



CASE STUDY

TAKAYASU ARTERITIS CONCEALED AS DILATED CARDIOMYOPATHY CASE REPORT WITH REVIEW OF LITERATURE

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ABSTRACT

Case report: We report a rare case of dilated cardiomyopathy in an 18-year-old girl who was later diagnosed to have Takayasu arteritis using doppler and computed tomography and subsequently treated by immunosuppressant therapy with prednisolone and methotrexate. **Discussion:** Takayasu 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 (Jameson *et al.*, 2012). Ultrasonography with doppler, computed tomography and Magnetic Resonance Imaging (MRI) can be used for the early diagnosis of the disease. **Conclusion:** In a case of dilated cardiomyopathy in young female screening for systemic vasculitis should be done as timely initiation of immunosuppressant therapy can reduce the development of other complications of Takayasu arteritis.

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INTRODUCTION

Takayasu arteritis is a form of large vessel granulomatous vasculitis (Ghosh *et al.*, 1999) affecting often young or middle-aged women of Asian descent. It mainly affects the aorta and its branches, as well as the pulmonary arteries. Dilated cardiomyopathy (DCM) is the most common type of cardiomyopathies in children (Ghosh *et al.*, 1999). 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 with left ventricle (LV) most commonly affected. In India, Takayasu arteritis as cause of DCM is, however, reported to be seen in only 5-6% of cases of Takayasu arteritis (Kerr *et al.*, 1994). Ultrasonography, computed tomography and MRI are the main imaging modalities which are to be used in the diagnosis of Takayasu arteritis.

Case report

An 18 year old young female presented with history of easy fatigability, breathlessness, weight loss, since two months associated with cough with whitish sputum for 3 days.

The patient was being treated as a case of idiopathic dilated cardiomyopathy for two months by digitalis and salt restricted diet. On clinical examination tachycardia, systolic murmurs with crepitations in the right basal zones of the lung were present. There were absent pulses in the left brachial, ulnar and radial arteries. Rest of the limbs showed normal pulses. ESR was 32mm/hr with positive C reactive protein. Two dimensional echocardiography revealed dilated cardiomyopathy with grade III diastolic dysfunction and ejection fraction of 30%. Subsequently the patient was referred to the Department of Radiology for doppler study of left upper limb. On doppler study there, long segment circumferential hypoechoic wall thickening involving the left subclavian (Fig. 1) and axillary artery causing 75% luminal narrowing with monophasic flow in the brachial, radial and ulnar arteries and reduced systolic velocities were noted. There was also associated circumferential hypoechoic wall thickening of the bilateral common carotid arteries (Fig 2) in their entire extent. However the bilateral external and internal carotid arteries showed normal wall thickness with normal flow pattern on doppler.

There was no evidence of calcification in the region of the wall thickening of the above mentioned arteries. Based on the above mentioned findings large vessel vasculitis was suspected and patient was advised for CT angiography.

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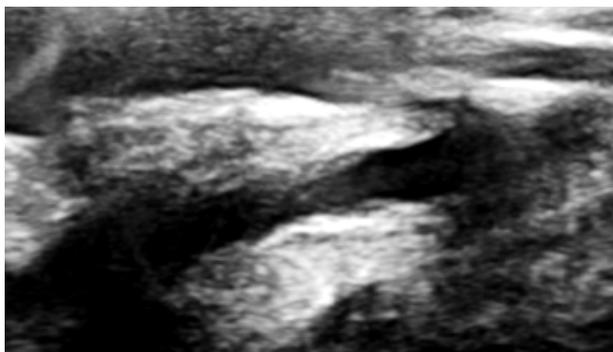


Fig. 1. Circumferential hypoechoic wall thickening involving the left subclavian (arrow)

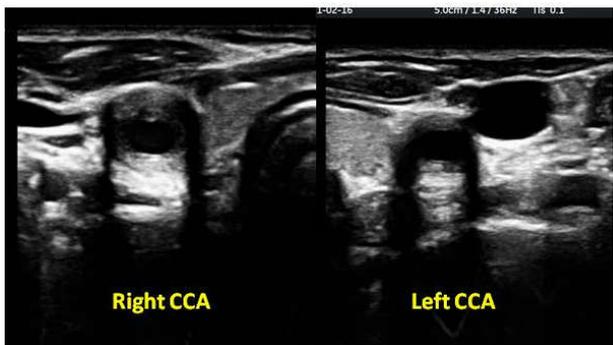


Fig. 2. Circumferential hypoechoic wall thickening of the bilateral common carotid arteries

On CT angiography there was circumferential wall thickening involving the ascending aorta (Fig 3a), aortic arch (fig 3b) and the descending thoracic (Fig 3c) & abdominal aorta (Fig 3d). There was also associated circumferential wall thickening involving the left subclavian artery from its origin with significant narrowing distal to the origin of the vertebral artery (Fig 4a,4b,4c).

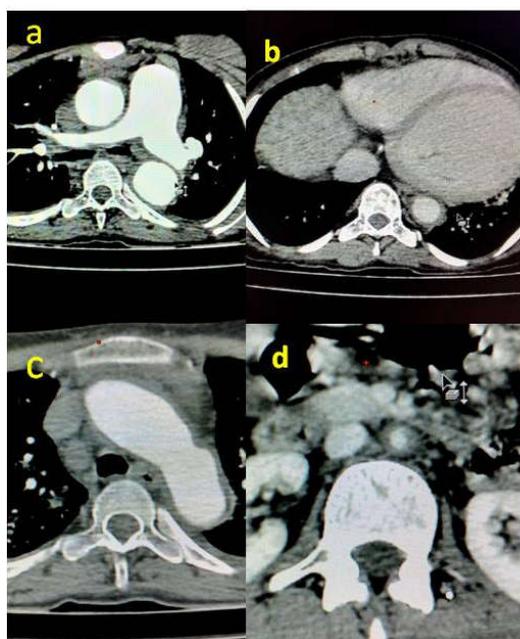


Fig. 3. CT angiography there was circumferential wall thickening involving the ascending (a), arch (b) and the descending thoracic (c) & abdominal aorta (d)

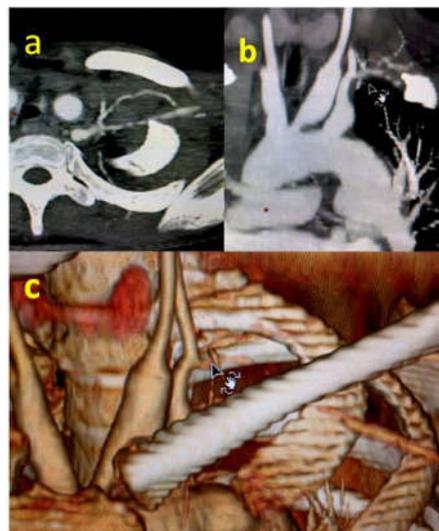


Fig. 4. Circumferential wall thickening involving the left subclavian artery in axial (a), coronal(b), 3D(c) from its origin with significant narrowing distal to the origin of the vertebral artery

Circumferential wall thickening without significant narrowing was noted involving the bilateral common carotid arteries up to the carotid bulb (Fig 5a, 5b), right brachiocephalic and right subclavian arteries. Bilateral brachial, radial and ulnar arteries were normal. There was cardiomegaly of left ventricular type (Fig 6). The entire pulmonary artery, coronary arteries and the bilateral renal arteries were normal. There was no evidence of aneurysm dilatation of the involved vessels.

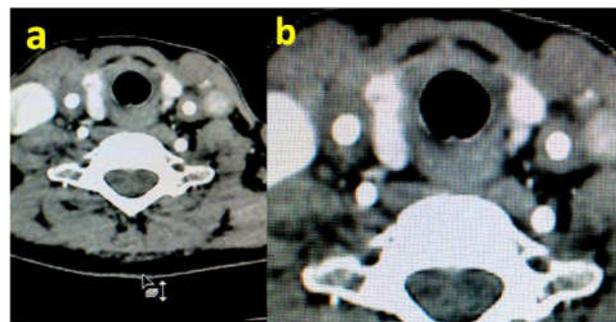


Fig 5. (a, b). Circumferential wall thickening without significant narrowing, were noted involving the bilateral common carotid arteries up to the carotid bulb



Fig 6. Cardiomegaly of left ventricular type

Based on the above mentioned findings and considering the age using 1990 criteria of American College of Rheumatology the diagnosis of Takayasu arteritis in active phase, was made. According to the New angiographic classification of Takayasu arteritis, the disease was classified as Type V Takayasu arteritis. The visualized lung parenchyma revealed tree in bud opacities with surrounding ground glass opacities in apical segment of the right lower lobe which were likely to be secondary to infective aetiology. The patient was subsequently treated with empirical antibiotics for the respiratory infection and then started on immunosuppressant therapy consisting of daily oral prednisolone (1mg/kg/d) and weekly oral methotrexate (0.15mg/kg/week). The patient could not be followed up.

DISCUSSION

Takayasu arteritis is a chronic inflammatory disease that involves the aorta, its branches and the pulmonary arteries. Takayasu arteritis (TA) is predominantly a disease of young adults in the second and third decades of life. The onset of illness may be earlier, including in childhood (Kerr *et al.*, 1994; Ladhani *et al.*, 2001) but rarely in infancy (Mitchell and Parisi, 1997). The female: male ratio has varied from 9: 1 in reports from Japan (Sekiguchi and Sazuki, 1992) to 1.3: 1 in India (Shrivastava *et al.*, 1986). 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 (Seko *et al.*, 1994). A number of lymphocytes (T cells and natural killer cells) infiltrate and incite the damage by liberating perforin on to arterial tissue which are reactive to particular antigen, heat shock protein (HSP)-65 which by itself is a major antigen of mycobacterium tuberculosis (Seko, 2000). TA involves mainly the elastic arteries (Kinare *et al.*, 1998). The disease may be patchy with normal skip areas in between, or diffuse along the length of the entire vessel. In the initial acute stage of the disease, exudative and granulomatous inflammation is seen, whereas fibrosis predominates later, but the two stages may co-exist (Hotchi, 1992).

Later in the disease process, nodular fibrosis in all layers of the artery is seen and the intima may become several times thicker than the tunica media obliterating the vessel lumen. The disease is diagnosed based on the American College of Rheumatology (ACR) 1990 diagnostic criteria. The disease is classified based on the site of involvement according to New angiographic classification of Takayasu arteritis, Takayasu conference 1994: (Johnston *et al.*, 2002)

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

The site of arterial disease determines its clinical presentation which usually includes diminished or absent pulses, vascular bruits particularly affecting the carotids, subclavian, and abdominal vessels, hypertension secondary to renal artery stenosis causing retinopathy, aortic regurgitation, neurological features including postural dizziness, seizures, and amaurosis. Pulmonary artery involvement is also seen. Other symptoms include dyspnoea, headaches, carotodynia, myocardial ischemia, chest wall pain, and erythema nodosum.

The presentation as DCM is rarely reported in 5-6% of cases and is due to involvement of coronary artery & severe hypertension (Jameson *et al.*, 2012). The progression of heart failure is associated with LV remodelling, 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 (Pieske, 2004). Death is due to either congestive heart failure or ventricular tachy- or bradyarrhythmias. The presence of systemic symptoms, raised ESR and worsening of vessel stenosis are considered evidence of active disease (Kerr *et al.*, 1994).

Therapeutic modalities include steroids, immunosuppressive agents, and antihypertensive drug therapy. 20-100% success rate of steroids have been reported in different studies.¹⁵ In the acute phase of TA, treatment with corticosteroids (1mg/kg/d) leads to clinical remission in 60% of cases (Kerr *et al.*, 1994). Immunosuppression with Cyclophosphamide (1-2mg/kg/d), azathioprin (1-2mg/kg/d) or methotrexate (0.15-0.35 mg/kg/week) may be tried in resistant cases, or in order to reduce steroid dosages (Kerr *et al.*, 1994). 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. Percutaneous transluminal renal arterial dilatation is done in case of renal artery involvement.

Conclusion

In a case of dilated cardiomyopathy in young female, screening for systemic vasculitis should be done as timely initiation of immunosuppressant therapy can reduce the development of other complications of Takayasu arteritis.

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REFERENCES

- Ghosh, S., Sinha, D.P., Ghosh, S., Mitra, D., Kar, A.K., Panja, M. 1999. Dilated cardiomyopathy in non-specific aortoarteritis. *Indian Heart J.*, 51:527-31.
- Hotchi, M. 1992. Pathological Studies on Takayasu arteritis. *Heart vessels.* (Suppl 7):11-17.
- Jameson, J.N., Kasper, D.L., Harrison, T.R., Braunwald, E., Fauci, A.S., Hauser, S.L. *et al.* 2012. Harrison's Principles of Internal Medicine. 18thed. New York: McGraw-Hill Medical Publishing Division; p. 1954-6.
- Johnston, S.L., Lock, R.J. and Gompels, M.M. 2002. Takayasu arteritis: a review. *J. Clin. Pathol.*, 55:481-486.
- Kerr, G.S., Hallahan, C.W., Giordano, J., Leavitt, R.Y., Fauci, A.S., Rottem, M. and Hoffman, G.S. 1994. Takayasu arteritis. *Ann Intern Med.*, 120:919-929.
- Kerr, G.S., Hallahan, C.W., Giordano, J., Leavitt, R.Y., Fauci, A.S., Rottem, M. and Hoffman, G.S. 1994. *Takayasu arteritis.* *Ann Intern Med.*, 120:919-929.
- Kinare, S.G., Gandhi, M.S., Deshpande, J.R. Mumbai: 1998. Non-specific aortoarteritis (pathology and radiology) monograph.
- Ladhani, S., Tulloh, R. and Anderson, D. 2001. Takayasu disease masquerading as interruption of the aortic arch in a 2 year old child. *Cardiol Young*, 1:244-246.
- Mitchell, C.S. and Parisi, M.T. 1997. Magnetic resonance imaging of Takayasu's aortitis in an infant. *J Am Osteopath Assoc.*, 97:607-609.
- Pieske, B. 2004. Reverse remodeling in heart failure - fact or fiction? *Eur Heart J Suppl.*, 6 Suppl:D66-78.
- Sekiguchi, M. and Sazuki, J. 1992. An overview of Takayasu Arteritis. *Heart Vessels.* (Suppl 7):6-10.
- Seko, Y. 2000. Takayasu arteritis. Insights into immunopathology. *Jpn Heart J.*, 41:15-26.
- Seko, Y., Minota, S. and Kawasaki, A. 1994. Perforin secreting killer cell infiltration and expression of -KD heat shock protein in aortic tissue of patients with Takayasu's arteritis. *J Clin Invest.* 93:750-758.
- Shrivastava, S., Srivastava, R.N. and Tandon, R. 1986. Idiopathic obstructive aortoarteritis in children. *Indian Paediatr.*, 23:403-410.
