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Pregnancy in Case Of Takayasu's Arteritis

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Abstract

Background

Takayasu's arteritis (TA) is a chronic, inflammatory, progressive, idiopathic arteriopathy, affecting young women of reproductive age group, causing narrowing, occlusion, and aneurysms of systemic and pulmonary arteries, the aorta and its branches. It needs a special attention during pregnancy. A Multi-disciplinary collaboration between rheumatologists, nephrologists, cardiologists, obstetricians and neonatologist is necessary to achieve optimal maternal and neonatal outcomes.

Case Report

Here a case reported where a patient 29 year old G2P1A0L1 with 24 weeks completed with intra uerine fetal death was a known case of takayasu arteritis with chronic hypertension having left and right renal artery stenosis. Following a multidisciplinary approach, she delivered a dead fetus following induction with dinoprost gel without any intra or postpartum complication.

Conclusion

Despite advancements in cardiovascular management and advent of new drugs, the optimal management for pregnant patients with this disease still remains elusive.

Introduction

Takayasu's arteritis (also known as Takayasu's disease, "aortic arch syndrome," "nonspecific aortoarteritis," "young female arteritis" and "pulseless disease") is a form of large vessel granulomatous vasculitis with massive intimal fibrosis and vascular narrowing, most commonly affecting often young or middle-age women of Asian descent, though anyone can be affected. Females are about 8–9 times more likely to be affected than males. Incidence is 13 cases per million population. It was first described by the Japanese ophthalmologists Mikito Takayasu and Onishi .It mainly affects the aorta and its branches, as well as the pulmonary arteries. Those with the disease often notice symptoms between 15 and 30 years of age. In pregnancy it most common in second and third trimester, so special should be taken on peripartum period because increase chances of hypertension, multiple organ dysfunction, and stenosis hindering regional blood flow leading to restricted intrauterine fetal growth and low birth weight in babies are more in this duration.

Case Report

A 29 Year old female G2P1AoL1 was admitted to Cardiac hospital as Pregnancy with 24 completed weeks with chronic hypertension with left and right renal artery stenosis. She was a known case of Takayasu Arteritis (taking T.Amlodipine 5mg twice in a day). At the time of admission, her Blood Pressure was 190/110 mmhg in Right Upper limb, So she was started T. Labetalol (200mg) twice in a day, T.Nifedipine R(20mg) thrice in a day, T.Hydralazine(25mg) thrice in a day. While doing renal artery Doppler and routine antenatal scan, fetal cardiac

activity was found absent so she was immediately transferred to Maternity hospital for further management. Patient was asymptomatic at the time of admission. She has completed 24 wk 1 day according to her last menstrual period. Her past menstrual history was regular moderate and painless. In her obstetric history she had one female child 3 yr old delivered by caesarean section. In her past history, on before 9 yr, patient had an episode of cough and cold for which patient consulted private practitioner where she was accidently diagnosed as a case of Takayasu Arteritis with Reno vascular hypertension. At that time, Doppler study showed left subclavian artery occlusion with right and left renal artery stenosis. For which she underwent PTA(Percutenous Transluminal Angioplasty) with stenting in both right and left renal artery. Again after 2 yr, Doppler ultrasound showed complete occlusion of left subclavian & left axillary with distally low velocity of left upper limb. Since then she was taking T.Amlodipine 5mg twice in a day. In this Pregnancy she was taking routine antenatal care regularily. She was booked and supervised throughout her pregnancy at the Maternity hospital, in liaison with cardiologists. She was reffered to cardiac hospital for left and right renal artery restenting.

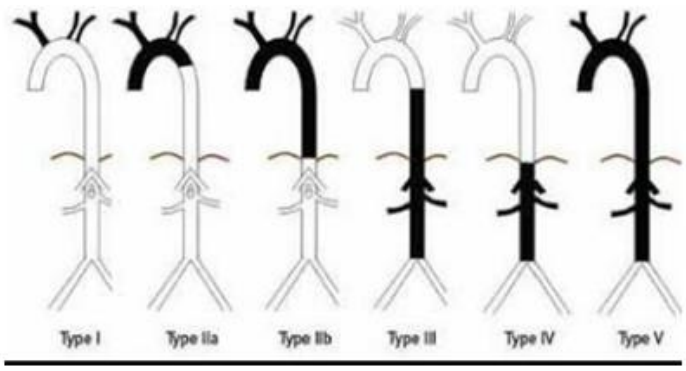
On admission at maternity hospital, she was asymptomatic. Her temperature was normal. Her Pulse in right hand was 98/min while in left hand pulse was feeble approx 10-12/min.Blood Pressure was 180/110 mmhg in Right Upper limb with no albuminuria, BP not recordable in left upper limb with 99% Spo2 on air. Per-abdomen examination showed a relaxed uterus, fundal height corresponding to 24 weeks with absent fetal heart sound with intact and non tender previous caesarean scar, with no active bleeding per vaginally, cervical os was closed. She was put on strict blood pressure monitoring. After thorough history taking, cardiology, and medicine references were done T. Labetalol (200mg) twice in a day, T. Nifedipine R(20mg) thrice in a day, T.Hydralazine(25mg) thrice in a day continued. All routine antenatal and specific blood investigations (INR, PT, and APTT) were normal. Echocardiography was performed, which shows Concentric LVH with Grade 2 MR with MVP, reduced left ventricular compliance with 55% ejection fraction Her fundus examination was normal. Her obstetric ultrasound showed a single intrauterine fetus with no cardiac activity with oligohydroamnios with AFI (4-5). The couple was counseled adequately about intra uterine fetal death and maternal high risk condition. After taking High risk consent she was induced with dinoprost gel twicely at 6 hour duration and tab. misoprost(50ug) was kept thricely on every 4 hour interval two times 700gms IUD Female child was delivered vaginally on next day. Patient's intrapartum and postpartum period were uneventful and she was discharged on 5th postpartum day with 94/min pulse and 160/100 mmHg blood pressure in right upper limb. She was adviced to continue T. Labetalol(200mg) twice in a day, T. Nifedipine R(20mg) thrice in a day, T.Hydralazine(25mg) thrice in a day, Pyridoxine 100mg thrice in a day for 5 days for breast milk suppression and ask to follow after 15 days in cardiac hospital for left and right renal artery restenting.

Discussion

Takayasu's arteritis is similar to other forms of vasculitis, including giant cell arteritis. This inflammation leads to arterial stenosis, thrombosis and aneurysms. Due to obstruction of the main branches of the aorta, including the left common carotid artery, the brachiocephalic artery, and the left subclavian artery. Takayasu's arteritis can present as pulseless upper extremities (arms, hands, and wrists with weak or absent pulses on the physical examination) which may be why it is also commonly referred to as the "pulseless disease". Involvement of renal arteries may lead to a presentation of renovascular hypertension. Some

people develop an initial "inflammatory phase" characterized by systemic illness with signs and symptoms of malaise, fever, night sweats, weight loss, joint pain, fatigue, and fainting. Fainting may result from subclavian steal syndrome or carotid sinus hypersensitivity. There is also often anemia and marked elevation of the ESR or C-reactive protein (nonspecific markers of inflammation). The initial "inflammatory phase" is often followed by a secondary "pulseless phase". The "pulseless phase" is characterized by vascular insufficiency from intimal narrowing of the vessels manifesting as arm or leg claudication, renal artery stenosis causing hypertension, and neurological manifestations due to decreased blood flow to the brain. Renal artery stenosis causes high blood pressure because normally perfused kidneys produce a proportionate amount of a substance called renin. Stenosis of the renal arteries causes hypo perfusion (decreased blood flow) of the juxtaglomerular apparatus, resulting in exaggerated secretion of renin, and high blood levels of aldosterone, eventually leading to water and salt retention and high blood pressure. The neurological symptoms of the disease vary depending on the degree; the nature of the blood vessel obstruction; and can range from lightheadedness to seizures (in severe cases). One rare, important feature of the Takayasu's arteritis is ocular involvement in form of visual field defects, vision loss, or retinal hemorrhage.

Classification of Takayasu Arteritis



Type	Vessel Involvement
Type I	Branches from the aortic arch
TypeIIa	Ascending aorta, aortic arch and its branches
TypeIIb	Ascending aorta, aortic arch and its branches, thoracic descending aorta
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

Effect of Takayasu Arteritis on Pregnancy Outcome

Pregnancy does not interfere with disease progression. The risk of Takayasu's arteritis associated with pregnancy, is mainly due to the consequences of arterial hypertension with pre-eclampsia, Preterm delivery, IUD, IUGR, heart failure and cerebral vascular events, secondary hypertension, Takayasu retinopathy. The major fetal risk is in utero death (IUD) but intra-uterine growth retardation is more frequent. Etiology of IUGR may be impaired placental blood flow. Incidence of IUGR and poor perinatal outcome is high when bilateral renal involvement is present. The risk is greatest during the third trimester and during the perinatal period. Fetal involvement is greatest in severe cases and in those treated late.

Diagnosis

Diagnosis is based on clinical manifestations, inflammatory markers (acute phase reactants), and arteriography demonstrating aortic stenosis and of its branches. Common features of active TA are fatigue, myalgia, arthralgia, and low-grade fever in initial stages and intermittent claudication, visual defects, and fainting attacks in later stages. Demonstration of vascular lesions in large and middle-sized vessels on angiography, CT scan, magnetic resonance angiography or FDG PET. Contrast angiography has been the gold standard. The characteristic finding is the presence of "skip lesions," where stenosis or aneurysms alternate with normal vessels

Management

Management of TA entails a combine approach with involvement of obstetricians, anesthesiologists,

cardiologists, rheumatologists, and neonatologist in a tertiary care center. The aims are control of inflammation, prevention and treatment of complications like hypertension and revascularization by percutaneous angioplasty, use of end prosthesis, or surgical correction for occlusive and stenotic lesions.

Preconception counseling is essential regarding dosage adjustment or cessation of cytotoxic drugs, folic acid supplementation in the periconceptional period, and optimal timing of pregnancy. Pregnancy should be ideally planned in remission phase. There should be an early booking with regular antenatal supervision. Along with routine antenatal visits, serial monitoring of blood pressure, renal function, cardiac status, and preeclamptic screening are vital in such patients. Fetal surveillance including daily fetal movement count, serial fetal biometry, biophysical profile, and fetal Doppler is also imperative as per requirements. Control of blood pressure is utmost important. Most people with Takayasu's arteritis respond to steroids such as prednisone (1 milligram per kilogram of body weight per day). Because of the significant side effects of long-term high-dose prednisone use, the starting dose is tapered over several weeks to a dose which controls symptoms while limiting the side effects of steroids. Promising results are achieved with mycophenolate and tocilizumab. But these are generally avoided in pregnancy or used after a meticulous assessment of the risk/benefit ratio for the patient.

Prevention is based on the initial work-up to identify the disease and possible complications, programming pregnancies and increasing surveillance during periods of risk, vaginal delivery is the preferred mode and epidural analgesia has been advocated for labor and delivery. In women with hypertension, delivery should be abbreviated by the use of outlet forceps. In women with stages IIb and III, cesarean section is preferred to prevent cardiac decomposition due to increased blood volume and blood pressure observed during uterine contractions and increased cardiac output observed during labor. Our patient was hemodynamic ally stable, we had induced labor with dinoprost gel.

Conclusion

Takayasu arteritis in pregnancy needs tight pre conceptional disease control, strict follow up and targeted treatment of high blood pressure. Early diagnosis and management can result in positive pregnancy outcome. A Multi-disciplinary collaboration between rheumatologists, nephrologists, and obstetricians is necessary to achieve optimal maternal and neonatal outcomes.

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