ORIGINAL RESEARCH PAPER

Obstetrics & Gynaecology

COARCTATION OF ARCH OF AORTA WITH COARCTATION OF ABDOMINAL AORTA WITH TAKAYASU'S ARTERITIS IN PREGNANCY: A RARE CASE REPORT

KEY WORDS:

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1.INTRODUCTION

Contemporary cohorts of patients with repaired aortic coarctation (CoA) have good long-term survival but relatively high cardiac complication rates, including re-coarctation in 34%, aortic aneurysms in 18%, hypertension in 32% and 13-fold increased risk for ischaemic stroke as compared with controls without congenital heart disease (1).

Pregnant women with repaired CoA are currently classified as mWHO II–III in the modified WHO classification (3). mWHO II–III represents an intermediate risk of maternal mortality and a moderate-to-severe risk of morbidity, corresponding with a cardiac event rate of 10%–19% during pregnancy. Women with unrepaired severe CoA are considered to be mWHO class IV (2).

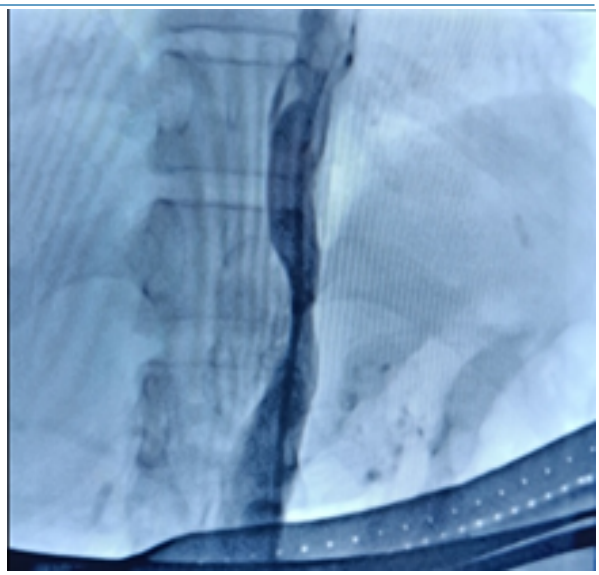
Takayasu's arteritis (TA), also known as pulseless disease/ aortoarteritis/"young female arteritis," is a rare chronic inflammatory progressive large vessel vasculitis (LVV) of unknown etiology afflicting women of childbearing age (3). It was first described by the Japanese ophthalmologists Mikito Takayasu and Onishi (4). Its incidence is reported to be 13 cases per million population (5). It is predominantly seen in the women of Asian origin (3). It leads to narrowing, occlusion, and aneurysms of systemic and pulmonary arteries in the body, affecting primarily the aorta and its branches (6).

Pregnancy as such has no effect on the evolution of the disease ; however, its peak incidence is in second and third trimesters. Thus, such patients warrant special attention during the peripartum period owing to the likelihood of development of complications such as hypertension, multiple organ dysfunction, and stenosis hindering regional blood flow leading to restricted intrauterine fetal growth and low birth weight in babies (7). An interdisciplinary collaboration of obstetricians , cardiologists, rheumatologists, and neurologists is often necessitated for an optimal maternal and fetal prognosis .

Here a case is described to enlighten the obstetricians on fetomaternal outcome and management of this infrequent, but not uncommon clinical entity encountered nowadays.

2. CASE REPORT

A 39-year-old primigravida was admitted in the hospital as pregnancy with pregnancy induced hypertension (on Tab labetalol 100mg TDS), at 32 completed weeks in labor pain and with leaking PV. In 5th month of Pregnancy she was diagnosed as a case of Coarctation of Arch of Aorta with Coarctation of Abdominal Aorta with Takayasu's Arteritis on treatment (taking prednisolone 5mg, aspirin 75mg, and clopidogrel 75mg OD.) She was a booked and supervised throughout her pregnancy at the govt hospital, in liaison with cardiologists. Her past and personal histories were reviewed thoroughly in the emergency department. MR Aortogram done earlier revealed Coarctation of Arch of aorta and abdominal aorta and small thrombus in left common carotid and left subclavian artery. She had surgical correction (Coarctoplasty) of Coarctation of Abdominal Aorta in 6th Month of Pregnancy. She had no history of other comorbidities like IBD and sinusitis



She gave history of myalgias, and fever off and on throughout her pregnancy, despite on corticosteroids . On admission, her BP was 150/90 in ankle and pulse was 90/min. There was no albuminuria. Per- abdomen examination showed a fundal height corresponding to 32 weeks with audible fetal heart sound and active uterine contractions. She was put on strict fetomaternal surveillance. PV examination revealed Os 2 cm dilated , 30% effaced with leaking and liquor was clear. After thorough history taking, cardiology references were done, her anticoagulants were stopped in v/o anticipated termination of pregnancy, and she was continued on labetalol 100mg TDS and continuous BP monitoring (non-invasive). All routine antenatal and specific blood investigations (INR, PT, and APTT) were normal. Though CRP was also normal, it was on a higher side of normal range (5.5 mg/dL). ESR was found to be on a higher side with value of 30 mm/1st hour (Normal <20 mm/1st hour). Echocardiography was done in 6th ANC month which showed LVEF of 60%. A renal Doppler was also done earlier and that was normal. Her obstetric ultrasound with Doppler velocimetry was normal.

The couple were counselled adequately about fetomaternal prognosis and their informed written consent was taken .Blood and Blood products were reserved and ICU bed was booked. she was given betamethasone for fetal lung maturity, following which LSCS was done at 32 weeks and she delivered a live baby weighing 2 kgs, who was transferred to nursery, and discharged after one week. Immediately After LSCS patient had continuous mucosal bleeding per vaginal with well maintained tone of the uterus. So RDP transfusion was done immediately. Post LSCS patient was transferred to ICU and was observed for 48 hours. she was discharged on prednisolone, Aspirin 75 mg, and Clopidogril. At the time of this writing, she is convalescing well, with both mother and baby doing fine, and following up periodically with cardiologists, and gynecologists.

3.DISCUSSION

Takayasu's arteritis is a LVV with aortic inflammation leading

to proximal occlusion and/or aneurysms of carotid, subclavian, pulmonary, iliac, and renal arteries (8). Mean age is typically reported between second and third decade of life (9). Its etiology remains primarily idiopathic. Autoimmunity, sex hormones, and genetic (predisposition of the human leukocyte antigen, (HLA BW52) factors have often been hypothesized as plausible factors causing it (10).

Various types of TA have been acknowledged in the past: type I (disease embroiling aortic arch and its branches), type II (lesions constrained to descending thoracic aorta and abdominal aorta), type III (patients with characteristics of types I and II), type IV (involvement of pulmonary artery), and type V (combined features of types IIb and IV) [10]. The above described patient was labeled as type II TA. The disease can also be classified into stages as per the presence of major complications such as hypertension, retinopathy, aneurysms, and aortic insufficiency (11): stage I (no complications observed), stage IIa (patients having only one of these complications), stage IIb (patients with only one of these complications, but the severe form), and stage III (more than one complication is present). The patient presented here was in stage Iib.

Pregnancy does not interfere with disease progression (1) but TA has several adverse implications on pregnancy like abortions, preeclampsia, IUGR, IUD, and abruption(12). More than 60% of patients have some kind of complications and the four most important ones are Takayasu's retinopathy, secondary hypertension, aortic regurgitation, and aneurysm formation. Hypertension is fairly common due to reduction in elasticity and narrowing of the arteries, besides abnormalities in functioning of aortic and carotid baroreceptors function (13). Blood pressure in such patients should be also measured in the lower extremity to pick up blood pressure discrepancies; like in our case BP recordings in lower extremity were higher than upper extremity. Besides, pulselessness of unilateral or bilateral radial arteries and vascular bruit should be checked in all cases of hypertension. Involvement of abdominal aorta is associated with adverse pregnancy outcomes, which was present in our case. Arterial ultrasound Doppler, quantifying the flow in the uterine arteries, is beneficial in evaluation of fetal well-being and growth in women with TA.

Diagnosis is usually 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. Many may be diagnosed after clinical examination, when one or more peripheral pulses are not palpable or blood pressures vary in two limbs. However, computed tomography or magnetic resonance angiography can detect TA even before the development of severe vascular compromise as in our case (14) Recently, 18 FDG-PET scan has been added as an adjunct imaging modality in the armamentarium of rheumatologists and cardiologists to diagnose LVV, with a pooled sensitivity and specificity of 70.1% and 77.2%, respectively (15). However, the gold standard for diagnosis still remains vessel biopsy (14) which could not be performed in our case.

Management of TA entails an interdisciplinary approach with involvement of obstetricans, anesthesiologists, cardiologist , 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 endoprosthesis, or surgical correction for occlusive and stenotic lesions.

Preconception counselling is essential regarding dosage adjustment or cessation of cytotoxic drugs, folic acid supplementation in the peri-conceptional 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 kick count, serial fetal biometry, biophysical profile, and fetal Doppler is important in such patients (16)

Antihypertensive drugs and antiplatelets can be started as per need, as was in the present case. TA may respond symptomatically to corticosteroid therapy (first line drugs) at a dose of 1-2mg/kg/bodyweight for 4 weeks followed by slow tapering. However, chronic use of corticosteroids could lead to suppression of adrenal gland activity with inadequate release of endogenous corticosteroids in moments of stress, such as surgeries (17).

Utilization of immunomodulatory agents like mycophenolate-mofetil, infliximab, tocilizumab, leflunomide, and abatacept has gained momentum in recent times for treat- ment of TA, especially in refractory cases (18); however their safety in pregnancy has not yet been established.

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, LSCS is preferred to prevent cardiac decompensation due to increased blood volume and blood pressure observed during uterine contractions and increased cardiac output observed during labor. Our patient was hemodynamically stable and she belonged to group IIb and so decision of LSCS was taken.

Patients with metallic valvular prosthesis should be maintained anticoagulated during pregnancy. The choice of medication should take into account the probable due date and reversibility of the method. Heparin should be discontinued 4 to 6 hours before anesthesia, and it can be reversed with protamine if the gravida goes into labor or in case of bleeding.

The overall five-year survival rate after diagnosis was 83.1%. Death typically is a consequence of congestive heart failure or cerebrovascular events. The survival is better in patients without a progressive course and in those below 35 years of age. Early diagnosis with proper medical or surgical management is essential for a good prognosis. A high index of clinical suspicion in patients presenting with pulseless peripheral vessels could be kept in mind to optimize the management following multidisciplinary approach.

4.CONCLUSION

Pregnancy with Coarctation of Arch of Aorta with Coarctation of Abdominal Aorta with TA presents as an onerous medical condition to manage for an obstetrician.

Competing Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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