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Case Report

Young male with cardio-vascular complication: culprit Takayasu arteritis

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ABSTRACT

Takayasu arteritis (TA), a rare type of granulomatous vasculitis characterized by inflammatory changes and stenosis in large and medium sized arteries with a strong predilection for the aortic arch and its branches, usually presented in young female. We are reporting a rare case of TA in 23-year-old male with cardiovascular complication in form of heart failure and hypertension with involvement of right supraclavicular artery and abdominal aorta in form of aneurysm and stenosis, respectively.

Keywords: TA, Severe LV dysfunction, DCM, Hypertension

INTRODUCTION

Takayasu arteritis (TA), a rare type of granulomatous vasculitis characterized by inflammatory changes and stenosis in large and medium sized arteries with a strong predilection for the aortic arch and its branches, usually presented in young female.[1] A rare case of TA with cardio-vascular complications in young male is being reported.

CASE REPORT

A 23-year-old male patient, diamond worker attended casualty for complain of dyspnoea on exertion and easy fatigability for 10 days.

On examination patient has discrepancies of blood pressure (Rt-180/20 and Lt-80/60) and weak pulse in left upper limb with pulsatile swelling in right supraclavicular region and patient has grade V systolic murmur in mitral area on auscultation.

Blood investigation s/o Hb: 4.5 gm%, TLC: 12900 cells, platelets: 6.24 Lakhs, S. Creat: 0.54 mg/dL, ESR: 73 mm, CRP: 18.5 mg/dL, IL6: 185.18 pg/dL, procalcitonin: 0.1 ng/dL, protein electrophoresis s/o hypergammaglobulinemia with hypoalbuminemia and ANA profile: Neg.

Chest X-ray showed calcified aortic changes and USG neck s/o dilatation of 4 cm segment of right subclavian artery with alteration of normal triphasic wave form pattern. 2D echo s/o DCM, EF 30% with severe LVD and concentric LVH, dilated aortic root and proximal aorta, moderate MR with mild pericardial effusion. CT aortogram showed aneurysmal dilation of right proximal subclavian artery with abrupt narrowing with heterogeneously enhancing wall thickening and calcification in descending aorta from D10 level and abdominal aorta leading to partial occlusion and celiac trunk occlusion S/o TA with vascular complications.

As per clinical findings, blood reports, radiodiagnosis and Ishikawa criteria (1 major and 4 minor with Obligatory criterion) diagnosis of TA was made.

Patient was transfused with 4 units of RCC and started on tab. prednisolone 1 mg/kg and tab. methotrexate with

treatment of Heart failure and hypertension after consulting cardiologist and rheumatologist.

Patient symptomatically improved with treatment and was advised regular follow up on discharge.

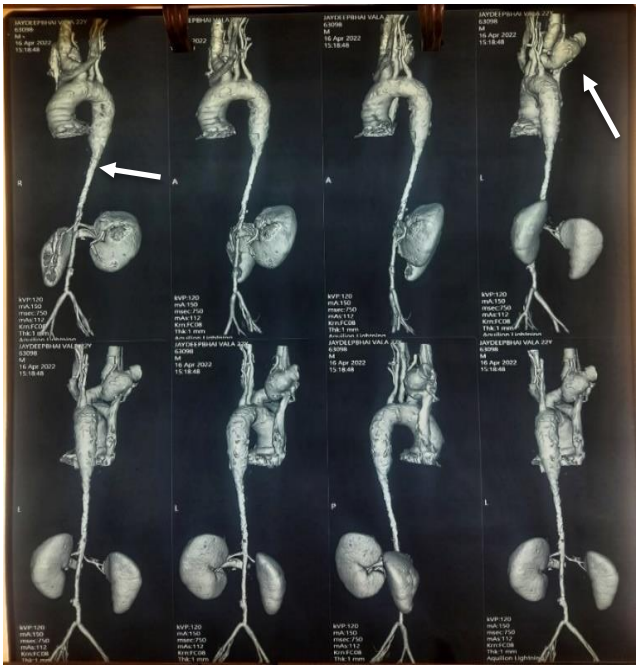

Figure 1: CT aortography with 3D reconstruction showing aneurysmal dilatation of right subclavian artery and abrupt narrowing of descending aorta (white arrows).

DISCUSSION

TA was first described by MIKITO Takayasu, which later on become popular as "pulseless disease."[2] TA is a rare type of granulomatous vasculitis characterized by inflammatory changes and stenosis, occlusion or positive remodelling in form of dilatation/Aneurysm in Large & medium sized arteries. It has strong predilection for the aortic arch and its branches, typically in young before 40s with female-to-male ratio of up to 10:1.[3]

Presentation is typically nonspecific and initially present as fever, carotidynia, malaise, anorexia, night sweat and weight loss. In chronicity limb claudication, BP discrepancies, hypertension, abnormality of pulse and arterial bruit develops. May complicate as heart failure, aortic regurgitations, limb ischemia, aortic stenosis, aneurysm, stroke, hypertension, IHD, MI or myocarditis.

Arteritic lesions demonstrate adventitial thickening and focal leucocytic accumulation in the media with intimal hyperplasia. Growth factor derived mesenchymal cell proliferation leads to intimal hyperplasia and fibrosis and subsequent arterial stenosis or occlusion.[3]

Diagnosis of TA depends principally on the physician including the disease in differential diagnosis with index of suspicion, must be high in young patients with discrepancies in blood pressure and unexplained acute phase response and hypertension.[3] Abnormal lab finding include CBC-Variable degree of anaemic, elevated ESR and CRP, protein electrophoresis (High globulin and low albumin levels). CT aortogram may show stenosis/occlusion. MR angiography is also helpful. Doppler USG of affected artery may show halo sign. Digital Subtraction angiography and 18-FDG PET are advance modalities for diagnosis.

In diagnostic criteria Ishikawa criteria is used.[4]

2 major / 1 major + 2 or more minor / At lease 4/9 minor criteria used.

Obligatory criterion-Onset of disease <40 years/at onset of characteristic signs and symptoms of 1 month duration

Major criteria included left mid-SCA lesion, right mid-SCA lesion.

Minor criteria includedRaised ERS, carotid artery tenderness, hypertension, aortic regurgitation or annulo-aortic ectasia, pulmonary artery lesion, left mid-common carotid lesion, distal brachiocephalic trunk lesion, descending thoracic aorta lesion, abdominal aorta lesions.

Treatment includes oral steroid, non-steroidal immunosuppressants (Methotrexate/ MMF/ azathioprine), Tocilizumab, anti-TNF therapy and other symptomatic therapy and treatment of complications. Once vascular inflammatory process under well control surgical management is advised if needed.[5-6]

CONCLUSION

TA should be considered in a young male patient with discrepancies in blood pressure, fatigue and dyspnoea. As finding are nonspecific and due to which these types of cases are often remain undiagnosed, clinician must consider diagnosis of TA in differential diagnosis. This case illustrates the consequences of delayed diagnosis.

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