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Moyamoya Disease Presenting with Stroke

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Moyamoya disease is a rare chronic and progressive cerebrovascular disease characterized by bilateral stenosis or occlusion of the arteries around the circle of Willis with prominent dilation of arterial collateral circulation. The estimated overall incidence is 0.086 per 100,000 in the US.

The patient is a 60 year old female with hypertension and diabetes who presented with slurred speech and right sided arm weakness starting this morning. Family reports the patient woke up with these symptoms at 8 am and last known normal the night prior. On exam the patient had 4/5 muscle strength in the right upper extremity, right sided facial droop, slurred speech, and diminished sensation of the R face, right upper extremity. Code stroke was activated, CT w/o contrast showed no evidence of infarct or hemorrhage. CTA brain showed suspected 65% stenosis of the distal left petrous segment. Without large vessel occlusion no intervention was performed and the patient was outside the window for TPa. The patient's symptoms resolved after one day. Two days later she began complaining of new left sided weakness, numbness, and facial droop. An MRI diagnosed Moyamoya syndrome with an acute infarction in the R middle cerebral artery involving the right corona radiata, right frontal and parietal centrum semiovale. Frequent neurological checks were performed with permissive HTN. Aspirin 81 mg daily + clopidogrel 75 mg daily x 3 days was initiated and on day 4, clopidogrel antiplatelet monotherapy was continued. Neurological symptoms improved but did not resolve with L sided weakness of upper and lower extremities. Patient was discharged home with scheduled neurosurgery follow up for definitive treatment with EC/IC bypass.

Moyamoya means puff of smoke in Japanese after the puffy and cloudy appearance of vessels on imaging. The disease was first described in Japan and has a higher prevalence in East Asian populations. It is associated with RNF213 gene on chromosome 17q25.3 as well as alleles of class II human leukocyte antigen. Clinical presentation is variable and includes TIA, ischemic stroke, hemorrhagic stroke, and epilepsy. Imaging is diagnostic with brain MRI or MRA. The disease is staged using the Suzuki stages showing initial narrowing of carotid fork to stenosis and eventual obliteration of cerebral arteries with circulation supplied by only the external carotid artery system. Early anticoagulation is used in patients who suffer ischemic strokes with secondary stroke prevention involving revascularization. Direct revascularization procedures include bypass of stenosed circle of Willis vessels. Indirect revascularization procedures increase development of collateral circulation to prevent repeated ischemia.