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Abstract 22-03

Title: Moyamoya Syndrome with Associated Hypercoagulable State

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Introduction/Background: Moyamoya syndrome describes a cerebrovascular susceptibility to reduced blood flow through both the internal carotid artery (ICA) and its proximal branches.1 Collateral vessels form to supply blood to brain that is distal to the stenosis, producing a characteristic "puff of smoke" on angiography. Epidemiology demonstrates a predominance in women, typically affecting young children and adults in their mid-forties.1 The initial presentation commonly involves stroke, TIA, or intracranial hemorrhage secondary to disrupted blood flow. Moyamoya presents either as primary disease or part of a larger syndrome, as an underlying disease process secondary to an existing condition.1 Related conditions include Neurofibromatosis type 1, Sickle Cell Disease, irradiative therapy for CNS lesions, and Down's Syndrome.1 In this case, we describe a patient presenting with a right MCA ischemic stroke subsequently diagnosed as Moyamoya syndrome secondary to Antiphospholipid Syndrome.

Description: Ms. D is a woman in her forties with hypertension, hyperlipidemia, uncontrolled diabetes mellitus type II (A1C 9.4), epilepsy, and prior stroke 1 month prior to presentation. She initially presented with left hemiparesis, right gaze deviation, and incoherent speech and received an NIHSS score of 3 upon initial evaluation for left lower facial droop and left hemianesthesia. Alteplase was contraindicated due to recent prior stroke and the patient was not a candidate for intra-arterial thrombectomy due to low NIHSS score. She was initiated on secondary stroke prevention with aspirin and atorvastatin. Over the course of her hospitalization, her symptoms worsened including a left homonymous hemianopia, left upper extremity drift, and extinction to bilateral simultaneous stimulation. She denied use of tobacco, alcohol, or recreational drugs and recounted a family history of stroke in her mother.
Imaging revealed an acute right frontotemporal infarction and posterior temporo-occipital infarcts with multiple subacute and chronic infarcts in the left cerebral hemispheres suggesting long standing cerebrovascular disease. Head CT angiogram revealed a "puff of smoke" in the left M1 distribution, raising concern for Moyamoya. Conventional angiography was pursued which confirmed the diagnosis of Moyamoya syndrome.
A hypercoagulable panel was collected to evaluate for the etiology of the multiple ischemic strokes in this relatively young female. Labs revealed an elevated anticardiolipin IgM, elevated Beta2 glycoprotein IgG, and positive Lupus anticoagulant. Hematology was consulted and recommended treatment with anticoagulation for Antiphospholipid Syndrome (APLS)2. Interestingly, this presented a therapeutic challenge as imaging revealed a trace occipital hemorrhage. However, the risks and benefits of therapy were weighed and the patient was ultimately initiated on therapeutic anticoagulation therapy with warfarin.

Discussion and Conclusion: Discussions between the Vascular Neurology and Hematology teams led to the conclusion that this patient's Moyamoya syndrome is likely multifactorial in

etiology, secondary to APLS and T2DM. This case describes a presentation of Moyamoya syndrome with an multifactorial etiology which contributes to existing knowledge of this syndrome's pathophysiology.3

References:

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