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Piyush Upadhyay, P.L Prasad, R.Madhok, D.C.Pant, A.V.S Rathee

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CASE REPORT

A Case of Rheumatic Heart Disease with Mid Aortic Syndrome possibly due to Takayasu Arteritis

Piyush Upadhyay[1], P.L Prasad[1], R.Madhok[2], D.C.Pant[3], A.V.S Rathee[1]

Department of Pediatrics[1], Department of Radiodiagnosis[2], Department of Cardiology[3]
Sri Ram Murti Smarak Institute of Medical Sciences, Bareilly, U.P, India

Abstract:
Takayasu Arteritis (TA) is a chronic, idiopathic and granulomatous vasculitis of the large arteries. It involves primarily the aorta, especially aortic proximal branches, and occasionally the pulmonary arteries. We report a 10 year old boy with rheumatic heart disease withmid aortic involvement probably due to TA. Complete Aortogram revealed narrowing of abdominal aorta, superior mesenteric artery and the two renal arteries.

Keywords: *Takayasu arteritis; rheumatic heart disease; endocarditis*
Corresponding author: *Piyush Upadhyay, Department of Pediatrics Sri Ram Murti Smarak Institute of Medical Sciences, Bareilly, U.P, India*
e-mail: piyushdrpeds.com@gmail.com

Introduction

Takayasu, a Japanese ophthalmologist, first described the clinical entity in 1908 in a young female with retinal changes. This inflammatory condition may result in stenosis, occlusion, dilatation or aneurysm of the involved arteries [1]. Takayasu's arteritis (TA) is the commonest cause of renovascular hypertension in Asian children [2,3]. Hypertension is the most common reason for heart failure, but may occur in the absence of severe hypertension [4]. TA in children is a serious illness and a mortality of 10-30% has been reported on followup [4-6]. The association of TA with rheumatic heart disease has not been reported earlier.

Case report

A 10 year old male child presented with ten days history of cough, fever, dyspnoea, palpitation and swelling all over the body. There was no history suggestive of chronic cough, hemoptysis, recurrent pneumonia, chest pain, syncope or transient ischemic attack. There was past history of 3 episodes of epistaxis and claudication in lower limbs for last 1 year. On physical examination, child had grade 4 pansystolic murmur in mitral area (clinically Mitral Regurgitation), just palpable spleen, bilateral renal bruit and fever. In the lower limbs pulses were low volumic and bilateral dorsalis pedis artery pulses were absent. Blood pressure in upper limbs were higher (116/90 mmHg and 114/90 mmHg in Rt. And Lt. Upper lims respectively) than that of lower limbs (90/60 mm Hg in both lower limbs; difference of around 24mm Hg in systolic values). His neurological and opthalamological examination were normal. Echocardiography was suggestive of rheumatic heart disease, severe mitral regurgitation (MR), mild aortic valve regurgitation (AR) and normal left

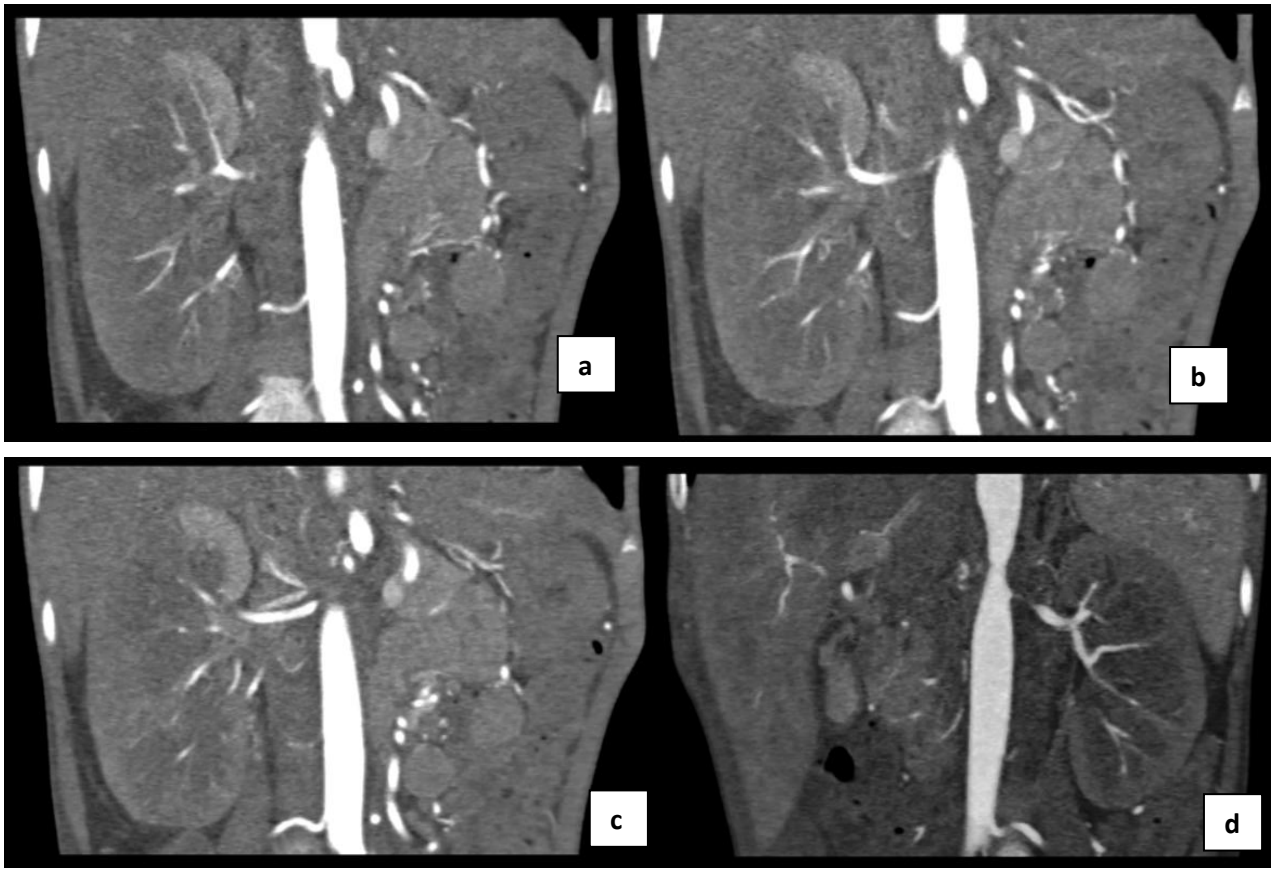

Figure 1 a, b, c, d. Complete aortogram revealed narrowing of abdominal aorta, superior mesenteric artery with bilateral renal artery narrowing.

ventricular function. The child was provisionally diagnosed as a patient with acquired heart disease of rheumatic origin with some aortic pathology. He was stabilised and investigated. Laboratory investigations showed microcytic hypochromic anemia with high total leukocyte count. The erythrocyte sedimentation rate (ESR) and C reactive protein were raised. There was documented evidence of 6 fold increase in anti-streptolysin O titres. Blood cultures came out to be sterile hence infective endocarditis was ruled out. Blood urea and serum creatinine were consistently high in serial reports. Urine examination revealed hematuria and albuminuria in serial reports. Mantoux test was negative. On chest radiography, mild cardiomegaly with prominent pulmonary vascular markings was present. Ultrasonography of the abdomen revealed enlargement of the liver and spleen with normal echotexture. The ultrasonography was also suggestive of bilateral renal parenchymal disease with no backpressure changes and normal perinephric spaces. After stabilising new findings were revealed. Review Echocardiography confirmed severe MR and mild AR with normal left ventricular function. Complete aortogram was done which revealed narrowing of abdominal aorta, superior mesenteric artery with bilateral renal artery narrowing (Figure 1a,b,c,d). Thus, he was diagnosed as having Rheumatic heart disease with Mid Aortic syndrome possibly due to TA.

Discussion

Very few cases of Takayasu arteritis in children have been reported in the medical literature. The presentation in the form of rheumatic heart disease alongwith involvement of the renal artery and abdominal aorta due to Takayasu arteritis has not been previously described in children. There is considerable variability in disease expression due perhaps to geographic differences

[7]. The disease has worldwide distribution but is more common in Japan, India and China [8]. Bacterial endocarditis may present itself with rheumatological symptoms in 28–42% of patients [9]. The presence of systemic symptoms, raised ESR and worsening of vessel stenosis are considered evidence of active disease [9]. Takayasu's arteritis has been linked to rheumatic fever and other streptococcal infections, rheumatoid arthritis, and other collagen vascular diseases [10]. A case of Takayasu Arteritis in a 17 year old female patient with rheumatic fever complicated with Sydenham's Chorea was published recently [11]. In that case patient presented at age of 6 years with rheumatic fever followed by chorea a month later. At the age of 16, she developed a blood pressure discrepancy between the arms and faint pulses. Computed tomography angiography revealed diffuse aortic involvement and narrowing of the arteries in that case. This association is rare and therefore of great interest. The co-presence of rheumatic fever and TA also raises the possibility of an immunological basis for the pathogenesis of the disease.

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