Pulseless Cardiomyopathy

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Abstract

Takayasu arteritis can have myriads of presentation, depending on site and extent of disease. As no age is immune for this disease, it is important to keep a high index of suspicion, otherwise the diagnosis can be missed. We hereby describe a case of Takayasu arteritis, which presented as dilated cardiomyopathy.

INTRODUCTION

Takayasu arteritis (TA) is a non-specific granulomatous inflammatory arteropathy of unknown origin that results in occlusive obliterans or less commonly aneurysmal dilatation of large and medium sized elastic arteries. TA being the commonest cause of renovascular hypertension in India, also causes dilated cardiomyopathy (DCM) in 5% of affected patients. It is also the commonest cause for middle aortic or mid aortic dysplastic syndrome, defined as coarctation of aorta located at the distal thoracic or abdominal aorta or both. We hereby report a case of middle aortic syndrome with renovascular hypertension and DCM secondary to TA.

CASE REPORT

A 12 years female child presented with complaints of shortness of breath, weakness and low-grade fever. The complaints were of 6 months duration with a recent increase in severity of symptoms. She had received different antibiotics and non-steroidal anti-inflammatory agents for her symptoms, which did not improve. Evaluation revealed cardiomegaly and an echocardiogram was performed which revealed DCM. Then she was treated with angiotensin converting enzyme inhibitors, digoxin, and diuretics, for 4 months prior to hospitalization. This treatment also failed to relieve her symptoms.

Clinical Examination

Thin built, febrile with temperature 100°F, dyspnoea grade 4 (New York Heart association grading), and pulse–110/min in both upper limbs. Pulsations were absent in femoral, popliteal, and dorsalis pedis in both the lower limbs. Blood pressure was 150/110 in both the upper limbs and could not be recorded in lower limbs. Common carotid pulsations were equal on both sides, while abdominal aortic pulsations could not be felt. Bruit was audible over abdomen. Chest - fine crepitations heard in both lung fields extending up to lung apex, Heart - JVP was engorged (10cm). SI soft, mid systolic murmur best audible at apex and radiating toward the axilla was heard. Another systolic murmur at the left sternal edge, with inspiratory accentuation was also audible. Abdomen - mild tender hepatomegaly. CNS and other systems examination were normal. Fundus was normal. A clinical diagnosis of TA with renovascular hypertension and congestive heart failure was made; severe congenital coarctation of aorta was kept as the second closest diagnosis, however the presence of congestive heart failure pointed against it.

Investigations

Blood sugar fasting-80 mg/dl, Creatinine-1.1 mg/dl, Urea 30 mg/dl, Hemoglobin-9.4 gm/dl, ESR-10 mm 1st hour, Mantoux test was negative, LDH- 200 IU/Lt, Lipid profile, ANA, dsDNA, CRP, ASLO were all normal. PA chest radiograph – enlarged cardiac shadow, with diffuse alveolar shadows consistent with pulmonary edema, ECG showed sinus tachycardia with left ventricular hypertrophy, Ultrasonography – hepatomegaly with normal size kidneys. 2D Echocardiography (M mode) showed DCM, with severe left ventricular systolic dysfunction, ejection fraction – 25%, moderate mitral regurgitation, and severe tricuspid reguritigation. Peripheral Doppler was suggestive of significant strecture of abdominal aorta at the level of renal arteries with the involvement of both renal arteries. Contrast enhanced helical computer tomogram of abdomen and reconstruction of angiographic and venographic images showed circumferential wall thickening of lower descending aorta and abdominal aorta resulting in long segment smooth tapering stenosis upto L3 level, maximum at the D11 level (residual luminal diameter 3mm) (Fig. 1), with moderate stenosis of superior mesenteric artery and bilateral renal arteries.

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just distal to their origin (Fig. 2). Computer tomogram of thorax showed normal arch of aorta and its branches with all four cardiac chamber enlargement. We planned for conventional angiography, cardiac catheterization, vessel biopsy and renal angioplasty, but patient’s attendants refused these invasive procedures.

In view of available investigations diagnosis of TA type III, with stenosis of aorta (distal thoracic and abdominal), bilateral renal, and superior mesenteric arteries, with DCM was made. Patient was given prednisolone 40mg/kg/day, and methotrexate 2.5 mg/week, (increased to 7.5 mg/week), anti hypertensives, digoxin and diuretics. She was discharged and advised to consult department of cardiology whenever willing for interventions.

**DISCUSSION**

Mikito Takayasu first described Takayasu arteritis or pulseless disease, in 1908. It is a chronic inflammatory disease mainly affecting aorta and its primary branches, leading to vessel stenosis, occlusion, or less frequently aneurysm formation. One century has passed since the first description of TA, but still we are not close to its etiology. Some workers suggested that *Mycobacterium tuberculosis* is causative factor for TA, while some others are of opinion that autoimmunity (the suspected antigen may not be tuberculosis) is the primary culprit. The most accepted concept is that “pathogenesis of TA starts in genetically susceptible individual, with perhaps a specific humoral milieu, followed by exposure to a yet unidentified antigen, leading to immune response that targets the large vessels.”

TA commonly affects patients in 2nd and 3rd decade of life. Females are affected more commonly than males, with a frequency varying in different geographical areas, in India the ratio being 2:1. In adults TA usually involves aortic arch while in children abdominal aorta is most commonly affected. Characteristics features of TA includes diminished or absent pulses (84-96% of patients), vascular bruit (in 80-94% patients), hypertension (in 33-83% patients), generally due to renal artery stenosis, which is seen in 28-75% patients, congestive heart failure (due to hypertension and dilated cardiomyopathy), and pulmonary artery involvement in 14-100% patients. In active stage TA is treated by steroids and a success rate of 20-100% has been reported in different studies. Cyclophosphamide and methotrexate are used as steroid sparing agents and in resistant cases. Obstructive lesions need revascularization techniques such as angioplasty and surgery.

The present case had weakness, easy fatigability, and pyrexia of unknown origin. These seem to reflect nonspecific, constitutional symptoms of TA. Hypertension in this case was due to renal artery stenosis. The cause of DCM here is difficult to define as both hypertension and TA could act as its causative factors. Absence of pulses in lower limbs, presence of hypertension and presence of abdominal bruit were the clues toward diagnosis. Modern imaging techniques have aided greatly in diagnosis of TA and this may replace conventional angiography in coming time.

The purpose of reporting this case is to highlight the role of detailed physical examination including detailed vascular examination, aided by correct use of modern imaging to diagnose TA.
Acknowledgement

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References


Announcement

Lectureship (2007)

1. Unichem Lectureship in Gastroenterology (2007)
2. Dr. Yodh Memorial and Gwalior Conference Training Fellowship (2007) (General Medicine)
4. Shree Krishnaji Govind and Mrs. Pramalabai Bate Memorial Lectureship in Asthma and Bronchitis (2007)
6. Dr. Shurvir Singh Trust Visiting Professorship (2007) (General Medicine)

The selected candidate has to deliver his/her lecture at the institution of his/her choice in the year 2006. The candidate has to get a notification in writing from the Institution that he/she has delivered the lecture.

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The selected candidate has to deliver his/her lecture at the Annual Conference of API 2008.

Please note that all the above lectureships/orations need prescribed application forms which are available from the API Office and on website apiindia.org. The complete application forms for the above Lectureship should reach to Dr. Sandhya Kamath, Hon. General Secretary of API, Unit No. 6 and 7, Turf Estate, Opp. Shakti Mill Compound, Off. Dr. E Moses Road Near Mahalaxmi Station West, Mumbai 400 011 not later than 10th January, 2007.

II. Orations

Suggestions are invited from members for the following assignments so as to reach Dr. Sandhya Kamath, Hon. General Secretary not later than 10th January, 2007.

3. Dr. P.J. Mehta Oration (2008)
4. Dr. GS Sainani Oration (2008)
5. Ranbaxy Oration (2008) for Infectious Diseases

Please note that Orations No. 1,2,3 and 4 are for any subject in General Medicine.

All the above orations, Persons are selected from the recommendations received from members of the Association. The recommendations for the above assignments must be accompanied with reasons for recommending a particular person showing the value of his/her research and eight copies each of three of his/her best publications. All relevant papers in connection with the suggestions, such as the bio-data, list of publications etc., should be submitted in 8 sets by the proposer. The recipient of the above oration should deliver a lecture pertaining to his/her work at the Annual Conference in January, 2008.

A person who has received oration in the past is not eligible for any oration.

All lectureships, orations and awards are open to eminent persons from the discipline of medicine and allied subjects such as Pharmacology, Biochemistry, Pathology and Physiology. The orator in the discipline of Medicine should preferably be a member of API.

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