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CASE REPORT

TAKAYASU'S ARTERITIS LEADING TO DILATED CARDIOMYOPATHY: A RARE PRESENTATION

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ABSTRACT

An eight year old female child presented with limb pain and features of congestive cardiac failure, found to have absent pulse in left upper limb and hypertension in right upper limb, investigated and found to have dilated cardiomyopathy with diffuse stenosis of aorta and renal arteries. The case was diagnosed as Takayashu's arteritis with dilated cardiomyopathy and was managed conservatively with oral steroids, to which she responded well.

INTRODUCTION

Takayasu's arteritis (TA) is a chronic granulomatous inflammatory disease of large vessels which affect mainly aorta and its major branches and also coronary, pulmonary and renal arteries [Johnston SL, 2002; Numano F, 2005]. The disease is also called 'pulse less disease', since peripheral pulses are often absent due to vascular obstruction. Takayasu's arteritis leads to progressive fibrosis and narrowing of lumen, occasionally it destroys the arterial media inducing aneurysm formation. It affects mainly females in second decade of life from south East Asia with a median delay of approximately 15 months between onset and diagnosis [Cassidy JT, 2005]. More recently, however, it has been shown that Takayasu's arteritis can affect both the sexes, any age and all ethnic groups, with different phenotypes in different countries; moreover, in each subgroup of patients, it shows different patterns of vascular involvement, clinical manifestations and prognosis [Vanoli M 2005, Mwipatayi BP 2005, Moriwaki R 1997].

Dilated cardiomyopathy is the most common type of cardiomyopathy in children [Ghosh S, 1999].Different types of cardiomyopathy have different causes and affect the heart in different ways. In India, Takayasu's arteritis as a cause of dilated cardiomyopathy (DCM) is, however, seen in only 5 – 6% of cases of TA while reno-vascular hypertension is observed in 28 – 75 % and congestive cardiac failure in 75 %, are reported[Ghosh S, 1999].The presentation as cardiomyopathy is rarely reported and is due to involvement of coronary artery, severe hypertension and cardiac failure, while all these are bad prognostic factors [Ghosh S, 1999;Greidanus PM, 2006].

The case

An 8 years old girl child presented with dyspnoea on exertion for 3 months which got aggravated during 6 days before admission, limb pain for 3 months, generalised body swelling and weakness for 15 days. On examination child was conscious, cooperative and tachypnoec. Facial puffiness and bilateral pedaloedema were noted. Pallor, icterus, clubbing and cyanosiswere absent. Pulse in right upper limb was 136/ min with good volume and normal rhythm but was absent in left upper limb. Blood pressure in right upper limb was 180/108 mm Hg but not recordable in left upper limb. Bruit was present in left suprasternal area and abdomen. On systemic examination, chest air entry was equal bilaterally and fine crepitation was noted. On CVS examination, JVP was raised, apex was down and out, S1 S2 werenormal, a systolic murmur grade II/VI was noted in apical area and left parasternal region. On abdominal examination, liver was palpable 5 cm below right costal margin and was tender. Blood investigation shows normal haemogram with hemoglobin 14 gram%, TLC 14600 cells/cmm, platelets count 4 lakh/cmm , raised acute phase reactants with ESR 24 mm at first hour and CRP was positive. Kidney function test (Urea 26mg%, creatinine 0.7 mg %) was normal. Liver function test showedfeatures of mild hepatitis (serum bilirubin 1.2mg/dl (direct 0.6, indirect 0.6), ALT 289U/L, AST 211U/L, GGT 202 U/L) with normal serum

protein. Serum electrolytes were normal (sodium 136 mmol/l, potassium 3.6 mmol/l, calcium 7.2 mg% and phosphorus 2.8 mg%. Thyroid function test were normal (T 3 - 1.68 nmol/l , T4-7.66 µg/dl, TSH – 3.64 µIU/ml), ASO titre normal , ANA was negative , tuberculosis workup done and were found negative. Urine culture and blood culture showed no growth, chest X-ray showed cardiomegaly with pulmonary congestion. ECG showed tachycardia, non specific ST – T changes (LV strain pattern) and LVH.

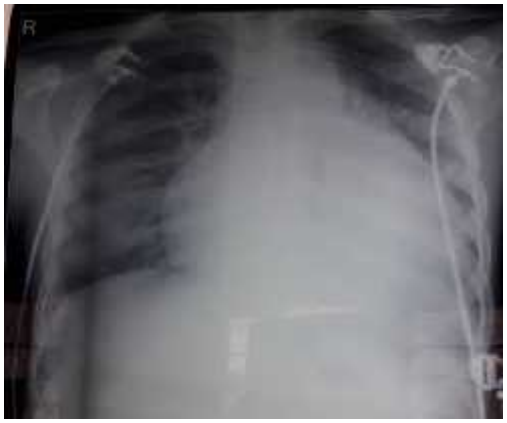


Fig 1 Chest X-Ray showing cardiomegaly with pulmonary congestion

Echocardiography showed Dilated cardiomyopathy with dilated LV cavity and global wall hypokinesia with severe LV systolic dysfunction (ejection fraction 33%) , moderate MR , Mild TR , Mild + AR, Gradient noted in descending thoracic aorta .USG showed bilateral renal arteries stenosis at ostium, long segment narrowing of left subclavian artery with thickened vessel wall and hepatomegaly.

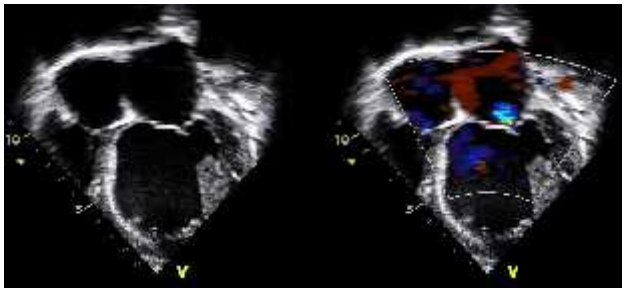


Fig 2 Echo showing dilated left ventricle, mitral regurgitation and tricuspid regurgitation

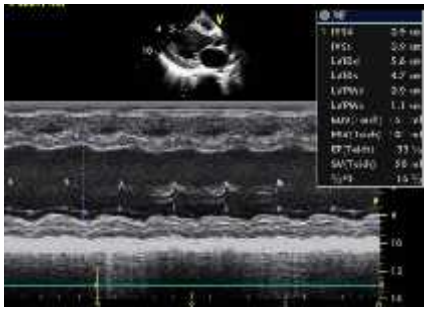
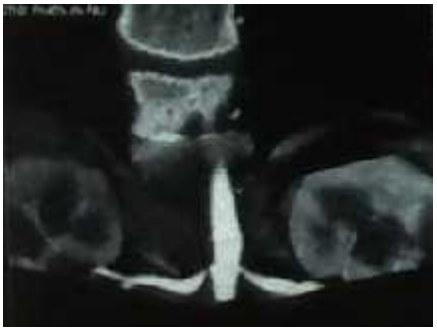


Fig 3 M mode: Dilated LA,LV with EF 33%

CT angiogram (Post-contrast) showed mild stenosis of distal arch of aorta, moderate to severe stenosis of descending aorta and abdominal aorta, both renal arteries stenosis, significant stenosis of left subclavian artery, mild thickening of coronary arteries with multiple epicardial collaterals, LV cavity was grossly dilated with global wall hypokinesia.

A



B

Figure 4A and 4B, CT angiogram showing stenosis of distal aortic arch, descending aorta, abdominal aorta (A) and bilateral renal artery (B).

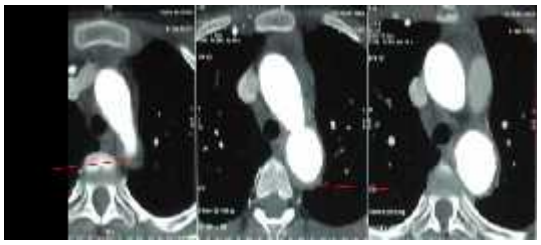


Fig 5 CT Angiogram showing circumferential wall thickening of distal aorta

With these finding a diagnosis of Takayasu's arteritis (Type III), dilated cardiomyopathy, CCF and reno-vascular hypertension was made. The child was treated with anti-congestive measures and with antihypertensive. Prednisolone was started in view of activity of the disease. On follow-up; prednisolone was tapered off at 6 months because of absence of fresh lesions and acute phase reactants returning to normal. Hypertension has persisted even on three antihypertensive drugs. Correction of renal artery stenosis, like angioplasty or stenting was necessary and hence, referred to higher centre.

DISCUSSION

TA is a chronic inflammatory disease mainly affecting aorta and its major branches leading to vessel stenosis, occlusion or less frequently aneurysm formation. Even after a century since the first description of TA by MikitoTakayasu in 1908, the aetiology is still unclear. Infections, autoimmunity and genetic factors have been considered as etiological factors.

Worldwide incidence is estimated at 2.6 cases per million per year [Sen PK, 1971]. In India, mean age of onset of symptoms

was 24.0 (± 8.8) years with a female preponderance (63%) [Subramanyan R, 1989].

TA mainly affects young adult in 2nd& 3rd decade of life but it rarely may affect infants. Female are more commonly affected than male, with a frequency varying in different geographical area. The female: male ratio varied from 9:1 in Japan [Sekiguchi & Sazuki, 1992] to 1.3:1 in India [Shrivastava *et al* 1986]

In the acute phase of Takayasu's arteritis, histology is characterised by a pan-arteritis extending from the adventitia to the media which is infiltrated by lymphocytes and occasional giant cells with neovascularisation. In the chronic phase, there is thickening of the whole vassal wall, the adventitia becomes fibrotic, the media is fragmented because of the destruction of elastic fibres and the proliferation of the intima causes a reduction of the vessel lumen. The development of arterial aneurysms, less frequent than stenosis, is probably due to the lack of fibrotic tissue or to a localised weakness of the intima. The inflammatory process can invade the surrounding structures mimicking the inflammatory aneurysm.

Clinical presentation of TA is nonspecific. The clinical course is divided in to an early active inflammatory phase and late chronic phase. The active phase lasts week to months and may have a relapsing and remitting course. It is characterized by constitutional symptoms like fever, malaise and weight loss, night sweat, headache, dizziness, arthralgia and skin rashes. The late chronic phase is due to arterial stenosis and or occlusion and ischemia of organ and depends upon site of arterial involvement which usually includes diminished or absent pulses (84-96 %) [Kerr GS, 1994], vascular bruit (80 - 94 %) [Sen PK, 1971]. Hypertension (33 - 83 %) generally due to renal artery stenosis which is seen in 28 to 75 % patients [Kerr GS, 1994], congestive cardiac failure (28 %),(due to hypertension and DCM) and pulmonary artery involvement in 14 to 100 % patients is observed.

The diagnosis of TA is based on characteristic finding of diseased aorta and its major branches seen on angiography. The disease is classified as: Type I- Aortic arch involvement, Type II- Thoraco-abdominal involvement, Type III- Diffuse involvement, Type IV- Pulmonary artery involvement, Type V- Aneurysm formation [Satsangi DK, 2007]

In our case involvement of left subclavianartery, distal arch, descending aorta, abdominal aorta and renal artery indicates stage III disease.

Dilated cardiomyopathy present in our case is rarely reported and occurs in only 5 – 6 % of the cases of TA. The cause of DCM is difficult to define as both hypertension and TA itself could act as its causative factors.

The treatment of TA begins with pharmacological control of acute arteritis in order to induce clinical remission, followed by management of vascular abnormalities. Another aspect is the treatment of vasculitis complications such as hypertension and thrombosis. Active disease is treated with steroid and immunosuppressive agents with about 20-100% success [Yadav MK, 2006].

In our case, the patient responded well to prednisolone at a dose of 1 mg/kg/day and was tapered off after 6 months after achieving remission. For progressive disease and non-responder of steroid therapy orcases with steroid toxicity, immunosuppressive therapy like methotrexate, cyclophosphamide or biologic agents might become necessary.

CONCLUSION

In children with hypertension, heart failure with dilated cardiomyopathy, the possibility of vasculitis syndrome should be considered because early diagnosis of Takayasu's arteritis and institution of immunosuppressant have a positive bearing on prognosis.

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