TAKAYASUS ARTERITIS PRESENTING WITH DILATED CARDIOMYOPATHY – A RARE CASE REPORT

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ABSTRACT

Takayasu’s arteritis is a large vessel vasculitis in which the inflammatory process involves aorta and major branches. The cause is largely unknown. Dilated cardiomyopathy (DCM) is, however, reported to be seen in only 5-6% of cases of Takayasu arteritis. We report a rare case of takayasus arteritis presenting with dilated cardiomyopathy.

Keywords: Dilated cardiomyopathy,Takayasu’s arteritis

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Case Report

Introduction

Dilated cardiomyopathy (DCM) is one of the cardiomyopathies, a group of diseases that primarily affect the myocardium (the muscle of the heart). Different cardiomyopathies have different causes and affect the heart in different ways. In DCM, the heart becomes Weakened and enlarged and cannot pump blood efficiently. The decreased heart function can affect the lungs, liver, and other body systems.
In DCM, a portion of the myocardium is dilated, often without any obvious cause. Left ventricular (LV) or right ventricular systolic pump function of the heart is impaired, leading to progressive cardiac enlargement and hypertrophy, a process called remodeling.\[^1\] About one in three cases of congestive heart failure (CHF) is due to DCM.\[^1\] About 25-35% of patients have familial forms of the disease,\[^1\] with most mutations affecting genes encoding cytoskeletal proteins,\[^2\] while some affect other proteins involved in contraction.\[^2\]

Takayasu’s arteritis (also known as “aortic arch syndrome,” “nonspecific aortoarteritis,” and the “pulseless disease”) is a form of large vessel granulomatous vasculitis\[^1\] which causes massive intimal fibrosis and narrowing of arteries, affecting often young or middle-aged women of Asian descent. It mainly affects the aorta and its branches, as well as the pulmonary arteries. Females are 8-9 times more affected than males.\[^1,3\] Patients often notice the disease symptoms between 15 and 30 years of age. Takayasu’s arteritis is similar to other forms of vasculitis, including giant cell arteritis.\[^3,4\] Due to obstruction of the main branches of the aorta, including the left common carotid artery, the brachiocephalic artery, and the left subclavian artery, Takayasu’s arteritis can present as pulseless upper extremities (arms, hands, and wrists with weak or absent pulses on the physical examination), commonly referred to as the “pulseless disease” and renal arteries are involved. In India, Takayasu arteritis as cause of renovascular hypertension is reported in 28-75%, CHF in 76%, aortic regurgitation in 20-24\(^\%\)^\[^5\] and DCM is, however, reported to be seen in only 5-6\(^\%\) of cases of Takayasu arteritis.\[^4\] The presentation as DCM is rarely reported and is due to involvement of coronary artery, severe hypertension, and cardiac failure are bad prognostic factors.\[^1,2\]

**Case Report**
A 18-year-old female presented with complaints of breathlessness on exertion since 11 months and aggravated since 5 days prior to admission, sudden in onset, New York Heart Association classification Grade 3, aggravated on lying down and relieved at rest.

On general physical examination, pulse rate was 110 beats/ min of right radial artery, regular in rate, rhythm. Left upper limb arterial pulsations were absent. There was no radiofemoral delay. Blood pressure was 100/60 mmHg of right arm in the supine position. Nodular lesions presented on the nose, biopsy was suggestive of trichoepithiloma. Jugular venous pressure was raised.

On examination, cardiovascular system revealed apex beat was shifted downward and outward. Heart sounds were normal, tachycardia was present. On respiratory system examination, bilateral basal crepitations were present. Per-abdomen examination suggestive of hepatomegaly, liver span-15 cm. On central nervous system, examination patient was conscious and oriented. B/L sensory neural deafness was present lt>rt

**Investigations**
Hemogram revealed hemoglobin of 12.1 g%, total leucocyte count was 8900 cells/cumm, platelet count was 4.14 lakhs/ cumm. Erythrocyte sedimentation rate was 51 mm at the end of 1st h. C-reactive protein was positive. Renal function tests were normal. Blood urea was 31 mg%, and serum creatinine was 0.9 mg%. Serum electrolytes were normal. Serum sodium was 136 mmol/L, serum potassium was 5.1 mmol/L, Anti-nuclear antibodies were negative, and anti-ds-DNA was negative.

Electrocardiography showed sinus tachycardia with LV hypertrophy chest X-ray was suggestive of cardiomegaly Twodimensional echo revealed DCM, global LV hypokinesia, severely depressed LV systolic function LV-ejection fraction-30\(^\%\) . Color arterial Doppler of right left extremity showed monophasic flow noted in infraclavicular subclavian artery with narrowing of left subclavian artery. Monophasic flow noted distally in axillary, brachial, radial, and ulnar artery. The peak flow velocities are reduced. Circumferential
wall thickening of left subclavian, axillary and b/l common carotid arties  Findings are suggestive of large vessel vasculitis

**Figure 1:** Electrocardiogram of the patient showing left ventricular hypertrophy

![Electrocardiogram](image1)

**Figure 2.** chest x ray showing dilated cardiomyopathy

![Chest X-ray](image2)
Figure 3: Two-dimensional echocardiography of patient showing dilated cardiomyopathy

Computed tomography-angiography of aorta Showed

1. Irregular circumferential wall thickening involving the ascending arch and the ascending aorta causing significant luminal narrowing distal part of abdominal aorta luminal diameter -4.2mm
2. Circumferential wall thickening involving the left subclavian artery from its origin with significant narrowing distal to the origin of vertebral artery.
3. Irregular circumferential thickening of wall involving right brachicephalic, right subclavian vein common carotid up to the level of carotid bulb
4. Bilateral vertebral arteries are normal
5. Cardiomegaly of left ventricular type
6. Features are suggestive of large vessel vasculitis.
Patient has been started on oral prednisolone and methotrexate apart from heart failure therapy patient showed improved symptomatically with wait gain and decreased heart failure symptoms.

**Discussion**

Takayasu arteritis is the most common cause of renovascular hypertension in India. Indian origin aortoarteritis is a chronic granulomatous, necrotizing vasculitis, predominantly affecting the aorta with its branches.\(^6\)

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\(^6,7\)

The disease is classified based on the site of involvement\(^8\)

- Type I: Aortic arch involvement
- Type II: Thoracoabdominal involvement
- Type III: Diffuse involvement
- Type IV: Pulmonary involvement
- Type V: Aneurysmal type.

The site of arterial disease determines its clinical presentation.

In our patient, there is involvement of descending thoracic aorta, right subclavian artery Type III-Takayasu’s arteritis.

The presentation as DCM is rarely reported and is due to involvement of coronary artery, severe hypertension, and cardiac failure are bad prognostic factors\(^9,10\). The progression of heart failure is associated with LV remodeling, which manifests as gradual increases in LV end-diastolic and endsystolic 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.\(^11\) Death is due to either CHF or ventricular tachy-or bradyarrhythmias.
Therapeutic modalities include steroids, immunosuppressive agents, and antihypertensive drug therapy. 20-100% success rate of steroids have been reported in different studies.\textsuperscript{12} Cyclophosphamide and methotrexate are often needed to control intense inflammatory response. In addition, balloon dilatation or stenting is often necessary.\textsuperscript{13} Drug therapy can slow down progression of cardiomyopathy and in some cases even improve the heart condition. Standard therapy may include salt-restricted diet, diuretics, and digitalis.\textsuperscript{11}

**Conclusion**

With this case report, we want to emphasize that young female patient with hypertension, heart failure, and/or renal failure should be screened for systemic vasculitis, since only an anti-inflammatory approach can avoid or at least reduce other complication of vasculitis.

**References**