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Cardiovascular Manifestations of Takayasu's Arteritis: A Case Study of Severe Aorto - Arteritis Type IIA

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Abstract: ***Background: 1) Takayasu arteritis is an inflammatory and stenotic disease of medium & large sized vessel with strong predilection for aortic arch. 2) It is caused by intimal proliferation, fibrosis, scarring of arterial tunica media results in blood flow compromisation of involved organs. Case Presentation: A 30 year old female presented with 2 months history of continuous headache associated with dizziness, devoid of phonophobia, photophobia, fever, weight loss, chest pain and palpitation. 2) On general examination, pulse was feeble in left upper limb with radio - radial delay and feeble pulses in both lower limbs with discrepancy of blood pressure in all four limbs. 3) On cardiovascular system examination, grade 3/6 systolic murmur heard at aortic & pulmonic area, radiating to bilateral carotid artery and precordium and continuous murmur heard at inter - scapular area. 4) 2D - echo suggestive of severe juxtaductal coarctation of aorta with Gmax of 79mmhg. 5) CT aortogram suggestive of severe narrowing in aortic isthmus 20mm distal to left subclavian artery, severe osteal stenosis involving left common carotid and subclavian artery. 6) Patient was treated with revascularization procedure (PTA) & anti - inflammatory therapy with glucocorticoid and mycophenolate mofetil. Conclusion: 1) Coarctation of aorta was suspected with thorough clinical examination and early diagnosis was possible even in the absence of typical features of vasculitis that makes favourable outcome.***

Keywords: Takayasu arteritis, Coarctation of aorta, Radio - radial delay, Murmur, Revascularization procedure

1. Introduction

- Takayasu arteritis is an uncommon disease with an estimated annual incidence rate of 1.2 - 1.6 cases per million most prevalent in adolescent girls.
- It is an inflammatory and stenotic disease of medium & large sized vessel with strong predilection for aortic arch.
- It is caused by intimal proliferation, fibrosis, scarring of arterial tunica media, results in blood flow compromisation of involved organs.

2. Case Description

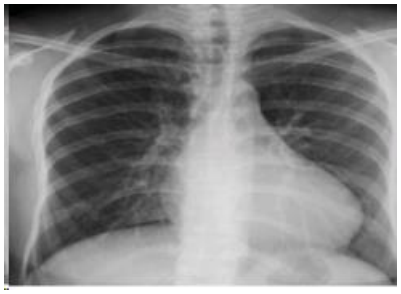
- A 30 year old Hindu, married female presented with 2 months history of continuous headache associated with dizziness, devoid of phonophobia, photophobia, loss of consciousness, fever, weight loss, chest pain, palpitation and breathlessness.
- Her family history, past history and personal history was not significant. Her obstetric history was G2P2A0L2.
- General examination was suggestive of feeble pulses in left upper limb with radio - radial delay, normal carotid pulse and feeble pulse in both the lower limbs with discrepancy of blood pressure in all four limbs.
- Right upper limb (180/80mmhg) Left upper limb (140/100mmhg)
- Right lower limb (110/70mmhg) Left lower limb (100/64mmhg)
- Patient was fairly built and well nourished, with body mass index of 22.5 and arm span is 150cm.

Crdiovascular System Examination

- Inspection: Apex impulse was visible at 5th intercostal space lateral to mid - clavicular line. Visible suprasternal pulsation was present. No precordial bulging, dilated veins and scar marks in chest and back.
- Palpation: Heaving apex, apex beat at 5th intercostal space lateral to mid - clavicular line. No palpable thrill or parasternal heave noted.
- Percussion: Normal
- Auscultation: S1S2 were normal. Significant finding was grade 3/6 ejection systolic murmur at aortic & pulmonic area with radiation to bilateral carotid artery and precordium. Continuous murmur heard at inter scapular area.
- Rest other systems examination were normal.

Investigation

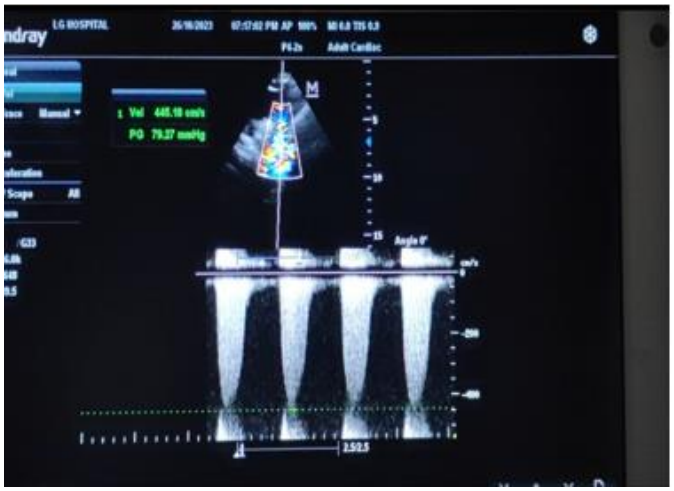
Haemoglobin – 10.4
MCV - 74
Creatinine - 0.89
Potassium – 4.2
Trop I - 0.001
ESR - 120
CRP – 4.8
Urine examination - Normal
USG Kidney – Normal size with preserved cortico - medullary differentiation
Fundus examination – Normal
Electrocardiogram – Suggestive of left ventricular hypertrophy
X - ray – Suggestive of irregular notching of inferior margin of posterior ribs

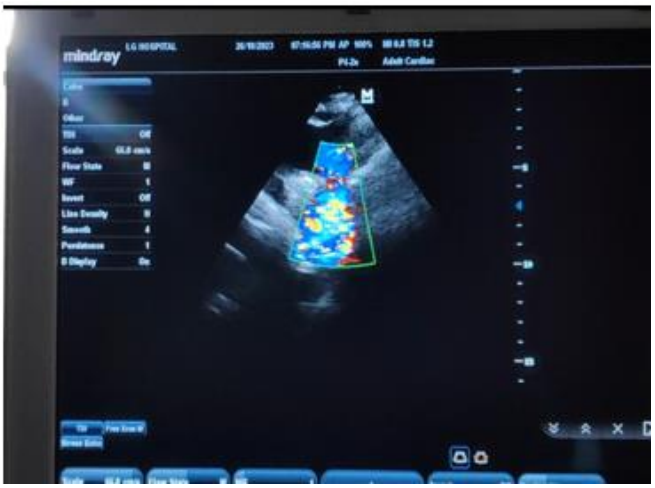


Specific Investigations

2D Echo: Left ventricular hypertrophy without any RWMA, Normal left ventricular function with normal valves. Severe juxtaductal coarctation of aorta with Gmax (79mmhg), Gmin (41mmhg), Vmax (4.3), AVVTI (92 cm/s) C. W/P. W/Doppler: Trivial MR, Trivial TR, Trivial AR.

Continuous wave doppler Colour doppler

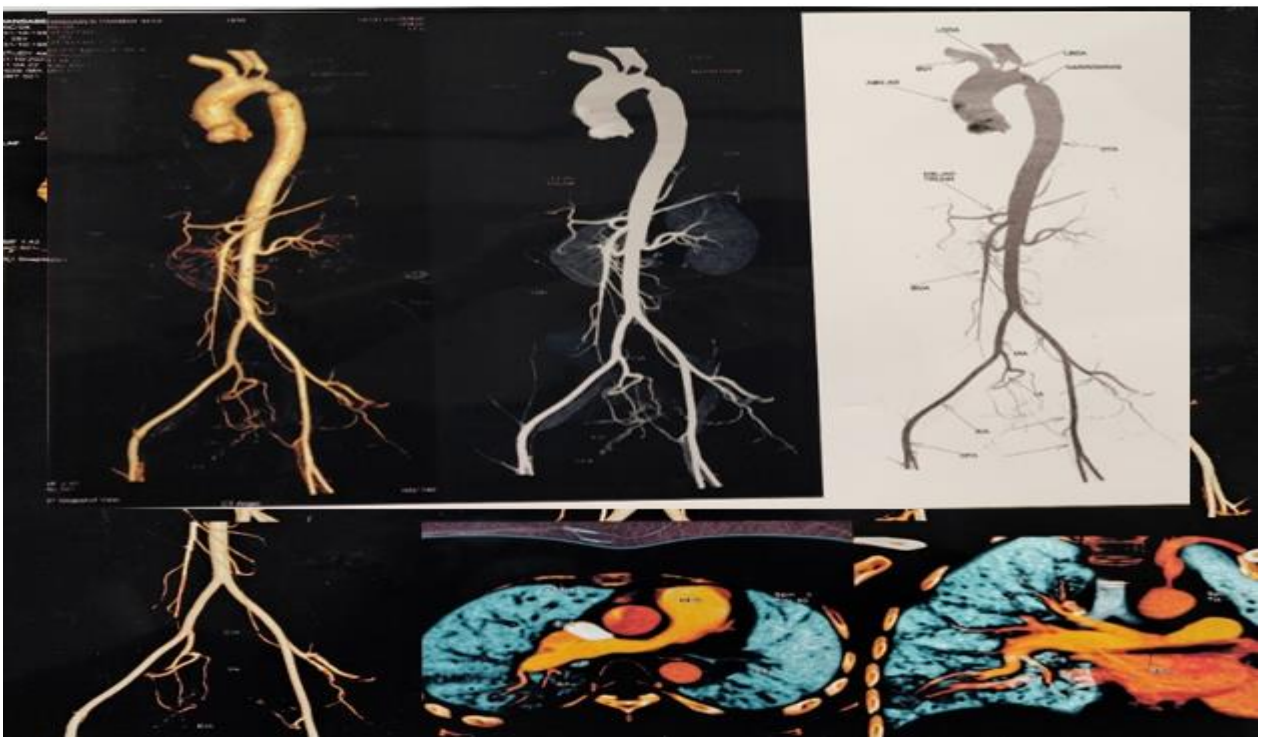




The coarctation of aorta was our provisional diagnosis so CT - Aortogram was done to confirm the diagnosis.

CT Aortogram:

1) Ascending Aorta - Normal proximally with increased intimo - medial thickness in distal aorta. No luminal narrowing.
2) Arch - Circumferential increased intimo - medial thickness involving entire arch and isthmus resultant severe liminal narrowing. (Approximately 20mm from left subclavian artery origin)
3) Arch vessels –
   Left Common carotid artery – Severe (80 - 90%) osteal stenosis due to increased intimo - medial thickness (~5mm).
   Left subclavian artery – Severe (80 - 90%) osteal stenosis due to increased intimo - medial thickness (~4mm).
4) Left pulmonary artery – Complete chronic occlusion of left pulmonary artery from origin with collateral reforming branches from hilar level.



All this findings were suggestive of AORTO - ARTERITIS P/O TAKAYASUS AORTO - ARTERITIS TYPE IIA with pulmonary artery.

Treatment

Patient was treated with PTA (percutaneous transluminal angioplasty) and glucocorticoids and later with steroid sparing agent mycophenolate mofetil.

3. Discussion

Takayasu arteritis is an inflammatory and stenotic disease of medium and large sized arteries characterized by a strong predilection for the aortic arch and its branches; pulmonary artery may also be involved.

The disease is a pan arteritis with inflammatory mononuclear cell infiltrate and occasionally giant cell. There is marked intimal proliferation and fibrosis, scarring and vascularisation of media, disruption and degeneration of elastic lamina.

Takayasu arteritis is systemic disease with generalized as well as vascular symptom. Generalized symptoms are fever, malaise and night sweat. Vascular symptoms are according to involvement of particular vessels.

There is an angiographic classification of Takayasu arteritis mentioned below:

Type	Vessel involvement
Type I	Branches from the aortic arch
Type II a	Ascending aorta, aortic arch and its branches
Type II b	Ascending aorta, aortic arch and its branches, thoracic descending aorta
Type III	Thoracic descending aorta, abdominal aorta, and/or renal arteries
Type IV	Abdominal aorta and/or renal arteries
Type V	Combined features of types II b and IV
Involvement of the coronary or pulmonary arteries should be designated as C (+) or P (+), respectively	

The combination of glucocorticoid therapy for acute signs and symptoms and arterioplastic therapy for stenotic vessel has markedly improving outcome and reduced morbidity.

Complication of this disease include aortic aneurysm, cerebral aneurysm, hypertension, cardiomyopathy and stroke.

These individuals need prophylaxis for endocarditis if they undergo any invasive procedures.

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