

Takayasu Arteritis: A Cause of Necrotizing Giant Cell Aortitis

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Introduction

Takayasu arteritis (TAK) is a large-vessel vasculitis which primarily affects the aorta and its main branches. It shares some histologic and clinical features with giant cell arteritis.

Case presentation

21-year-old African American female who presented to the emergency department with 3-day history of severe chest and upper back pain. Labs showed a positive ANA 1:80 in a speckled pattern, RF 62, RNP 1.9, ESR 50 and CRP 1.0. The other autoimmune serologies were negative. Chest radiograph showed dilatation of the thoracic aorta without mediastinal widening. CT angiogram of the chest, abdomen and pelvis demonstrated diffuse dilatation of the thoracic aorta extending from the aortic root through the mid to distal descending thoracic aorta, greatest in the proximal aortic arch where there was moderate to severe dilatation. She underwent emergent aortic valve sparing root replacement, replacement of the ascending aorta, total aortic arch replacement and right axillary conduit placement by cardiac surgery. The biopsy of the aorta was consistent with necrotizing giant cell aortitis. She was started on prednisone and azathioprine for Takayasu arteritis with improvement in symptoms.

Discussion

Takayasu arteritis primarily affects the aorta and its main branches. Inflammatory process leads to narrowing, occlusion, or dilation of the arteries. Predominantly affects young women. The symptoms may include limb claudication, blood pressure discrepancy between extremities, diminished pulses, angina and neurologic symptoms. Imaging is done with either CTA, MRA or PET-CT. Diagnosis is made based on typical symptoms and specific vessel imaging findings. Biopsy of the large arteries shows findings like giant cell arteritis. Systemic glucocorticoids are the mainstay of therapy. No specific steroid sparing agent has been well proven to be effective, but options include methotrexate, azathioprine and TNF inhibitors. Vascular intervention may be necessary for stenosed or occluded arteries and for the management of aneurysmal disease.