

An unusual case of acute heart failure

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ABSTRACT

Takayasu's arteritis is a large vessel vasculitis of unknown etiology affecting predominantly young women. This case report describes an atypical presentation of this disease in a 49-year-old Caucasian woman who presented in acute congestive heart failure. Workup showed occlusion and stenosis of multiple branches of the aorta, including the left subclavian, right renal, celiac artery, and superior mesenteric artery. She had moderate aortic regurgitation, an aneurysm of the ascending aorta, and a severely reduced ejection fraction of <20%. Clinical and radiographic improvement occurred following prompt immunosuppressive treatment with corticosteroids.

Keywords: Takayasu arteritis, vasculitis, heart failure

INTRODUCTION

Takayasu's disease is a chronic inflammatory disease of predominantly large arteries, including the aorta and its main branches, the pulmonary arteries, and the coronary vessels. The disease is named after a Japanese ophthalmologist, Mikito Takayasu, who first described the disease in a young woman of Asian descent.¹ Below we describe a rare and late presentation of the disease as acute congestive heart failure in a Caucasian woman.

CASE REPORT

A 49-year-old Caucasian woman was admitted to the hospital with progressively worsening cough and shortness of breath of 5 days' duration. Her cough was mildly productive with white mucoid sputum. She had exertional dyspnea without orthopnea or paroxysmal nocturnal dyspnea. She also had associated fatigue and nausea without accompanying fever or chills.

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Her past medical history was significant for hypertension; however, she was noncompliant with her medications. Her past surgical history included an appendectomy and cholecystectomy. Family history was significant for coronary artery disease in her mother and hypertension in multiple family members. She had history of chronic tobacco abuse and occasional alcohol use.

On presentation to the hospital, she was alert, oriented, tachypneic, and in mild distress. Vital signs showed a heart rate of 86 beats per minute and hypoxemia with O₂ saturation in the 70s on room air. Physical examination revealed a feeble left radial pulse but normal pulses in her other extremities. A significant blood pressure discrepancy was noted between her arms.

The blood pressure recorded in her right arm was 130/98 mmHg; blood pressure recorded in her left arm was 90/64 mmHg. Chest auscultation revealed diminished breath sounds bilaterally with rales and rhonchi. A chest radiograph demonstrated bilateral infiltrates and a moderate right sided pleural effusion. T wave inversions from V4 to V6 were seen in the electrocardiogram. She had a mildly elevated leukocytosis (12.09 k/ μ L), a normal hemoglobin of 12.5 g/dl, and a platelet count of 232 k/ μ L. Comprehensive metabolic

profile was within normal limits. Significant laboratory values included an elevated B-type natriuretic peptide of 34,258 pg/mL and troponin of 0.13 ng/mL.

Transthoracic echocardiogram (TTE) showed severe left ventricular systolic dysfunction with ejection fraction of less than 20%, severe hypokinesia of the left ventricle, moderate aortic regurgitation, and mild mitral regurgitation. Left heart catheterization and angiogram showed a 4.4 cm aneurysm of the ascending aorta and irregular mural thickening of the aortic wall suggestive of a vasculitic process (Figures 1–3). She also had total occlusion of the left subclavian artery, the right renal artery, and the celiac artery and thrombosis of the superior mesenteric artery (Figure 1). Additional laboratory tests included an elevated high sensitivity CRP (24.4 mg/dl) with slightly elevated ESR (23 mm/hr) and negative autoimmune markers. Based on the significant blood pressure differences between the arms (>10mm Hg), the



Figure 2. Left heart catheterization showing the aneurysm of the ascending aorta.

elevated inflammatory markers, and arteriographic narrowing and occlusion of multiple branches of aorta, a diagnosis of Takayasu's arteritis was made. She was promptly started on high dose corticosteroids with prednisone at 60 mg/day. She improved

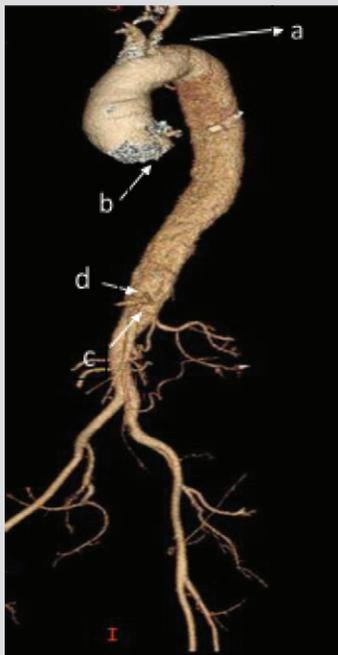


Figure 1. Angiographic study showing total occlusion of the left subclavian artery (a), an ascending aorta aneurysm (b), and occlusion of the right renal artery (c) and celiac axis (d).



Figure 3. Angiographic study showing the irregular wall thickening of the aorta.

clinically in response to the corticosteroids and later follow up at 2 months showed more improvement in her symptoms and in her ejection fraction (now 40%).

DISCUSSION

The diagnosis of Takayasu's arteritis (TA) is often delayed or even missed because this disease has a non-specific clinical presentation and no specific laboratory tests. A detailed and thorough physical examination is often very helpful in this diagnosis as absent or diminished pulses and BP discrepancy are diagnostic markers of the disease.² This case highlights the advanced age and rare presentation of the disease as acute congestive heart failure in a Caucasian woman. The disease is usually seen in the second and third decade of life.³ In a clinical study of 107 patients, the majority of patients (77%) presented between 11 to 30 years of age.⁴ The most common symptoms in this disease in two cohorts of TA patients were diminished or absent pulses leading to limb claudication and BP discrepancies, which was seen in 84%–88% of cases.^{5,6} Some patients with TA develop congestive heart failure, the main causes being increased afterload due to renovascular hypertension and aortic regurgitation. Myocardial ischemia induced by myocarditis, accelerated atherosclerosis, or severe pulmonary hypertension can also contribute.⁷ The elevated troponin in this patient may be due to an associated myocarditis.

Aortic regurgitation and heart failure have been reported in about 25% of patients.^{8,9} Aortic regurgitation associated with ascending aorta aneurysm is relatively rare in TA.^{10,11} Aneurysms are more commonly described in Asian populations than in North American populations.¹² Takayasu's arteritis, if left untreated, can have substantial morbidity with life threatening vascular complications. Imaging techniques, such as computerized tomography, magnetic resonance angiography, fludeoxyglucose positron emission tomography-computerized tomography, and contrast-enhanced ultrasonography, can help with this diagnosis.¹³ Vascular interventions with immunosuppressive therapy with corticosteroids and biological agents, especially in refractory cases, are the therapeutic mainstays of this disease.^{13,14} Surgical

indications include claudication of extremities affecting daily activities, hypertension with critical renal artery stenosis, cerebrovascular ischemia or critical stenoses of three or more cerebral vessels, cardiac ischemia due to coronary artery involvement, and moderate aortic regurgitation.¹⁵ Review of literature has shown that fewer than 20% of the patients undergo surgery at some point of their disease course.^{16,17} Surgical outcomes are better when done during periods of inactivity. Procedures during acute inflammatory stages have a seven times greater chance of failure.¹⁸ The mean survival rate in a longitudinal study of 106 patients for 20 years was 73.5%, the main cause of death being congestive heart failure.¹⁹ Takayasu's arteritis is a progressive disease of systemic vasculopathy and if untreated is potentially fatal. It requires an interdisciplinary approach to management and long term follow up for satisfactory patient outcomes.

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