2 7 ANNUAL O MEETING BOSTON, MASSACHUSETTS OCTOBER 7-11,2017

Moya Moya Disease: Single Centre Experience of 136 Patients

Ashish Suri; Manmohan Singh MCh; Shashwat Mishra MCh; Bhawani S. Sharma

Introduction

Moya Moya disease or "puff of smoke" is a progressive disease with occlusion of cerebral vasculature, especially around the Circle of Willis.

Methods

We analysed the records of 136 patients with Moya Moya disease evaluated and treated at our institute from 2001 to 2016. Adult and pediatric data were tabulated separately. Ischaemic and hemorrhagic presentation, surgical treatment with direct, indirect or combined revascularization and long term outcome as per follow up records were analysed with respect radiological parameters.

Results

A total of 136 patients were admitted, evaluated and managed at our tertiary care centre. Surgical treatment with direct, indirect or combined revascularisation was performed in 107 patients and 132 procedures i.e. hemispheres. Male to Female ratio was 1:1.43. There was a bimodal age distribution; 5-15 years and 25-40 years; 76 patients were in paediatric <14 years age group. Children presented with 100% ischaemic presentation; hemiparesis=78%, seizures=30%, TIA=16%, painless visual deterioration=18%, developmental delay=6% and headache=2%. Adults presented with ischaemic presentation in 62% and hemorrhagic in 18%; hemiparesis=70%, seizures=8%, TIA=10% and headache=24%. 80 indirect revascularisation procedures enchaphaloduro-arteio-myo-synagiosis – EDAMS or enchaphalo-myo-synagiosis - EMS were performed and 52 direct revascularization ie STA-MCA bypass procedures were performed. Improvement in hemiparesis occurred in 85% patients. Minor complications occurred in 8% patients -deterioration of motor power due to progression of infarct (1.8%), seizures (3.7%), wound infections (1.8%), subdural hematomaacute (0.9%) and chronic (0.9%).

Conclusions

Stroke in young children should raise a high index of suspicion for Moya Moya disease. Early diagnosis and treatment can prevent irreversible brain damage. Direct vascularization (ST-MCA) results immediate restoration of cerebral circulation and should be attempted whenever feasible. Indirect vascularisation (EDAMS) is simple but effective method of revascularisation.

Learning Objectives

Stroke in young children should raise a high index of suspicion for Moya Moya disease. Early diagnosis and treatment can prevent irreversible brain damage. Clinical presentation in paediatric patients is predominantly ischemic and in adults is both ischaemic and hemorrhagic. Direct vascularization (ST-MCA) results immediate restoration of cerebral circulation and should be attempted whenever feasible. Indirect vascularisation (EDAMS) is simple but effective method of revascularisation.

References

- 1. Sahoo SS, Suri A, Bansal S, Devarajan SL, Sharma BS. Outcome of revascularization in moyamoya disease: Evaluation of a new angiographic scoring system. Asian J Neurosurg. 2015 Oct-Dec;10(4):252-9.
- 2. Garg AK, Suri A, Sharma BS. Ten-year experience of 44 patients with Moya Moya disease from a single institution. J Clin Neurosci. 2010 Apr;17(4):460-3. Epub2010 Feb 26. IF-1.165;CI-10