Takayasu’s Disease Presenting with Pain Chest, Prolonged Pyrexia and Pleural Effusion

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Abstract

Takayasu’s disease (TD) is a diffuse arteriopathy recognised by various names viz. Takayasu Arteritis, Takayasu’s Disease, Takayasu Syndrome, Pulseless Disease, Non-specific Aortoarteritis, Reversed Coarctation, Aortic Arch Syndrome, Aortitis Syndrome, Young Female Arteritis, Idiopathic Arteritis and Martorell Syndrome. Though described about a century ago and with many eponyms, TD yet remains a challenging problem regarding etiopathogenesis, clinical presentation and management. We present a case of TD with pyrexia of unknown origin (PUO), angina and left sided pleural effusion.

INTRODUCTION

Takayasu’s disease is typically characterised by a tetrad of non-specific constitutional symptoms, absent pulses in one or more of the limbs, reversed coarctation with lower limb hypertension and renal failure/renovascular hypertension. Cardiac or pulmonary symptoms though recognised are rare on initial presentation.

We present a report of a young woman misdiagnosed initially because of presentation with fever, pleural effusion and angina-like symptoms.

CASE REPORT

A 32 years female presented with recurrent fever, shortness of breath and angina on effort. The ailment started about five years prior to consultation with chronic febrile illness. Evaluation revealed left side pleural effusion and was treated with antituberculous drugs. Fever persisted and antituberculous therapy was discontinued after 3 months by the patient herself. She consulted many doctors for her febrile illness and was given a variety of antibiotics and NSAIDs, but without a satisfactory response. She had full term pregnancies at age of 25, 27 and 29 which were uneventful.

She consulted a gynecologist for irregular periods and was referred to a cardiologist for chest pain. She had come with multisystem complaints including pain and throbbing sensation on right side of neck, palpitation, slow pulse left side, fullness of abdomen, polymenorrhea, intermittent low grade fever (99-100°F) and pain left chest with radiation to left arm on effort. This had been present for 3 years prior to hospitalization but increased in intensity recently.

Clinical Examination

Thin built, non-toxic look, not dyspneic, BP 140/80 mm Hg (Rt) – 90/70 (Lt) in upper extremity 150/86 mm Hg in lower extremity.

The right common carotid pulsation was more marked with bruit over it, right radial and brachial pulses felt, left radial and brachial absent, left femoral diminished, left carotid diminished, abdominal aorta pulsation well marked. Chest–dullness left base with diminished breath sounds. Heart – Early blowing diastolic murmur left sternal edge and short systolic murmur localised to aortic area.

Abdomen – Mild hepatosplenomegaly. CNS and other system examination – no abnormality detected, Fundus – normal.

Investigations

Blood sugar fasting - 71 mg/dl, Creatinine - 1.3 mg/dl, Urea - 24 mg/dl, Hb-9.2 gm, ESR-60 mm, T3 – 1.00 ng/ml, T4 6.00 mg/dl, TSH – 5.50 mIU/ml, HBsAg – Negative, HIV – Negative, PCR for TB-Negative, Mantoux test – Negative, Blood culture × 3 samples – Negative, Malarial parasite-Negative, Lipid profile – Normal, Pleural fluid – exudative lymphocytes 300/μm, protein – 3.2 g/dl, LDH – 210 IU/l, AFB –ve, Gram Stain –ve, ANF, anti-double stranded DNA – negative. ECG – LVH with ST-T changes. PA Chest Radiograph – Enlarged cardiac shadow with dilated aorta, small left pleural effusion. Echocardiogram – Thick aortic valve with moderate aortic regurgitation : LV ejection fraction ~45%, AV gradient ~30 mmHg. Ultrasound abdomen – Contracted small left kidney with thin echogenic cortex.
with loss of corticomedullary distinction vascular Doppler – Extensive narrowing throughout the length of left carotid, compromising the lumen. The false lumen measured 6mm and the true lumen 108mm. Markedly dilated right carotid and innominate artery: the abdominal aorta and branches – normal (Fig 1). (i) CT scan of Cranium – normal study; (ii) CT Angiogram of Circle of Willis – Hypoplastic A-1 segment of left anterior cerebral artery with good filling of A-2 segment through anterior communicating artery.

Cardiac catheterization and angiography - Right side – PCW pressure - a 34, v 41, Mean 26 PA – 67/31 : RV 61/0; RA – a8 V6 Left side – LV 208/17; AO 208/74; DA – 201/76. Right brachiocephalic dilated : Right subclavian – Obstruction in mid segment 80% and 100% distal, Right carotid – normal; Left subclavian and left carotid – not visualized (Fig.2); Renal artery – normal. Coronary artery – RCA – normal; LCA - Left main ostial stenosis 70% without any reflux of dye with post stenotic dilation (Fig. 3), LAD – normal, LCX – normal, Global Hypokinesia, LVEF – 40%.

Fig. 1 : Vascular doppler of aorta and main branches showing extensive narrowing throughout the length of left carotid compromising the lumen. The false lumen measured 6mm and the true lumen 108 mm.

Fig. 2 : Angiographic findings of main aortic vessels.

Fig. 3 : Coronary angiogram showing left main ostial stenosis.

The patient was placed on Prednisolone 60 mg daily with Methotrexate 2.5 mg weekly increasing the dose to 7.5 mg weekly a month. At the end of 3 months she had significant clinical improvement in clinical symptoms. She refused vessel biopsy and further intervention.

**DISCUSSION**

Takayasu’s Disease (TD) is an idiopathic inflammatory disease of large elastic arteries occurring in young with occlusive lesion in aorta and its branches and pulmonary artery and its branches.

In 1908 Mikito Takayasu,1 a Japanese ophthalmologist reported a case of retinal arteriovenous anastomosis and absent upper extremity pulse. But the first description of non-specific arteritis was mentioned by Savory and Kusmaul in 1856. In spite of recognition of this disease for more than a century and numerous publications in medical literature, etiopathogenesis of TD remains a mystery. Several workers supported the role of *Mycobacterium tuberculosis* as a causative factor for TD.2,3 However, Sagar et al4 concluded that autoimmunity may have a role in pathogenesis of TD, the antigen may not be *M. tuberculosis* and direct involvement of aorta by *M. tuberculosis* appears unlikely. The existing knowledge about pathogenesis of TD suggested that “it starts among genetically predisposed individual with perhaps specific hormonal milieu, followed by exposure to an unidentified antigen leading to immune response targeting large vessel.”5

The clinical presentation of TD may be non-specific constitutional symptoms like fever, weight loss and...
lethargy. Lack of specificity of symptoms often delays the correct diagnosis. Clinical manifestations like hypertension, renal failure, intermittent claudication, amaurosis fugax, syncope, TIA, Raynaud’s phenomenon have been reported by Panja and Mondal.

The present case had initially prolonged fever of unknown etiology and pleural effusion, treated by antituberculous drugs with no response. Investigation for tuberculosis like Mantoux test and PCR were non-supportive for tuberculous infection. Aspiration and culture of the pleural fluid revealed an exudative effusion. AFB were negative on both Ziehl Neelsen staining and culture. Blood culture and urine culture were negative for microbial growth. Pain in neck was due to carotidynia. Aortic regurgitation was sequelae to dilated aorta and aortic valvulitis. Her coronary angiogram showing left main disease supported her recent onset angina. Moderate LV dysfunction (LVEF - 40%) is due to coronary artery disease and/or aortic regurgitation. She has symptomatic improvement since she was put on steroids and methotrexate. Pain in neck was due to carotidynia. Aortic regurgitation was sequelae to dilated aorta and aortic valvulitis. Her coronary angiogram showing left main disease supported her recent onset angina. Moderate LV dysfunction (LVEF - 40%) is due to coronary artery disease and/or aortic regurgitation. She has symptomatic improvement since she was put on steroids and methotrexate. Pulmonary involvement (pulmonary hypertension and/or pleural effusion) and CAD are recognized manifestations of Takayasu’s arteritis found in 9-25% of the disease. However, they are rarely the presenting features of the disease.

While the presentation of Takayasu’s arteritis may be protean, our patient presented with a triad of non-specific symptoms – weight loss with low grade fever, effort angina (without any family history of known risk factor) and unexplained left pleural effusion. This presentation has been rarely recorded in the literature.

In a female presenting with weight loss, pleural effusion and angina like chest pain, one should look for features of Takayasu’s disease, when tuberculosis and/or connective tissue disease have been ruled out. This will enable a prompt diagnosis by appropriate investigations and treatment so that at least some of the long term consequences may be alleviated.

It is concluded that in a case of PUO, the clinician must pay attention to detailed vascular examination for early detection for arteritis. Vascular Doppler, arteriography and MR angiography are of paramount importance in the early diagnosis of this condition.

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REFERENCES


Announcement

21st Annual National Conference of Indian Rheumatology Association

The 21st Annual National Conference of Indian Rheumatology Association (IRA) is being organized during December 1-4, 2005 in Hyderabad.

The abstracts of scientific papers are invited, prizes will be given for best papers and poster presentations.

Registration fee is Rs. 1400/- (Rs. 1200/- for IRA members, Rs. 750/- for postgraduates and accompanying persons) before 31st August 2005. The draft should be drawn in favor of IRACON2005, payable at Hyderabad.

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