TAKAYASUS ARTERITIS COMPLICATING PREGNANCY-A CASE REPORT
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Abstract: Takayasu arteritis is a rare disease involving chronic inflammation of large vessels Aorta and its branches, with their predilection towards young women of oriental origin (1). The uniform global incidence is 1-2 million. Females make up 80-90% of patients with this disease (2). We present here a rare case of Takayasu arteritis in pregnancy who suffered due to its complications, identified, diagnosed, treated and recovered.

Keyword: Takayasu arteritis, Pulseless disease, Aortoarteritis, panvasculitis.

INTRODUCTION:
Takayasu's Arteritis is an uncommon panvasculitis involving chronic inflammation of larger arteries in the body, leading to narrowing, occlusion and aneurysms of the systemic and pulmonary arteries (1). It predominantly affects young females of reproductive age group. (M:F ratio 1:8) (2).

The disease was first described by two Japanese scientists, Takayasu and Onishi in 1908 (2). The evolution of the disease is not affected during pregnancy; however, one should be careful with the peripartum conditions of these patients, since they can develop complications such as hypertension, cerebral ischemia, near complete occlusion of systemic and pulmonary arteries, CCF, IUGR etc. (1). Since aorta and its branches are most commonly involved, this is also called as "Aortoarteritis", "Pulseless disease", "Aortic arch syndrome".

CASE REPORT:
A 26 years old postnatal mother P1L1 delivered by Labour Natural referred to our institution on second postnatal day as a case of acute renal failure with absent pulses in both upper limb, suspected Takayasu’s Arteritis for further evaluation and management. Patient gives history of decreased urine output for two days. H/o intermittent pain in both upper limb.

Past history:
At 16 years of age patient had low grade fever for which she went to a private hospital, informed that her pulses are absent in both upper limb and she needed further evaluation. But patient didn’t turn up then. Obstetric history:
She didn’t attend her routine antenatal visits. Not immunised. All three trimester said to be uneventful. Advised evaluation at tertiary centre. Patient didn’t turn up.

Examination:
On admission here, detailed general examination and abdominal examination was done. Radial, Brachial pulses absent - in both upper limb. Right side carotid pulse - feeble with bruit, thrills and pulsation. BP recorded in left lower limb at ankle found to be systolic 100mmHg.

Nephrologist, Cardiologist and Physician opinion obtained. Patient was anaemic with haemoglobin of 8.9gms/dl. Blood urea-110, serum creatinine- 5.4, platelet count-32,000, serum uric acid-11.4, liver enzymes mildly elevated, Proteinuria of 1+ seen. Suspicious of HELLP syndrome. ESR and C-reactive protein increased. Anti-Nuclear antibody – negative. Peripheral smear showed picture of microangiopathic haemolytic anemia. Ultrasound of abdomen showed Grade 1 medical renal disorder with minimal pleural effusion. Ultrasound Doppler of both upper limb showed- stenosis of bilateral common carotid artery origin and stenosis of bilateral subclavian artery.

CT-Angiogram revealed 1. complete occlusion of Subclavian artery on right side, 2. diffuse severe stenosis (70% diameter narrowing) of Common carotid and Subclavian artery on left side, (fig 1, fig 2) 3. moderate pleural effusion and splenic abscess. Echocardiogram showed normal ejection fraction of 67%, normal aortic valve roots and no aneurysm nor dilatation. Normal cardiac status. Fundus examination of eye showed left eye ocular ischemic syndrome secondary to left carotid insufficiency with pre retinal haemorrhages and tractional retinal detachment.

Treatment: Patient underwent five cycles of hemodialysis for acute renal failure, recovered with normal urea and creatinine levels. She was transfused with one unit of packed cells and three units of platelet. Hemoglobin and platelet count returned to normal after few days. She was treated with aspirin, atorvastatin, phenytoin for brain ischemic lesions and glucocorticoids for vasculitis.
The disease is a panarteritis with inflammatory mononuclear cell vasculitis with more predilection towards aorta and its branches.

Takayasu's arteritis is a chronic granulomatous necrotising DISCUSSION:

fig 2 -CT ANGIOGRAM

manifestations (erythema nodosum, ulcers). (3) (6) (anorexia, nausea) and skin (dyspnoea, haemoptysis and pleurisy), gastrointestinal symptoms and symptoms such as malaise, arthralgia, mild synovitis, weakness, myalgias, weight loss and low grade fever.

PULSELESS PHASE:

Types Anatomical location
1 - Aortic arch and its branches 2 - Thoracic aorta and abdominal aorta 3 - Combination of 1 and 2 4 - Pulmonary artery Symptoms come under the following phases: (3) EARLY PHASE/ PRE -PULSELESS PHASE: Non specific symptoms such as malaise, arthralgia, mild synovitis, weakness, myalgias, weight loss and low grade fever.

occlusive phase:

usually involves common carotid artery (visual defects, stroke, TIA), vertebral artery (dizziness, visual defects), subclavian artery (arm claudication), aorta (aortic regurgitation, CCF), renal artery (hypertension), iliac artery (claudication). (3) (6) Most cases are diagnosed in the occlusive phase.

DIAGNOSTIC CRITERIA: (3)

Diagnosis of Takayasu is largely based on the combination of 1. clinical manifestation, 2. laboratory evaluation, 3. diagnostic imaging. The gold standard investigation for confirming Takayasu is CT-Angiogram or MRI-angiogram. The patient must meet 3 out of the following 6 criteria of American college of Rheumatology classification to be diagnosed as Takayasu's Arteritis,

1. Age less than 40 years
2. Claudication of extremities
3. Decreased brachial artery pulse
4. BP difference of >10 mmHg between both arms
5. Bruit over subclavian/carotid/aorta
6. Arteriogram abnormality: occlusion or narrowing in aorta and its main branches.

GRADES OF SEVERITY OF DISEASE: (2)

I No complication
IIa one complication (mild)
IIb one complication (severe)
III two or more complications

Overall maternal mortality rate of women with Takayasu's arteritis is 4-8% and further pregnancy is not advised in group IIb & group III. COMPLICATIONS: (2, 5)

arterial hypertension with superimposed pre-eclampsia and its sequelae such as eclampsia, HELLP syndrome

_ congestive cardiac failure
_ cerebrovascular events
_ retinopathy etc
_ Fetal risks- IUGR, IUD, Prematurity

LABORATORY FINDINGS:

Elevated ESR, raised CRP, mild anemia, elevated immunoglobulin levels. (4)

The investigation of choice for the diagnostic evaluation of Takayasu is - CT or MRI Angiography(3) (4). Echocardiography & duplex Doppler ultrasonography are the first investigations which shows wall thickening, stenosis, calcifications, occlusions and pulsatility. (7)

CT-angiography and MRI-angiography show concentric wall thickening, aneurysms, thickened aortic valvular cusps, signal alterations within and adjacent inflamed vessels, mural thrombi (7)

CONCLUSION:

The patient presented here comes under Type I classification (involving aortic arch branches) and grade III severity, since the patient had more than one complications (acute renal failure, brain infarct, retinopathy).

Glucocorticoids being the first line of treatment in Takayasu's arteritis, patient was treated with prednisolone for four weeks. Patient had renal failure for which she underwent five cycles of hemodialysis. She was also transfused with packed red cells and platelets. For brain infarct she was put on aspirin, atorvastatin, phenytoin. Patient responded well to all these treatment, recovered from the complications and discharged on 42nd post natal day. She was advised no further conception and to adopt contraception. She was also emphasized on periodical follow up with continuation of steroids.

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