

A Rare Case of Non-Ischemic Dilated Cardiomyopathy Revealing a Takayasu Arteritis

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Abstract

Background: Takayasu arteritis is a rare, systemic, inflammatory large vessel arteritis of unknown aetiology. It predominantly involves the aorta and its major branch arteries. It commonly occurs in women younger than 50 years of age. It can affect the cardiovascular system and renovascular hypertension and aortic regurgitation are the most prevalent manifestations. Dilated cardiomyopathy is rare and commonly due to renal artery stenosis or coronary artery involvement, otherwise, it is extremely rare.

Case Presentation: We are reporting a case of a 37-year-old woman presenting to the Emergency Department with acute stage IV NYHA dyspnea and orthopnea. Symptoms and signs were suggestive of acute decompensated heart failure which was managed medically using IV furosemide and nitrates. Transthoracic echocardiogram revealed a dilated cardiomyopathy, LVEF at 28%. Coronary artery angiography showed a distal chronic complete obstruction of left anterior descending artery with a subocclusion of the origin of the first diagonal artery, lesions which are not explaining the cardiomyopathy. Duplex ultrasound did not find any renal artery stenosis but a severe left post-vertebral subclavian artery stenosis. Laboratory tests are showing increased inflammation, no arguments for connective tissue disease. Takayasu arteritis was confirmed using the American College of Rheumatology criteria and patient was prescribed corticosteroids and cyclophosphamide along with heart failure therapy. This attitude improved her symptoms and quality of life as well as survival.

Conclusion: It is important for cardiologists to think of Takayasu arteritis as a potential cause of non-ischemic dilated cardiomyopathy, especially in a young woman, as immunosuppressive therapy can improve symptoms and LV remodeling, prevent LV systolic function deterioration and reduce cardiovascular events.

Keywords: *Takayasu Arteritis; Young Woman; Dilated Cardiomyopathy; Acute Decompensated Heart Failure; Inflammation*

Abbreviations

NYHA: New York Heart Association; LV: Left Ventricle; EDD: End-Diastolic Diameter; LVEF: Left Ventricle Ejection Fraction; RV: Right Ventricle; ADHF: Acute Decompensated Heart Failure; WBC: White Blood Cells

Introduction

Takayasu arteritis is a rare, systemic, inflammatory large vessel arteritis of unknown aetiology. It is defined as “granulomatous inflammation of the aorta and its major branches” by the Chapel Hill consensus conference on the nomenclature of systemic arteritis [1]. It predominantly involves the aorta and its major branch arteries, and (less frequently) the pulmonary arteries [2]. Because of its predilection for the brachiocephalic vessels, this arteritis has been labeled pulseless disease and aortic arch syndrome. Takayasu arteritis commonly occurs in women younger than 50 years of age [3]. The disease has been reported in all parts of the world, although it appears to be more prevalent in Asians [6]. The diagnosis of TA is often delayed because many patients manifest non-specific symptoms.

The disease can affect the cardiovascular system and manifests as valvular disease (aortic regurgitation), aortic root disease, coronary artery disease, or constrictive pericarditis [7]. Renovascular hypertension is reported in 28 - 75% and aortic regurgitation in 20 - 24% [6]. Dilated cardiomyopathy is less often seen and is either due to secondary hypertension from renal artery stenosis, coronary artery involvement, or due to an extremely rare primary myocarditis [8]. Its prevalence is 5% of all Takayasu arteritis patients [13] and it is worsening the prognosis. However, non-ischemic dilated cardiomyopathy being the presenting manifestation of the disease is extremely rare, and described in only one case report [4], at least to our knowledge.

We are reporting a very rare case of non-ischemic dilated cardiomyopathy revealing a Takayasu arteritis in a young woman.

Case Presentation

We are reporting a case of a 37-year-old woman, with a history of dyspnea on moderate exertion of 6 months, presenting to the Emergency Department with acute stage IV NYHA dyspnea and orthopnea. Physical examination finds a regular rhythm, HR at 105 bpm, absent radial pulse in the left arm, BP in right upper limb at 130/80 mmHg and unmeasurable in left upper limb, SpO₂ in room air was 89%, crackles heard in both pulmonary bases, bipedal oedema. Symptoms and signs were compatible with an acute decompensated heart failure (ADHF). Electrocardiogram showed sinus rhythm, inverted T waves in left precordial leads. Transthoracic echocardiogram showed a dilated LV (index EDD of 34 ml/m²), severely impaired LVEF at 28%, Global longitudinal Strain (GLS) at 6.1%, mitral inflow pattern was restrictive (E/A = 3.2), mild secondary mitral regurgitation, enlarged left atrium with an area of 25 cm², RV function was normal with a mild secondary tricuspid regurgitation with a maximum gradient of 47 mmHg, inferior vena cava was dilated at 23 mm with decreased inspiratory collapse, systolic pulmonary artery pressure was estimated at 62 mmHg (Figure 1). Laboratory tests showed WBC count at 15000/mm³, plasma fibrinogen level at 6.5 g/l, C-reactive protein at 90 mg/L, normal procalcitonin level, high first hour sedimentation rate at 74mm, high sensitivity cardiac Troponin I level was at 5672 pg/ml, calcemia and TSH levels were normal. Testings for hepatitis virus B and C and HIV were negative. Plasma antinucleus antibodies, rheumatoid factor, antineutrophil cytoplasmic antibodies levels were within normal range. ADHF was managed using IV furosemide and nitrates. Coronary angiography with normal right radial access was performed: distal chronic complete obstruction of left anterior descending artery with a subocclusion of the origin of the first diagonal artery, not explaining the LV dilation and dysfunction. An upper limb Duplex ultrasound was performed showing a severe stenosis of left distal subclavian artery (Figure 2). Extracranial cerebral arteries Duplex revealed an increased intima-media thickness in right internal carotid artery, no stenosis nor aneurysms were shown. Duplex ultrasound of renal arteries has not shown a renal artery stenosis. All arguments were compatible with type III Takayasu arteritis according to American College of Rheumatology criteria [10] with subclavian, carotid and coronary artery involvement associated with a myocarditis. The patient was prescribed heart failure medications (ACEI, beta-blocker, furosemide) and immunosuppressive therapy (cyclophosphamide, corticosteroids). At 6 months follow-up, symptoms improved and the patient recovered a good quality of life, inflammation regressed, high sensitivity cardiac troponin I level was normal. Echocardiography showed normal left ventricle filling pressures, index EDD of 32 mm/m², LVEF and GLS levels improved with values respectively of 40% and -12%. We attribute the clinical response to immunosuppression along with decongestive therapy as inflammatory response regressed, left ventricle systolic and diastolic function improved.

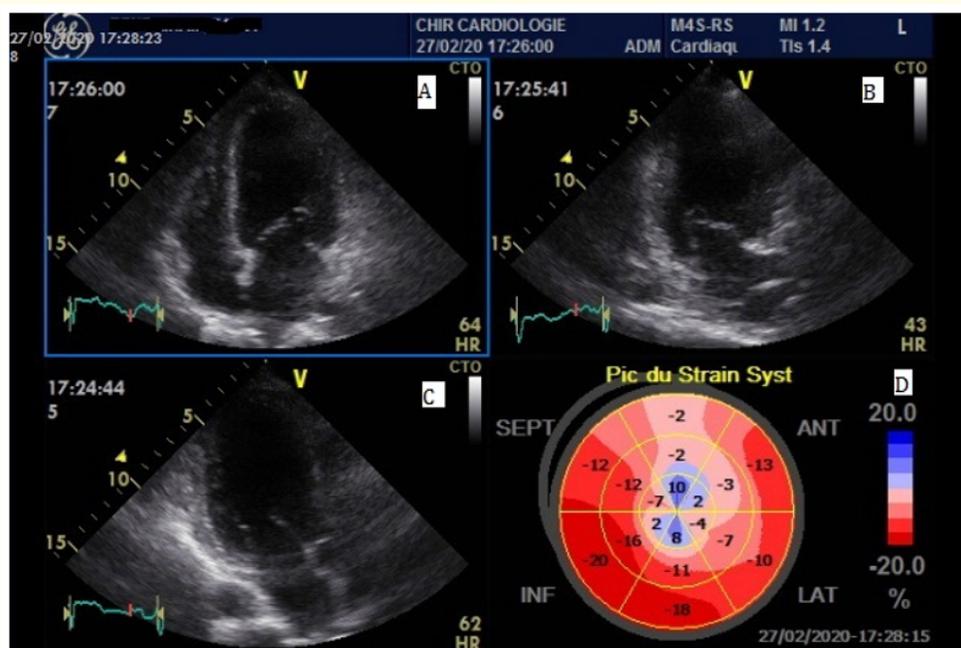


Figure 1: A, B, C: 2D transthoracic echocardiogram showing a dilated left ventricle. D: 2D Speckle Tracking Imaging showing a depressed longitudinal systolic function (GLS = -7.5%).

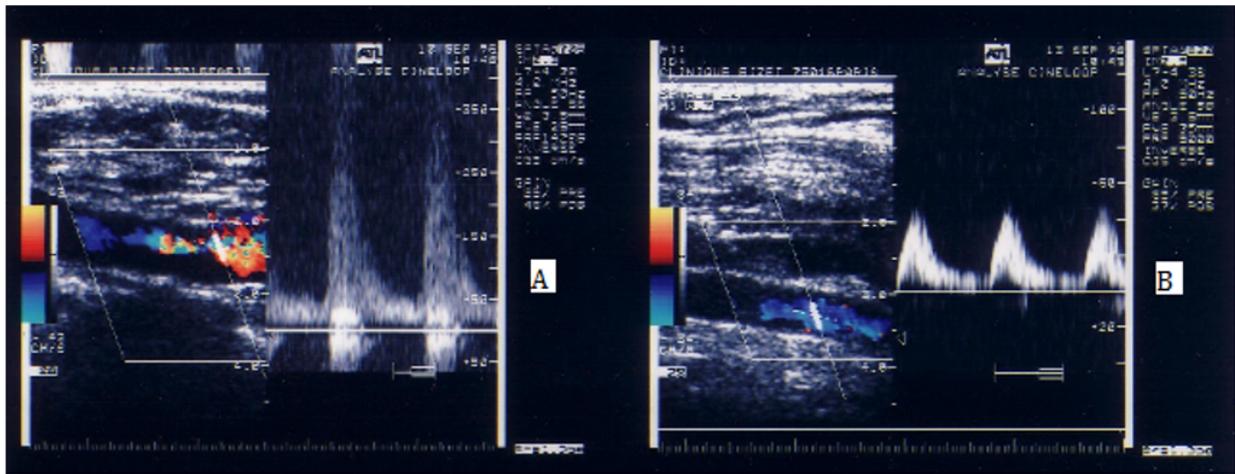


Figure 2: A: A long non-atherosclerotic severe stenosis of the post-vertebral left subclavian artery.
 B: A demodulated flow in the left axillary artery confirming the severity of the stenosis of the subclavian artery.

Discussion

Takayasu arteritis is an inflammatory disease of large and medium-sized arteries, with a predilection for the aorta and its branches. It has an incidence rate of 2.6 cases per million individuals per year in the United States. The *HLA-B5* genetic locus is linked with susceptibility [11]. It is predominantly a disease of young females in their second and third decades. Although this disorder is more frequent in Asians, it occurs worldwide and no race seems to be immune [8]. It is characterized by inflammation of the arterial wall. Though the exact pathogenesis of the arteritis is still unknown, tuberculosis, streptococcal infections, rheumatoid arthritis, and other collagen vascular diseases have been debated as its etiology in the past. Recently, more emphasis has been given on an immunopathological cause. Diagnosis can be challenging, because the non-specific signs and symptoms can mimic infection, malignancy, thrombotic disorders, and connective tissue disease. Clinical symptoms are associated with the affected vessel. Limb claudication, absent pulses, and unequal blood pressures are typical symptoms of large-vessel vasculitis.

Dilated cardiomyopathy (DCM) is one of the cardiomyopathies, a group of diseases that primarily affect the myocardium. It has different causes and is classically classified into ischemic and non-ischemic dilated cardiomyopathy. Non-ischemic dilated cardiomyopathy has several and varied etiologies (infectious, metabolic, genetic disorders...). Dilated cardiomyopathy is seen in 5% of cases in Takayasu arteritis and it is more frequently associated with renovascular hypertension or coronary artery involvement, rarely due to primary myocarditis [8]. In our patient, there wasn't any renal artery stenosis and distal coronary artery involvement couldn't explain the dilated cardiomyopathy. Then, the dilated cardiomyopathy is thought to be due to primary myocarditis as there was an important biological inflammatory syndrome, an important elevation of cardiac troponin I and no significant coronary artery disease. Histopathological study (on 3 autopsy cases) showed non-specific inflammation of myocardium with lymphocyte/mononuclear cell infiltration and normal coronary vessels [8]. Histopathological study was not performed in our case.

Coronary artery stenosis has been reported in Takayasu arteritis. It is due to the extension of the inflammatory process and intimal proliferation in the ascending aorta. Seventy-three percent of occlusive coronary artery diseases are localized around the coronary ostium, followed by non-ostial proximal lesions (18.5%) [12,13]. Distal coronary artery involvement has been found in our patient but never has been reported, at least to our knowledge. However, it does not explain the dilated cardiomyopathy.

Cardiac involvement carries a worse prognosis in Takayasu arteritis patients and treatment with immunosuppressive agents, corticosteroids and heart failure medications allows to improve cardiac remodeling, prevent LV systolic function deterioration, reduce future major cardiovascular events and improve quality of life and symptoms. In some cases, as we reported, immunosuppressive therapy allows to improve systolic and diastolic function along with heart failure therapy.

Conclusion

This case presentation shows the importance to think of Takayasu arteritis as a cause of non-ischemic dilated cardiomyopathy, especially in young woman. It can be explained either by secondary hypertension due to renal artery stenosis or by primary myocarditis. Such a diagnosis is associated with a bad prognosis with a risk of cardiovascular death and hospitalization for heart failure. Medical therapy using immunosuppressive agents along with immunosuppressive drugs allows to improve systolic and diastolic left ventricle function, prognosis, symptoms and quality of life.

Conflict of Interest

None to declare.

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