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INITIAL EXPERIENCE WITH INDIRECT REVASCULARIZATION SURGERY IN MOYA-MOYA DISEASE

Neurosurgery

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ABSTRACT

Moyamoya disease (MMD) is a rare idiopathic progressive vaso-occlusive disease characterized by irreversible condition of main blood vessels to the brain as they enter into the skull. MR Angiogram and CT angiogram are the modalities to diagnose this disease. Surgical revascularisation after initial stabilisation of patient is helpful in decreasing neurological symptoms in MMD. **Materials And Methods:** We identified 4 patients with MMD treated with EDAS at department of neurosurgery in our hospital. Diagnostic criteria for Moyamoya disease (MMD) was based on following findings in MRA or cerebral angiography :- (1) stenosis of supraclinoid ICA or proximal portions of ACA / MCA (2) abnormal collateral artery formation beyond occlusive artery (3) bilateral findings in both above criteria **Results:** EDAS resulted in better outcomes and no subsequent paralytic episodes in 6 month follow up period . **Conclusion:** Recurrent brain ischemic episodes in Moyamoya disease often leads to progressive neurological worsening in patients. Thus early diagnosis and surgical revascularisation is effective in preventing neurological attacks and subsequent morbidity.

KEYWORDS

INTRODUCTION

Moyamoya Disease (MMD) is a vaso-occlusive condition of distal Internal carotid Artery (ICA) or bilateral Anterior Carotid Artery (ACA) or Middle Carotid Artery(MCA) or any combination of these arteries leading to compensatory collateral artery formation .MMD is generally seen in pediatric population of 5-10 year age and in adults of 40-50 year age[1]. Pediatric population often present with ischemic stroke whereas hemorrhagic stroke is more common in adult[2].

The incidence of MMD exhibits significant regional differences, with a high incidence in East Asia and a low incidence in other regions. The prevalence of moyamoya disease (MMD) is about 10.5/100,000 in Japan. Incidence of Moya moya disease in India is poorly reported. But with increasing awareness there are increased reported cases of this disease across many centers in India too.

Moya moya disease often runs in families. It is often found associated with genetic disorders like sickle cell anemia, neurofibromatosis-1, Down's syndrome, congenital heart defects, antiphos- pholipid syndrome, renal artery stenosis, and thyroiditis[3] Prevelance of Moya moya disease in down's syndrome is estimated to be around 3.8%[4].

Case Description

Case1

9 year old male child affected with Down's syndrome referred to Neurosurgery department at our institute with repeated episodes of transient weakness in right upper and lower limbs for a month with frequency of attacks being 4-5 days/week.

MRI brain was suggestive of chronic infarction in internal carotid artery. Four vessel cerebral angiography was suggestive of bilateral supraclinoid Internal carotid artery(ICA) stenosis. ICA being filled via P-comm collaterals from Posterior cerebral artery and external carotid artery (ECA), complete occlusion of right vertebral arteries in v4 segment with collateral formation from branch of External carotid artery.

Antiplatelet therapy with aspirin was initiated and Encephalo-duro-arterio-synangiosis (EDAS) + encephalo-myo-arterio-synangiosis(EMAS) surgery was planned. Patient underwent successful surgery and was discharged on antiplatelets. On 6 month post-op followup the patient had no episodes of stroke or transient ischemic attacks.

Case 2

A 5 year old male patient presented to our hospital with complain of weakness in left upper and lower limb. Weakness lasted for around four to five minutes with complete gain in power thereafter. Patient had history of similar episodes for three times in last year. MRI Brain was suggestive of multiple acute non hemorrhagic infarction in right high parietal lobe, bilateral frontal lobe and right centrum semiovale. MR angiography showed severe segmental luminal narrowing in supracavernal segments of bilateral internal carotid artery (right>left). Moderate to severe diffuse narrowing was seen in intracranial part of rest of bilateral Internal carotid artery. Digital Substracion Angiography (DSA) was suggestive of complete stenosis of bilateral supraclinoid ICA. Patient underwent EDAS with EMS on right side. On 6 month follow up patient had no similar episodes of stroke or transient ischemic attacks

Case 3

A 6 year old female presented with complaints of multiple episodes of seizure and. Transient ischemic attacks causing weakness in left upper limb lasting for 2-3 minutes followed by full recovery.MRI Brain showed acute on chronic infarct in right ICA territory . MR Angiogram showed flow in Right MCA was attenuated and no flow in distal branches. Severe luminal narrowing of bilateral ACA .Moderate focal luminal narrowing of right supraclinoid ICA was seen . Diffuse moderate narrowing of right PCA was seen. Multiple intra parenchymal collaterals in capsuloganglionic-thalamic region (right >left) and meningeal collaterals was seen. EDAS with EMS was subsequently planned on 2 weeks followup after the acute event. Six month post surgery , patient had no episodes of TIA or strokes.

Case 4

10 year female presented with complains of weakness over right upper and lower limb for around a month associated with recurrent episodes of weakness over last 3 to 4 years. On examination patient was conscious but disoriented. Gait was hemiplegic and tone was decreased on right side with power 2/5 in right upper and lower limb. MRI brain with MRA was suggestive of multiple acute on chronic infarcts along left ICA territory with bilateral supraclinoid IGA stenosis with multiple collateral formation. Patient was started on antiplatetet drugs. EDAS +EMS was subsequently planned on 4 weeks followup after the acute event. Six month post surgery patient had no episodes of stroke or transient ischemic attack.

Surgical Treatment

Re-vascularisation procedure in our institution was done if the patient met this criteria - (1) diagnosed MMD (2) MMD patients presenting with ischemic / hemorrhagic symptoms

EDAS with EMS was preferred re vascularisation procedure for MMD at our institution . Procedure involves suturing of parietal branch of superficial temporal artery(STA) beneath dura in ischemic territories . The donor artery with strip of galea is detached from the fascia. Two

burr holes are made at proximal and distal ends of arterial bridge and the burr holes are connected to make an oval bone flap . The dura and arachnoid is then opened . The target artery is then sewn to pia mater with 10-0 prolene suture. Arachnoid may act as barrier for revascularisation so suturing of pia with arachnoid matter is done . The bone is then replaced after cutting out entry and exit points for envisage artery.

Clinical Followup

After discharge , outcome was made sure by clinical visits or telephone. Clinical outcomes was done by dividing into 3 categories. (1)- excellent :- no further ischemic attacks (2)- good :- ischemic attack but with decreased frequency (3)- poor :- ischemic attacks but with same frequency

Background And Discussion

Moyamoya disease (MMD) is a cerebrovascular disease of unknown etiology causing progressive steno-occlusive lesions in the supraclinoid internal carotid artery and/or its main branches in the circle of Willis. To compensate decreased blood flow , collateral vessels develops resembling puff of smoke around the area of occlusion hence the japenese term 'Moyamoya'.[5]

Clinical features of moyamoya disease may include monoparesis, hemiparesis, dysarthria, aphasia, headache, convulsions, and involuntary movements[5]. Children of moyamoya disease having bilateral occlusive lesions and history of recurrent stroke may develop intellectual and executive problems[6]. Acute episodes warrant symptomatic management in form of antiepileptic , antiplatelet drugs and supportive treatment. Surgery remains the mainstay in treating the disease.[5]

Since pediatric MMD is characteristically more progressive than in adult patients, revascularization surgery is indicated in most children with MMD.[7]

Surgical approach for treatment for MMD includes (1) direct Revascularization (2)indirect Revascularization and (3) combined revascularisation procedures.[2]

Direct techniques include STA-MCA bypass, STA-ACA (low performance) and OA- PCA bypass (visual disturbances) Direct techniques are difficult to perform however, are associated with better outcomes and improved revascularisation[2]. Vascular diameters of the anastomosing artery in one of the factors for consideration of direct procedure.One of the complications associated with direct revascularisation procedure in post-op period is hyperperfusion syndrome.

Indirect Revascularization procedures include encephalo-arterio-synangiosis(EAS) , encephalo-myo-arterio-synangiosis(EMAS), encephalo-duro-synangiosis(EDS), encephalo-duro-arterio-synangiosis(EDAS) , encephalo-duro-arterio-myo-synangiosis (EDAMS) as well as various combinations of these[8]. Advantage of Indirect revascularisation is that it is easy to perform and complication of hyperperfusion is rare however recovery in patients is slow compared to direct procedure[8].

In Encephalo-duro-arterio-synangiosis (EDAS) , STA is harvested with galea and periosteum and margins of galea and dura are sutured together to cover brain with arterial flap. Advantage with EDAS is that it has predictable outcome with immediate improvement of blood flow. Vessel thrombosis and hyperperfusion vessel disease may occur as a complication of this procedure.

Arachnoid may act as barrier for revascularisation during EDAS so suturing of pia with arachnoid matter was introduced. This further improved revascularisation seen as excellent post operative outcomes in the patients[8].

RESULTS

All the four patients operated in our institution showed excellent outcomes with no subsequent ischemic episodes post-operatively.

Most common complication post-operatively was subdural hematoma seen in one out of three patients which responded well to conservative management.

Other complications like epidural hematoma , seizure , sub-cutaneous effusion were not seen in our study .

CONCLUSION

MMD is an increasingly identifiable cause of recurrent neurological ischemic episodes in paediatric population. Early diagnosis and surgical intervention is warranted to prevent these episodes and subsequent morbidity. EDAS is an effective revascularisation procedure with promising post operative outcomes. Since arachnoid may act as barrier for revascularisation, we modified the surgery to include suturing of pia with arachnoid. This resulted in increased the success rate of revascularisation .

Limitations

As our study included only four patients, for further validation of these results replication on a larger sample size is needed. Also moya moya disease and its surgical outcomes in adult population was out of scope of this study.

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