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Moya Moya Disease: A Rare Cause of Ischemic Stroke in Children: About Two Cases

Meryem Raoui[1*], Yousra Sekkat[1], Daoud Bentaleb[1], Meryem Harmak[1], Dalal Laoudiyi[1], Kamilia Chbani[1], Siham Salam[1]

[1]Radiology Division, Ibn Rochd University Hospital, Abderrahim Harouchi Mother and Child Hospital, Casablanca 20250, Morocco

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***Corresponding author:** Meryem Raoui
Radiology Division, Ibn Rochd University Hospital, Abderrahim Harouchi Mother and Child Hospital, Casablanca 20250, Morocco

Abstract | **Case Report**

Moya Moya disease (MMM) is a chronic cerebral arteriopathy of undetermined etiology, characterized by steno-occlusive lesions of the terminus of the intracranial internal carotid arteries and the proximal part of the arteries of the polygon of Willis, with development of a fine anomalous compensatory vascular network. It affects both children and young adults, and is a frequent cause of stroke in children. Imaging plays a fundamental role in diagnosis, therapeutic strategy and follow-up. Non-invasive, non-irradiating MRI plays a fundamental role in monitoring this disease. The prognosis for MMM is guarded: in the absence of treatment, symptoms worsen and the risk of stroke increases. Surgical revascularization techniques appear to improve cerebral perfusion. We report the case of two children with Moya-Moya disease.

Keywords: Moya-Moya Disease, Child, Vasculitis, AVCI, Angioscanner, Angio-MRI.

INTRODUCTION

Moya Moya disease is a rare cerebrovascular pathology characterized by progressive stenosis of the cerebral arteries, mainly the internal carotid arteries and their main branches.

It leads to the formation of a collateral network of small vessels, which, on angiographic imaging, appears as a "smoke cloud", hence its name of Japanese origin "Moya Moya" meaning "smoke cloud".

This condition is a rare cause of ischemic stroke, particularly in children.

PATIENT AND OBSERVATION

Observation 1:

- A 2-year-old male child was admitted 48 hours ago with sudden onset of left hemiplegia.
- The patient underwent cerebral MRI, which revealed patchy and nodular cortico-subcortical fronto-parietal, periventricular and lenticulostriate lesions on the right in T1 hyposignal, T2 hypersignal and FLAIR, with diffusion restriction. **(figure 1,2)**
- A right fronto-parietal cortical thickening following the shape of the sulci was also noted in T1 hypersignal, FLAIR with diffusion restriction.
- Angiographic sequences showed: **(figure 3)**
  - Sténose occlusion of the termination of the right ACI with irregular stenosis of the proximal portion of the homolateral ACM.
  - Aspect irregular stenosis of the left ACM, which remains permeable.
  - Développement of bilateral lenticulostriate ollateral circulation.
  - Absence anomaly on venous angiographic sequence.

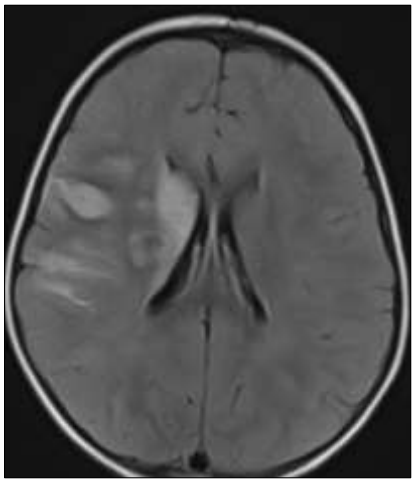

Figure 1, 2: Axial section T2 FLAIR sequence

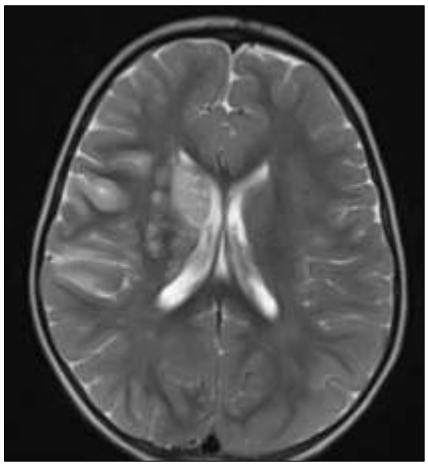

Figure 1, 2: Axial section T2 sequence

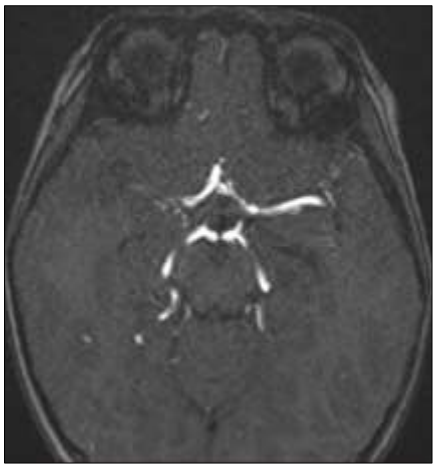

Figure 3: Axial section Angiographic sequence

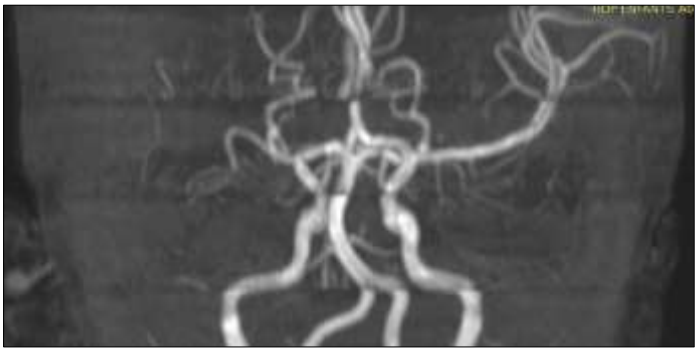

Figure 4: Angiographic sequence

This led us to suggest a sub-acute ischemic stroke of the entire sylvian territory due to Moya Moya vasculitis.

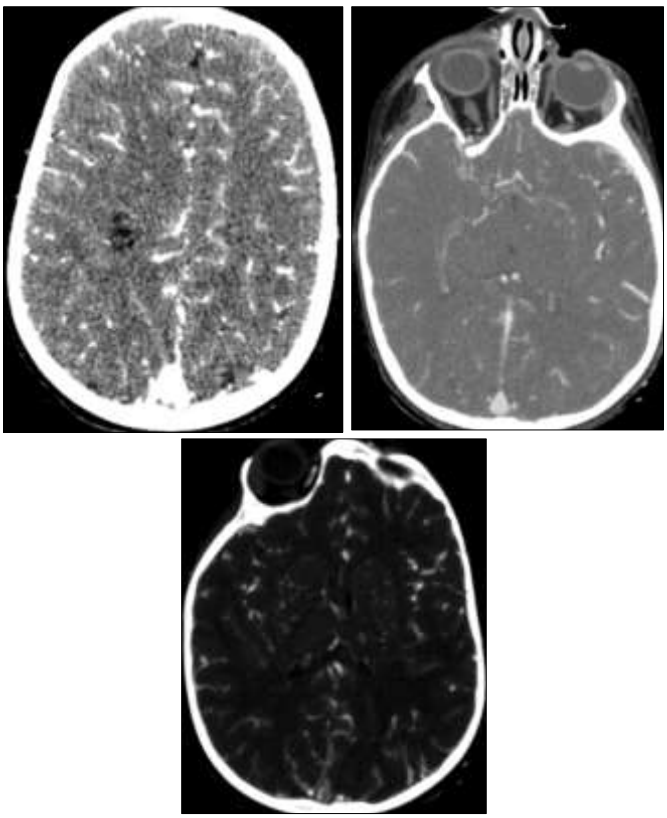

Figure: 1, 2, 3

Observation 2:

- A 3 years-old girl with a history of stroke and transient ischemic attack underwent cerebral angioscanning for suspected Moya Moya disease.
- The cerebral angioscanner revealed **(figure 1,2,3)** a small aspect of the internal carotid arteries with bilateral occlusion of the initial portion of the ACM over a distance of 6.8mm on the right and 7mm on the left.
- The development of a large network of collaterals around the occluded segments described above was also observed, enabling repermeabilization of the downstream ACMs.
- The downstream cirulation is small but permeable.
- The distal parts of the M1 portion are sparse and irregular, more marked on the right.
- The distal portions of the right ACM are sparse compared with the left.
- The PCAs, ACAs and vertebral arteries are of normal caliber, permeable and normo opacified.
- There was also a deep right fronto-parietal hypodensity area of similar density to that of the CSF.
- In view of these findings, the diagnosis of Moya Moya cerebral angiopathy complicated by a right fronto-parietal sequellar ischemic lesion was evoked.

DISCUSSION

The incidence of ischemic stroke in children is estimated at between 1.2 and 2.7/100,000 per year, with Moya Moya accounting for 10-15% of the total [1], characterized by progressive occlusion of the distal portions of the supraclinoid internal carotid arteries, and/or the proximal portions of the middle and/or anterior cerebral arteries, with the development of an irregular suppletive vascular network creating a "smoke cloud" appearance, Moya Moya in Japanese.

These vessels are pathological, fragile and may present dilatations and microaneurysms, leading to the risk of rupture and intracerebral and/or ventricular hemorrhage.

The term Moya Moya refers only to the distinctive findings on cerebral arteriography, irrespective of the cause.

By definition: Moya Moya disease is distinguished from Moya Moya syndrome, which is associated with other diseases such as: neurofibromatosis type 1, arteriosclerosis, trisomy 21, neoplasia, head trauma, meningitis, previous radiotherapy, sickle cell disease and autoimmune disease.

In Moya Moya disease, arteriographic findings are pathognomonic.

Involvement is bilateral, although severity may vary from one side to the other.

However, 40% of patients initially present with unilateral disease, which may progress to bilateral disease.

Moya Moya is the most common pediatric cerebrovascular disease in Japan, with a prevalence of around 3/100,000 [1].

Although the cause and pathogenesis of Moya Moya disease are still poorly elucidated, genetic factors play a major role: the familial incidence of affected first-degree relatives in Japan is 10%, and 6% in a recent series in the USA [1].

The Moya-Moya pathogen most likely involves both genetic and environmental factors. The pathophysiology is poorly understood, but is linked to chronic idiopathic arteriopathy, with progressive thickening of the cerebral artery walls.

This leads to the production of angiogenic factors responsible for the formation of replacement neovessels of three types: 1) Moya Moya vessels: formed via intraparenchymal perforating arteries; 2) leptomeningeal collateral vessels of the posterior cerebral artery: gives the "Ivy sign" on MRI and transdural collateral vessels of the middle meningeal artery, superficial temporal artery, ethmoidal artery and/or occipital artery.

Patients with Moya Moya present with signs and symptoms resulting from changes in the circulation of the internal carotid artery, which can be classified into two groups: ischemic lesion producing TIAs or DVAs, or consequences of compensatory phenomena to this ischemia: hemorrhage due to vascular fragility, aneurysm, unilateral transdural dilatation [2].

In terms of imaging, CT scans can reveal signs of ischemic or hemorrhagic stroke. ANGIO-CT shows stenoses and occlusions of the internal carotid arteries, and/or the proximal portions of the anterior and middle cerebral arteries.

It allows visualization of the suppletive vascular network: the Moya Moya network in the form of serpentine supernumerary vascular structures at the location of the polygon of Willis [3].

Angiomagnetic resonance imaging (MRI) can be used as a 1st-line examination, with high sensitivity and specificity.

It highlights parenchymal and vascular lesions. Ischemic lesions are visualized as T1 hyposignal, T2 hypersignal, FLAIR and diffusion variable according to age.

Hemorrhagic lesions have a variable T1 and T2 signal depending on age, and a constant T2* hyposignal.

Vessels of Moya Moya are visualized on the T2* sequence as punctiform images in signal void, and serpiginous.

In the final stage of the disease, there are signs of brain atrophy usually beginning in the frontal lobes [4]. Arteriography is a reference examination for diagnosis.

Diagnosis is based on the presence of bilateral stenosis and/or occlusion of the carotid siphons and/or proximal portions of the ACM and ACA. Arterial time opacification is the basis for Suzuki and Takaku's classification of 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: increase in the number of vessels; Grade IV: reduction in the number of vessels; Grade V: significant decrease in the number of vessels; Grade VI: disappearance of Moya Moya vessels.

Transcranial Doppler is a non-invasive means of monitoring changes in blood circulation.

Moya's prognosis is difficult to predict because the natural history of the disease is poorly known.

The surgical indication should be discussed fairly quickly, as early intervention can quickly improve the patient's condition and prevent ischemic recurrence [5].

CT angiography or MRI with MRA sequences or even conventional angiography is often performed one year after surgical treatment, and then annually for evolutionary control [5].

CONCLUSION

- Moya Moya is a rare chronic cerebrovascular disorder, idiopathic or secondary, and a significant cause of pediatric stroke.
- Diagnostic delay is common, due to its polymorphic symptomatology. -Angioscanner and MRI angiography are currently the gold standard for initial diagnosis and monitoring.
- Arteriography remains the gold standard for accurate diagnosis and lesion assessment.

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