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SURGICAL OUTCOME OF MOYAMOYA DISEASE IN EIGHT PATIENTS FROM NORTH WEST INDIA

Neurosurgery

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ABSTRACT

Moya Moya disease is a rare form of chronically progressive disorder affecting bilateral supraclinoidal internal carotid arteries with extensive collateral at the base of brain. We will discuss our experience of 8 patients treated surgically.

PATIENTS AND METHOD- All the 8 patients are reviewed retrospectively. Out of 8 patients 3 presented with hemorrhagic stoke underwent either exclusion of distal aneurysm or clot evacuation followed by STA-MCA bypass with EDAMS. Rest of the patients presented with either recurrent TIA/stroke or recurrent seizure had undergone STA-MCA bypass with EDAMS. Modified Rankin score was calculated at time of admission and at 6 month to 1 year follow up.

RESULTS- The incidence of Moyamoya disease is equal in children and adult in this study with slight predominance of male over female. Modified Rankin score showed improvement in postoperative neurological status. None of our patient suffered recurrent symptoms.

CONCLUSION- STA-MCA bypass with EDAMS technique is an every effective in treating Moya-Moya disease. So all the patient who are presented with acute stoke and suspected to have Moyamoya disease should be thoroughly evaluated.

KEYWORDS

INTRODUCTION

Moya Moya is a Japanese term use to describe hazy rust like a puff of cigarette drift in the air(1). Moya Moya first described by Suzuki and Takaku for an idiopathic, rare chronically progressive stenosis or occlusion of bilateral supra clinoidal internal carotid artery with extensive collateral from base of skull. Numerous entities mimicking Moyamoya disease appearance are called as Moyamoya syndrome(2). Moyamoya disease had variable distribution world wide, it is more common in japan and Korea(3,4). It is the important cause of childhood stroke nearly affecting 6% of pediatric stroke. This disease has bimodal age of distribution. Most patients presented in first decade of life and second peak around 3rd or 4th decade(5). TIA and ischemic stokes are more common in children(6). Hemorrhages are common in adult but can present in children(7). Hemorrhage occur either due to rupture of microaneurysm or fragile neovascular vessel. Most of patient diagnosed with the help of magnetic resonance angiography or computed tomography angiography. Digital subtraction angiography is a gold standard investigation(8).

Mainstay of the treatment is revascularization of ischemic area of brain. Surgical revascularization includes either direct STA-MCA bypass and indirect include EDAMS and many other techniques(9). We retrospectively review our surgical experience of treating Moyamoya disease.

METHOD –

All the cases are selected who undergone revascularization procedure for Moyamoya disease at Mahatma Gandhi medical college from 2017-2019. Some of the patients are presented to us in the emergency department with acute stroke and some are referred to us for refractory symptoms. Demographic data, mode of presentation, baseline modified Rankin score were recorded. Those patients who are presented with ischemic stroke had undergone MRI scan to look to perfusion deficit. All the patients either presented with ischemic or hemorrhagic stroke had undergone computed tomography angiography or digital subtraction angiography.

Those who presented with aneurysm bleed had undergone exclusion of aneurysm first and then STA-MCA bypass with EDAMS in second surgery. Those who presented either with ischemic stroke or TIA had undergo STA-MCA bypass with EDAMS. This procedure was planned to prevent further episode of bleed and infract in these patients.

Post operative Modified Rankin score done at the time of follow up. Primary end point of the study is either the recurrent stroke or progressive neurological deficit.

RESULT-

We have included 8 patients in our study, out of which mostly are male, male to female ratio was 5:3. In our patients the youngest patient was of 6 years and the eldest patient was of 45 years of age. Maximum patients presented in second decade of life. Except one patient most of the patients having bilateral involvement. Four out of eight patients are presented in Suzuki grade III (figure 1), reset of the patient are either grade II or grade IV. Five patients presented with either infarct or TIA and related complication (figure 2). Three patients presented with intracranial bleed, two out of three patients shows intracranial aneurysm one at distal anterior cerebral artery(figure 3) and another at branches arising from posterior choroidal artery. Rest of clinical feature are described in table 2. Seven out of eight patients undergone combine procedure(STA-MCA bypass with EDAMS). Five out of seven undergone bilateral procedure. Modified Rankin score was calculated at the time of admission and at 6 month to 1 year follow up. Mean score at time of admission was 2.87 and at follow was 1.62, there is no history of recurrent stroke or deterioration of clinical symptoms in any of our patients in mean follow up of 1 year.

Table 1 Age and sex distribution

	Male	Female
Children	3	1
Adult	2	2

Table 2 clinical feature

Infarct	3
Bleed	1
Aneurysmal bleed	2
TIA	2
Seizure	2
Vision loss	2
Speech	1

Sensory loss	2
Motor weakness	1
Headache	2

Table 3 mean of modified Rankin score in preop and at follow up at 6 month -1 year

	Pre op	Post op
Children	3	2
Adult	2.75	1.25
Overall	2.87	1.62
Follow up event	-	0

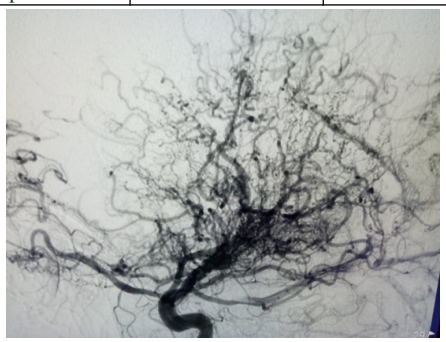

Figure 1 19 years old male patient presented with repeated transient attack of headache with aphasia. Digital subtraction angiography shows stenosis of bilateral ICA with appearance of Moyamoya vessel

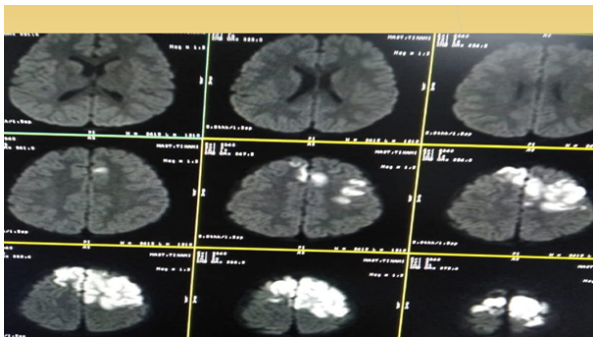

Figure 2 6year male child presented with recurrent episode of seizure with weakness of right side of body. MRI showed diffusion restriction in bilateral frontal lobe. Later Computed tomography angiography was done showing stenosis in bilateral ICA, ACA and MCA territory

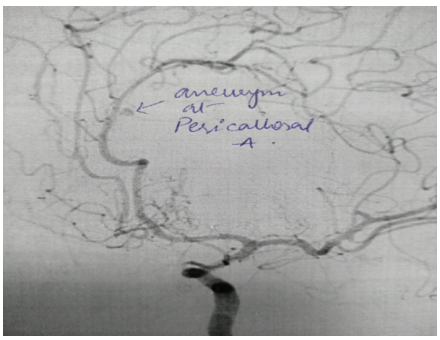

Figure 3 45 years old male patient presented with sudden onset of headache with altered sensorium. Computed tomography show interhemispheric hematoma with intraventricular bleed. DSA was done showing stenosis of bilateral ICA with aneurysm arising from branch of Distal anterior cerebral artery

DISCUSSION -

There are only few studies had been conducted in India. The exact incidence of Moyamoya disease in India remain unknown till now. We are the in the few centers in this region of India who provide treatment of this disease.

Shirvastava et al had reported a series of 26 patients with equal number of adult and children. They also showed female predominance of disease with female to male ratio of 3.3:1(10). In our study we found similar incidence of disease among adult and children but we summited slight male predominance than female, may be due to differential pattern of referral to a private hospital. Largest study published from India is by Sadashiva et al of 70 cases in which 54 patients are children and 16 patients are adult. They also showed female predominance with female to male ratio is 1.8:1(11).

In this series the most common symptoms present in patients are related to infarct or TIA. In our series 3 patients presented with bleed in which 2 are adult and 1 is child. In these patients 2 presented with aneurysmal bleed and both of the aneurysm are located at the distal branching point. Most of the Indian studies showed predominance of symptoms related to infarct which is similar to your experience except one study by Garg et al (9-11,13). Most of the aneurysm arising in Moyamoya are pseudoaneurysm arising from anterior or posterior choroidal artery and most of the major artery aneurysm occur in Vertebrobasilar system(12) resulted from hemodynamic stress suffered by posterior circulation due to the collateral suppling the anterior circulation. In our patient we see two aneurysm one arising from the branch of posterior choroidal artery and second arising from branch of distal anterior cerebral artery . in both cases we done exclusion of aneurysm from circulation as both aneurysm arising from the terminal branches suppling non eloquent part of brain. Both of patients recovered without any postoperative deficit. The patient with distal anterior cerebral artery aneurysm received STA-MCA bypass with EDAMS 3 moths later after the index surgery. Similarly Garg et al reported 11 aneurysm in 44 patients in which 5 aneurysm was coiled and 1 was clipped and out of 11 aneurysm 10 raised from posterior circulation(13).

In our study modified Rankin score was observed before the surgery and 6 month after the procedure. We had reported significant improvement in modified Rankin score in our patients. These improvement of more in pediatric patients then in adult patients. Sadashiva et al reported series of 70 patients in which 108 procedure was done. In their series 58 combined procedure and 50 indirect procedure was done. They noticed that combined procedure was associated with better outcome in compare to indirect procedure and pediatric patients had better outcome in compared to adult patients. Sahoo et al reported an experience of 33 case in which 40 procedure was done. 36 cases indirect procedure and 4 cases the combined procedure was performed. They also reported improvement in modified Rankin score in 20 month of follow up (14).

CONCLUSION

The incidence of Moyamoya is rare in this part of country. Despite that number of patients identified suffering from this disease. Surgery had shown marked improvement in neurological status and prevent the further deterioration. So all the patient who are presented with acute stoke and suspected to have Moyamoya disease should be thoroughly evaluated.

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No

CONFLICT OF INTEREST-

No

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