

CASE REPORT

UNUSUAL PRESENTATION OF TAKAYASU ARTERITIS AS DILATED CARDIOMYOPATHY IN YOUNG MALE.

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ABSTRACT: Takayasu arteritis is a rare granulomatous systemic disease characterized by stenosis or obliteration of large & medium sized arteries mainly aorta and its main branches as well as pulmonary artery. Although this disease has variable presentation, but its presentation as Dilated cardiomyopathy is rare. Renal involvement is also less frequently reported in young males. We report a case of 20 yr old male who presented with features of congestive heart failure, renal failure and incidentally detected with absent right radial pulse and hypertension .His CT angiography revealed occlusion of right subclavian artery, superior mesenteric artery & bilateral renal arteries with splenic infarct suggestive of Takayasu arteritis.

This case report illustrates a rare case of dilated cardiomyopathy and renovascular hypertension with renal failure secondary to Takayasu Arteritis which responded with systemic corticosteroids.

KEY WORDS: Absent pulse, Heart Failure, Malignant Hypertension, Renal failure.

INTRODUCTION: Takayasu arteritis (TA) is a rare chronic inflammatory vasculitis that affects primarily the young female in 2nd and 3rd decade .It occurs worldwide with wide geographical variation, seen mainly in Japan, South East Asia, India and Mexico [1].

It mainly affects the aorta & its main branches and has variable and nonspecific clinical presentation due to vessel inflammation leading to wall thickening, fibrosis, stenosis, thrombosis, end organ ischemia & aneurysm formation thus leading to delayed diagnosis and treatment. It is the common cause of Reno vascular hypertension [2].Carotid intimal thickness, C-reactive protein, & elevated ESR are important marker for disease activity, which is followed up in TA [3]. We report a rare case of Takayasu arteritis presented as dilated cardiomyopathy and Reno vascular hypertension

CASE REPORT: A 20 year young male admitted with history of low grade fever and progressive shortness of breath for 20 days, which rapidly worsened to orthopnoea (N Y HA class iv) in one week with severe upper abdominal pain .There was no history of joint pains, limb pain on

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walking (claudication), contact with tuberculosis, jaundice, diabetes mellitus, hypertension or any cardiac disease in past. On physical examination he was tall thin built, febrile, tachypnoeic with respiratory rate of 36/min. His pulse was 108/min in left radial and absent pulses in right upper limb both brachial & radial. Lower limb and common carotid pulsations were equal on both the sides. Blood pressure in left upper limb was 210/150 mm of Hg, not recordable in right upper limb & was equal (230/160 mm of Hg) in lower limb. He had signs of volume overload like raised JVP, facial puffiness, bilateral pitting pedal edema and tender hepatomegaly. Respiratory system examination revealed fine bibasilar crepitations upto mid scapular area. On auscultation of cardiovascular system S₃ gallop, and pansystolic murmur was present in mitral area. Right subclavian bruit and bilateral renal bruit was noted. Ophthalmological examination show normal visual acuity with mild pallor of optic disc.

ON INVESTIGATIONS: He was anemic (hemoglobin 8.6gm%, normocytic normochromic). Urinalysis revealed microscopic hematuria (6-8 RBC/hpf) & proteinuria (3+) though his 24 hr urinary protein was 5mg/l. Renal function tests were deranged with urea 165mg/dl & s. creatinine of 2.75 mg/dl which increased up to 295 mg/dl & 7.7mg/dl respectively. His CRP (1.2mg/dl), corrected ESR (26) were elevated. His ANA, HIV, HB_sAG, VDRL, MOUNTX TEST and ASO titer were negative. Serum electrolytes & Liver function tests were also normal. X-ray chest showed- cardiomegaly (CT RATIO: 65%) & pulmonary edema (Fig-1) Echocardiography with color Doppler: showed global hypokinesia with moderate LV dysfunction (LVEF 25%), moderate (GRADE II) mitral regurgitation and aortic regurgitation & trivial tricuspid regurgitation, suggestive of dilated cardiomyopathy. USG abdomen revealed bilateral renal parenchymal disease (grade I) and RENAL DOPPLER showed tardus parvus flow in main renal & interlobar arteries bilaterally (Fig-2.) CT ANGIOGRAPHY revealed occlusion of right subclavian artery with distal flow through collateral, narrowing of proximal part of left subclavian artery with good distal flow, irregular wall thickening of thoracic & abdominal aorta with focal ring like narrowing of abdominal aorta (50%), significant right renal artery narrowing at its origin & occlusion of proximal part of left renal artery and splenic infarct involving significant part of spleen. (Fig 3-4)

Patient was treated initially for congestive heart failure and renal failure with furosemide, digoxin and hemodialysis. Based on clinical findings and supportive investigations diagnosis of vasculitic syndrome-Takayasu arteritis was made and prednisolone in the dose of 1mg/kg/day together with antihypertensives was started. On discharge his BP was well controlled, s. creatinine came down from 7.76 to 1.86mg/dl, and urinary sediments disappeared. Surgery and other interventions were not done as his inflammatory markers were on higher side. On follow up after 2 months his renal function tests returned to normal & cardiac function improved markedly with LVEF of 45% in Echocardiography.

DISCUSSION: Takayasu arteritis is a chronic progressive inflammatory disease of large & medium sized arteries seen mostly in Asian and Hispanic origin with female preponderance. It was first reported by Japanese Ophthalmologist Mikito Takayasu (1908) in a young female with retinal changes. Later Shimzu and Sane (1928) described it as "pulse less disease". Females are affected more commonly than males, with varying ratio as high as 1:9 in Japan to as low as 1:1.2 in Israel where as recent series reported ratio of 1:6.4 in India [1] In adults TA usually involves aortic arch while in children abdominal aorta is most commonly affected. In Japan proximal aortic involvement with features of

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“reverse coarctation” is seen, where as abdominal aorta involvement with Reno vascular lesion(middle aortic syndrome) is seen in Southeast Asia and Africa [4] .In India Takayasu arteritis as cause of Reno vascular hypertension is reported in 28%-75%, Congestive heart failure in 76%, aortic regurgitation in 20-24% and dilated cardiomyopathy only in 5%.^[5]The presentation as dilated cardiomyopathy is rarely reported ,and is due to involvement of coronary artery .Severe hypertension and cardiac failure are bad prognostic factors.^[6-8]

Our case is a young male presented with Reno vascular hypertension, congestive heart failure and dilated cardiomyopathy with absent arterial pulsation in right upper limb. According to ACR (1990) criteria ^[9] presence of three out of six criteria is required for the diagnosis and our case fulfilled FIVE criteria. It was angiographically confirmed as type V takayasu arteritis (According to new angiographic classification of Takayasu arteritis 1994) .The incidence of type V TA as reported by Panja et al in largest series is only 10%. ^[1]

CONCLUSION: With this case report we want to emphasize that any young patient with 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 which can be devastating.

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Fig 1 x-ray chest showing cardiomegaly & pulmonary edema

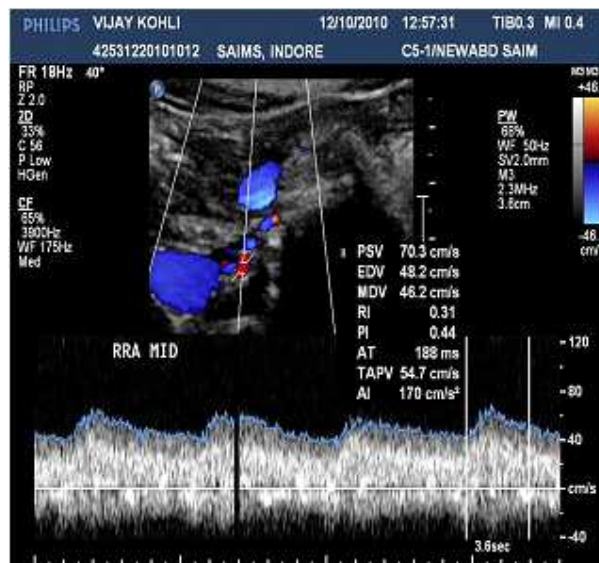


Fig- 2 On spectral Doppler, Right renal artery shows dampening of systolic flow with increase acceleration time suggesting tardus and parvus type flow

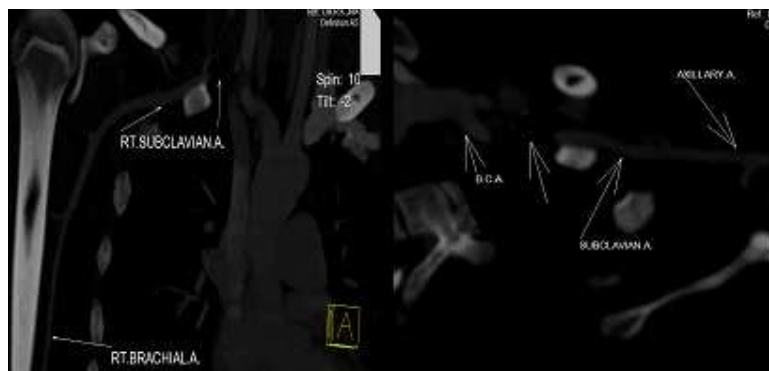


Fig- 3 Occlusion of right (complete) and left (70%) subclavian artery

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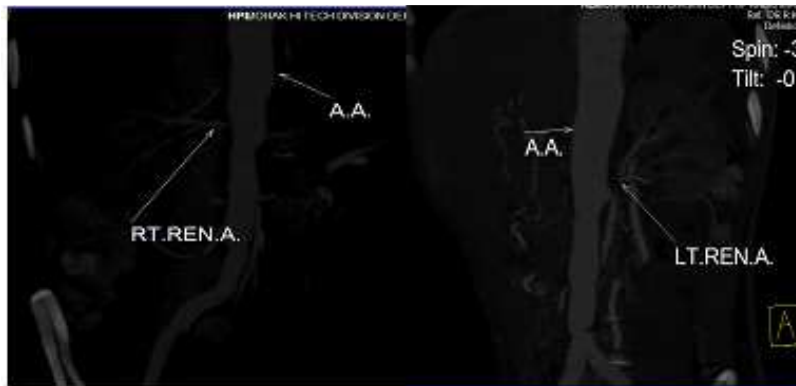


Fig -4 Focal significant narrowing of renal arteries at its origin