AN INTERESTING PRESENTATION OF TAKAYASU ARTERITIS
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Abstract :
Takayasu arteritis or pulseless disease is a chronic granulomatous necrotizing vasculitis predominantly affecting the aorta and its branches. The most common presenting vascular symptoms were claudication, reduced or absent pulse, carotid bruit ,hypertension , carotidynia, light headedness and asymmetrical arm blood pressures.Rarely, Takayasu arteritis presents as congestive cardiac failure in 2 percent cases .Here we describe a case of Takayasu arteritis (TA) which presented with congestive cardiac failure and dilated cardiomyopathy and Pulmonary artery involvement.

Keyword :
Takayasu arteritis ,Dilated cardiomyopathy , Congestive cardiac Failure.

CASE REPORT:
A 24 year old female presented to our hospital with complaints of difficulty in breathing, fatigue, giddiness, palpitation, swelling of both legs , facial puffiness of 3 months duration. There was no history of fever, chest pain or syncope. There was no significant past medical or surgical history. She had regular menstrual cycles, married since 4 years, but nulliparous.

On examination she was tachypnoeic with a respiratory rate of 32/min, bilateral Pedal edema extending up to the knees, Jugular venous pressure was raised. Radial Pulse in her right upper limb was 96 beats per minute, low volume and distal pulses was found to be absent in left subclavian ,axillary, brachial &radial arteries and both lower limbs . Blood pressure recorded in right upper limb was 162/100 mm hg and was not recordable in the other three limbs. Superficial temporal, common carotid pulsations were equal on both sides. No subclavian or carotid bruit but bruit was heard in right flank. Examination of the cardiovascular system revealed a hyper dynamic apical impulse with loud pulmonary component of second heart sound and a pan systolic murmur in mitral and tricuspid area. Bilateral fine end inspiratory crepitations were heard at the lung bases. Abdominal examination revealed tender hepatomegaly . She had no focal neurological deficits. Fundus examination was normal. Based on these clinical findings , diagnosis of vasculitis with renovascular hypertension and congestive cardiac failure was made.

INVESTIGATION:
Laboratory investigation revealed normal hemoglobin of 10.8 mg/dl and Erythrocyte sedimentation rate of 55 mm for one hour. Blood Sugar was 86 mg/dl, Blood Urea was 20 mg/dl, Serum Creatinine was 1.2 mg/dl. Urine routine showed albumin (++), Sugar –nil / Deposits: 3-5 pus cells / 1-2 epithelial cells. Liver Function test was normal. Cardiac enzymes and lipid profile were normal. ECG showed Sinus Tachycardia with Poor progression of R waves V1-V5. X ray chest PA view showed Cardiomegaly.

Mantoux test was negative. CRP was 14.1. Rheumatoid Factor was negative. Serological test for Venereal Disease Research Laboratory (VDRL), Antinuclear antibody (ANA ) titers ,anti-ds DNA and ANCA were negative. 24 hour urine VMA was 5.2mg. Overnight Dexamethas on suppression test showed value 32 nmol/lt. Thyroid profile was normal.

Echocardiogram showed Global hypokinesia with Ejection fraction of 39% with Moderate Mitral and Tricuspid Regurgitation with Mild pericardial effusion and moderate pulmonary hypertension , with features consistent of Dilated cardiomyopathy. She was subjected to Coronary Angiogram, revealed Normal coronary arteries.

USG abdomen showed contracted Right kidney (6.9 x 3.4 cm), left kidney (10.2 x 4.4cm). USG Doppler study of renal vessels showed increased peak systolic velocity in Main right Renal artery, Parvus Tardus flow in right interlobar arteries suggestive of right renal artery stenosis. Doppler study of arteries revealed absent flow in left subclavian , brachial, monophasic flow in radial and umar arteries and reduced and monophasic flow in both femoral, popliteal,posterior tibial , dorsalis pedis arteries.

DTPA SCAN [Tc -99m diethylene –triamine –penta acetiac acid scan] showed Total GFR of 89 ml/min. Relative renal function of Left kidney was 76% and Right kidney was 24%. Reduced perfusion and reduced function of right kidney suggestive right renal artery stenosis .Near normal perfusion and function of Left Kidney with no obstruction to drainage.
Takayasu arteritis affects patients in 2nd and 3rd decade of life. Females are affected more commonly than males ratio being 2.5:12. Two stages of the disease process have been described. A systemic or “pre-pulseless” phase characterised by nonspecific symptoms followed by a sclerotic or pulseless phase during which vascular insufficiency develops with diminished pulses, especially in the upper limbs and bruit over diseased arteries. The most common presenting vascular symptoms were claudication (35%), reduced or absent pulse (25%), carotid bruit (20%), hypertension (20%), carotid tenderness (20%) light headedness (20%), and asymmetrical arm blood pressures (15%).

In Takayasu Arteritis, the myocardium can be affected secondary to Systemic Hypertension or involvement of the coronary arteries, valves or pulmonary arteries. Our patient was a young female who presented with congestive cardiac failure, Dilated Cardiomyopathy and pulmonary artery involvement. Unusual presentation in takayasu arteritis is congestive cardiac failure, which constitutes only 2%. She also had rare finding of pulmonary artery involvement which is seen in 5% of cases only.

American College of Rheumatology formulated Criteria for diagnosis of Takayasu’s Arteritis

The presence of three or more of the six criteria is sensitive (91%) and specific (98%) for the diagnosis of Takayasu’s arteritis

Onset before age 40 yr
Limb claudication
Decreased brachial artery pulse
Unequal arm blood pressure (>10 mm Hg)
Subclavian or aortic bruit
Angiographic evidence of narrowing or occlusion of aorta or its primary branches, or large limb arteries

As per American college of rheumatology criteria for Takayasu arteritis, our patient had 3 out of 6 criteria.

New angiographic classification of Takayasu arteritis, Takayasu conference (Tokyo)19948

Type I involving the aortic arch and its branches,
Type II a involving ascending aorta ,aortic arch and its branches ,
Type II b involving ascending aorta , aortic arch, and its branches, thoracic descending aorta
Type III involving thoracic descending aorta, abdominal aorta and or renal arteries.
Type IV involving abdominal aorta and or its renal arteries.
Type V involving the combined features of type II b and IV.

According to this classification system, involvement of the coronary or pulmonary arteries should be designated as C (+) or P (+), respectively.

Congestive cardiac failure as intial presentation in Takayasu arteritis is rare, constituting only 2% of cases, as in our case. Congestive heart failure and renal failure are the most common causes of death. Aortic regurgitation is important because it frequently progresses and may lead to left ventricular dilation with secondary mitral regurgitation and congestive heart failure. Angina can develop as a result of coronary artery disease. Takayasu Arteritis of the coronary arteries most often produces ostial lesions but can also produce either diffuse vasculitis of the coronary arteries or aneurysms. Myocarditis also causes potentially reversible congestive heart failure. Pericarditis is very rare. Although Takayasu Arteritis (TA) of the pulmonary arteries is rare, Pulmonary involvement in Takayasu Arteritis (TA) is usually secondary to vasculitis of the large- or medium-sized pulmonary arteries.

Discussion:
Takayasu arteritis is a granulomatous systemic idiopathic disease of the aorta and its primary branches that may lead to segmental stenosis, occlusion, dilatation or aneurysm formation. Mikito Takayasu first described Takayasu arteritis or pulseless disease, in 1908. The exact pathogenesis of the arteritis unknown. Though tuberculosis, streptococcal infections, rheumatoid arthritis and other collagen vascular diseases had been debated for its etiology in the past, recently more emphasis has been given on immunopathological cause.
The most characteristic findings of pulmonary artery involvement are stenosis and the occlusion of lobar and/or main pulmonary arteries. Pulmonary artery involvement is seen in 5% and correlates with the degree of involvement of brachiocephalic vessels.

The aim of treatment is to relieve inflammation in the arteries in order to induce and maintain prolonged remission and minimise damage or treatment-related morbidity. Prednisone is usually given at 1 mg/kg daily for 1–3 months and later tapered to an alternate day schedule. Medication should be continued for at least 1 year following clinical remission. Methotrexate (0.3 mg/kg/week), cyclophosphamide (2 mg/kg/day), and mycophenolate mofetil (2 g/day) orally combined with corticosteroids have been used. TA is the form of vasculitis most frequently requiring revascularization procedures.

Percutaneous angioplasty, surgical reconstruction and repair or bypass grafting of stenosed arteries are among the surgical procedures most commonly used in Takayasu Arteritis (TA).

**Conclusion:**

This case is presented because of rare presentation of takayasu arteritis as congestive cardiac failure and dilated cardiomyopathy. This case highlights the enigmatic nature of Takayasu arteritis, and reiterates that a high index of suspicion is required in clinical practice to make an early diagnosis. So early diagnosis and prompt treatment of takayasu arteritis will provide patients better quality of life.

**References**