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Discovery of a Moya-Moya Vascular Disease during an Inaugural Epileptic Seizure Leading to the Discovery of Ischemic Sequel: A Case Report and Review of the Literature

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Abstract **Case Report**

Moya-Moya disease is a vascular disease. In its typical form, it is characterized by a narrowing of the distal internal carotid artery that extends to the proximal segments of the middle and anterior cerebral arteries, resulting in the formation of collateral vessels. We report on a 49-year-old female patient who initially presented with epileptic seizures and was diagnosed with this condition.

Keywords: Moya Moya – Secondary- Epileptic seizure.

INTRODUCTION

Moya Moya disease is a vascular disease. In its typical form, it is characterized by a narrowing of the distal internal carotid artery that extends to the proximal segments of the middle and anterior cerebral arteries, resulting in the formation of collateral vessels. These arise from parenchymal, perforating, leptomeningeal and other transdural anastomoses. These collateral vessels have a characteristic appearance on angiography forming a smoke cloud appearance called Moya Moya network. We report on a 49-year-old female patient who initially presented with epileptic seizures and was diagnosed with this condition.

CLINICAL OBSERVATION

The patient was a 49-year-old woman with no previous history of any particular pathology who presented to the emergency room with epileptic seizures. The clinical examination, particularly the neurological examination, was unremarkable.

First, a brain scan without injection of contrast was performed and came back without any particularity apart from bilateral calcifications of the lenticular nuclei (Figure 1). As there was no answer as to the cause of the seizures, an off-line brain MRI revealed rare non-specific T2-FLAIR hyper signals in the supratentorial, bi-hemispheric and peri-ventricular white matter, as well as sequential lesions in the occipital region, the ventricular crossroads and the left anterior frontal region (Figure 2).

On arterial MRA sequences, there is bilateral distal stenosis of the terminal segment of the internal carotid arteries with a sparse appearance of the cerebral arteries with a network of anastomotic collaterals of the base making the classic "smoke volute" appearance and a sparse appearance of the right posterior peri-callosal artery and the left posterior cerebral artery (Figure 3).

An injected CT scan showed the signs described above as well as leptomeningeal enhancement with an "Ivy-sign" appearance, typically found on acquisitions after contrast injection (Figure 4). (Figure 4)

The diagnosis retained was that of a Moya Moya type vasculopathy discovered during an inaugural epileptic attack.

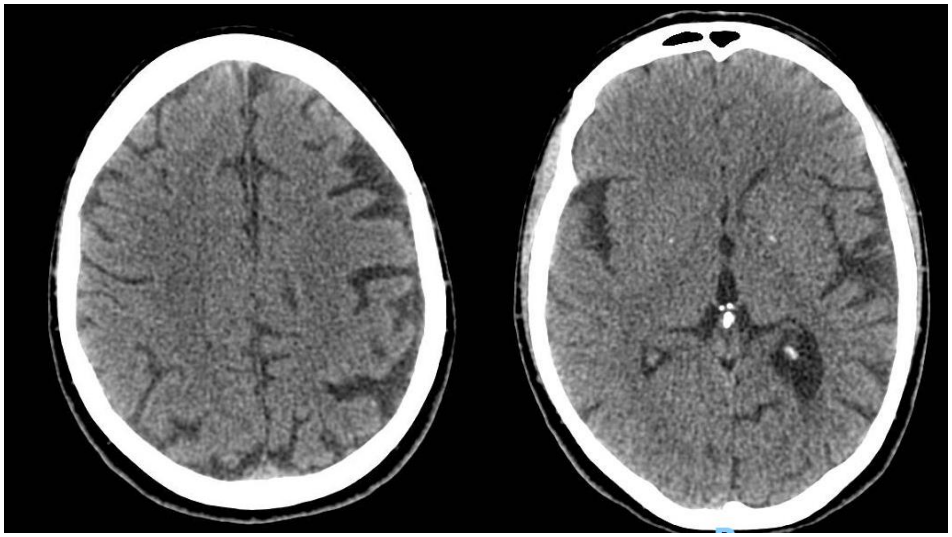

Figure 1: Cerebral CT scan without injection of contrast medium in axial sections revealing no notable abnormality apart from bilateral calcifications of the lenticular nuclei

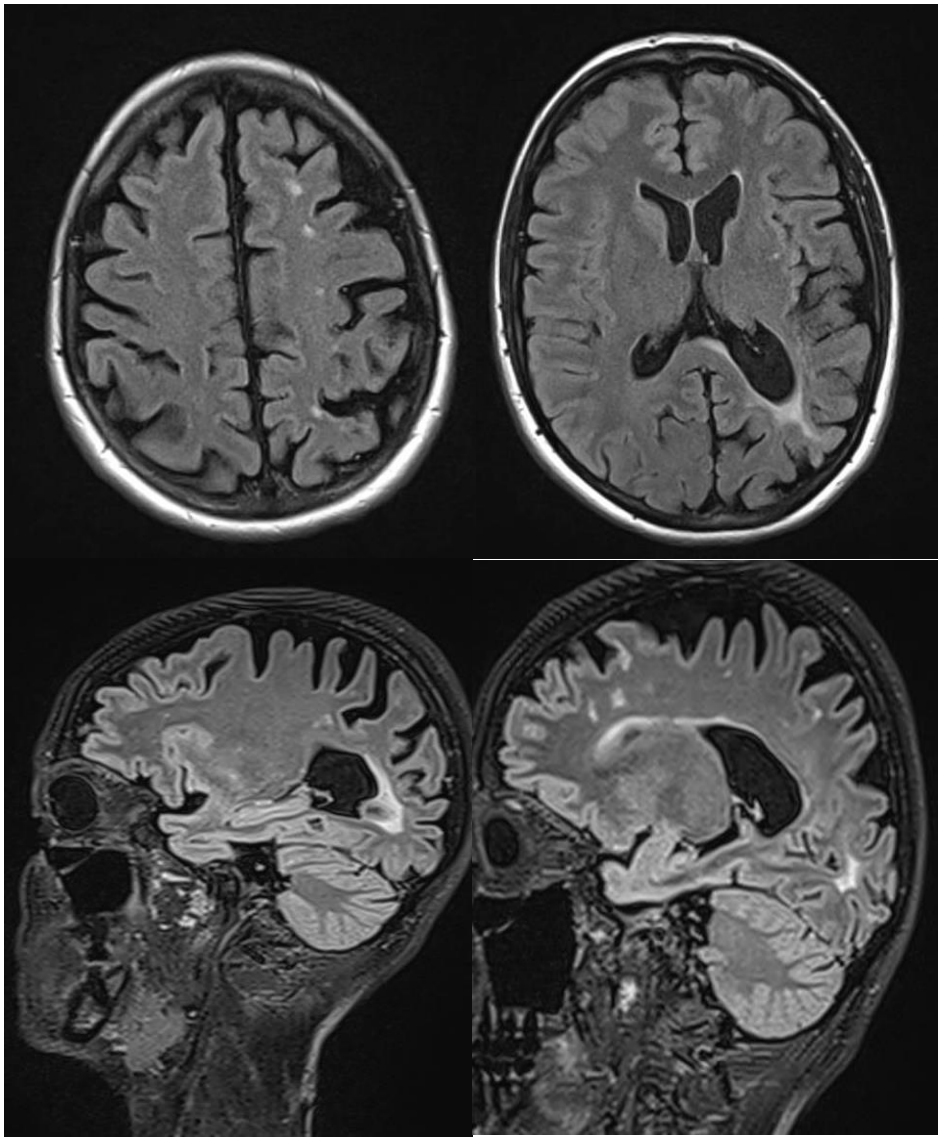

Figure 2: Brain MRI in FLAIR sequence in axial and sagittal sections showing sequelae of the occipital region, the ventricular crossroads and the left anterior frontal region

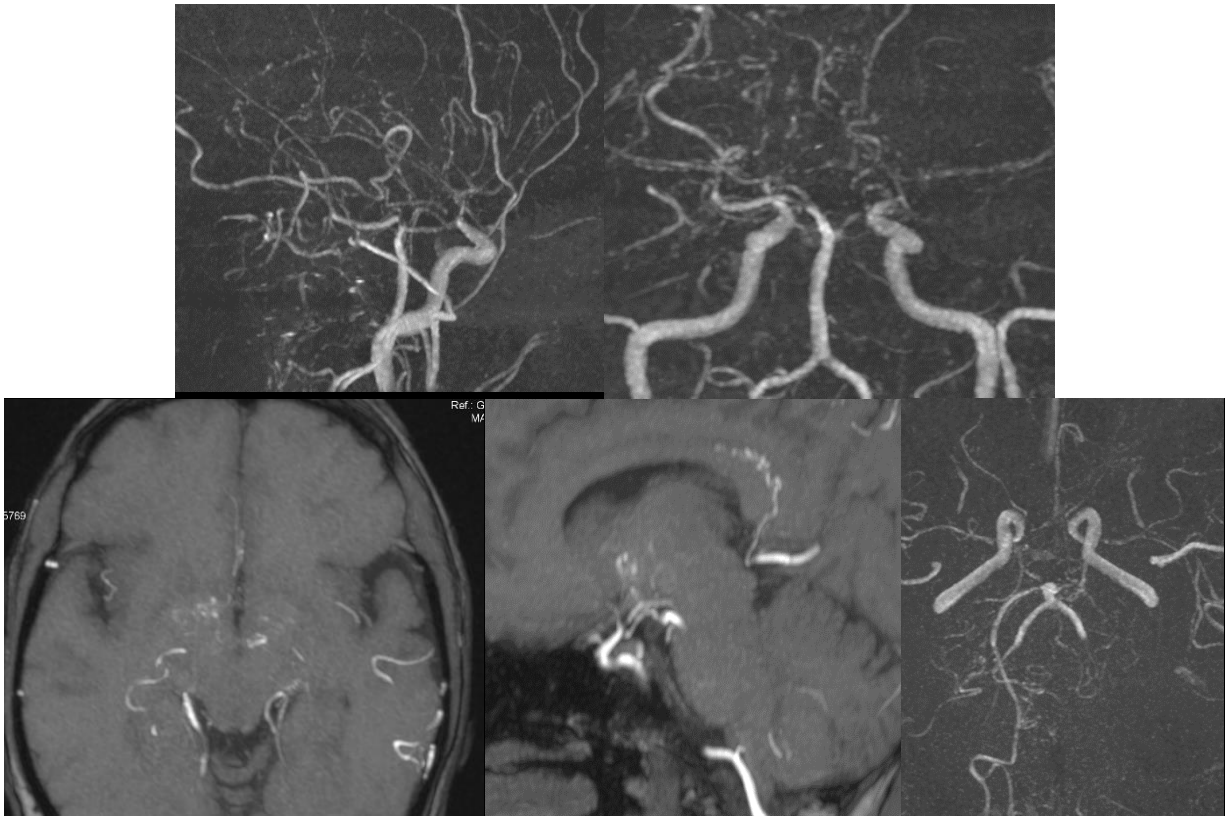

Figure 3: Arterial MRA demonstrating bilateral distal stenosis of the terminal segment of the internal carotid arteries with impingement and small aspect of the cerebral arteries with a network of anastomotic collaterals of the base realizing the classic aspect of "smoke volute" and right posterior peri-callosal as well as a small aspect of the left posterior cerebral artery

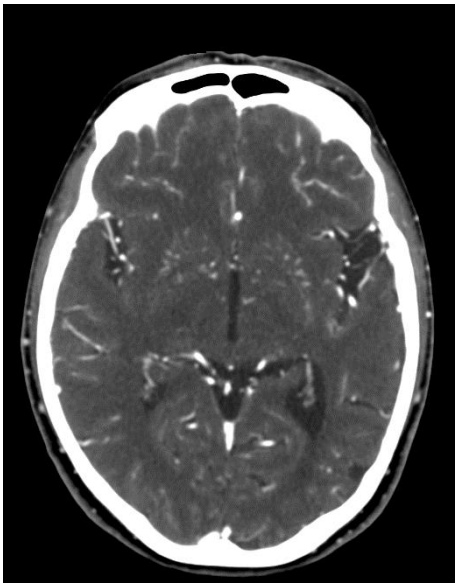

Figure 4: Ivy sign

DISCUSSION

In Japanese, the term "Moya Moya" means smoke cloud; language used to describe the angiographic appearance of a network of collaterals induced by progressive arterial stenoses or occlusions in the basal segment of the intracranial arteries, usually the anterior (A1) and middle (M1) cerebral arteries and the terminal portion of the carotid arteries. In our case, we observed involvement of the left posterior cerebral artery, which appeared to be very small.

This typical appearance is thought to be unique to Asian children [1]. Indeed, according to the literature, this syndrome was first described in Japan. There are two age peaks: children around 5 years of age

and adults around 40 years of age, with women being twice as affected as men. It is the most common pediatric cerebrovascular disease in Japan with a prevalence of about 3/100,000 [2]. Our patient was not of Asian origin and has no ancestry with this syndrome.

The pathogenesis of this condition is not well understood, but the literature reports a number of factors that may be involved, including genetic and environmental factors. There is a hypothesis of a chronic idiopathic arteriopathy, with progressive thickening of the cerebral artery walls, which leads to the production of angiogenic factors responsible for the formation of new replacement vessels, which are of three types: 1) Moya Moya vessels: formed via intra parenchymal perforating arteries; 2) lepto meningeal collateral vessels of the posterior cerebral artery: gives the "Ivy sign" (figure 4) and transdural collateral vessels of the middle meningeal artery, superficial temporal artery, ethmoidal artery and/or occipital artery [3].

Clinically, the symptomatology is varied, ranging from transient ischaemic attack to haemorrhagic attack due to rupture of the anastomotic network found in the pathology [1]. The disease can also be discovered, as in our observation, during recurrent epileptic seizures, notably in 20% of cases, in particular after haemorrhage or cerebral infarction [4]. Our patient had sequelae of ischaemia which would obviously be the cause of the seizures and in this context, the vascular involvement is probably of secondary origin given its non-Asian origin and the late discovery which obviously is more in favour of a secondary origin. Thus, according to the literature, a distinction is made between moya moya of primary origin, which is the prerogative of Asian subjects, particularly in Japan and Korea, and moya moya of secondary origin, which may be part of a syndrome such as neurofibromatosis type 1, or in inflammatory states, pre-thrombotic states, premature ageing, congenital mesenchymal defects and suprasellar irradiation in children [1].

Radiologically, several modalities contribute to the diagnosis. On CT scan, punctiform contrast in the basal ganglia and an abnormal arterial network in the base of the brain are demonstrated. The cerebral angioscanner shows an abnormal polygon of Willis with collateral arterial network. In our patient, after injection of contrast medium, a gyriform contrast pattern was found, indicating leptomeningeal contrast.

Magnetic resonance imaging has an important role in the diagnosis. In typical cases, a hyper-signal appearance of the small vessels, cortical ischemia and white matter is observed on T2. The FLAIR sequence shows a hyperintense appearance of the cortical sulci called the leptomeningeal ivy sign. In T2*, signs of haemosiderosis are found in case of previous haemorrhage and sometimes asymptomatic microbleeds in adults. The diffusion sequence is useful for the detection of ischaemia and should always be correlated with the FLAIR sequence for lesion dating. Lenticulostriate collaterals present as pumiceous basal ganglia contrast and/or thin cisternal vascular structures with a reticular appearance. Angio-MRI shows stenosis of the distal part of the internal carotid arteries and the proximal segments of the Polygon of Willis. The anterior circulation would be the most affected and posterior involvement is more rare. What makes our observation special is the fact that the posterior circulation is affected. Venous angiography can also reveal venous vasculopathy [5].

Angiography is useful in the study of collaterals and the evaluation of the stages of evolution described above.

In total, three diagnostic criteria should be considered: the presence of stenosis or occlusion of the terminal internal carotid artery or of the proximal segments of the anterior and middle cerebral arteries (classically); an abnormal vascular network or hypointensities of the grey nuclei; and finally, the bilateral character, bearing in mind that atypical cases of unilateral involvement are possible.

CONCLUSION

Moya Moya disease is a rare condition that can reveal cerebral ischaemia. Seizures may be the initiating sign. Several imaging modalities contribute to the diagnosis. MRI and angiography are currently the gold standard for diagnosis showing stenosis of the distal part of the internal carotid arteries and the proximal segments of the Polygon of Willis. Posterior circulation can be abnormal like in our clinical observation.

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