Unusual cause of dilated cardiomyopathy in an adolescent girl

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Dilated cardiomyopathy (DCM) is one of the cardiomyopathies where left ventricular (LV) or right ventricular systolic pump function of the heart is impaired with progressive cardiac enlargement and hypertrophy.1 About one-third of cases of congestive heart failure (CHF) is due to DCM.1

Takayasu's arteritis (also known as ‘aortic arch syndrome’ and the ‘pulseless disease’) is a form of granulomatous vasculitis involving mainly the aorta and its branches,1 which causes extensive intimal fibrosis and narrowing of arteries. Females are 8–9 times more affected than males.1,2 Due to obstruction of the main branches of the aorta, Takayasu's arteritis can present as pulseless upper extremities. Renal arteries are also involved.

In India, Takayasu's arteritis as cause of renovascular hypertension is reported in 28–75% cases.2 DCM per se, however, is reported in only 5–6% of cases of Takayasu's arteritis.3

Case report

A 13-year-old female child presented with complaints of severe shortness of breath, weakness, pedal swelling and low-grade fever since last seven days. The complaints were of six months duration with a recent increase in severity of symptoms.

On examination she was thin built, febrile with temperature 100°F, dyspnoea grade 4 (New York Heart association grading), and pulse 110/min in left radial artery, regular in rate and rhythm, BP 110/64mmHg in left arm in supine position. Right upper limb pulsations were absent.

Investigations

Blood sugar fasting-80mg/dl, Creatinine-0.6mg/dl, Haemoglobin-10.5gm/dl, ESR-90mm first hour, TLC-7900, N55L34, Mantoux test was negative, ASO titre-123(<200), CK-MB-16, Total bili 1.7, albumin-3.3, SGPT-39, SGOT-47, CRP-16.6 (high), RA factor-5.4(<15).

ANA, anti-ds DNA, p ANCA and c ANCA were negative.

PA chest radiograph—enlarged cardiac shadow, with diffuse alveolar shadows consistent with pulmonary edema (Figure 1).
ECG showed sinus tachycardia with left ventricular hypertrophy, Left axis deviation, left atrial enlargement and prominent p-waves.

Ultrasonography whole abdomen—mild hepatomegaly with normal size kidneys, mild ascitis.

2D Echocardiography (M mode) showed dilated cardiography, with severe left ventricular systolic dysfunction, LVIDD -56, LVIDS-52, ejection fraction–16%, moderate mitral regurgitation and severe tricuspid regurgitation (Figure 2).

Color arterial Doppler of right upper extremity showed monophasic flow noted in subclavian artery with narrowing of right subclavian artery. Monophasic flow noted distally in axillary, brachial, radial and ulnar artery.

The peak flow velocities are reduced. Findings are suggestive of obstruction in the aortic arch region.

MDCT scan of Aortic arch and thoracic and abdominal aorta shows NO significant stenosis or dilatation seen in aortic arch or ascending thoracic aorta. Descending thoracic aorta and abdominal aorta shows long segmental areas of diffuse luminal narrowing and associated concentric wall thickening. Left subclavian artery and left axillary artery shows diffuse luminal narrowing with non-visualisation (cut off) of luminogram in distal two-thirds of right subclavian and right axillary artery.

Segmental concentric wall thickening with associated luminal narrowing seen in both renal arteries with mild post-stenotic dilatation further s/o aortoarteritis (Figure 3).

Incidentally, all four chambers were dilated, ie, DCMP with dilated MPA (main pulmonary artery) (Figure 4).
Figure 2: Echo Doppler study showing dilated all four chambers of heart with poor contractile function.
Figure 3: MDCT angio of thoracic and abdominal aorta shows diffuse luminal narrowing involving renal arteries.

Figure 4: MDCT scan showing dilatation of all four chambers (DCM) including main pulmonary artery.

Discussion

Exact pathogenesis of the Indian origin aortoarteritis is still unclear, tuberculosis, streptococcal infections, rheumatoid arthritis and other collagen vascular diseases have been attributed as its etiology in the past. Recently, more emphasis has been given on an immunopathological cause.3,4

The primary presentation of Takayasu’s arteritis as DCM with biventricular failure is rarely reported and may be due to inflammatory process directly involving the myocardium, coronary arteries or severe hypertension secondary to renovascular involvement.5,6

Therapeutic modalities include steroids and immunosuppressive agents. Cyclophosphamide and methotrexate are often needed to control intense inflammatory response. In addition, balloon dilatation or stenting is often necessary. Drug therapy can slow down progression of cardiomyopathy and in some cases even improve the heart condition.

Conclusion

With this case report, we want to emphasise that the young female patient with heart failure, dilated cardiomyopathy and/or hypertension, renal failure should be screened for systemic vasculitis, as it is potentially correctible with timely immunosuppressive treatment.
Competing interests: Nil.

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