Takayasu's Arteritis: Spectrum of disease

Pablo Zertuche MD[1], Ginger Janow MD[2], Kristin Wong MD[1]
[1]Rutgers New Jersey Medical School, Departments of Medicine & Pediatrics, [2]Hackensack University Medical Center, Department of Pediatrics

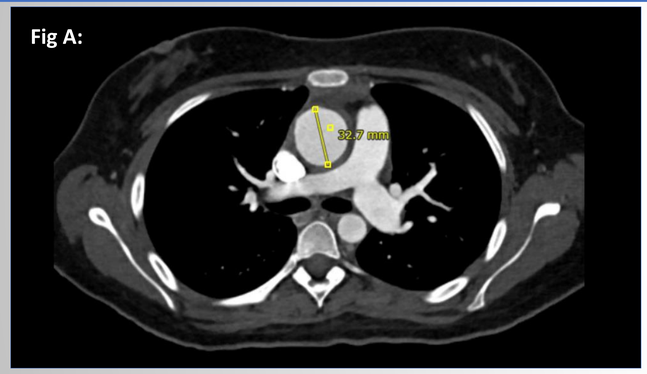
CASE 1: Presentation

A 17 year old woman presented to the ED with intermittent worsening chest pain described as heaviness and tightness with exertion for the past 2-3 weeks.

She had 3 recent ED visits where she was found to have elevated inflammatory markers, normal cardiac enzymes, normal EKG and echocardiogram that showed a small circumferential effusion. She was treated for suspected pericarditis with NSAIDs and then colchicine without improvement.

Troponin was mildly elevated to 0.74 (normal less than 0.01). Echocardiogram showed mild aortic dilatation and mildly decreased LV function (when compared to previous study). An EKG showed diffuse elevation of ST segments.

CASE 1: Imaging



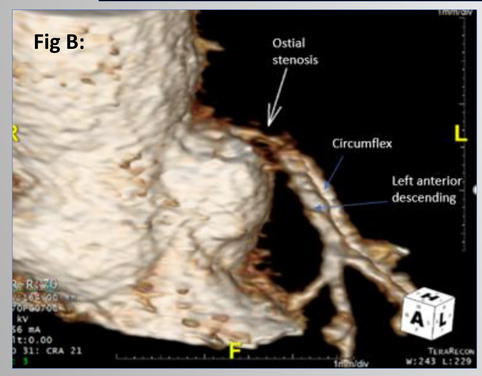


Fig A: CT Angiogram showing mild dilatation in the ascending aorta with circumferential mural thickening and inflammation and high-grade stenosis of the left main coronary artery. Rest of vasculature normal.

Fig B: 3D Reconstruction showing high-grade stenosis of the left main coronary artery.

CASE 1: Clinical course

Patient underwent cardiac catheterization which showed ostial stenosis of the LAD. She then had ascending aortic replacement and CABG x3 with LIMA to OM, RIMA to LAD, and SVG to RCA. Patient was started on tocilizumab and methotrexate and pulsed with corticosteroids.

Symptoms returned after a few months with repeat left heart catheterization showing re-stenosis. Currently pending further intervention.

INTRODUCTION

Takayasu arteritis (TAK) is a vasculitis of the aorta and its major branches that affects mostly women of young age, with up to 30% of patients presenting during childhood[1]. Because of the heterogenic vascular involvement, clinical presentation is highly variable, and early in the disease symptoms can be non-specific. Timely diagnosis is challenging which leads to high morbidity and mortality secondary to vascular damage. We present 2 cases of variable presentation and morbidity in adolescent women.

CASE COMPARISON

	Case 1	Case 2
Symptom Duration	Weeks	Months
Primary Symptoms	Chest pain	Light headedness, claudication
Primary Exam Findings	None	BP difference and bruits
Vascular Involvement	Localized	Extensive
Major Complication	Cardiac	None
Treatment	Steroids, MTX, tocilizumab	Steroids, MTX, tocilizumab
Prognosis	Poor	Reserved

DISCUSSION

Symptoms in TAK vary depending on location of vascular involvement and morbidity and mortality can be worse in patients with lower burden of disease but anatomically critical lesions. During the inflammatory stage of the disease, treatment is required to prevent vascular damage which includes stenotic and occlusive lesions.

Once vascular lesions develop, surgical or endovascular intervention might be necessary for complications such as uncontrolled renal hypertension, coronary ischemia, limb ischemia, severe aortic regurgitation, etc. These procedures have high risk of complications, which worsen with higher disease activity, including re-stenosis.[2]

A large Canadian cohort shows that relapses and major complications within a year of diagnosis are similar in pediatric and adult patients (39% and 30% in children and 28% and 43% in adults respectively). At last follow up (2.6 years for children and 3.9 years for adults) the rate of inactive disease was 67% and 88% respectively.[1]

CONCLUSION

It is important to keep a high index of suspicion in patients whose symptoms could be attributed to ischemia from TAK in order to minimize inflammatory damage with timely treatment and prevent morbidity and mortality.

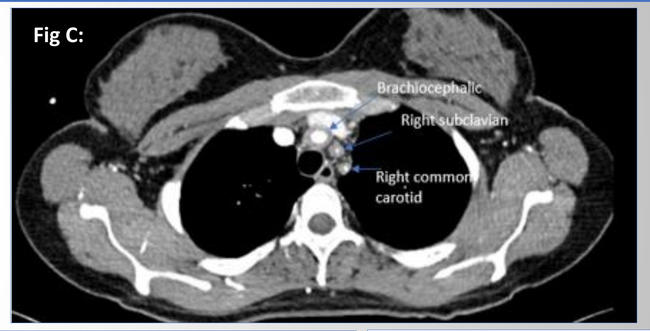
CASE 2: Presentation

A 19 year old woman presented to the ED complaining of worsening bilateral calf pain after walking 2 blocks and shoulder pain with activity for 5-6 months. Also complaining of intermittent dyspnea, dizziness and hand numbness and tingling. Patient has had several ED visits for lightheadedness and was treated for hypotension with IV fluids several times.

On physical exam, patient had absent bilateral radial and left dorsalis pedis pulses and a bounding right dorsalis pedis pulse. Bruits were noted on the abdomen, bilateral neck and the right popliteal area.

Blood pressures: LA: 68/42, RA: 78/56, LL: 86/58, RL: 105/68

CASE 2: Imaging



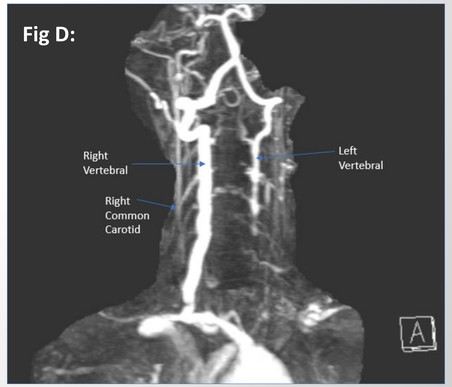


Fig C: CT angiogram showing diffuse wall thickening with moderate to severe narrowing of the proximal right common carotid and proximal subclavian arteries.

Fig D: MRA showing severe stenosis of the right and left common carotid and internal carotid arteries, and a very prominent right vertebral artery providing most of the intracranial arterial blood supply. Arteritis also found throughout the aorta, arch vessels, pulmonary arteries, and renal arteries with complete occlusion of the superior mesenteric artery.

CASE 2: Clinical course

Patient was started on tocilizumab and methotrexate and pulsed with steroids. Discharged on steroid taper with normalization of inflammatory markers and improvement of dizziness, claudication and arm pain. Currently patient is back to her daily activities and stated "I feel alive for the first time in years."

References:

1. Aeschlimann FA, Barra L, Alsolaimani R, et al. Presentation and Disease Course of Childhood-Onset Versus Adult-Onset Takayasu Arteritis. *Arthritis & Rheumatology*. 2019;71(2):315-323. doi:10.1002/art.40690
2. Di Santo M, Stelmaszewski EV, Villa A. Takayasu arteritis in paediatrics. Cardiology in the Young. 2018;28(3):354-361. doi:10.1017/S1047951117001998