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Takayasu Arteritis: An Exceptionally Advanced Clinical Presentation

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ABSTRACT

Takayasu Arteritis (TKA) is granulomatous inflammation of the proximal aorta and its main branches, characterized by granulomatous inflammation. As compared to Giant Cell Arteritis (GCA), diagnosis of TKA is a clinical challenge owing to vague initial symptoms and impossibility to biopsy major vessels, therefore, clinical and radiological parameters are the main diagnostic standards. Ishikawa's guidelines were the first clinical guideline regarding the diagnosis of TKA, however, guidelines by the American College of Rheumatology are the mainstay of clinical diagnosis. The development of TKA has been associated with Mycobacterium tuberculosis infection and with rheumatic fever in case series, but the exact trigger factor is unknown to the best of our knowledge. Damage caused by TKA is irreversible due to ischemic changes, therefore active disease status, and end-organ damage presenting as renal artery stenosis, severe hypertension, and shrunken kidneys are the poor prognostic factors. TKA is the most common cause of non- atherosclerotic vascular stenosis. We present a case of a young female, who presented at 16 years of age with involvement of ascending and descending aorta that required coronary artery bypass grafting at age of 17 and later on required Percutaneous Coronary Stenting to Left Main Stem too. Due to worsening hypertension and shrinking kidneys, she required renal artery stenting. In the course of progression, she developed radial artery and carotid artery stenosis, however, the disease progress started to halt, once she was started on glucocorticoids.

Keywords: Takayasu Arteritis (TKA); Giant Cell Arteritis (GCA); Hypertension; Cardiology

INTRODUCTION

Takayasu arteritis, also known as the pulseless disease is a large vessel vasculitis characterized by granulomatous inflammation involving the aorta and proximal aortic offshoots.[1] Although originally reported in young Asian females, disease epidemiology details are quite restricted, and have been found to have worldwide distribution. Takayasu arteritis has variable presentation posing an extreme clinical challenge to diagnose, initially presenting with non-specific systemic inflammatory symptoms, including night sweats, fever, weight loss, and fatigue in earlier disease.[2] Advanced disease presentation is owing to the vascular narrowing and occlusion of the aorta and its

primary offshoots.[3] Takayasu arteritis was first reported in Japanese literature in 1830,[4] but it was not until 1988 when the first clinical criteria for diagnosis of Takayasu arteritis was developed by Ishikawa.[5] Later on the diagnostic guidelines were described by the International Chapel Hill Consensus Conference (CHCC) in 1994,[6] and revised in 2012.[7] However, American College of Rheumatology guidelines published in 1990 still is the most clinically validated guidelines to distinguish Takayasu arteritis from other vasculitides[8] which include one radiologic and 5 clinical parameters.

CASE DETAILS

We present a case of 16 YO F presented to the cardiology clinic with palpitation, weakness, and episodic chest pain with exertion. The patient denied any significant past medical or family history. On examination, the patient was hypertensive, with a parasternal ejection systolic murmur and blood pressure difference on 30 systolic between upper and lower extremities. Resting EKG was normal and Echocardiogram showed supravalvular aortic stenosis with no gradient and relative narrowing of descending aorta with a systolic gradient of 130mmHg. The patient was started on atenolol, atorvastatin, and Lisinopril. An Aortogram revealed supravalvular aortic stenosis, dilated and calcified ascending aorta. The descending aorta was irregularly narrowed with tubular narrowing from the origin of the left subclavian artery up to the abdominal aorta with mild localized obstruction showing a trans-aortic pressure gradient of 32mmHg. Cardiothoracic surgery consults advised to continue medical management.

A month later, the patient presented with new-onset exertional chest pain radiating to the left arm, and headache despite good medication compliance. EKG was suggestive of ST depression in anterior and lateral chest leads with left axis deviation, with raised Troponin T. Angiogram showed moderate ostial stenosis of the left main stem, with the normal distal flow to LAD, LCX, and tight stenosis of Right coronary artery which was small caliper non-dominant vessel. Bilateral Renal artery stenosis was also detected during the angiogram, but her renal function tests were normal. CT Coronary angiogram revealed porcelain aorta involving ascending aorta and arch of aorta, thickened and narrowed descending aorta with moderate stenosis of right subclavian artery, total proximal occlusion of the celiac trunk, tight proximal stenosis of Superior Mesenteric Artery, tight proximal stenosis of Right and left renal artery with normal internal mammary arteries. Transthoracic Echocardiogram showed turbulent flow across ascending aorta with a pressure gradient of 25mmHg, with narrowing of descending aorta with peak systolic gradient of 120mmHg. Positive laboratory findings included elevated erythrocyte sedimentation and C-reactive protein. Blood cultures, venereal disease research laboratory tests, and autoimmune serological findings were negative. The patient underwent off-pump coronary artery bypass graft surgery where the left internal mammary artery was anastomosed with the left anterior descending artery with the uncomplicated postoperative course.

The patient remained stable in medical therapy for 3 years subsequently with aspirin, clopidogrel, atorvastatin, atenolol, Lisinopril, and amlodipine. She gradually started to experience worsening blood pressure control that was managed with amiloride, furosemide, and glyceryl trinitrate. CT Angiogram for graft status showed a patent graft

with bilateral proximal renal artery stenosis. Repeat coronary angiogram revealed Left main stem tight ostial stenosis, tight right renal artery stenosis with moderate left renal artery stenosis. In the next two months, the patient again started to experience episodic chest pain and exertional shortness of breath. An echocardiogram revealed turbulent flow in descending aorta with an Aortic Pressure Gradient of 35mmHg. Medical management was continued with methyldopa, trimetazidine, doxazosin, and telmisartan, but due to progressive symptoms, PCI to left main stem to left circumflex was performed. Two months later, the patient underwent right renal artery stenting due to worsening hematuria and poor blood pressure control. The patient had escalating treatment for worsening blood pressure control with doxazosin, isosorbide dinitrate, nebivolol, amlodipine, valsartan, furosemide hydralazine, and nifedipine with inadequate control. Follow-up Renal angiogram revealed patchy calcification of infrarenal aorta with patent right renal artery lumen developing mild in-stent restenosis.

The patient developed another episode of hypertensive emergency with BP escalating to 210/110. EKG suggested RV strain. Examination revealed a weakened right radial pulse. Cardiac enzymes were positive. Blood pressure was controlled with amlodipine and carvedilol. The patient was referred for vascular surgery consult for radial artery before proceeding for PCI to coronaries. Rheumatology workup showed elevated ESR and CRP. The abdominal sonogram revealed splenomegaly, bilateral shrunken kidneys with normal renal Doppler. The patient was diagnosed to have large vessel vasculitis involving aortic offshoots. The patient was started on low-dose corticosteroids and low-dose methotrexate, with stabilization of ESR and CRP.

Over a year, the patient developed worsening claudication of upper extremities with dizziness, exertional chest pain with lower extremities claudication with a claudication distance of 40 steps. Cardiac enzymes were elevated. Echocardiogram revealed trace TR, Trace MR with moderate concentric LV Hypertrophy with grade II diastolic Dysfunction and Ejection fraction of 65%. CT angiogram revealed a normal caliber of the aorta with bilateral renal arteries with no evidence of stenosis as shown in following figures. Carotid Doppler showed a circumferential thickness of bilateral carotid arteries with a focal area of calcifications, which were hemodynamically non-significant. The patient was discharged to follow up with continued medical therapy.

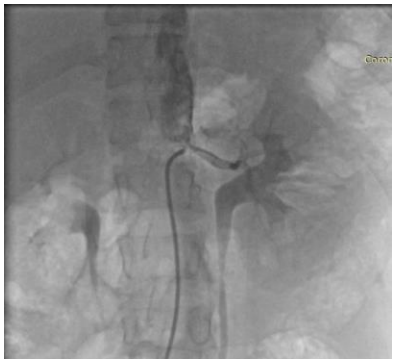

Figure 1: Renal Artery Angiogram with stent in situ.

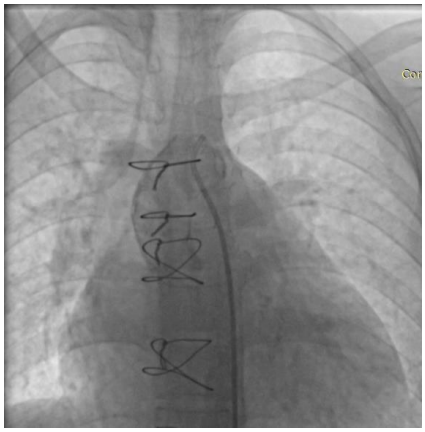

Figure 2: Aortogram showing aortic arch calcification.

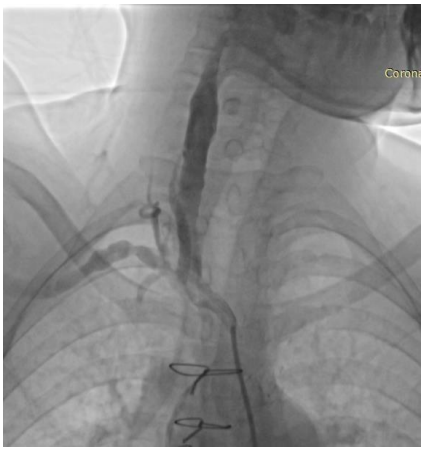

Figure 3: Right Subclavian artery stenosis with stenosiscarotid stenosis.

Figure 4: Radial artery

DISCUSSION

Takayasu arteritis is a rare disease, with worldwide incidence estimates around 2.6 cases per million per year,[9] and prevalence varies between 2.6-6.4 cases per million population, being highest at 40 per million in Japan and lowest at 0.9 per million in the US.[10] Female patients tend to involve the proximal part of the aorta mainly the arch and the proximal branches, whereas male patients involve the distal part of the aorta involving the abdominal aorta and branches[11] Because of the rarity of the disease, and biopsy of major vessels involved is impossible, etiology of the disease cannot be ascertained. Radiology remains the most reliable method to diagnose the disease. CT and MRI may reveal homogenous concentric thickening of the arterial wall, with stenosis and aneurysms. PET scan shows active metabolic disease at foci of Arteritis.[12]. Mycobacterium Tuberculosis' 65-kd Heat shock protein has been reported in a case series as a possible etiology of Takayasu arteritis because of elevated level of IgG, and IgA titers against M tuberculosis extract than the patients without Takayasu arteritis.[13] Severe renal vasculature involvement in the Takayasu arteritis requiring endovascular stenting in the absence of atherosclerosis is a rarely documented incidence,[14] whereas, in the presented patient, the patient did not only require endovascular stenting, she developed in-stent thrombosis too. Involvement of proximal aorta with stenosis of proximal coronary arteries especially aortic Ostia is a known complication with Takayasu arteritis. It has been reported that proximal anastomosis improves clinical outcomes in patients requiring bypass grafting.[15] Damage to vessels in Takayasu arteritis is irreversible, therefore the early start of treatment before the onset of Pulseless status, improves the patient's long-term prognosis.[16] It is therefore recommended by the American College of Rheumatology to start non-glucocorticoids along with the non-glucocorticoid immunosuppressant as initial therapy.[17] Sun et al found in their study that patients presenting with renal insufficiency (HR 2.37, 95% CI 1.76-15.83, P = 0.03), bilateral renal artery involvement (HR 6.95, 95% CI 1.18-21.55, P = 0.01), and severe stenosis (> 75%; HR 4.75, 95% CI 1.08-11.33, P = 0.05) predict adverse outcomes. It was also found that patients who did not have preoperative treatment had a higher chance of restenosis of renal arteries (41.46% vs 16.67%, P < 0.01) and worsening hypertension post-procedure (25.93% vs. 10.53%, P < 0.01). Active disease at the time of revascularization was another independent predictor of adverse outcomes following revascularization.[18]

CONCLUSION

Takayasu arteritis along with the Giant cell arteritis causing inflammation to the walls of major vessels consequences of which can result in vascular stenosis, aneurysms, and ischemic damage to end organs, which is irreversible in most cases. Early diagnosis leading to early treatment is the cardinal step of the management process with dictates the long-term prognosis of the disease. Therefore, the development of diagnostic criteria with good sensitivity and specificity should be established to help the clinician diagnose the disease in the early phase. Essentially, active disease, with worsening hypertension, renal insufficiency, and end- organ damage are worse prognostic markers.

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