45-year-old male presenting with acute right hemiparesis and aphasia

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Acute Stroke in Moyamoya Disease



CT non-contrast



Left paramedian hypodensity, concerning for acute infarct



CT angiogram



Right M1 occlusion

Left M1 occlusion versus high-grade stenosis



CT angiogram

opacification of the

Lack of

right MCA

Left M1 occlusion versus high-grade stenosis

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CT angiogram

Right ACA high grade stenosis



Left ACA high grade stenosis



CT perfusion



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Left ICA angiogram



Left ICA bifurcation with high-grade stenosis in both left ACA and left MCA



MRI DWI







DWI hyperintensity corresponding to ADC hypointensity = diffusion restriction, as may be seen in the setting of acute ischemic infarction



Moyamoya

Moyamoya disease is an idiopathic, progressive, non-inflammatory, non-atherosclerotic, vaso-occlusive disease characterized by progressive narrowing of distal ICA/proximal circle of Willis vessels with secondary collateralization.

The term moyamoya disease (MMD) should be reserved for an <u>idiopathic</u> condition.

Moyamoya syndrome can be due to numerous conditions causing arterial occlusion of the circle of Willis with resultant collateralization. May have similar appearance to Moyamoya disease.



Moyamoya

Epidemiology

- Bimodal distribution
 - 5-10 years and second peak during 4th decade
- Most frequent cause of stroke in Asian children

Presentation

- Most common is ischemic stroke followed by TIA and intracranial hemorrhage
- Less frequently present with seizure



Imaging Findings

Cerebral angiogram

 Small abnormal net-like vessels proliferate giving characteristic "puff of smoke" sign

CT without contrast

- 50-60% of affected children show anterior>posterior atrophy
- Stroke (children)
- Intracranial hemorrhage (more common in adults)
- Well circumscribed, hypodense lesions measuring simple fluid density

CT with contrast

- Enhancing dots (enlarged lenticulostriate arteries) in basal ganglia
- Abnormal netlike vessels at base of brain

CT perfusion

- Depicts penumbra and infarct core in acute ischemia



Imaging Findings

MRI

- T1: multiple dot-like flow voids in basal ganglia
- T2: increase sign in small vessel cortical and white matter infarcts, collateral vessels = net-like filling defects in basal cisterns
- Flair: Bright sulci = leptomeningeal poison ivy sign, slow-flowing engorged pial vessels, thickened arachnoid membranes, correlates with decreased cerebral vascular reserve
- DWI: helpful for "acute on chronic" disease
- T1+C: enhancing "dots" in basal ganglia and net-like thin vessels in cisterns, leptomeningeal enhancement



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