

Takayasu arteritis in pregnancy: A case series

Dr.S.Nivedha Priya , Prof Dr.K.S.Rajeswari

¹(Post graduate, Department of obstetrics and gynaecology,Sri Ramachandra university/Sri Ramachandra Institute of Higher education and Research, chennai ,India)

²(Professor, Department of obstetrics and gynaecology,Sri Ramachandra university/Sri Ramachandra Institute of Higher education and Research, chennai ,India)

Abstract:

Takayasu arteritis is a rare, chronic ,inflammatory, progressive, idiopathic arteriopathy ,affecting young women of reproductive age group causing narrowing, occlusion and aneurysm of systemic and pulmonary arteries, especially aorta and its branches. Takayasu arteritis has higher preordance for women in reproductive age group in second and third decade of life. The diagnosis is based on combination of clinical history, physical examination, clinical suspicion, vascular imaging technique. The etiology is unknown and treatment is aimed at controlling the inflammatory process and preventing secondary sequelae particularly systemic arterial hypertension. An interdisciplinary collaboration of Obstetricians ,cardiologist and neurologist is necessary to improve maternal and fetal prognosis.

Key Words: Takayasu arteritis, idiopathic arteriopathy, interdisciplinary collaboration.

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I. Introduction

Takayasu arteritis is a chronic inflammatory arteriopathy affecting large vessels predominantly affecting aorta and its main branches(1,2).Takayasu arteritis is a chronic, progressive inflammatory, idiopathic disease that cause narrowing, occlusion and aneurysm of systemic and pulmonary arteries affecting mainly aorta and its branches(3,4,5). It affects women predominantly (male: female =1:4) (6). In general it's incidence is 13 per million population ,predominantly seen in Asian origin .The diagnosis is based on combination of clinical history, physical examination, clinical suspicion ,vascular imaging technique. The etiology is unknown and treatment is aimed at controlling the inflammatory process and preventing secondary sequelae particularly systemic arterial hypertension (7).Takayasu arteritis has higher preordance for women in reproductive age group in second and third decade of life. Pregnancy is a favourable state if disease is in remission state. Clinical presentation varies depending on sites of constriction of blood vessels and so typing.The evolution of disease is not affected during pregnancy however one should be careful with peripartum complications in these patient. Since these patients are prone to develop HTN, multiorgan dysfunction, stenosis that hinder regional blood flow and therefore, monitoring of BP and IUGR (8,9) is mandatory .An interdisciplinary collaboration of Obstetrician, Cardiologist, Rheumatologist, Neurologist is often necessitated for optimal maternal and fetal prognosis.

II. Case series

1. A 23 years old ,booked outside, Primi, 27 weeks, chronic hypertensive since 16 years of age on multiple antihypertensive medication ,known case of thoracoabdominal aortic aneurysm for which mesh repair done in 2013 (5 years before presentation) referred from outside hospital for uncontrolled BP (220/110 mmhg) .Both limbs BP were recorded. Growth scan showed IUGR with Doppler changes. Patient was started on Tab. Wysolone 20 mg OD, Tab. Azathioprine 100 mg HS as per rheumatologist opinion. ECG,ECHO – normal. Even with multiple medications BP was not under control .urine albumin-3+. Fundus –normal. Patient was taken up for lscs in view of uncontrolled hypertension with IUGR and Doppler changes. Intraoperative period was uneventful .She delivered a boy baby of weight 830 gms with APGAR of 5/10,8/10.Post operatively patient shifted to ICU for BP monitoring, after stablising BP in ICU for two days ,patient stepped down to HDU and patient was discharged on POD 10 with Tab. Nifedipine 20 mg TDS, Tab. labetalol 300 mg TDS, Tab. Prazosin 7.5 mg BD and Tab.azathioprine 100 mg HS.

2.A 28 years old ,booked ,primi ,37 weeks 3 days ,Rh negative ,known case of Takayasu arteritis since 10 years of age on Tab. Prednisolone 10 mg OD .CT aortogram showed occlusion of left common carotid artery with reformation of external carotid artery and with reformation of internal carotid artery, occlusion of left subclavian artery origin, occlusion of proximal right subclavian artery .Patient's Prednisolone dose was stepped

up to 20 mg OD and started on Tab Azathioprine 100 mg HS since pregnancy after Rheumatologist opinion. Patient was under regular follow up with obstetrician, cardiologist and rheumatologist. Fetal growth monitored with serial scans. Patient suddenly had high BP of 160/110 mmhg at 37 weeks 3 days, urine albumin-2+,and she was started on Tab.Labetalol 100 mg TDS and she was under observation in hospital. Fundus -normal. Patient decided for LSCS at 38 weeks. Rheumatology opinion obtained advised to proceeded with lscs and to change tablet wysolone to Inj.hydrocortisone before and after surgery every 6 th hourly for first 24 hours .Patient taken up for Elective LSCS under spinal anaesthesia at 38 weeks and she delivered a girl baby of 3.03 kg with APGAR of 8/10,9/10 ,baby's blood group -B positive and hence inj.anti D given to the mother. Patient shifted to Icu for BP monitoring and was on observation for 1 day, stepped down from ICU and patient was discharged with Tab. Prednisolone 12 mg OD and Tab.Azathioprine 100 mg OD on POD 8.

3. Mrs.X ,22 years old ,outside booked , primi @ 39 weeks 2 days known case of Takayasu arteritis on Tab.wysolone 20 mg OD for 6 years and Tab.Azathioprine 50 mg OD started since pregnancy. known case of old pulmonary tuberculosis treated at 8 years of age, Patient had elevated BP at 36 weeks, PIH profile-normal, urine albumin-nil, .fundus-normal, patient started on Tab.labetalol 100 mg BD , patient was admitted for safe confinement at 39 weeks 2 days . Bilateral upper and lower limb doppler, carotid study were done. Carotid artery Doppler showed long segment circumferential thickening of bilateral common carotid artery. ECHO - normal with concentric LVH with EF-65%. Rheumatology opinion obtained, On admission ESR,CRP -normal and they advised to change Prednisolone to inj.hydrocortisone 100 mg iv at the time of induction . Patient was induced with 1 dose of PGE2 gel,patient has persistently high BP (230/90 mmhg) with no imminent sign.In view of unfavourable cervix with persistently high BP ,patient was taken up for LSCS .She delivered a female baby of 2.8 kg.Patient shifted to ICU and started on Nitroglycerine and labetalol infusion after stablising the BP ,patient shifted to HDU on POD 2 and patient discharged on POD 5 with Tab,Amlodipine 5 mg OD.

4.Mrs.B ,32 years ,booked ,G2P1L1,36 weeks 2 days ,previous lscs, steroids covered at 34 weeks.known case of Takayasu arteritis since 10 years on Tab.Prednisolone 20mg OD ,Tab.Azathioprine 100 mg BD came with the complaints of elevated BP of 160/100 mmhg .PIH Profile- normal . ,ECG- normal sinus rhythm. CT Arteriogram-occlusion of left common carotid artery occlusion of left subclavian artery ,stenosis at origin of celiac artery and bilateral renal artery stenosis. Urine protein- 2+, Nephrology opinion obtained for deranged renal function test and they advised termination of pregnancy in view of deteriorating RFT. Rheumatology opinion obtained advised stress steroid coverage and to proceed with LSCS. Patient was planned for repeat elective LSCS at 36 weeks 4 days. Intraoperative period were uneventful.She delivered a live boy baby of 2.67 kg with APGAR of 8/10,9/10 . Postoperatively patient shifted to HDU for observation. BP maintained with Tab.lobet 100 mg bd and patient discharged in good condition on postoperative day 5 on Tab.wysolone 20 mg OD.

III. Observation & Result

Table no 1: Age distribution.

Age	Total Number
20-25 years	2
26-30 years	1
31-35 years	1

Table no 2: Gravida.

Gravida	Total Number
Primi	3
Multi	1

In the Study, 3 were Primigravida and 1 was multigravida

Table no 3: Duration of the disease.

Duration	Total Number
<5 years	0
6-10 years	2

>10 years	2
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In the Study 2 had Takayasu arteritis for 6 to 10 years of duration and 2 had the arteritis for >10 years.

Table no 4: Type of Takayasu arteritis.

Types	Number (n=4)
Type 1	2
Type 2	0
Type 3	1
Type 4	1
Type 5	0

In the study, 2 belongs to type 1 Takayasu arteritis, 1 belong to type 3 and 1 belong to type 4 Takayasu arteritis.

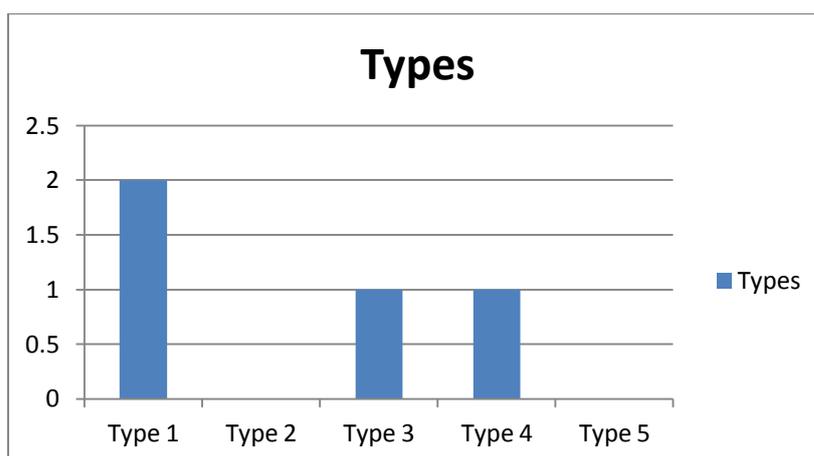


Figure 1: Bar chart showing types of Takayasu arteritis.

Table no 5: Angiographic Abnormality.

Angiographic abnormality	No of cases
Renal artery stenosis	1
Subclavian artery stenosis	2
Celiac artery stenosis	1

Among the 4 cases, 1 had renal artery stenosis, 2 had subclavian artery stenosis and 1 had celiac artery stenosis.

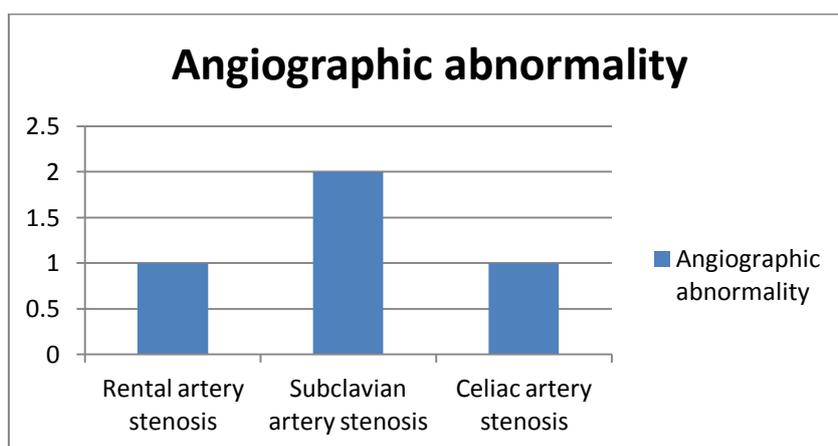


Figure 2: Bar chart showing angiographic abnormality

Table no 6: Maternal presentation.

Maternal complication	Numbers
Chronic hypertension with superimposed preeclampsia	1
Gestational hypertension	2
Pre eclampsia	1

Table no 7: Mode of delivery

Mode of delivery	Total number
NVD	0
LSCS	4

In the study, all 4 of them delivered by LSCS

Table no 8: Fetal outcome

Fetal outcome	Total number
IUGR	1
Preterm delivery	2

Among the 4 patients, 1 delivered a healthy term baby, 1 had IUGR fetus and 2 had preterm delivery.

IV. Discussion

Takayasu arteritis also known as pulseless disease or aortoarteritis or young female arteritis is a chronic inflammatory progressive large vessel vasculitis (10).It leads to narrowing ,occlusion and aneurysm of systemic and pulmonary arteries in the body affecting primarily the aorta and its branches(11,12).Worldwide incidence is around 13 per million, relatively more common in South East Asian countries(2).In the above study 3 of them were primi and 1 multiparous women and in the study all were diagnosed before pregnancy.The pathophysiology has been described as chronic inflammatory disease affecting aorta and its branches in progressive pattern which lead to secondary HTN, retinopathy ,cardiac failure ,stroke and death at early age (13).Histopathologic inspection show mononuclear infiltration of adventitia, occurs early in disease (14).As inflammatory process continues, panarteritis occur ,fibrosis of media and thickening of intima lead to compromise of vessel lumen ,resulting in vessel stenosis. Angiography remains gold standard for diagnosis. Recently 18 FDG-PET scan added as adjunct imaging modality in armamentarium of rheumatologist ,cardiologist to diagnose, with pooled sensitivity and specificity of 70.1% and 77.2% respectively and also multidetector CT angiography is a reliable tool in non-invasively depicting both luminal and mural lesion in the aorta and its main branches. Aim in the treatment of Takayasu arteritis involves control of HTN ,control of inflammation, prevention of renal failure, arterioplasty approach for stenotic vessels. Occlusive and stenotic lesion might require revascularisation by percutaneous angioplasty or use of endoprosthesis for surgical correction (18).Patient with metallic prosthetic valves should be maintained on anticoagulants.

The American college of rheumatology - classification criteria for diagnosing Takayasu arteritis (15)

- Age under 40 years at disease onset
- Claudication of extremities
- Decreased brachial arterial pulses.systolic BP difference more than 10 mmhg between arms
- Bruit over subclavian artery or aorta
- Angiogram abnormality: occlusion or narrowing of entire aorta it's primary branches or large arteries in proximal upper or lower extremities that is not due to arteriosclerosis,fibromuscular dysplasia or other causes.

For Takayasu arteritis 3 out of 6 criteria must be fulfilled .If these criteria are satisfied there is 90.5% sensitivity and 97.8% specificity.

According to **new angiographic classification of Takayasu arteritis**, 5 types of disease identified that depend on angiographic findings and vessel involvement.(16)

- Type1- Involves the branches of aortic arch
- Type2a-Involves ascending aorta ,aortic arch and its branches Type 2b- affects ascending aorta ,aortic arch and its branches and thoracic descending aorta.

- Type 3-Involves thoracic descending aorta,abdominal aorta and/or renal arteries.
- Type 4- Involves only abdominal aorta and/or renal arteries
- Type 5-combined features of type 2B or IV

Additionally involvement of coronary and pulmonary arteries should be indicated as C(+) or P(+).This classification helps us to allow comparison of patient's characteristic according to involved vessels and they are helpful for planning surgeries but they offer little information on prognosis.In the study 2 of them belongs to type 1 ,1 belongs to type 3 and 1 belongs to type 4 disease.

Clinical features of Takayasu arteritis-

- fatigue,
- myalgia,
- arthralgia,
- low grade fever in initial stage ,
- intermittent Claudiacation,
- visual defect,
- fainting attack.

Clinical features depend upon the site of pathology.Few patients are asymptomatic and more than 60% have some kind of complications. HTN caused by reduction in elasticity and narrowing of aorta and its branches ,besides abnormalities in function of aortic and carotid baroreceptor function.

Effect of disease on pregnancy:

In pregnant patient with Takayasu arteritis hypertensive complications such as exacerbation of chronic hypertension, retinopathy, arterial aneurysm, cardiovascular problems ,stroke, rarely aortic dissection and fetal complications like abortion ,IUGR fetal death have been reported in 60-90% of these cases (17).Involvement of abdominal aorta and renal vessels could be the cause for higher incidence of IUGR (29.5%) and poor perinatal outcome. Pulmonary hypertension is common feature with high maternal mortality. Type 1 and 2 have favourable outcome. In the present study all four patients had hypertension,1 was IUGR ,and 2 patients had preterm delivery.

Effect of pregnancy on disease:

Pregnancy does not interfere with disease progression usually.

Aim in treatment of Takayasu arteritis involves control of HTN, prevention of renal failure, arterioplastic approach for stenotic vessels (19) .Management include corticosteroids (glucocorticoid 1 mg/kg /bodyweight for 4 weeks and then taper off) is a main stay of treatment with remission rate of 60%,Immunosuppressive therapy in resistant cases, angioplasty and bypass surgeries. Patient with metallic prosthesis should be started on anticoagulants during pregnancy.

Preconceptional counselling:

Preconceptional counselling is essential regarding dosage adjustment cessation of cytotoxic drug ,folic acid supplementation in periconceptional period and optimal timing of pregnancy.

Antenatal care:

There should be early booking with regular antenatal checkup. Along with routine antenatal visit,serial monitoring of BP, renal function ,cardiac status and preeclampsia screening are vital in such patient. Bp should be measured in all four limbs and also peripheral pulses should be checked for any disparity. Fetal surveillance including DFMC, serial Fetal biometry, BPP and Fetal doppler is also imperative as prerequisite (23). BP monitoring can be difficult in patient with pulseless peripheral arteries. It is better preferred to use non invasive technique and the Invasive technique should be reserved for cases in which non invasive BP monitoring not possible, in case of prolonged surgeries and uncontrolled BP, since complications of arterial catheterisation are most common in patient with peripheral vasculopathies (20).Patient with metallic valvular prosthesis should be maintained on anticoagulation during pregnancy. For those patients on oral steroids, should be changed to Injection form before induction or before surgery and it can be converted back to oral form after 24 hours of delivery.

Mode of delivery:

Vaginal delivery could be indicated for patient with group 1 and 2 disease as long as epidural analgesia is used for pain relief and delivery (2nd stage), second stage of labour should be cut short using forceps.(21). In group 2 b and 3 ,one should preferred LSCS since increased blood volume and BP observed during contraction as well as increased cardiac output observed during labour led to cardiac decompensation (22).In the study all four underwent LSCS in view of persistently high BP and regional anaesthesia was given for all.

If planning for delivery, for patient on anticoagulant therapy .Heparin should be discontinued 4-6 hours before anaesthesia and it can be reversed with protamine if gravida goes in to labour or in case of bleeding .Patient with prophylactic dose of Enoxaparin should receive last dose 12 hours before anaesthesia (23).

Anaesthesia:

Regional anaesthesia is the technique of choice because it allows monitoring of brain perfusion through Patient's level of consciousness .Hydration should be guided by patients cardiovascular condition and it can be initiated at time of blockade.(24).Combined anaesthesia can be good alternative to epidural anaesthesia but with greater need of fluid replacement and vasopressor(20).

Four most important complication:

- Takayasu retinopathy
- Secondary hypertension (most common due to reduced elasticity and narrowing of arteries besides abnormality in functioning of aortic and carotid baroreceptor).
- Aortic regurgitation
- Aneurysm formation

Differential diagnosis of Takayasu arteritis:

- Marfan's syndrome,
- Ehler Danlos syndrome,
- Fibromuscular disorder ,
- some autoimmune condition like SLE ,behcet disease, temporal arteritis.

Prognosis :

Type of Takayasu arteritis affects the Fetal and maternal outcomes. Involvement of abdominal aorta is associated with adverse pregnancy outcomes. The overall 10 years survival rate is approximately 90%,however this rate can be reduced in the presence of major complications(25)

Cause of death in Takayasu arteritis - by exacerbated BP and its complication. Abruption, heart failure, rarely aortic aneurysm and cerebral hemorrhage .

V. Conclusion

Takayasu arteritis should be suspected as one of the differential diagnosis when a young female presents with hypertension. The importance of careful palpation of all peripheral pulses cannot be over emphasised. Regular antenatal visit, frequent BP monitoring, cardiac follow up is mandatory. Takayasu arteritis should be managed by multidisciplinary approach .All antenatal women should be subjected for regular growth scans and fetal doppler from 20 week of gestation.

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