between the degree of frontal lobe brain changes and paranoia [13].

For use of antidepressants in patients with MMD, selective serotonin reuptake inhibitors are preferred due to data on improved motor coordination after ischemic injury, relative lack of effects anticholinergic side effects compared to tricyclic antidepressants and low impact on blood pressure compared to some serotonin-norepinephrine reuptake inhibitors [14].

Moyamoya and Psychosis

The clinical presentations are very variable. Several case reports describe patients with insomnia, disorganization, restlessness and internal preoccupations, persecutory delirium of auditory and visual hallucinations, treated as schizophrenia, and on radiological investigations MMD was demonstrated [15] .Acute psychiatric symptoms in the setting of known MMD may also signal new ischemia. One report describes an eight-year-old boy with MMD who presented to outpatient psychiatry with irritability, delusions of persecution, poor autonomy, insomnia, and auditory and visual hallucinations. MRI then revealed new ischemic changes in the distribution of the left middle cerebral artery [16].

Two clinical picture close to our observation: Mrs. A is a 54-year-old Chinese-American woman who presented to the emergency room for management of a psychotic clinical picture. She could not provide a coherent story and appeared withdrawn, selectively mute and confused. The psychiatrist requested paraclinical and radiological examinations and thus a diagnosis of MMD was made on the results of a brain MRI [7]. A 44-year-old man from Bangladesh presented with acute psychotic episodes of visual, auditory and tactile hallucinations, paranoia, somatoform disorders, and non-epileptic seizures. He had a history of mental disorders for three years and was later admitted to hospital with a headache and hemiparesis secondary to intraventricular hemorrhage. Neuroimaging showed characteristic features of MMD [5].

Patients with MMD exhibit high susceptibility to neuroleptics, with frequent reports of extrapyramidal symptoms and neuroleptic malignant syndrome, particularly with first-generation agents [15]. Impaired basal ganglia function due to abnormal vessel collateralization or ischemia may account for this tenderness, which is similar in dementia with lewy bodies. Antipsychotics, however, are effective in reducing hallucinations and paranoia [4, 13]. Second-generation agents, such as quetiapine, are preferred given the lower risk of extrapyramidal complications. Dosage reduction after prolonged resolution of symptoms seems reasonable, particularly if symptoms occurred before surgery. Antipsychotic treatment may also help with motor tics if present [4].

CONCLUSION

Transient ischemic events including MMD can easily be confused with multiple psychiatric clinical pictures, highlighting the need for thorough screening for precipitating triggers and characterization of symptoms and multidisciplinary management. Therefore, in the presence of any atypical psychiatric presentation, clinical and radiological investigations should be performed to look for other etiologies.

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Ethical statement: Authors stated that the study did not require approval from ethics committee since it is a study of the type: case report. The patient was informed of the conditions related to the study and gave his written and informed consent, respecting the anonymity and confidentiality of the data.

Data sharing statement: Data supporting the findings and conclusions are available upon request from corresponding author.

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