Research Papers

Moya Moya Cases Treated with Encephaloduroarteriosynangiosis

Parimal Tripathi, Varsha Tripathi, Ronak J. Naik and Jaimin M. Patel

From Gujarat Cancer & Research Institute, Ahmedabad; Sterling Hospital, Ahmedabad and Harsh Neurosurgical Institute, Ahmedabad, Gujarat, India

Correspondence to: Dr. Varsha P. Tripathi, 409 Sangita Complex, Opp.Doctor House, Ellis bridge, Ahmedabad 380006, Gujarat, India, E-mail: varshapt@yahoo.co.uk.

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Objective: *To study neurological outcome of Moya Moya disease treated surgically with Encephaloduroarteriosynengopsis (EDAS).* ***Design:*** *Prospective observational study.* ***Settings:*** *Community and General with tertiary care facility.* ***Subjects:*** *Eight children diagnosed with Moya Moya disease by Magnetic Resonance Angiogramover 4 years of period were selected for EDAS. Children who were not able to sustain surgery excluded from study.* ***Methods:*** *Treatment modality selected were surgery in form of EDAS. After surgery subjects were followed up forminimum of 2 year period to know neurological out come. Outcome was reported as poor, fair, good and excellent. No nstatistical analysis performed due to small sample size.* ***Results:*** *After surgery no episode of stroke or TIA was obereved in any patient during 2 year follow up period and all patients are living without any new neurological deficit.* ***Conclusion:*** *Long term outcome of EDAS is promising.*

Keywords: *Encephaloduroarteriosynangiosis, Moya Moya Disease, Magnetic Resonance Angiogram.*

MOYA Moya disease is a rare idiopathic disorder that leads to irreversible blockage of the major blood vessels to the brain. In Japanese it means "puff of smoke" which refers to collateral circulation. It can lead severe functional impairment or even death(1,2). It is important to recognize and treat them early. Magnetic Resonance Angiography (MRA) is better than other diagnostic modalities(3,4). Once the vascular occlusion begins, it tends to continue despite any known medical management. The treatment goal is to improve blood flow to hypoperfused cerebral regions by surgery. Encephaloduroarteriosynangiosis (EDAS) has shown excellent postoperative results.

Subjects and Methods

The patients reported in this study were seen by us during 2001-2005. Eight clinically suspected children of Moya Moya disease, 3 boys and 5 girls, diagnosed with MRA(3) and underwent EDAS. None of the patients had underlying autoimmune disease, neurofibromatosis or meningitis. All the patients underwent necessary investigations to find out etiological association. The follw up was upto two years to find out clinical improvement and occurrence of complication.

Results

Details of patient are summarized in *Table I.*

It was seen that one patient had recurrent Transient Ischemic Attack (TIA) for one year, 4 had recurrent stroke with hemiparesis in 2 of them, one had single episode of stroke, one had aphasia with hemiparesis and one had refractory multifocal simple partial seizures.

The duration of symptoms ranged from one week to two years. In Magnetic Resonance Imaging (MRI) showed multiple infarcts in the region of the blocked arterial territory. MRA, Bilateral Internal Carotid Artery was involved in all patients by definition and one patient had complete block. Middle Cerebral Artery (MCA) was also involved in all patients and was bilateral in 6 patients. Anterior Cerebral Artery (ACA) was involved in 6 patients and was bilateral in three. Posterior Cerebral Artery (PCA) was involved in only one patient. EDAS was done in all patients using superficial temporal artery: bilateral

in 7 patients and unilateral in one patient. In bilateral EDAS group 2 patients had two stage surgery while rest had one stage surgery. One stage surgery is preferable over two stages to avoid chance of missing patient after first stage.

On follow up examinations, no patient had episode of TIA or stroke. Speech was recovered in patient with expressive aphasia. Patients who had hemiparesis showed improvement in power to grade III-IV. Patient who had complete block of bilateral ICA had episodes of tremors with severe headache and right upper limb simple partial seizure till nine months after surgery which resolved spontaneously. No complications were observed in remainder.

Discussion

The exact etiology of Moya Moya disease is unknown(3). Peak age of onset in the Asian population is bimodal; with first peak in the first decade and a second peak in the fourth decade of life (range is 6 months to 67 years). In our case series the age range is 3-14 years. The female-to-male ratio is 1.8:1, which is also observed in our series. It occurs primarily in Japanese, followed by other Asian population and is least common in Caucasians.

Moya Moya disease is characterized pathologically by intimal thickening in the walls of the terminal portions of the internal carotid vessels bilaterally which may contain lipid deposits. The ACA, MCA, and PCA may show varying degrees of stenosis or occlusion. Numerous small vascular channels can be seen around the circle of Willis(4).

MRI was suggestive of multiple lacunar infarcts in all patients in blocked territory. MRA was done in all of them and Digital Subtraction Angiography (DSA) in one child. Reports have shown good results with MRA in the diagnosis of Moya Moya disease(3,5). The diagnostic criteria for Moya Moya disease were proposed by the research committee on spontaneous occlusion of the circle of Willis(3); occlusion or stenosis of the terminal portion of the ICA and proximal portion of the ACA and MCA, abnormal vascular network at the base of the brain, and no underlying disease. If these findings are present bilaterally, the diagnosis is definite Moya Moya disease otherwise the diagnosis is probable Moya Moya disease. According to this 5 patients had definite Moya Moya disease and 3 patients had probable Moya Moya disease (*Fig.1,* PreEDAS MRA).

Pharmacological therapy for Moya Moya disease is disappointing. In cases of severe stroke, intensive care unit monitoring is indicated. If the patient has had an ischemic stroke, consider anticoagulation or antiplatelet agents to prevent further strokes, especially in stenotic vessels where further infarction can occur if occlusion progresses.

Surgery is the mainstay of the therapy. Patients with poor clinical condition may not be ideal operative candidates. Revascularization should be performed under nonemergent conditions(6,7). All surgeries have in common the concept of a blood and oxygen "starved" brain reaching out to grasp and develop new and more efficient means of bringing blood to the brain. Encephaloduroarteriosynangiosis (EDAS), also known as Pial Synangiosis(8) was first described by Matsushima, *et al*.(9). Superficial temporal artery is sewn to the inside edge of the dura such that it remains in contact with the exposed cortex. Over time, angiogenesis results in the formation of small arteries to the brain. The results reported by Matsushima, *et al.*(9). Using EDAS in 38 pediatric patients (70 hemispheres) showed #100% revascularization of brain with many patients showing improvement in symptoms(10). Suzuki, *et al*. reported a series of 21 young patients with Moya Moya disease, showing that EDAS resulted in increases in hemispheric and cortical flow, particularly in patients with TIA symptoms, and new cortical vessels were noted as early as two weeks after EDAS. We operated all patients with EDAS and obtained 100% revascularization. EDAS was done bilateral in 7 patients and unilateral in one patient. Surgeries were not done under EEG monitoring as it is not indicated.

Routine postoperative care was taken with special reference to unmask intracranial bleeding(11). Any changes in neurological examination should be evaluated with emergent CT. We routinely perform MRA at three to six months post-EDAS, to document extent of the cerebral cortex neovascularization, and to assess regression of Moya

TABLE I–*Clinical Profile of Patients with Moya Moya disease*

Sr.No.	Age	Sex	Presenting features	Duration of symptoms	MRI	MRA / DSA	Surgery	Outcome
1	10yr	F	B/L TIA since 1 yr. Rt. sided TIA more severe increased since 1 month	1 yr	Multiple lacunar infarcts more on Lt. side	MRA narrowing of B/L distal ICA, proximal ACA, MCA with collaterals, Puff of smoke (collateral better on Rt side)	EDAS-U/L Lt. side	Symptoms free3 months, Penetrating vessels on post surgical scan, did not come for Rt sided surgery
2	8 yr	F	Expressive aphasia, Lt. hemiparesis, Tremors	2 wks	Rt. MCA, +multiple lacunar infarcts	ICA B/L;Rt ACA MCA, PCA; Lt. MCA	B/L two stage done after 6wk, first on Lt. then on Rt. after 3 months	Speech recovered, residual Lt. hemiparesis, mobile, penetrating vessels on scan
3	7 yr	F	Recurrent stroke on Lt. side, presented with partially recovered hemi paresis	6 mo	B/L lacunar infarct	B/L ICA; Rt. ACA, MCA; MCA; Lt. MCA	Two stage B/L at the interval of 4 months	Mild weakness of Lt. UL Grade III to IV grip power, Not affording for scan
4	7yr	M	Recurrent U/L stroke Lt. side, Rt. Handed	8-9 mo	B/L infarct	B/L ICA MCA ACA	One stage surgery	Normal, Not affording for MRI
5	14 yr	F	Rt facial Rt. hemi paresis stroke two episode	15 days	Parietal infarct, Lt sided, small peri-ventricular infarcts	B/L ICA MCA ACA	One stage surgery	Normal
6	6yr	F	Refractory multifocal simple partial seizure	2 yrs	Multiple small peri-ventricular infarcts	B/L complete block of ICA, Multiple collaterals, DSA–B/L complete block of ICA, Multiple collaterals	B/L EDAS one stage	When emotionally stressed patient had tremors and severe headache, Rt. UL siimle partial seizure till 9 months. Increased vasculatures on scan. 3yrs after surgery child is going to school. Normal intelligence.
7	3 yrs	M	Single stroke Lt. side	1 wk	CT: Rt. parietal developing infarct. MRI: Rt. parietal infarct	B/L ICA; Lt. MCA ACA; Rt. ACA	B/L EDAS one stage	Normal
8	6 yrs	M	Alternating stroke+ seizure	2 yrs	Left sided infarct MCA territory; Rt. side recent infarct	B/L ICA MCA ACA	B/L EDAS one stage	Operated 1 month back, symptom free on first follow up.

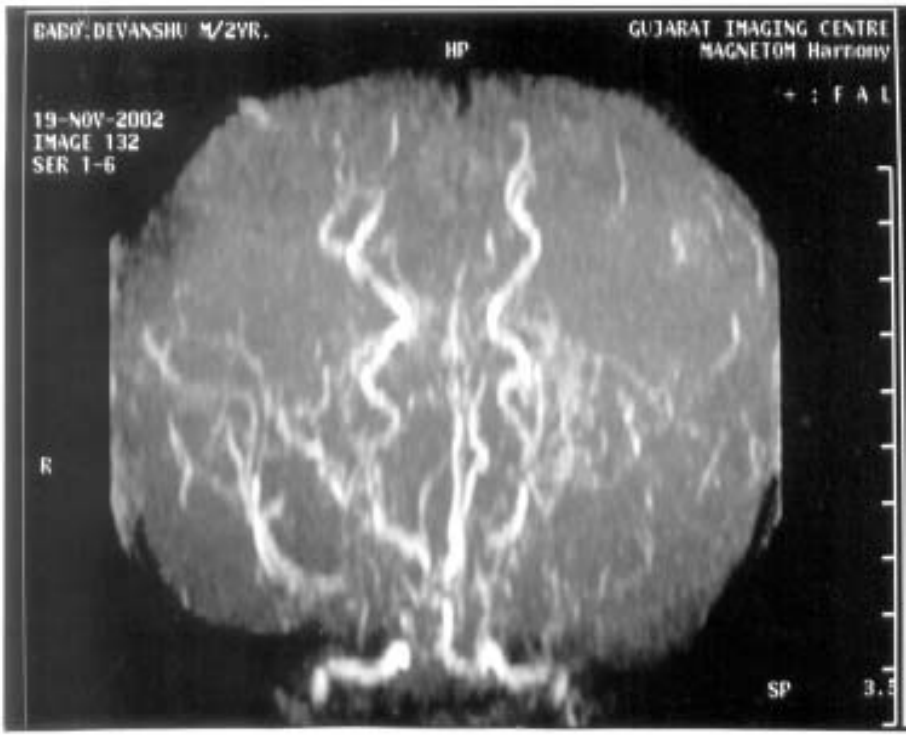

Fig. 1. MRA preoperative–impaired cerebral blood flow.

Moya vessels(10,12). (*Fig. 2*, Post EDAS MRA). All patients had satisfactory collaterals from STA and regression of Moya Moya vessels on postoperative MRA.

An experienced pediatric neurologist evaluated patients' neurological status after EDAS. It was divided into four categories(6) poor, indicating neurological deficit after surgery; fair, indicating unchanged symptoms after surgery; good, indicating improvement of minor neurological deficits; and excellent, indicating complete recovery of neurological deficits and disappearance of TIAs. All the patients had excellent postoperative recovery.

No immediate complications were noted in any patients. Patient with complete block of bilateral ICA had episodes of tremors with severe headache and right upper limb simple partial seizure till nine months after surgery which resolved spontaneously. After three years of surgery child is going to school and has normal intelligance. No episode of stroke or TIA was observed post operatively and during two year of follow up in any patient. Speech recovered in patient with expressive aphasia. Patients with hemiparesis had improvement of power up to grade III-IV. The seizure was aborted in patient with refractory seizure. While symptoms may seem to improve almost immediately after surgery, it would probably take three to six months before new vessels can be seen on MRA. No mortality has been observed in this series. Thus long-term outcome of treatment with EDAS seems to be promising.

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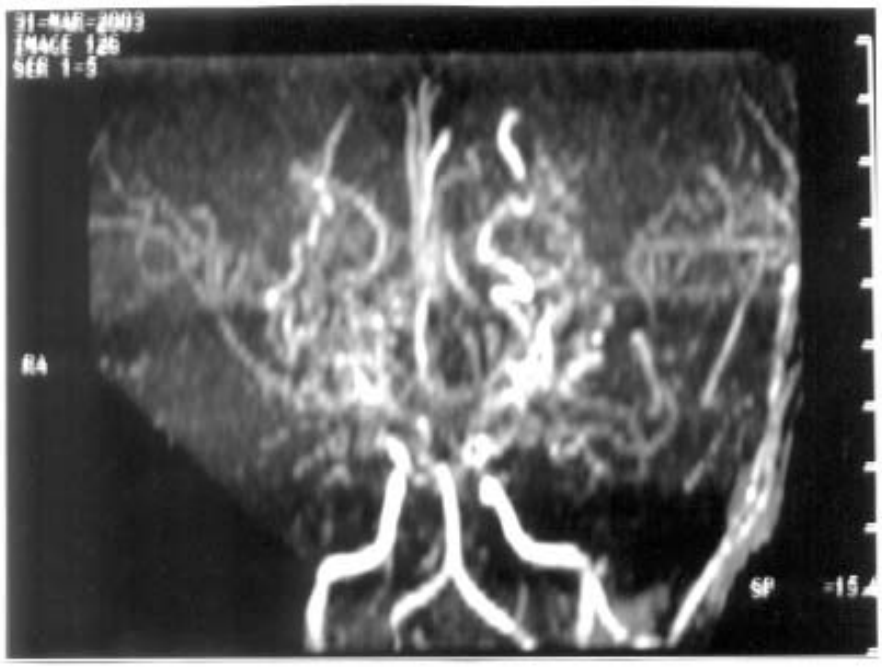

Fig. 2. MRA 6 months postoperative–improved cerebral blood flow.

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