

Rare Case of Takayasu's Arteritis with Severe Aortic Regurgitation: a Case Report

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ABSTRACT:- Takayasu's arteritis occasionally called as pulseless disease is a rare chronic inflammatory condition which usually affects large blood vessels in the body and its branches . It usually occurs in young women of age below 40 with a female to male ratio as 8:1. We are presenting with a rare and interesting case of woman aged 29 years with a past medical history of Coronary artery disease and hyperthyroidism since 3 years, unstable recurrent angina, syncopal attack, acute aortic regurgitation, anemia and paratracheal lymphadenopathy. Now, the patient presented complaints of recurrent on and off chest pain for 2 days, arm claudication and chest heaviness. Her diagnostic workup revealed multiple manifestations supporting Takayasu's arteritis.

KEYWORDS:- Takayasu's arteritis, Aortic regurgitation, Aortic aneurysm, Percutaneous Transluminal Coronary Angioplasty, Hypokinesia

I. INTRODUCTION

Takayasu's arteritis is a well known yet a rare form of large vessel vasculitis. It is also called as pulseless disease with chronic inflammation of arteries affecting large vessels like aorta and its main branches. Vessel inflammation leads to wall thickening, fibrosis, stenosis and thrombus formation. (1)

Majority of cases are observed in Asia, Africa, Latin America. Due to the poor prognosis patients often experience claudication, absence of pulses, hypertension, MI, Cerebrovascular accidents. It is usually associated with granulomatous panarteritis of aortic arch and its primary branches ascending aorta, thoracic descending aorta and abdominal aorta. It is usually idiopathic, but a recent study in Turkish TA patients showed the influence of HLA-B*52 in the disease progression. This might indicate the role of genetic factors in the disease severity. (2) TA is characterized by T-Lymphocyte, monocyte and macrophage infiltration of the arterial wall, leading to intimal myofibroblast proliferation and fibrosis of the media and adventitia. (3)

This report is on a 29 year old woman who presented with unstable recurrent angina, arm claudication and syncope. She also has a past medical history of CAD and Hyperthyroidism. A detailed careful examination, appropriate laboratory and diagnostic tests were performed to determine this case of Takayasu's arteritis. The main finding was pulse discrepancy of 20mm Hg between upper limbs, vascular bruits and anginal symptoms.

II. CASE REPORT

A 29 year old Asian woman was admitted to our hospital because of left sided chest pain, claudication of arms and several formless symptoms like malaise, weakness and fatigue. Symptoms had presented occasionally since she was 26 years old. A relapse of these symptoms occurred 1 week before admission. Three years ago, patient had undergone Percutaneous Transluminal Coronary Angioplasty to Left main coronary artery ostium with placement of primary stent. Thus she was on anticoagulants and anti-platelets aggregatory drugs. She was also on anti-hyperthyroids.

Present physical examination showed palpitation, a remarkable blood pressure discrepancy of 20mmHg between right and left arm. Blood pressure was undetectable in the left arm, but was 80/60 mm Hg in the right arm. Both left radial and brachial pulses were impalpable. She was afebrile with tachycardia. Pallor and lymphadenopathy was present. CVS-S1, S2 heard, S1 Normal, no S3, S4. Chest breath sounds were bilaterally equal. No creps or wheeze. No intra-cranial pathology was found.

ECG showed normal sinus rhythm, Left ventricular hypertrophy, T wave inversions from V2 -V6. Chest X-Ray was normal. ECHO test showed severe aortic regurgitation, LVEDD/ESD: 46/30, LVEF:63%, RWMA: Hypokinesia of mid apical septum. CT-Thorax showed significant wall

calcification involving distal aortic arch and thoracic descending aorta. Maximum diameter of the descending aorta is 3.7cm. Multiple mildly enlarged lymph nodes noted in right paratracheal region. Largest measures 1.7*0.3 cm. No cardiomegaly or pleural effusion.

Angiogram-thoracic and abdominal aortogram shows normal contrast opacification. No evidence of dissection. Doppler ultrasound of left ventricular artery-Shows mild luminal irregularities of sub-clavian artery and proximal axillary artery. Doppler shows spectral broadening. Angiographic diagnosis showed normal LAD (Left Anterior Descending artery), LMCA-short.

Blood investigations showed Hb-9.7,WBC-11,400,Platelets-3.25,Sr.Creatinine 0.7, Sodium-141,Potassium-3.8,SGPT-25,SGOT-19,FBS-70,Total cholesterol-112.Stat high sensitive Trop-I was 14.8. TFT shows T3-13.4, T4-2.5,TSH-0.064.

She was primarily given with Inj.Efcorlin, Inj.Tramadol, T.Clopilet,T.Aztor,T.Ecospirin as stat medicines. She was then treated with anti-anginals including T.Nicorandil, C.Cytogard, T.Imdur. Immunosuppresants and Corticosteroids were initiated for the management of Takayasu's arteritis. T.Wysolone(prednisolone) which is a glucocorticosterid and T.MMF(Mycophenolate mofetil) which is an immunosuppresant was prescribed.Since the patient was anemic T.Livogen was given.Routine use of low dose aspirin was advised.

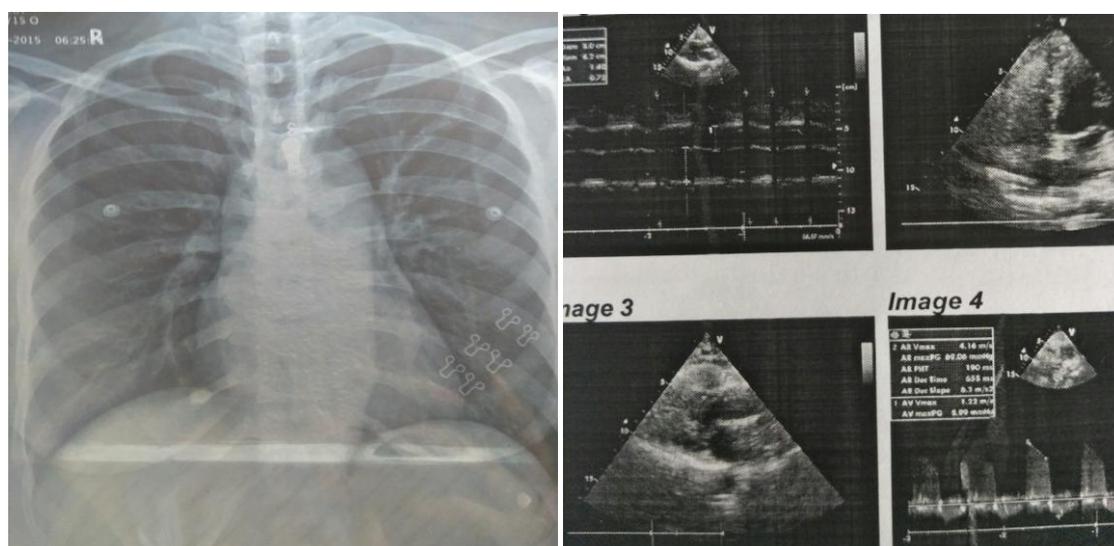


Fig:image showing X-ray and echocardiogram respectively

III. DISCUSSION

Takayasu's arteritis is a large vessel vasculitis characterized by granulomatous inflammation of the vessel walls with an unknown etiopathogenesis.(4). It occurs most commonly in young females.Since its a rare condition, its epidemiological data is limited.Though the disease distributed worldwide, its more common in Asians.The incidence of TA was estimated to be 1-2/million in Japan and 2.2/million in Kuwait.(5) American Rheumatological society considers 3 of the following 6 criteria for a definite diagnosis of Takayasu arteritis.

1. Onset before 40 years
2. Claudication of the extremities
3. Decrease in the brachial pulse in one or both arms
4. Difference of 10 mmHg or more in BP measured in both arms
5. Audible bruit on auscultation of the aorta or subclavian artery
6. Narrowing at the aorta or its primary branches on arteriogram (6)

Our patient met with 5 out of 6 criteria.

Clinical course of the disease is divided into early active inflammatory phase and late chronic phase.It is characterised by systemic disease which last for weeks to months and has a remitting and relapsing course.Late chronic phase is the result of occlusion or atrial stenosis ,ischemia of organs

Based on vessel involvement,TA can be classified into 4 types

Type I involves the aortic arches and its branches

Type II involves thoraco-abdominal aorta and its branches

Type III involves vessels of both type I and type II

Type IV involves pulmonary arteries.(7)

Cardiac features are present in 40% of patients. Even though the patients don't have risk factors of atherosclerosis, they present with atheromatous aorta.

Medical management varies according to the severity of disease. Two most important criteria for the treatment are controlling inflammatory process and hypertension. In active disease, pharmacological treatment with corticosteroids(1mg/kg/day) are preferred till remission is achieved. For those who don't achieve remission with corticosteroids immunosuppressants such as methotrexate (0.15-0.35mg/kg/week),cyclophosphamide (1-2mg/kg/day) and azathioprine (1-2mg/kg/day) are considered. Combination of the above is given if a relapse happens again.(8)

Angioplasty of the aorta with or without stenting is successfully proven.

In regions of severe stenosis or occlusion, graft has been used to bypass the area of occlusions.(9)

Percutaneous Transluminal Coronary Angioplasty is performed in few cases of severe stenosis. Information about the outcome is limited. The success of angioplasty and stenting has made surgery a less preferable approach except in cases of large aneurysm and undilatable fibrotic stenosis.(10)

In 25% of cases,aortic regurgitation has significantly resulted in worst outcomes.In some cases, surgical intervention with valve or root replacement is considered.But,its difficult to manipulate cases with fragile and inflamed tissue which can lead to valve detachment or anastomatic aneurysm formation. Thus, we wait for the reduction of vascular inflammation using immunosuppressive therapy before opting for surgery.(11)

IV. CONCLUSION

This case report emphasize on the diagnostic and therapeutic methods of Takayasu's arteritis. From the above data, we conclude that its a rare disease, most commonly seen in Asian young women.

Poor prognosis of this condition can lead to further complications like aortic aneurysm.In cases of severe aortic regurgitation, surgery is preferably delayed until reduction of vascular inflammation is achieved with immunosuppressive therapy.

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