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Case Report

Takayasu's Arteritis Presenting as Dilated Cardiomyopathy: A Rare Case Report

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Abstract

Takayasu's arteritis is a rare, idiopathic large vessel vasculitis that affects large arteries, mainly the aorta and its branches. The coronary, pulmonary and renal arteries are also affected in the progression of the disease. We report a rare case of dilated cardiomyopathy in a 42 years old woman who was later diagnosed to have Takayasu's arteritis using magnetic resonance angiography (MRA). She was subseaquently treated by immunosuppressant therapy with prednisolone and methotrexate along with symptomatic treatment with gradual improvement of ventricular systolic dysfunction. Dilated cardiomyopathy (DCM) is reported to be seen in only 5-6% of cases of Takayasu's arteritis. In a case of dilated cardiomyopathy screening for systemic vasculitis should be done if evidence of peripheral vascular disease is present, as timely initiation of immunosuppressant therapy can help in symptom-free survival.

Keywords: *Takayasu's Arteritis, Cardiomyopathy*

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Introduction

Takayasu's arteritis (TA) is a chronic granulomatous inflammatory disease of large vessels which affects mainly the aorta and its major branches along with the coronary, pulmonary, and renal arteries.[1] It is common in Asia and usually affects adolescents and young woman. However, recently, TA has been shown to affect both the sexes, any age, and all ethnic groups[2]. Diminished or absent pulses of upper extremities are found in most of the patients, hence, it is also called pulse less disease[5]. There is a vessel inflammation leading to wall thickening, fibrosis, stenosis, thrombosis, end organ ischemia & aneurysm formation. It has variable and nonspecific clinical presentation thus leading to delayed diagnosis and treatment. It is a cause of Reno vascular hypertension[6]. Ultrasonography, computed tomography and MRA are the main imaging modalities which are used in the diagnosis of Takayasu's arteritis. C reactive protein & elevated ESR are important marker for disease activity, which are used for patient follow up in TA[7]. It may cause dilated cardiomyopathy (DCM) in small proportion (5%) of affected patients resulting in congestive cardiac failure[8]. However, congestive cardiac failure in TA is mainly due to hypertension and aortic regurgitation. We had reported a rare case of Takayasu's arteritis presenting as dilated cardiomyopathy and reno vascular hypertension.

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Case Report

A 42 year old female presented in 2013 with the complaints of several episodes of exertional respiratory distress and palpitation with whole body swelling and orthopnea for last 4 years. For this she took repeated admission in different hospitals and diagnosed as a case of congestive cardiac failure (NYHA class- IV) with dilated cardiomyopathy (DCM). She was treated accordingly but still she had recurrent symptoms. The medical history was positive for generalized fatigue, low grade fever and occasional multiple joint pain. There was also history of pain in both upper limbs that worsened on exertion resembling ischemic pain which

was gradually progressive and associated with coldness and weakness as well as occasional neck pain, headache and dizziness.

On general examination, she was dyspneic. Her pulse was absent in both upper limbs but all other peripheral pulses were present and feeble. Pulse rate was 100/min, regular. Blood pressure was not recordable in both upper limbs. There was a bruit over the left subclavian artery. JVP was raised and ankle edema was present bilaterally. On cardiovascular system examination, apex beat was shifted. Palpable P_2, parasternal heave and epigastric pulsation were present. First heart sound was soft in apical area and pulmonary component of the second heart sound was loud. There was pansystolic murmur in apical area and also a systolic murmur in left parasternal area. Bilateral basal crepitation was present. On GIT system examination there was tender hepatomegaly and ascites. Other systemic examination including fundoscopy was normal.

On investigation, ESR (113 mm in 1st hr) and CRP (24 mg/L) were elevated. Liver function and renal function test were normal. Serum ANA was negative. USG of W/A revealed congestive hepatomegaly and ascites. CXR P/A view showed gross cardiomegaly (Fig. 1A). ECG showed sinus tachycardia with LVH with strain pattern. Doppler echocardiography showed severe LV systolic dysfunction (EF-20%) with dilated cardiomyopathy with pulmonary hypertension. MRA of great arteries revealed left subclavian artery and left vertebral artery occlusion. On the basis of clinical manifestations and angiographic findings, the diagnosis of congestive cardiac failure (NYHA class- IV) with DCM with severe LV systolic dysfunction with pulmonary hypertension with Takayasu's arteritis was made. The patient was given oral steroids on tapered doses and started on methotrexate, digoxin, spironolactone and frusemide. After starting medication, patients' condition gradually improved. Since then (from 2013), in each year she had regular thorough follow-up as an in-house patient in cardiology department of Dhaka Medical College Hospital with continuation of methotrexate weekly and two courses of steroid at a tapered dose. During this follow-up period her liver function and renal function were always within normal limit. She was found hypertensive on 2014 by recording BP on lower limb and treated with antihypertensives. Her ventricular systolic dysfunction gradually improved on serial echocardiogram from EF-20% (on 2013) to EF-45% (till now). Her ESR gradually decreased from 113 mm in 1st hr in 2013 to 28mm in 1st hr in 2022 with improvement of her symptoms and signs.

On her last follow-up in 2022, we did coronary and peripheral angiogram. Coronary angiogram revealed noncritical triple vessel coronary artery disease. Peripheral angiogram revealed right subclavian artery is 100% occluded after the origin of the right vertebral artery. Left subclavian artery is 100% occluded at the origin (Fig-2). Left vertebral artery is 100% occluded with normal right vertebral artery (Fig-3). Left common iliac artery had 40-50% stenosis at the origin (Fig-4). Left renal vessel is 50-60% stenosed at the origin (Fig-5). Right renal vessel is hypoplastic with rudimentary right Kidney. Patient was also diagnosed having diabetes on this occasion. So, we diagnosed the case as a Takayasu's arteritis (type V) with DCM with renal artery stenosis with noncritical triple vessel coronary artery disease with HTN with DM. On discharge, patient was given methotrexate weekly with folic acid, vasodilating â-blocker, spironolactone, lipid lowering agent and antidiabetic drugs with insulin.

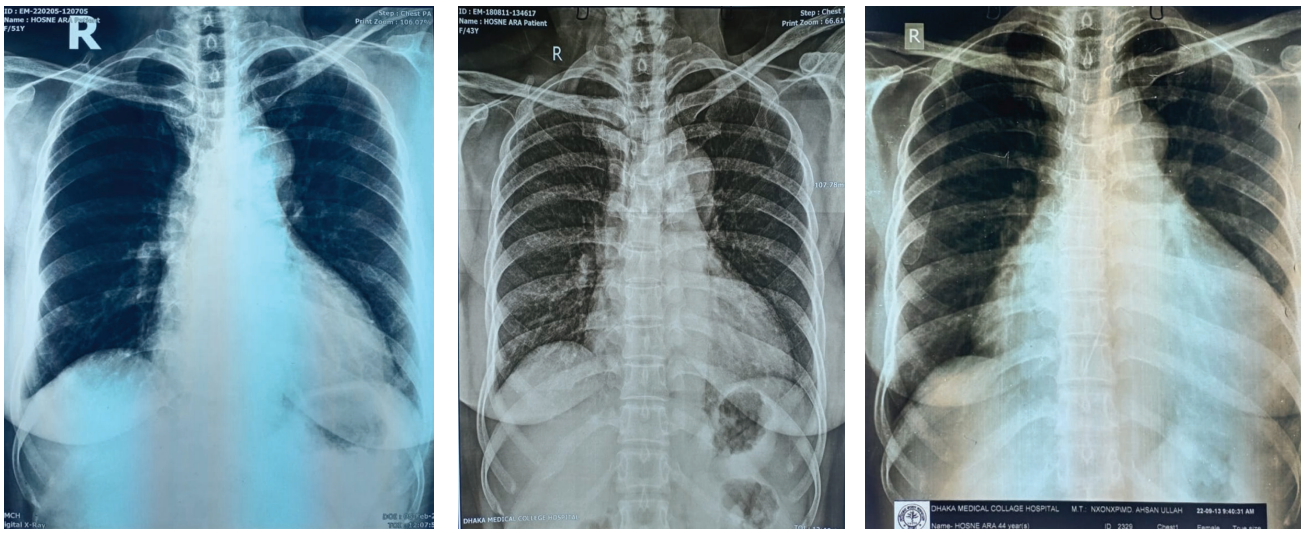

Fig-1: *Gradual decreasing of cardiomegaly on serial CXR-P/A view from 2013 (A) to 2022 (C)*

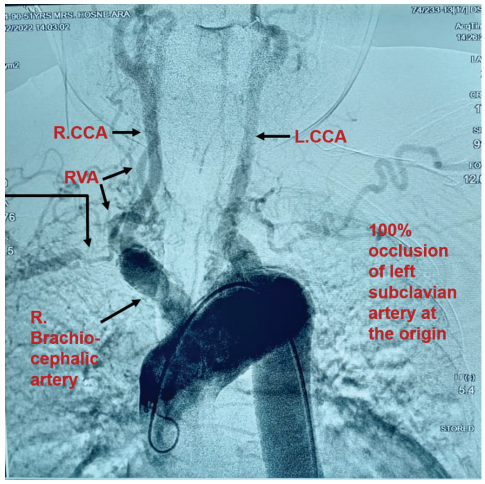

Fig-2: Angiographic view of aortic arch vessels

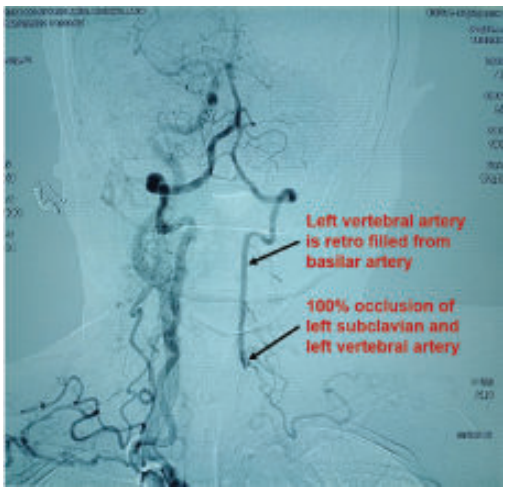

Fig-3: Digital subtraction angiography (DSA) of vertebral artery

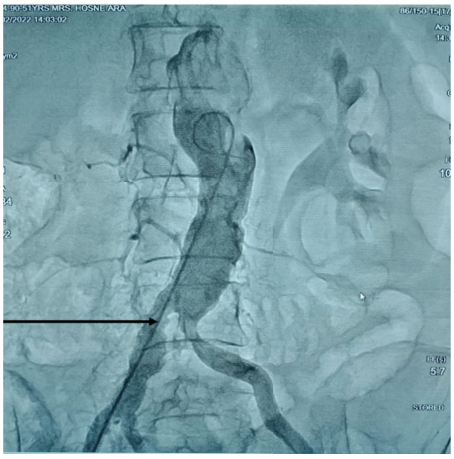

Fig-4: Angiographic view of lower limb vessels

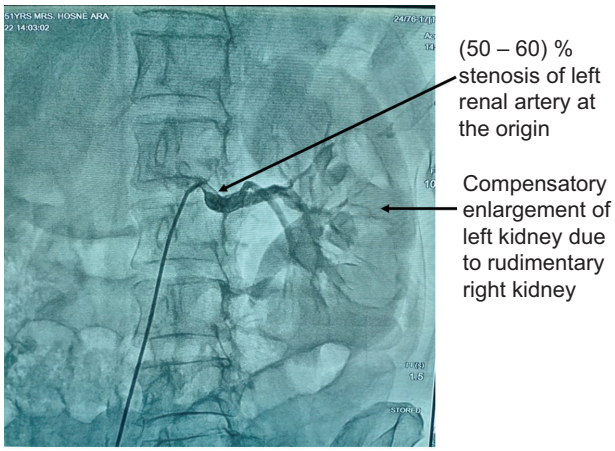

Fig-5: Angiographic view of left renal vessels

It was angiographically confirmed as type V Takayasu's arteritis. The incidence of type V TA as reported by Panja et al in largest series is only 10%.[1] The American Rheumatological Society considers three of the following six criteria necessary for a definite diagnosis of Takayasu's disease:[13]

1. Onset before 40 years
2. Claudication of the extremities
3. Decrease in the brachial pulse in one or both arms
4. Difference of 10 mm Hg or more in blood pressure measured in both arms
5. Audible bruit on auscultation of the aorta or subclavian Artery
6. Narrowing at the aorta or its primary branches on arteriogram

In case of this patient, five out of six criteria were met.

The disease is classified based on the site of involvement according to New angiographic classification of Takayasu arteritis, Takayasu conference 1994:[1]

- **Type I:** Branches from the aortic arch
- **Type IIa:** Ascending aorta, aortic arch and its branches
- **Type IIb:** Ascending aorta, aortic arch and its branches, thoracic descending aorta
- **Type III:** Thoracic descending aorta, abdominal aorta, and/or renal arteries
- **Type IV:** Abdominal aorta and/or renal arteries
- **Type V:** Combined features of types IIb and IV

Our patient was diagnosed as TA type V, as there was involvement of bilateral subclavian artery (Type IIb) and left common iliac and Renal artery (Type IV).

The presentation as DCM is rarely reported in 5-6% of cases and is due to involvement of coronary artery & severe

hypertension.[14] The progression of heart failure is associated with LV remodeling, which manifests as gradual increase in LV end-diastolic and end-systolic volumes, wall thinning, and a change in chamber geometry to a more spherical, less elongated shape. This process is usually associated with a continuous decline in ejection fraction.[15] Our patient presented with dilated cardiomyopathy with severe LV dysfunction.

Therapeutic modalities include steroids, immunosuppressive agents, and antihypertensive drug therapy. Different studies have reported 20-100% success rate of steroids. Cyclophosphamide and methotrexate are often needed to control intense inflammatory response. Drug therapy can slow down progression of cardiomyopathy and in some cases even improve the heart condition. In our patient, there was gradual improvement of ventricular systolic dysfunction after adding steroid in tapered dose and weekly methotrexate along with heart failure treatment.

Conclusion

With this case report we want to emphasize on taking detailed history and doing meticulous clinical examination for appropriate and early diagnosis of systemic vasculitis. We should also screen for systemic vasculitis in the case of dilated cardiomyopathy in young female, as timely initiation of immunosuppression therapy can improve the cardiac condition and prolong the survival.

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