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Case Report

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A Case of Moyamoya Physiology Presenting as Abdominal Pain

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Case Introduction

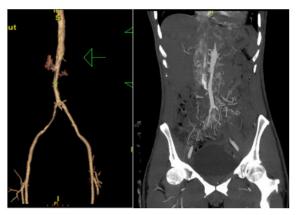
33 y.o. left handed male with a medical history significant for hypertension (HTN) who presented for further evaluation of abdominal discomfort. He reported a history of chronic postprandial periumbilical discomfort ongoing for eighteen months. He denied any diarrhea, hematochezia, melena or vomiting. His review of systems noted a weight loss of 30 lbs in less than a year that he attributed to loss of appetite secondary to pain. He was diagnosed with uncontrolled HTN in 2013. Family history is significant for hypertension in parents when in their 60s. Patient does not smoke, consume alcohol or use illicit drugs.

Pertinent Exam Findings

On exam, patient was noted to have an abdominal bruit, carotid bruit and 4/6 systolic heart murmur loudest in the second intercostal space. Cognition was intact, and no focal neurological deficits were observed.

Pertinent Workup

CT angiography of abdomen showed extensive noncalcified soft tissue plaques causing severe stenosis of the distal aorta, extending into bilateral common iliacs, moderate stenosis of right common iliac, occlusion of the celiac arteries, superior and inferior mesentaric arteries, right hypogastric and two right renal arteries and an irregular, highly stenotic left renal artery. Given the extent of disease and carotid bruit on examination, vascular imaging of brain and chest was performed. CT angiography head and neck showed 50% stenosis of the left and right common carotid arteries and occluded bilateral Internal Carotid Arteries (ICA). The right vertebral artery was patent, and left vertebral artery was occluded. The entire intracranial circulation was supplied by the right vertebral artery and collateral flow through the external carotid arteries. Multiple collateral arteries were present along the neck, suggesting chronic ICA stenoses. CTA head in addition showed that bilateral M1 Middle Cerebral Artery (MCA) segments and A1 Anterior Cerebral Artery (ACA) segments were not seen. Prominent bilateral lenticulostriate collaterals were present. An extensive workup for hypercoagulability, vasculitis and malignancy was unremarkable (Figure 1).



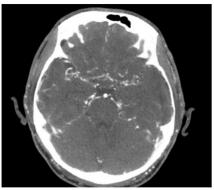


Figure 1: Moyamoya physiology- Images show Reconstruction of Abdominal Aorta and Multiple collaterals noted on CTA abdomen and CTA head.

Discussion

The findings were consistent with an atypical presentation of systemic large vessel vasculopathy, with significant extracranial and intracranial vasculopathy and imaging features consistent with Moyamoya physiology.

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