The Mechanics of Moyamoya Disease

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Introduction

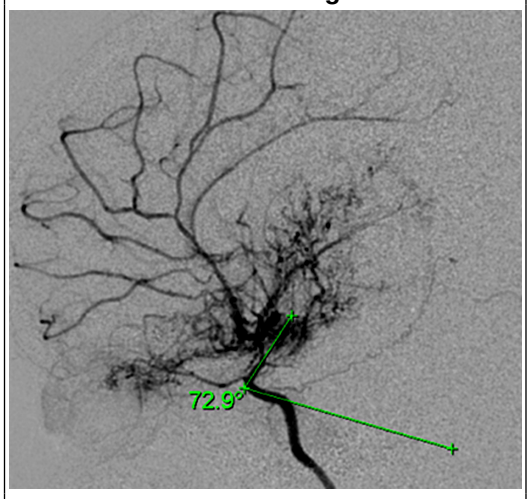
Moya moya disease (MMD) is characterized by progressive stenotic occlusion of the distal internal carotid arteries (ICA) and proximal anterior and middle cerebral arteries. Despite several associated conditions described, the exact aetiology of MMD has not been elucidated. The authors investigated if MMD could be caused exclusively by mechanical stresses induced by blood flow against a susceptible intracranial vasculature.

Methods

The pre-operative digital subtraction angiography (DSA) images of 54 patients diagnosed with MMD were reviewed. The angle subtended by the horizontal cavernous segment of the ICA and the supraclinoid segment (CAV-SCL angle) of the ICA in lateral view angiograms was measured and compared with age-matched controls.

The DSA image of the ICA from the cervical segment to the bifurcation into the anterior and middle cerebral arteries of a control subject was extracted. The (CAV-SCL angle) of the ICA in true lateral view was progressively increased using a software, to obtain four models of increasing angles viz. original, 600, 900 and 1100. The models were subjected to simulated blood flow studies using computational fluid dynamics (CFD) methodology.

CAV-SCL angle

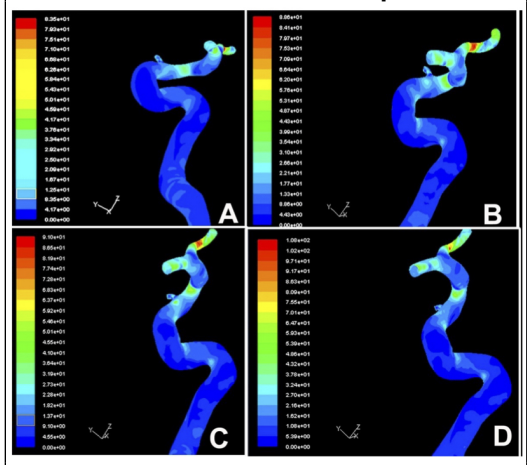


The angle subtended by the axes of cavernous and supraclinoid ICA was measured in lateral angiograms.

Results

The (CAV-SCL angle) of the ICA was greater in MMD patients compared to controls. The CFD methodology simulated blood flow in the ICA model. As the CAV-SCL angle was progressively increased, the wall shear stress increased in the vicinity of the ICA bifurcation. The high wall shear stress induces the formation of endothelial cushions. Erratic cushion formation acting on a 'swaying' supraclinoid ICA and hemodynamic disruption of the internal elastic lamina causing changes in the tunica media could account for the pathogenesis of MMD.

The wall shear stress plot



Wall shear stress increases as the CAV-SCL angle is increased. A. Original B. 60 degrees C. 90 degrees and D 110 degrees

CAV-SCL angle observations

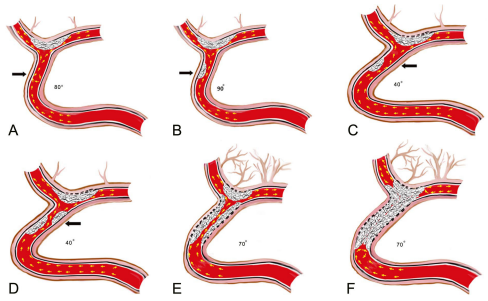
	Observed angles	Mean and SD of Cavernous-supraclinoid ICA angle measured among		P-value
	(number)	Patients with MMD	Patients without MMD	
All	105	69.76(26.44)	40.5(15.84)	<0.0001
Adult	43	70.14(31.39)	37.93(20.49)	<0.0001
Minor	62	69.5(22.65)	42.29(11.43)	<0.0001

The mean CAV-SCL angle was significantly greater in the MMD group compared to the age-matched control group.

Conclusions

Moya moya disease occurs in the setting of a unique interplay of mechanical factors acting on a susceptible weak vasculature. The 'Mechanical Theory' of Moya moya disease could explain the pathogenesis of this enigmatic disease.

"Mechanical theory" of Moyamoya disease



A to E shows the varying CAV-SCL angle. B. With increasing angle, blood flow becomes turbulent and a new endothelial cushion develops at the site of blood stream impingement marked by the bold arrow.Streamlined blood flow is restored. C. The angle reduces as the SCL segment of the ICA is mobile and slumps down to reduce flow. A new impingement zone develpos. D. A new endothelial cushion develops and streamlined flow is restored. E. The endothelial proliferation is dysregulated. Cerebral perfusion is compromised. Retrograde blood flow is established via pial collaterals. Lenticulostriate perforators proliferate to improve perfusion. The ICA blood stream may dislodge endothelial cushion emboli. The internal elastic lamina is disrupted. F. The advanced stage of MMD with total obliteration of lumen of distal ICA and proximal ACA and MCA. Extensive collaterals develop in an attempt to augment blood flow to the brain.