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PEDIATRIC MOYA MOYA SYNDROME WITH FAMILIAL HISTORY AND ENCEPHALO DURO ARTERIO MYO SYNANGIOSES. - A RARE CASE REPORT.

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ABSTRACT Moya Moya disease (MMD) is a type of chronic cerebrovascular occlusion disease, which frequently occurs in East Asian populations, including pediatric and adult patients, and may lead to ischemic or hemorrhagic stroke, headache, epilepsy or transient ischemic attack. To date, the underlying mechanisms of MMD have remained to be fully elucidated, but certain studies have indicated that genetic factors may be an essential component of its development. Cerebral angiography is the best approach for diagnosing MMD. However, with technological advances, non-invasive techniques are increasingly used to accurately evaluate MMD. MMD is commonly treated via surgery, and an increasing number of patients are benefitting from the intra- and extra-cranial revascularization. The present article provides a case report of a pediatric moyamoya disease.

KEYWORDS : Moya Moya Disease(mmd), Cerebral Stroke, Superficial Temporal Artery(sta), Revascularization.

INTRODUCTION

MoyaMoya disease (MMD) is a progressive cerebrovascular occlu sive disease of the bilateral internal carotid arteries that leads to a compensatory abnormal vascular network at the base of the brain. Its average annual incidence of 0.54 per 100,000 population, but it is the most common pediatric cerebrovascular disease in East Asia. Suzuki and Takaku named it MoyaMoya, which means a puff of smoke that is the angiographic representation of collaterals. In children, unilateral involvement occurs about 18% and progress to bilateral involvement within two years. The clinical signs of MMD mainly include two types: Cerebral ischemia and cerebral hemorrhage. These two types of symptom differ in their distribution between pediatric and adult patients. Most of the pediatric patients present with progressive cerebral ischemia, including transient cerebral ischemic attacks and cerebral infarctions. Mental decline or seizures may be the first symptom in children. In half of the cases in adults, intracranial hemorrhage is the first symptom, while ischemic symptoms first occur in the other half. [1] Patients with MMD had a genetic mutation in the genes RNF213 on chromosome 17q25, ACTA2 on chromosome 10q23.3, GUCY1A3 on chromosome 4q32. [2] We report a case of an 8-year-old female child who presented with left upper limb monoparesis, delayed milestones and diagnosed as MMD. Cerebral revascul arization surgery Encephalo Duro Myo Arterio Synangioses lead to favorable surgical outcomes.

CASE REPORT

8-year-old girl presented with a history of multiple episodes of GTCS since childhood followed by dysarthria for 2 years along with weakness of left upper limb. Family history revealed and elder sister diagnosed with a similar disease and now bed-bound. The patient had dysphasia. However, she was able to walk unsupported. There was a history of second-degree consanguinity. She was born full-term vaginally with Apgar scores of 8 and 9. She was afebrile with the following vitals: BP 90/50mm of Hg, HR 123 bpm and RR 24 per minute. Her weight was 18 kg (85th centile) and height was 108 cm (>97th centile). Pupils were equal and reactive at 4 mm. She had a normal cranial nerve examination, including sharp disc margins on fundoscopy. She moved all three extremities with left upper limb hemiparesis 4/5 and symmetrical 3+ deep tendon reflexes in the lower limb and 2 + in the upper extremities. Plantar responses were extensor bilaterally, without ankle clonus. Both electrocardiography and bedside electroencephalography were standard for age. The sickle cell test was negative. Renal ultrasonography with Doppler flow study showed no evidence of renal artery stenosis. Genetic testing was deferred in view of financial constraints. MRI brain with MRA contrast showed an abrupt cut off of cavernous portion and supra clinoid portion of ICA bilaterally with attenuation and narrowing of cavernous and supra-clinoid internal carotid arteries bilaterally. Numerous small punctuate vessels noted surrounding the basal cisterns were seen. The main segments of ACA, MCA, PCA were poorly visualized. The patient was diagnosed with bilateral moyamoya disease.

In view of more ischemic changes in the right cerebral hemisphere, the patient was planned for right indirect revascularization procedure initially. Under general anesthesia, the patient was positioned in the right lateral position. STA was marked preoperatively using the Doppler hand probe. Question mark incision was given. The superficial temporal artery was isolated and held separately with elastic markers. Temporalis muscle was separately elevated. Burr holes made and right fronto-temporo-patietal craniotomy was done. Dura was opened in a cruciate pattern and dural edges inverted. STA was placed over the pia after giving multiple stab incisions over the pia. Temporalis muscle was next sutured to the dural edges in a watertight fashion. The inferior edge of the bone flap was nibbled so as to provide atraumatic entry of the temporalis muscle. Bone flap fixed with mini plates,no:10 drain was put and subcutaneous tissue and skin were sutured. The patient made uneventful post-operative recovery. Follow up next six months revealed improvement in the fine motor movements of the left upper limb. She is being planned for a similar procedure on the left side and a follow-up angiography.

DISCUSSION

Moyamoya disease (MMD) is an idiopathic disease with a progressive nature leading to recurrent stroke due to occlusion of the terminal internal carotid arteries [3]. Although a recent genetic study identified a possible susceptibility gene [4], the pathogenesis of MMD has not been fully defined. Revascularization surgery for symptomatic MMD is considered the standard treatment for preventing further stroke [4,5]. The main objective of surgery is to augment intracranial blood flow using an external carotid system by either direct bypass or pial synangiosis. It can be achieved readily by either extracranial-intracranial bypass or vasculogenesis using indirect pial synangiosis for symptomatic patients. In particular, surgical revascularization to prevent ischemic stroke is an effective treatment for patients with MMD with an ischemic presentation[6].

Pediatric MMD is characteristically more progressive than in adult patients, and revascularization surgery is indicated in most children[7]. Early diagnosis and intervention should happen before irreversible brain damage occurs.

Since the 1970s, direct bypass has been used in patients with MMD [8]. Following successful anastomosis between donor and recipient arteries, improvement in flow is achieved immediately after surgery. The vascular diameters of the superficial temporal artery (STA) and cortical arteries are important factors to determinants of a direct bypass. In the advanced stage of MMD, most of the cortical arteries have shrunk to a small-caliber and the vessel walls of patients with MMD tend to be more fragile. Post-operative hyperperfusion

syndrome is another considerable problem leading to neurologic deterioration, which often develops after direct bypass surgery. As a donor artery, the STA is selected in the majority of cases.

A direct bypass is a somewhat difficult procedure in young pediatric patients or adult patients with advanced MMD due to the small caliber of the recipient artery. In such cases, an indirect bypass has been effective. However, it takes more time to improve cerebral blood flow because neovascularization from connective tissue is not immediate.

The Indirect revascularization surgical procedures can be classified according to the various tissues covering the brain. Historically, Encephalo-Myo-Synangiosis (EMS) for MMD was introduced by Karasawa et al. in the 1970s[9]. In this operation, the deep temporal artery (DTA), supplying the temporalis muscle, becomes the leading supplier of neovascularization. The temporalis muscle or dissected inner fascia can be covered with the brain surface being sutured with dura.

Other indirect methods such as Encephalo-Myo-Arterio-Synangiosis (EMAS), Encephalo-Duro-Arterio-Myo-Synangiosis (EDAMS), and Encephalo-Galeo-Synangiosis (EGS) are performed as variants of EMS and EDAS [9-14]. Bi-frontal indirect bypass can be considered if the patient has frontal lobar hypoperfusion[15]

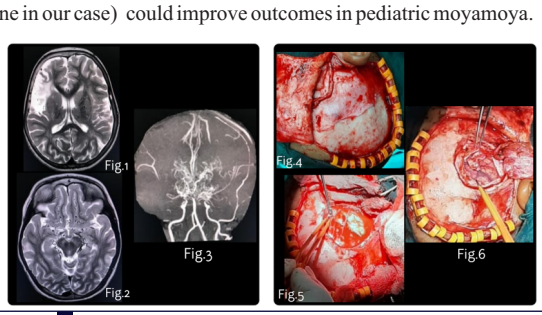
Since the cortical branches of the PCA are usually smaller in caliber, the use of an indirect bypass for PCA territories has also been well described [16,17]. The occipital artery is used as a supply vessel in this and other procedures that are similar to EDAS. Additional techniques include omental flap surgery and multiple burr hole surgery as rescue surgeries after failed revascularization. Neovascularization with these methods has been reported to be comparable to other methods [18-21].

Various studies have reported complications after revascularization surgery for MMD. Postoperative stroke with permanent neurologic deficits developed in 1.6%-16.0% [6,22,23-25] of patients and was more frequent in adults [26]. In addition, permanent neurologic deficits developed in 0.9%-8.0% of those with perioperative ischemic stroke [6,22,23,24]. However, the radiologic incidence of ischemic stroke was higher than symptomatic stroke. During surgery, maintaining euvolemic status and the appropriate blood pressure is important. Also, hypotension and hypercapnia can aggravate hypoperfusion during surgery. Maintaining the level of hemoglobin is also essential in for oxygen delivery capacity.

Hemorrhagic as a complication develops in 0.7%-8.0% of patients.([6,22,23-25] Postoperative epidural pain develops in 4.8% of pediatric[27]. Hyperperfusion syndrome (21.5%-50.0%)[28-31]. This phenomenon characteristically presents with transient neurologic deterioration with increased blood flow and mostly develops in patients with MMD after surgery [31].Scalp ischemia can lead to skin problems(17.6%-21.4%) [32,33]. In our present case, an indirect revascularization procedure was chosen as direct revascularisation could not be possible because of the small caliber of recipient's vessels.

CONCLUSION

At present, there is a standardized surgical approach for the treatment of MMD in children and numerous revascularization procedures have been used. They aim to prevent further ischemic injury by increasing collateral flow using external carotid circulation as a donor supply. Revascularization procedures can be divided into three main groups: Indirect (non-anastomotic) bypass techniques, Direct (anastomotic) bypass techniques or combined. Data proved that indirect revascula rization procedures like Encephalo Duro Arterio Myo Synangioses(as done in our case) could improve outcomes in pediatric moyamoya.



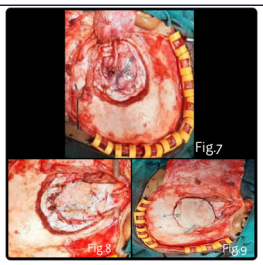


Figure:1	PRE OPERATIVE T2 WEIGHTED MRI.
Figure:2	PRE OPERATIVE T2 WEIGHTED MRI
Figure:3	PRE OPERATIVE MRA ANGIOGRAM.
Figure:4	SUPERFICIAL TEMPORAL ARTERY ISOLATED
Figure:5	CRANIOTOMY
Figure:6	DURAL OPENING
Figure:7	SUPERFICIAL TEMPORAL ARTERY PLACED OVER PIA
Figure:8	TEMPORALIS FASCIA SUTURED TO DURAL EDGES.
Figure:9	CRANIOTOMY CLOSED WITH MINI PLATES

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