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Description of a Probable Moya-Moya Disease in a Caucasian Subject

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ABSTRACT

Introduction: *Moya-Moya disease is a rare cause of stroke in young people, affecting most of the Asian population. It is a chronic cerebrovascular disease characterized by stenosis and progressive occlusion of the intracranial internal carotid artery termination and the proximal portion of the Willis polygon arteries. It is from prognosis reserved. Its diagnosis is essentially based on arteriography. Its treatment should be early and consists of a revascularization surgery. Through a singular observation the authors report a probable case of Moya-Moya, in a patient of French origin.*

Aim: *Describe the clinical peculiarities of a patient of French origin suffering from Moya-Moya disease.*

Observation: *A Caucasian of French origin aged 53-year-old presented a 3rd episode of sudden recent motor deficit, secondary to a left sylvian infarction. The stroke assessment carried out made it possible to highlight a probable Moya Moya disease thanks to an arteriography. It benefited a medical treatment, coupled with a motor rehabilitation.*

Keywords

Moya Moya disease, Young person, Europe, Arteriography.

Introduction

Moya-Moya disease is defined as an angiogenic disease associated with bilateral and progressive stenosis of the cerebral arteries located at the base of the brain with the development of a collateral network made of fragile neovessels, which gives smoke-cigarette appearance "(Moya Moya in Japanese) [1]. It has been described for the first in 1957 in Japan by Takeuchi and Shimizu [4].

It is a rare but serious etiology of stroke in young people, whose diagnosis and management have been refined in recent decades with MRI and surgery.

The authors report the observation of a young case, of French origin while relating it to the data of the literature finally to identify its clinical particularities.

Aim

Describe the clinical peculiarities of a patient of French origin suffering from Moya-Moya disease.

Observation

A Caucasian of French origin aged 53-year-old resident in Soissons is admitted to the Neurovascular Intensive Care Unit (ICUU) on 24/07/2019 for motor aphasia and right hemiparesis noticed on awakening. This is the third episode of recent acute neurological deficit in this patient. It presented a first Sylvian left superficial stroke in 2014 and a second Sylvian superficial left ventricular puncture in 2016, of undetermined etiology. There is no family history of stroke or other personal or family FRCV.

Neurological clinical examination revealed a total flasco-spastic right hemispherical pyramidal syndrome associated with motor aphasia and right HLH. Brain MRI showed left sylvian infarction with diffusion and FLAIR positive and distal occlusion in M1. Not thrombolyzed because certainly out of time (waking stroke), He was transferred to the Reims hospital for performing a thrombectomy. After the failure of the latter, he was admitted again in neurology at the Soissons hospital for the continuation of the care.

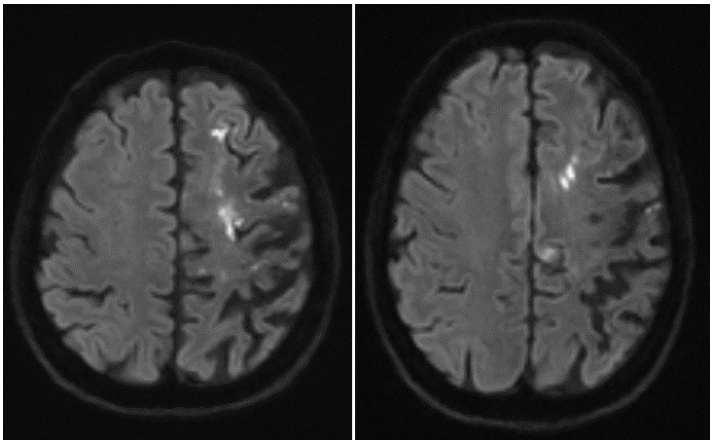
The complementary explorations with etiological aim of this 3rd recurrence did not show cardio-embolism nor of atherosclerosis. There was no evidence for systemic disease or antiphospholipid

syndrome. The thrombophilia assessment was not contributory. On the other hand, an inflammatory assessment showed accelerated VS at 29 then leading to angiitis. The CSF study revealed a clear CSF, with 2 lymphocytes with a protein content of 0.39 g / l, a glycorachia of 2.91 mmol / l and a chlorurachia of 131 mmol / l.

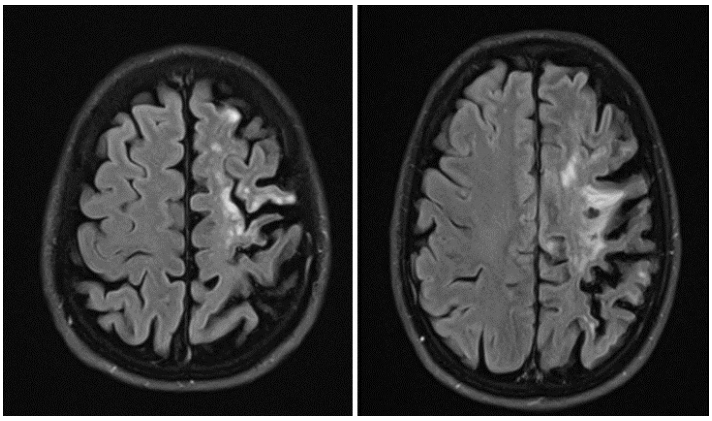
The arteriography performed showed, on the one hand, multiple chronic occlusions of left M3, M4 and left A3, A4 segments with a large network of collaterals giving a Moya-Moya pseudo appearance, and on the other a dysplasia of left internal carotid (CIG) in its intracavernous portion with ulcerated plaque appearance (probable additional dissection).

The diagnosis chosen was that of Moya-Moya disease associated with probable left cervical arterial dissection. Management was based on the administration of antiplatelet agents and statins with functional and speech therapy rehabilitation. A brain scan of control at 24H of hospitalization has objectified a stability of its lesions.

The short-term evolution was towards a minimal regression of aphasia. Rankin's score at the exit was 2. It keeps to this day a spastic hemiparesis and a lack of the word.



Figures 1 and 2: Cerebral MRI axial sections (diffusion sequence) left superficial sylvian infarction appearing hyperintense.



Figures 3 and 4: Axial MRI cuts (FLAIR sequence) superficial left superficial infarction appearing hyperintense.

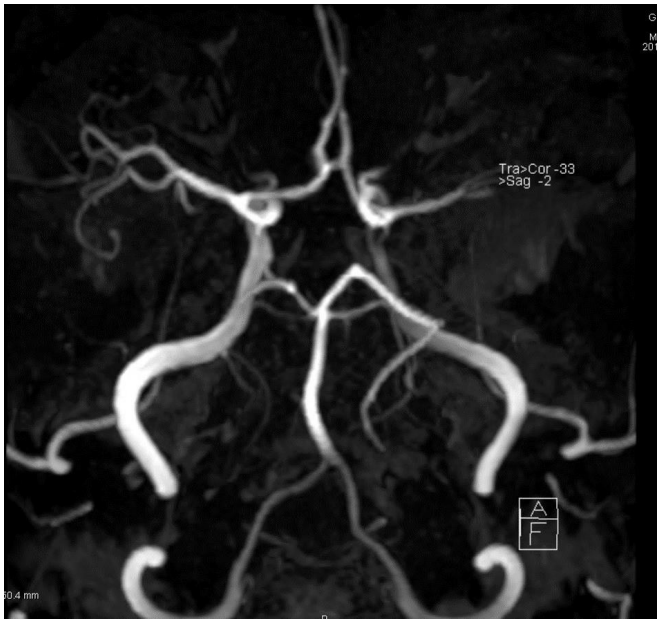
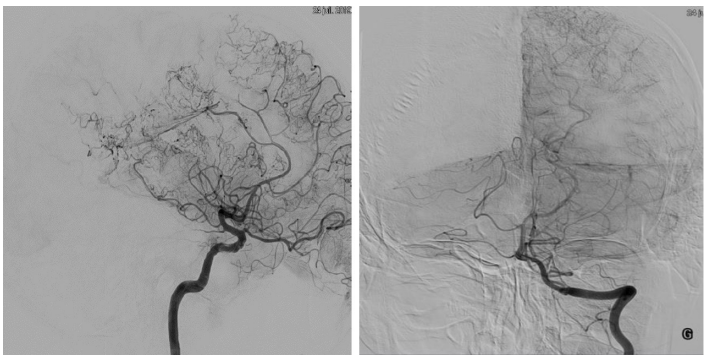


Figure 5: MRI brain scan TOF sequence: M1 occlusion on the left.



Figures 6 and 7: Arteriography brain: Moya-moya aspect on the left.

Discussion

Epidemiologically

Moya-Moya disease is a rare cerebrovascular entity and remains of unknown etiology. It is characterized by a stenosis of the internal carotid arteries with spontaneous development of a collateral vascular network at the base of the brain. The literal translation of the term Moya Moya means "a foggy puff of cigarette sailing in the air" in reference to the angiographic aspect found in the disease [9]. It is classically reported in Asian populations [1], but is described more and more in recent years in European populations [2,3]. Its prevalence is estimated at 1/30 000 in Japan against 1/300 000 in France [1] according to Fuzier in 2015.

It remains poorly known in non-Asian populations. Calviere et al. in 2009 in a Toulouse series of 12 cases reported an average age of 31.1 years [3]. Our subject, although young, is well above average reported in France as well as in other Western series. This difference could be explained by the delayed diagnosis observed in our patient because it is after a third ischemic episode that the disease was found. The female predominance is classically described but can also affect men [2,3] as illustrated in our observation.

The pathogenesis of this affection is indeterminate. There are family forms, but most often sporadic forms [1]. Its mode of transmission is of the autosomal recessive type (10% of cases) [1]. Our patient had no family history or FRCV that could lead to a hereditary character.

There are two types [1]:

- Moya-Moya disease limited to intracranial artery involvement. It can be idiopathic but there are family forms.
- Moya-Moya syndrome or secondary Moya-Moya, usually associated with a local or general condition (liver, heart), radiotherapy. It may appear during sickle cell disease or in patients with neurofibromatosis type I or Williams syndrome.

Clinically and paraclinically

The clinical presentation of Moya-Moya disease is very polymorphic, mainly represented by ischemic manifestations in children and haemorrhagic in adults. It can be asymptomatic or, on the contrary, manifest itself by focal signs: hemiparesis, hemichorea of headaches, convulsions, speech disorder and deterioration of higher functions [1]. Our patient presented, like classically reported signs in the literature, left hemiparesis associated with motor aphasia. Calviere in his series found ischemic strokes (CVA) as a mode of revealing the disease in ten cases [3]. Our patient has similar characteristics. Blanc, however, in a series of 10 cases published in 2015, showed that Moya-Moya disease manifested as ischemic stroke in 6 patients and haemorrhagic stroke in 2 others, while both had presented either transient ischemic attacks or exertional discomfort [3].

Imaging plays a central role in the diagnosis, the therapeutic strategy and the follow-up of the disease [4]. The diagnosis is radiological [1-3]: CT, angio-MRI, cerebral angiography: ischemic accidents of different ages, haemorrhages and abnormal vessels of the base. Conventional arteriography remains the reference exam [4]. It allows preoperatively to visualize intra and extra cranial vasculature. The diagnosis is based on the presence of stenosis and / or bilateral occlusion of carotid siphons and / or proximal portions of the middle carotid arteries (ACM) and anterior carotid arteries (ACA) [4]. The diagnosis is certain in the presence of bilateral anomalies, probable if they are unilateral [4]. It is thanks to this imaging technique that the diagnosis of Moya-Moya disease was highlighted in our observation. However, it remains a probable diagnosis, as evidenced by the work of Lahlou Mimi and Ouajdi [5,8] in view of the asymmetry of the lesions presented by our patient, but also because no other inflammatory or infectious cause has been highlighted. According to Ancelet et al. [6], catheter angiography and the techniques of nuclear medicine remain the standard examinations for morphological and hemodynamic evaluation. Nevertheless, noninvasive and non-invasive MRI is becoming increasingly important in management.

Therapeutically

Several therapeutic alternatives are discussed as evidenced by Thibaud et al. In their work [7]: therapeutic abstention, a drug treatment with or without surgery. The purpose of drug therapy is to prevent ischemic injury or treat a possible etiology. There is no curative treatment and each indication must be carefully asked to avoid its complications, especially hemorrhagic [7]. Thus, various drug classes can be proposed: platelet antiaggregants such as low aspirin doses, vasodilators, anticonvulsants, anticoagulants, corticosteroids, calcium channel blockers, nonsteroidal anti-inflammatory drugs [7]. Our patient benefited from the low-dose salicylic acid (75 mg) combined to atorvastatin 80 mg. Which is in line with most of the medications follow up elsewhere [3,4,7].

Therapeutic abstention or drug treatment (antiepileptics, calcium antagonists or platelet aggregation inhibitors) represent the two main management modalities of the patients followed in the Thibaud et al department. Only one patient was placed on calcium channel blockers (Nidrel®) whereas these have shown their efficacy in increasing cerebral perfusion in Moya Moya disease [7].

Surgery is also important for many authors in the treatment of Moya Moya disease [7,9,10]. Surgical techniques aim to establish collaterals by stimulating the formation of neovessels in order to restore blood flow in the ischemic areas. According to Jecko et al, surgery is a simple and effective method to prevent the recurrence of the manifestations of Moya-Moya disease and to stabilize or improve the neurological prognosis [9]. Despite its complexity and risks, surgery is retained by many authors [1]. It helps to prevent ischemic accidents, delay the development of arteriovenous malformations and improve symptomatology.

Thibaud et al. reported through two cases the association of a unilateral Moya-Moya network with an arteriovenous malformation, already described in the literature [7] (causal role of cerebral ischemia secondary to moya moya disease in the development of arteriovenous malformations).

Evolutionarily

The evolution can be interspersed with complications of type [1]: intellectual deterioration (50%), definitive neurological deficit (50-90%), death (4% in children, 10% in adults) by cerebral hemorrhage. Our patient did not remain on the sidelines of these data since he presented two recurrences on the ischemic mode. Calviere found just as in our observation, a cumulative probability of first recurrence of stroke was 50% at one and five years. At the end of the follow-up of its Toulouse series, four patients had no functional sequelae (Rankin score 0 or 1), seven had a moderate handicap (Rankin score 2-3) and one had died. Six patients had cognitive impairment suggestive of dysexecutive syndrome. Our patient also had a moderate disability with a Rankin score = 2. No cognitive impairment was reported in our observation, contrary to the usual data that highlights cognitive deterioration as a major complication of Moya Moya disease [1,3].

Conclusion

Moya Moya disease in young adult non-Asian is a rare disease but serious because of the high risk of stroke recurrence. Its diagnosis can be evoked on CT and MRI, but is essentially based on

arteriography. Its treatment should be early and consists essentially of a revascularization surgery. It is of reserved prognosis.

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