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Indirect revascularization in an Iraqi child with Moyamoya Disease

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ABSTRACT

Background: Moyamoya disease (MMD) is a rare cerebrovascular disease characterized by bilateral stenosis starting at the supraclinoid internal carotid artery (ICA), with the development of a collateral network of vessels. It is an established cause of stroke in the pediatric age group. Despite its increasing prevalence in various parts of the world, it remains largely underrecognized in the Middle East, particularly in Iraq. This is the first case of MMD in an Iraqi patient undergoing surgery.

Case description: A 12-year-old boy presents with a 3-months history of progressive behavioural changes. MRI revealed diffuse infarcts of different ages. MRA and CT angiography revealed extensive asymmetrical steno-occlusive changes of the supraclinoid ICAs extending into the anterior and middle cerebral arteries, with the development of a collateral network in the basal ganglia. Indirect revascularization of the right side by encephaloduroarteriomyosynangiosis (EDAMS) was performed. The clinical status of the patient improved during the follow-up and the MRA showed a re-establishment of the blood flow to the MCA.

Conclusion: MMD should be recognized as a cause of stroke or recurrent TIAs in the Iraqi population, particularly in pediatric patients. EDAMS is an effective revascularization procedure with good results in pediatric patients.

Keywords
internal carotid artery, stenosis, Moyamoya, revascularization, EDAMS

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INTRODUCTION

Moyamoya Disease (MMD) is a rare chronic idiopathic neurovascular disorder characterized by progressive bilateral steno-occlusive changes starting at the supraclinoid internal carotid artery (ICA) and extending distally to involve proximal parts of the anterior & middle cerebral arteries (ACA & MCA) [1]. The resulting hypoxia leads to the development of a compensatory network of dilated vasculature at the basal ganglia, giving the characteristic Moyamoya (Japanese for "puff of smoke") appearance on cerebral angiography [2].

First described in 1957 as “Hypoplasia of the bilateral internal carotid arteries” [29], the term “Moyamoya” was coined in 1969 by Suzuki and Takaku [2]. Originally, this disease entity was thought to be unique to East Asian populations, particularly Japan. However, it has

increasingly been identified in western populations [3, 4], and has become a recognized cause of stroke in pediatric patients [5]. Nevertheless, it remains a rare entity with varying prevalence across ethnic groups, ranging from 6 per 100,000 in Japan to a tenth of that in Europe [3, 4, 16-18]. It has a female predominance, with the female-to-male ratio ranging between 2:1 and 4:1 [16,19].

Some individual case reports and small series of MMDs have been reported in Middle Eastern individuals [6-14], but there are no large series or long-term studies available in the literature. Only one case of moyamoya syndrome in an Iraqi patient has been reported in the literature, in which surgical revascularization has not been performed [15]. To the best of our knowledge, this is the second case report of MMD in an Iraqi patient and the first to be successfully treated by cerebral revascularization.

Case presentation

A 12-year-old boy was brought by his parents, who described a 3-month history of progressive behavioral changes and decreased school performance. His physical and neurological examination was unremarkable. Routine blood and urine investigations were normal. Magnetic Resonance Imaging (MRI) showed signs of diffuse cortical and deep matter infarcts, as well as an old frontal infarct (Figure 1).

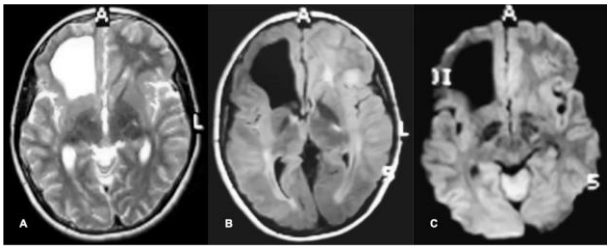

Figure 1. Axial MRI of the brain. **A-B:** T2-weighted & FLAIR images showing diffuse high signal intensities in the cortical and deep frontoparietal peri-ventricular regions. A large frontal porencephalic cyst, probably caused by an old infarct, can be seen as well. **C:** DWI image of the brain showing restricted flow in the abovementioned areas.

Magnetic resonance angiography (MRA) & computer tomographic angiography (CTA) (Figure 2) revealed asymmetrical steno-occlusive changes of the anterior circulation with an extensive deep collateral network at the basal ganglia. These modalities showed generalized stenosis of the intracranial right ICA with severe near-occlusive narrowing of its supraclinoid segment extending to the first few millimeters of the right ACA & MCA. The left supraclinoid ICA is completely occluded with obliteration of its proximal intracranial segments, and the proximal parts of the left ACA and MCA are completely occluded. The vertebrobasilar system and the posterior cerebral arteries (PCAs) were normal with no signs of stenosis, and the posterior communicating artery (Pcom) was intact on both sides, providing flow to the MCAs. As an endovascular facility was inaccessible at our center, no catheter angiography was performed.

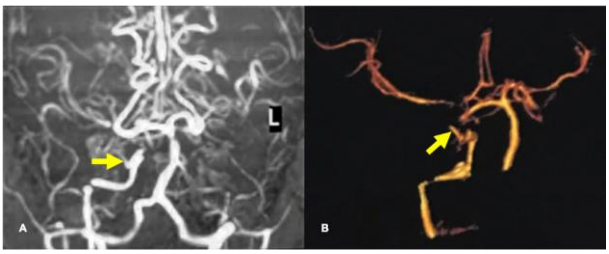

Figure 2. Brain MRA **(A)** and CTA **(B)** showing asymmetric stenosis of the anterior circulation with an extensive vascular network at the basal ganglia. The right ICA shows stenosis with near-occlusive narrowing of its supraclinoid segment extending to a few millimeters of the right ACA & MCA.

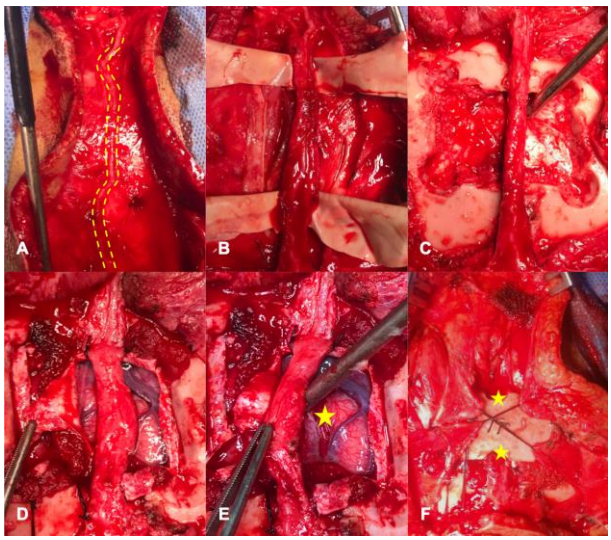

Figure 3. Revascularization by EDAMS. **A-B:** a linear incision is made along the course of the STA (dashes) with the aid of Doppler U/S and the artery is dissected and mobilized. **C-D:** a 5x5 cm craniotomy is created underlying the STA, and the dura opened and reflected in a cruciate fashion, with special care to preserve the middle meningeal artery. **E:** the STA and strips of temporalis muscle are sutured to the cortical surface adjacent to cortical MCA branches (star). **F:** reimplantation of the bone

flap after creating opposing notches (stars) to accommodate passage of the STA with its perivascular tissues.

A definitive diagnosis of asymmetrical MMD was made based on MRI and MRA criteria, and the patient underwent a right-sided extracranial-intracranial (EC-IC) bypass.

Indirect revascularization was performed by Encephalo-duro-arterio-myo-synangiosis (EDAMS), demonstrated in Figure 3. A linear incision was made along the course of the STA with the aid of Doppler U/S, after which the STA was dissected along with its perivascular tissue, and mobilized to allow safe drilling of 4 burr holes to make a 5 by 5 cm craniotomy. The dura was then opened and reflected in a cruciate fashion, with special care to preserve the middle meningeal artery (MMA). Afterwards, the STA was laid on & sutured to the cortical surface adjacent to cortical MCA branches. Strips of temporalis muscle were also reflected and attached to the cortex around the STA. The bone flap was re-implanted after preparation by creating opposing burr hole notches to accommodate the passage of the STA with its perivascular tissues.

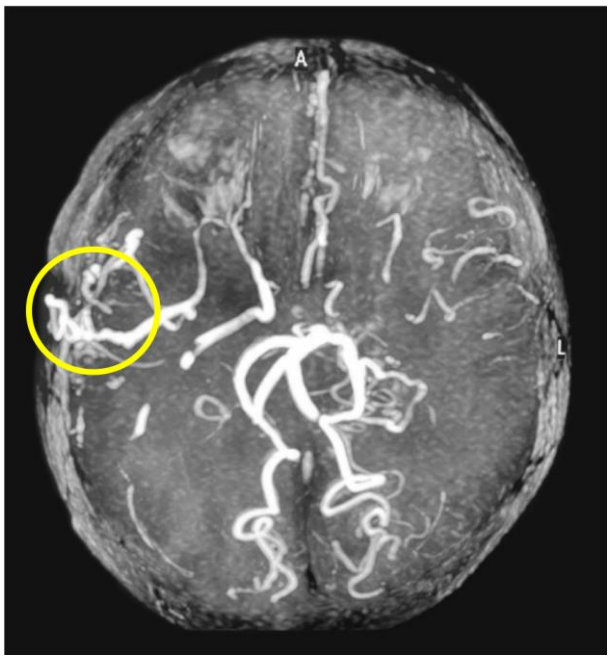

Figure 4. MRA performed 6 months following indirect revascularization (EDAMS) of the right side shows establishment of collateral blood flow from the STA to the frontal and parietal cortical territory of the MCA (circle).

There were no surgical complications, and the postoperative recovery was uneventful. The patient was discharged home with protective headgear at day eight postoperatively, and the parents were informed of the need to perform left-sided revascularization. At his six-month follow-up appointment, the parents reported a noticeable improvement in the patient's behavior and school performance. The follow-up MRA confirmed the re-establishment of collateral blood flow to the right MCA (Figure 4).

Discussion

Moyamoya disease versus syndrome

An important distinction when discussing moyamoya phenomena is differentiating moyamoya disease from moyamoya syndrome. MMD is characterized by bilateral, albeit sometimes asymmetrical, changes in the ICAs and eventually ACAs and MCAs. When these changes are coupled with certain well-documented associated conditions, or when the changes occur unilaterally, it is referred to as moyamoya syndrome [5]. Bilateral disease eventually develops in 40% of those with unilateral vasculopathy.

Our patient presented with bilaterally diseased cerebral circulation, with no associated risk factors and physical exam and laboratory investigations revealed no findings. Therefore, his condition is classified as moyamoya disease.

Presentation

MMD has a bimodal age distribution of disease onset (with peaks at ages 5-9 and 45-49) [19]. Pediatric patients are more likely to present with ischemic symptoms such as stroke, transient ischemic attacks (TIAs), or seizures. Hemorrhagic presentations are seen in both age groups, albeit at a much higher rate in adults [16, 20]. Our pediatric patient presented with ischemic symptoms that are consistent with established patterns in patient presentation.

Diagnosis

Diagnosis of MMD is generally based on clinical and radiological characteristics. MMD should be in the differential diagnosis of any patient presenting with neurological deficits or unexplained symptoms attributable to cerebral ischemia, particularly in the pediatric age group. Diagnosis can be confirmed by radiological evaluation, primarily with MRI, MRA and catheter angiography. Specific MRI sequences can detect cerebral infarction in its early and late stages, as was the case with our patient. FLAIR imaging also

enables the detection of chronic hypoxia, which manifests as linear high signals along the cortical sulci; the so called "ivy sign" [21]. A highly suggestive finding on T1 & T2-weighted images is the absence of ICA, ACA and MCA signal voids on the affected side, and the appearance of tortuous signal voids at the level of the thalamus and the basal ganglia, brought about by the development of collateral vessels in that region [22].

Catheter angiography is the most valuable tool for definitive diagnosis of MMD by detecting the steno-occlusive changes in the supraclinoid ICA extending to the ACA and MCA, and can visualize the leptomeningeal and/or basal collateral networks; the moyamoya or "puff of smoke" vessels. It also allows staging of the disease using the Suzuki grading system [2].

Due to concerns about cost, invasiveness and availability of catheter angiography, criteria have been established for diagnosing MMD based on MRI and MRA alone [23]. This criteria establishes a definitive diagnosis of MMD based on three conditions; namely the documentation of stenosis or occlusion at the terminal portion of the ICA and the proximal portions of the ACA and MCA on MRA, an abnormal vascular network in the basal ganglia seen on MRA or MRI, and the observation of the abovementioned two points bilaterally. In pediatric patients, the latter condition is not necessary for a definitive diagnosis. A staging system based on MRA findings alone has also been proposed [24], and is summarized in table 1. Using that system, our case would be categorized as grade II. This corresponds to grade III in the catheter angiography-dependent Suzuki grading system (stenosis of the ACA and MCA with patent Pcom and extensive collateral network at the basal ganglia).

Table 1. MRA-based grading system for MMD [24]

Vessel Changes	Score
ICA	
• Normal	0
• Stenosis of C1	1
• Discontinuity of C1 signal	2
• Invisible	3
MCA	
• Normal	0
• Stenosis of M1	1
• Discontinuity of M1 signal	2
• Invisible	3
ACA	
• Normal A2 and its distal signal	0
• A2 and its distal signal decrease	1
• Invisible	2
PCA	
• Normal P2 and its distal signal	0
• P2 and its distal signal decrease	1
• Invisible	2
Total	**0-10**

0-1: Grade 1; 2-4: Grade 2; 5-7: Grade 3; 8-10: Grade 4

Treatment

The mainstay of treatment for MMD is surgical revascularization using intact STA as an alternative source of blood flow. This has been shown to be a safe and effective treatment option that has reduced the incidence of strokes and TIAs in patients with MMD, with 96% of them having a 5-year stroke-free period and enhanced day-to-day activities [20, 23, 25].

There are no established surgical indications for patients with MMD, and some authors encourage surgical intervention in asymptomatic cases as neurological status at the time of surgery is stated to be the most significant predictor of long-term outcomes [20].

Surgery consists of direct STA-MCA bypass, indirect bypass techniques or a combination of both, with each modality having its own benefits and pitfalls. Direct revascularization provides an immediate augmentation of cerebral blood flow to the stenotic arteries, but due to the technical difficulties in anastomosing small-caliber vessels, its use is limited in pediatric patients. On the other hand, indirect revascularization is a less technically demanding technique, in which highly vascular tissues are approximated to the cortical surface to promote angiogenesis and enable the passive development of collateral EC-IC vessels. This offers excellent long-term outcomes comparable to those of direct revascularization, but improvement in cerebral blood flow is delayed and collateral vessels might take up to 3-4 months to develop [23].

Many techniques of indirect revascularization have been developed since the description of the disease in 1969, including encephalodurosynangiosis (EDS), encephalomyosynangiosis (EMS), encephaloduroarteriosynangiosis (EDAS), EDAMS, omental flaps transplantation, and placement of multiple burr holes [1].

We have reported the first case of MMD in an Iraqi patient to be successfully treated, using the EDAMS procedure of indirect revascularization. The aim of this procedure is to nourish the frontal and parietal cortical territories of the MCA. It combines the EDAS and EMS techniques thus maximizing the amount of vascular tissue involved in the synangiosis [26]. The very small size of the STA in our patient rendered direct bypass non-feasible. However, due to the long course of the vessel, its re-routing into the cortical surface was possible. Long-term follow-up data has shown EDAMS to be a safe and effective treatment modality for adults and older children with MMD [27, 28].

The reporting of such rare cases in the Iraqi population should warrant higher vigilance and the consideration of MMD as differential diagnosis in patients presenting with ischemic stroke, particularly children.

Conclusion

We present the second case of moyamoya disease in an Iraqi patient and the first to be successfully treated by indirect surgical revascularization, using the EDAMS technique. The article emphasizes the importance of recognizing this disease as a cause of stroke or recurrent TIAs in the Iraqi population, particularly in the pediatric age group.

Abbreviations

ACA: anterior cerebral artery
Acom: anterior communicating artery
CTA: computer tomographic angiography
DWI: diffusion-weighted imaging
EC-IC: extracranial-intracranial
EDAMS: encephaloduroarteriomyosynangiosis
EDAS: encephaloduroarteriosynangiosis
EDS: encephalodurosynangiosis
EMS: encephalomyosynangiosis
FLAIR: fluid attenuation inversion recovery
ICA: internal carotid artery
MCA: middle cerebral artery
MMA: middle meningeal artery
MMD: moyamoya disease
MRA: magnetic resonance angiography
MRI: magnetic resonance imaging
PCA: posterior cerebral artery
Pcom: posterior communicating artery
STA: superficial temporal artery
TIA: transient ischemic attack
U/S: ultrasonography.

Authors' contributions

S.S.H: Data collection
A.O.A: Manuscript drafting
Z.F.A: Manuscript revision
M.A.A: Manuscript revision

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