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Takayasu Arteritis in Pregnancy- A Case Report

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Abstract:
Takayasu arteritis (TA) is a rare, progressive, idiopathic vasculitis of aorta and its main branchesleading tostenosis, occlusion and aneurysm of the involved arteries. It is prevalent among young women in the reproductive age group. TA has an adverse effect on pregnancy outcome and there is an increased risk of complications. The overall prognosis is good when it is diagnosed early and patients receive regular antenatal follow up to prevent and manage the complications and to monitor the feto-maternal well being.Adopting a multidisciplinary approach for the management has shownimprove maternal and fetal outcome. We described here a case of pregnancy with TA involving the descending thoracic aorta and abdominal aorta with significant left subclavian artery stenosis and mild bilateral renal artery stenosis.

Keywords: *Takayasu arteritis, Vasculitis, pregnancy, maternal outcome, fetal outcome, complications*

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I. Introduction:

Takayasu arteritis, also known as pulseless diseaseis a systemic, chronic inflammatory arteriopathy involving the large arteries. It is rare and predominantly seen in women of Asian origin, in the second and third decades of their life[1]. About 80-90% of the cases are reported in women of child-bearing age, hence it is sometimes referred to as 'young female arteritis'.

The worldwide incidence is about 1.2-2.6 cases/million per year[2].

Itprimarily involves the aorta and its majorbranches like carotid, subclavian, pulmonary, iliac and renal arteries. The affected vessels demonstrate narrowing, occlusion and aneurysm of varying severity[2].

The etiology is often idiopathic, however literature suggests the involvement of autoimmune factors[3,4].

Although the available studies do not have sufficientdata to suggest that the disease worsens or improves with pregnancy, it is found that Takayasu arteritis adversely affectsthe pregnancy outcome in the form ofabortion, superimposed preeclampsia, fetal growth restriction, intra-uterine fetal demise and placental abruption[5-7].

II. Case Report:

A 25 year old $G_2P_1L_1$with 39 weeks period of gestation presented for antenatal care.

She was diagnosed with Takayasu arteritis- left subclavian artery stenosis, 5 years prior to the present antenatal visit. She had undergone percutaneous angioplasty and stenting of left subclavian artery.

She was on Azathioprine (50 mg BD), Prednisolone (5mg daily), and Clopidogrel (150mg daily) prior to conception

First pregnancy –Full term vaginal delivery, two and a half years ago, uneventful.

On examination, she was afebrile with a blood pressure of 120/70mmHg in both arms and pulse rate of 92 beats/min. Unequal radial pulsation on both sides noted.

Cardiac examination revealed normal first and second heart sounds. No bruit.

Per abdomen examination: Fundal height corresponded to term with full flanks,longitudinal lie, cephalic presentation and liquorwas clinically adequate.

Fetal heart rate- 150bpm, regular.

Investigations:

*Blood parameters:*ESR – 60mm/hr, Hb – 11.8 gm/dL, TC – 11,100 cells/mm3, Platelets- 2.17lakhs/mm^3.

2D ECHO- Normal valves and cardiac chambers with normal left ventricular function (Ejection Fraction- 62%)

CT angiogram: Circumferential thickening of left subclavian artery and left vertebral artery with significant luminal narrowing (upto 80%). Circumferential thickening of descending thoracic artery with mild luminal narrowing. Circumferential thickening of descending abdominal artery with narrowing of the lumen (upto 40%),extending upto the level of the origin of inferiormesenteric artery, beyond which the aorta showed fairly

normal caliber upto its bifurcation. The common iliac, external and internal iliac arteries showed fairly normal caliber and luminal filling.
Severe stenosis of the coeliac artery and superior mesenteric artery originnoted (upto 90%). Mild wall thickening noted in the bilateral renal artery origin (upto 40 %). The kidneys showednormal enhancement and morphology.

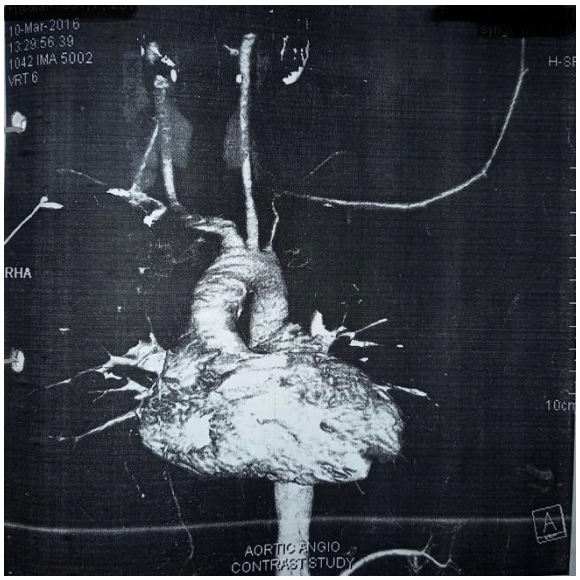


Figure 1

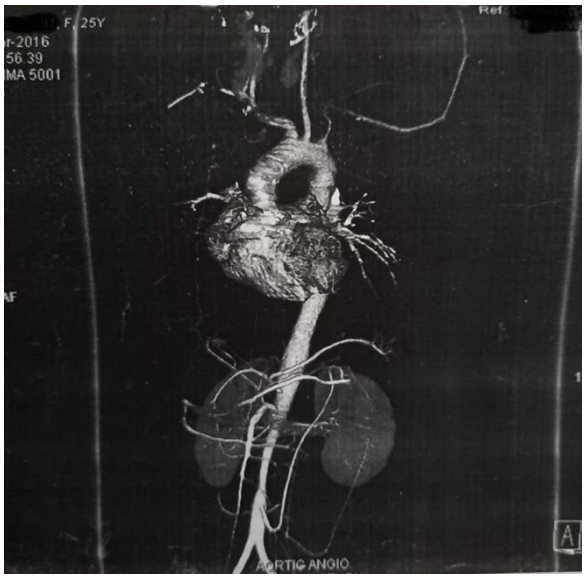
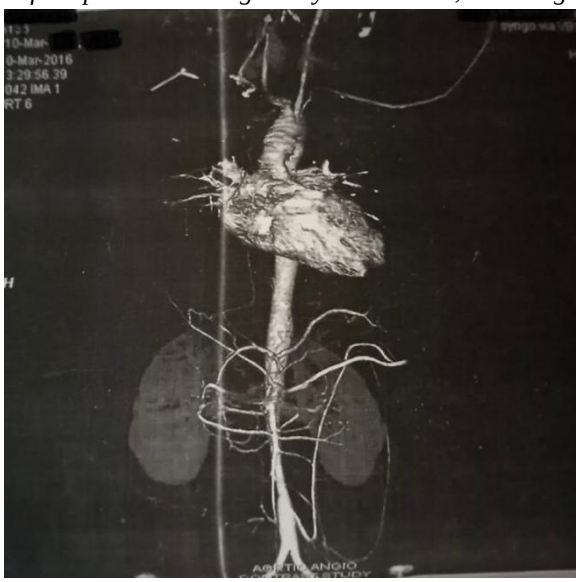


Figure 2

Figure 3

Figures 1,2,3: CT Angiogram of the patient showing Takayasu arteritis, involving descending thoracic aorta,



descending abdominal aorta.

*Management:*Multidisciplinary approach was adopted in the management. Emergency lower segment caesarean section was performed in view of non reassuring fetal status.
Postoperatively, she was closely monitored and re-started onAzathioprine and Aspirin.In addition, Enoxaparin sodium 40mg given for 5 dayspostoperatively.Post operative period was uneventful and patient was discharged.

III. Discussion:

Takayasuarteritis is a rare, chronicgranulomatous giant cell vasculitispredominantly involving the aorta and its main branches.Takayasu Arteritis was first described in 1908 by two Japanese Ophthalmologists, Takayasu and Onishi, whoobserved retinopathy and absentperipheral pulses in a 21 year old woman[1].Theexact etiology remains primarily idiopathic, but certain factors like autoimmunity, sex hormones (more common in

young females), andgenetics (demonstratedby the predisposition of the human leukocyte antigen – HLA BW52) have been implicated[8]. It is prevalent among young women in their reproductive years.

The clinical presentation is variable as it dependsupon the involved artery and the degree of stenosis.Symptoms can range from limb claudicationand numbness to vertigo, visual changes, angina, and seizures. The pathognomonicclinical signs are diminished or absent pulse (80-90%), difference in blood pressure, bruit over the carotid, subclavian or aorta (80-95%),retinopathy and hypertension[2].

The long term prognosis is usually good, with a 5 year survival rate of 90%. However, research has shown that these patients have a high prevalence of residual morbidity.Hypertension is common and is seen in about 90% of the affected[9].

According to the American college of rheumatology 1990 criteria, 3 out of the following 6 criteria should be fulfilled todiagnose Takayasuarteritis[10]

- Age under 40 at disease onset
- Claudication of extremities
- Decreased brachial arterial pulses
- Systolic blood pressure difference of more than 10mmHg between arms
- Bruit over subclavian arteries or the aorta
- Angiogram abnormalities: occlusion or narrowing ofentire aorta, its primary branchesor large arteries in the proximal upper or lower extremities.

These criteria have a 90.5% sensitivity and 97.8% specificity for the diagnosis of Takayasu arteritis.

The criteria for diagnosing active disease is as follows[13]

- Features of vascular ischemia or inflammation (such as vascular pain, claudication, diminished or absent pulses, bruit), asymmetric blood pressures in either upper or lower limbs or both
- Elevated ESR
- Systemic features, such as fever, musculoskeletalpains (without an identifiable cause).

According to the new angiographic classification of Takayasu arteritis, there are five types based on the vessel involved and angiography findings[12]

- TypeI: Aortic arch and its branches
- TypeIIa:Ascending aorta, aortic archand its branches
- TypeIIb:Ascending aorta, aortic arch and its branches, thoracic descendingaorta
- TypeIII: Thoracic descending aorta,abdominal aorta and/or renal arteries
- TypeIV: Abdominal aorta, and/or renal arteries
- TypeV: Combined features of type IIb and IV.

The involvement of coronary or pulmonary arteries should be indicated as C(+) or P(+) respectively.

In addition to the Type of disease, Ishikawa defined clinical groups based on the natural history of the disease and complications encountered. The 4 important complications considered for the grading are retinopathy, secondary hypertension, aortic regurgitation, and aneurysm, each being classified as mild/moderate or severe at the time of diagnosis. Accordingly,four grades of the disease are described as follows which are helpful in prognostic and treatment assessment[13].

- Group I: Uncomplicated disease with/without pulmonary artery involvement.
- Group IIA: Mild/moderate single complication together with uncomplicated disease.
- Group IIB: Severe single complication together with uncomplicated disease.
- Group III: Two or more complications together with uncomplicated disease.

Our patient upon presentation had Type V disease graded under Group I.

Although pregnancy hasn't shown to alter the disease progression, a 13 fold increase in obstetric complications have been observed in pregnant women with Takayasu arteritis in comparison to normal pregnancy[14].

Type I and II disease are usually associated with good feto-maternal prognosis with fewer complications.

Type III, IV, V are shown to have higher incidence of secondaryhypertension, IUGR and poor perinatal outcome.Higher incidence of IUGR can be attributed to the involvement of abdominal aorta and renal vessels.

High risk of maternal mortality is associated with complications such aspulmonary hypertension, abruption, cardiac failure, aortic aneurysm and cerebral haemorrhage[15].

In addition to the clinical criteria, Takayasu arteritis can be diagnosed with MR/CT angiography. 18 FDG PET canbe used as adjuvant diagnosing modality. However, gold standardfor diagnosis is vessel biopsy[16].

Interdisciplinary approach is needed for the management ofpregnancy with Takayasu arteritis.
Pregnancy should be planned in the remission phase with regularantenatal visits, serial monitoring of BP, renal function, cardiac status and screening for pre-eclampsia. Fetal monitoring should be done by daily kick count, growth scan, biophysical profile andDoppler studies.
The principles of treatment arecontrol of hypertension, prevention of renal failure and arterio-plastic approach to the stenosed vessels[9]. Management includes antihypertensives, antiplatelets, immunomodulatory drugs such as Tocilizumab, Leflunomide, angioplasty and bypass surgery of the affected vessels. Corticosteroids are the main stay of treatment with remission rates upto 60%[17].
Vaginal delivery is preferred in type I and IIa. Second stage of labour is cut short by instrumental delivery. Epidural anaesthesia is ideal in these women during labour and delivery as it maintains the hemodynamics.
In patients with stage IIb and III, LSCS is preferred over vaginal delivery in order to prevent cardiac decompression. Again, epidural anaesthesia is preferred over regional anesthesia as regional anesthesia is associated with sympathetic blockade and subsequent hypotension, especially in patients with stenosed arteries and compromised regional circulation[18].
The long term prognosis is good. Approximately 20% have monophasic self-limiting disease.

IV. Conclusion:

Takayasu arteritis should be considered as a differential diagnosis in a young female presenting with hypertension. Type of Takayasu arteritis affects the clinical course of the disease, management, maternal and fetal prognosis.Pregnancy does not appear to exacerbate the disease, however, there is an increased incidence of obstetric complications. Hence, early diagnosis and strict control of hypertension is prudent.Careful assessment, prevention and management of thecomplications, regular antenatal followup to assess feto-maternal wellbeing is needed. Preferably, a multidisciplinary approach involving obstetrician, cardiologist, rheumatologist and anaesthetist is adopted to improve the maternal and fetal outcome.

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