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Case Report

Moyamoya: A Cause of Pediatric Stroke

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Abstract

Moya Moya disease (MMM) is a chronic cerebral artery condition of unknown cause, identified by the narrowing and blockage of the end sections of the intracranial internal carotid arteries and the proximal arteries of the circle of Willis, leading to the development of a fine, abnormal compensatory vascular network. This disease affects both children and young adults, often causing strokes in children. Imaging is critical for diagnosis, treatment planning, and monitoring. Angiography is the primary diagnostic tool, while non-invasive, non-irradiating MRI plays a key role in disease monitoring. Surgical revascularization techniques have shown promise in improving cerebral blood flow.

Keywords: *Moya-Moya Disease; Stroke; Imaging*

Introduction

Moya Moya disease is an angiogenic disorder characterized by the narrowing of the far end of the internal carotid artery and its extension into the initial segments of the middle and front cerebral arteries. It is characterised by the development of these occlusions leading to the formation of substitute vessels. These vessels originate from parenchymal, perforating, leptomeningeal collaterals and other transdural anastomoses. On angiography, these collateral vessels display a unique pattern that looks like a cloud of smoke, commonly known as the Moya Moya network. Its aetiology remains poorly understood and accounts for 10 - 15% of strokes.

Case Report

A 9-year-old child, who has sickle cell disease has been hospitalized in the intensive care unit due to encephalopathy resulting from multiple cerebral vascular accidents in the context of sickle cell disease. The patient has exhibited a persistent delay in awakening, maintaining a stable Glasgow Coma Scale (GCS) score of 9 - 10 for the past 3 weeks.

A cerebral MRI has been requested. And it showed ischemic strokes of varying ages in the territories of the left middle cerebral artery and the right posterior cerebral artery (Figure 1), and areas with T1 hyperintensity indicative of infarction (Figure 2). And a hyperintensity in diffusion consistent with diffusion restriction (Figure 3).

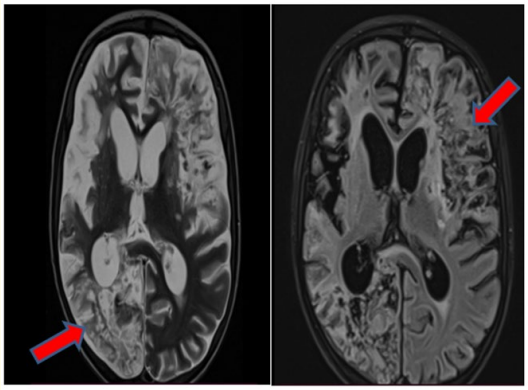

Figure 1: *Axial T2 and axial T2 flair showing ischemic strokes of varying ages in the territories of the left middle cerebral artery and the right posterior cerebral artery.*

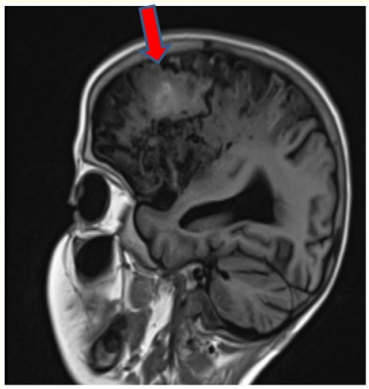

Figure 2: *Sagittal T1 image revealing areas with T1 hyperintensity indicative of infarction.*

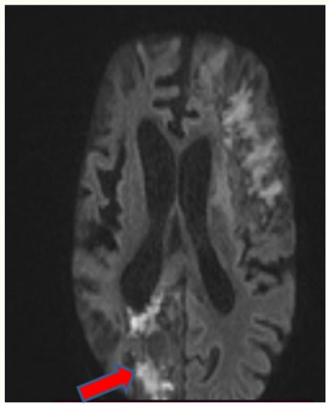

Figure 3: *Axial diffusion imaging revealing a hyperintensity in diffusion consistent with diffusion restriction.*

After Gadolinium injection it showed narrow flow voids in the bilateral MCA and multiple collateral vessels favoring a diagnosis of MMD (Figure 4).

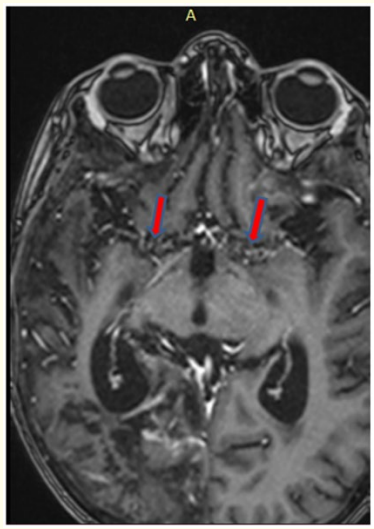

Figure 4: *MRI axial shows narrow flow voids in the bilateral MCA and collateral vessels.*

Discussion

Cases have now been documented worldwide [1,2]. However, the highest incidence of cases is observed in Asia and other regions with non-Caucasian populations [3].

In children, the typical presentation often involves recurring episodes of cerebral ischemia, clinically manifested by focal neurological deficits, sensory disturbances, and seizures [4].

The constriction of cerebral vessels appears to be a response of brain blood vessels to diverse external triggers, injuries, or genetic anomalies [5].

However, more than half of the children diagnosed with this disease exhibit Moya Moya syndrome without an identifiable cause. Once the blockage process initiates, it tends to persist despite known medical interventions unless surgical treatment is employed [6]. Magnetic Resonance Imaging (MRI) not only identifies infarcted areas but also provides a direct view of collateral vessels, presenting as multiple small flow voids at the brain's base and basal ganglia. Magnetic resonance angiography (MRA) is employed to validate the diagnosis and observe the anatomy of the affected vessels. It typically reveals the narrowing and blockage of proximal cerebral vessels, along with extensive collateral flow through smaller arteries, exhibiting the characteristic appearance likened to a "puff of smoke" [7].

Radiological studies are employed to confirm a suspected diagnosis of Moyamoya disease. The initial evaluation of a child suspected of having Moyamoya disease often starts with a head computed tomography (CT) scan to rule out more common conditions such as tumors or hydrocephalus. Typically, findings on a head CT in pediatric patients with Moyamoya include areas of reduced density indicating previous infarctions, especially in regions supplied by the middle cerebral artery, as well as in the basal ganglia, deep white matter, and

periventricular areas [8-10]. While uncommon in children, head CT imaging may also detect hemorrhagic conditions such as intracerebral, intraventricular, subarachnoid, and subdural hemorrhages [8,11,12]. Depending on the extent of previous infarctions, cerebral atrophy and encephalomalacia might also be evident. Recent developments in CT angiography have expanded its application to both diagnosing Moyamoya disease and assessing neovascularization following surgical bypass procedures [13].

Magnetic resonance imaging (MRI) has established itself as a dependable diagnostic method for detecting Moyamoya disease. Acute cerebral infarctions are easily discernible using diffusion-weighted imaging, while chronic infarctions can be identified through both T1- and T2-weighted imaging [14].

Fluid attenuated inversion recovery MRI is utilized to infer cortical ischemia and is believed to indicate sluggish flow in the poorly perfused cortical circulation in pediatric Moyamoya patients [15,16]. The most indicative MRI findings for Moyamoya disease typically include diminished flow voids in the internal carotid artery, anterior cerebral artery, and middle cerebral artery bilaterally, accompanied by prominent flow voids in the basal ganglia and thalamus, indicating collateral formation of Moyamoya vessels [17,18].

Fluid attenuated inversion recovery MRI which demonstrates linear high signal intensity following a sulcal pattern (ivy sign) has been used to infer cortical ischemia and is felt to represent slow flow in the poorly perfused cortical circulation in children with moyamoya Magnetic resonance angiography (MRA) has been used to accurately characterize both the stenosis and basal collateral formation associated with moyamoya disease [17]. The MR findings most suggestive of moyamoya disease remains diminished flow voids in the ICA, ACA, and MCA bilaterally, with concurrent large flow voids in the basal ganglia and thalamus representing collateral moyamoya vessel formation. In recent times, MRI/MRA has been proposed as a dependable alternative to conventional angiography for diagnosing Moyamoya disease, owing to its convenience and lower procedural risks, particularly in the pediatric population.

Conventional angiography continues to be regarded as the gold standard for diagnosing and planning surgical interventions for patients suspected of having Moyamoya disease. A thorough angiographic examination should encompass imaging of both external carotid arteries, internal carotid arteries, and one or two vertebral injections. Emphasizing the visualization of bilateral external carotid arteries is essential for preoperative planning to safeguard against collateral disruption during surgical revascularization. Typical angiographic findings consist of supraclinoid ICA, proximal anterior cerebral artery, and middle cerebral artery stenosis, accompanied by basal ICA, leptomeningeal, and transdural collaterals, resulting in the characteristic “puff of smoke” appearance.

Although no randomized controlled trials have directly compared surgical and medical treatments in patients with Moyamoya disease (MMD), surgical revascularization is widely accepted as the only effective treatment option. Medical management may involve the use of drugs such as antiplatelet agents, typically administered to prevent thrombosis. Surgical procedures are typically categorized into three main groups: direct bypass procedures, including superficial temporal artery (STA) to middle cerebral artery (MCA) anastomosis; indirect bypass procedures, such as encephaloduroarteriosynangiosis (EDAS) and encephalomyosynangiosis (EMS); and combined bypass procedures. Direct bypass surgeries can be technically challenging, especially in pediatric patients with smaller cortical arteries, but they can lead to immediate improvement in cerebral hemodynamics postoperatively. Indirect bypass surgeries stimulate spontaneous angiogenesis between the brain surface and the vascularized donor tissues, although this process typically takes 3 to 4 months for the collateral vessels to fully develop. The prognosis of patients with MMD appears to be influenced by age and the type of presentation [19]. In a study conducted in Japan, EDAS was shown to be an effective procedure, benefiting 75% of patients experiencing transient ischemic attacks within one year [20].

Conclusion

Despite its rarity, Moyamoya disease is gaining global recognition as a cause of pediatric cerebrovascular events, warranting consideration in any child presenting symptoms of cerebral ischemia. Timely diagnosis in pediatric cases is crucial, as the neurological

status at treatment initiation predicts long-term outcomes more effectively than age. Diagnosis typically relies on MR and conventional angiographic imaging, with associated clinical conditions and syndromes also assessed due to their impact on Moyamoya disease development and progression risk. Untreated Moyamoya disease advances, often resulting in permanent neurological and cognitive impairments. While no curative medical treatment exists, surgical revascularization procedures have shown sustained improvement in children. Given the success of both direct and indirect revascularization approaches, individualized management is paramount, aiming to halt and possibly reverse the chronic ischemic state.

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