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CASE REPORT

Pulseless and Ambulatory Pregnant Woman: An Obstetric and Medical Challenge

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Introduction

Takayasu's arteritis is a chronic inflammatory arteriopathy affecting large vessels predominantly the aorta and its main branches [1, 2]. It is a rare disease, relatively more common in Japan, India, Asia and Mexico [3]. It has a higher preponderance for women in reproductive age groups, in the 2nd and the 3rd decades of life [4–6]. The etiology

remains speculative, but the current literature suggests autoimmune basis [2, 7, 8]. The disease can vary remarkably in terms of sites of constrictions of blood vessels and hence the clinical presentation [9, 10]. Optimal management of pregnancy in this condition cannot be too standardized or generalized due to rarity and variation in disease profile. Therefore, experience sharing forms an important aspect of learning to manage such patients. We share one such case here and the lessons we learnt from it.

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Case Report

A second gravida (G2P1L0), aged 26 years, visited in Antenatal clinic at 8 weeks of pregnancy. Her first pregnancy had ended in intrauterine demise in the 7th month (no details available). On examination, she was short with medium built (Ht-150 cm, Wt-47 kg, BMI-21.7 kg/m). Her bilateral radial pulses were not palpable, and her BP

could not be recorded in either arm by auscultatory method using the aneroid BP instrument in the Antenatal clinic. Patient was admitted for complete evaluation. The BP taken using a digital manometer (NIBP) was very high (170/100–210/130) with a wide variation in the readings in the four limbs. Aortic bruit could be heard in the midline over abdomen. No carotid bruit was found. The clinical suspicion of aortoarteritis was confirmed by Doppler study (Figs 1–3). She was started on folic acid, antihypertensives (alphamethyldopa 250 mg TDS and labetalol 50 mg BD) and ecosprin 75 mg OD. Regular follow up was advised fortnightly. At 25 weeks, she developed superimposed

preeclampsia and also mild IUGR (lag of 3 weeks); Doppler study showed a high systolic/diastolic ratio in umbilical arteries. She was advised to stay hospitalized for further evaluation and monitoring, but she left against medical advice. Patient reported back to the hospital at 29 weeks with decreased fetal movements. USG revealed severe IUGR (lag of 5 weeks) with moderate oligohydramnios. Fetal Doppler showed absent diastolic flow in umbilical arteries and preferential flow in middle cerebral arteries. Patient was taken for emergency LSCS on the same day. A preterm female baby weighing 660 g was born, was kept in NICU and fortunately did not require

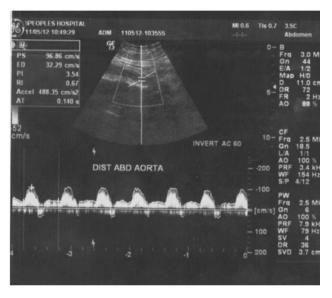


Fig. 1 Color and spectral Doppler study showing very high velocity turbulent flow in abdominal aorta (below epigastrium) suggestive of distal narrowing

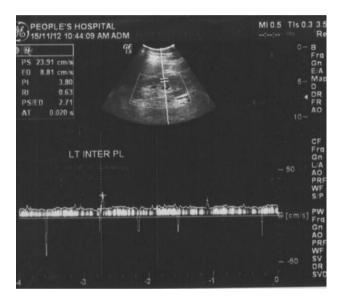


Fig. 3 Color and spectral Doppler study showing significant narrowing at the origin and hilum of left renal artery

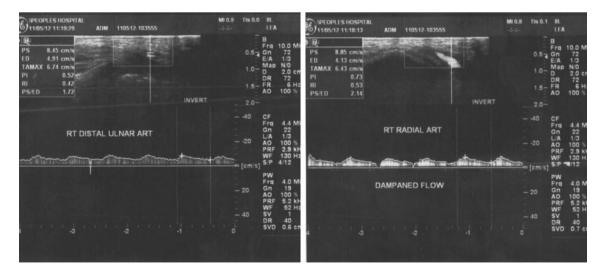


Fig. 2 Color and spectral Doppler study showing bilateral radial and ulnar arteries showing dampened flow and significant arterial narrowing

significant ventilatory support (only BUBBLE C PAP for 48 h). The patient recovered well postoperatively. Hypertension persisted beyond 6 weeks, patient continues to be on two antihypertensive medications: telmisartan 40 mg OD and Amlodipine 5 mg BD. Patient was advised CT Angiography for further evaluation of disease, which she did not comply with for financial reasons.

Discussion

TA is a rare, chronic, giant cell vasculitis which primarily involves the aorta, its main branches, and coronary and pulmonary arteries. The disease causes various clinical conditions such as arm claudication, decrease artery pulses, visual loss, stroke, aortic regurgitation, HT, and CCF. Clinical presentation can be varying depending on site or level of vessel stenosis or occlusion [9, 10].

According to new angiographic classification of TA, five types of disease can be identified [11]. The presence of three or more of these criteria demonstrates a sensitivity of 90.5 % and specificity of 97.5 %.

Type Vessel involvement

Type I Branches from the aortic arch

Type IIa Ascending aorta, aortic arch, and its branches

Type IIb Ascending aorta, aortic arch and its branches, and thoracic descending aorta

Type III Thoracic descending aorta, abdominal aorta and/or renal arteries

Type IV Abdominal aorta and/or renal arteries

Type V Combined features of types IIb and IV

Types I and II of TA are commonly symptomatic. Patients with TA types III, IV, and V may remain asymptomatic and hence undiagnosed. Being type IV TA, the reported case remained undiagnosed till her second pregnancy. The diagnosis was picked up through meticulous general examination.

Earlier literature has suggested that most of the pregnant patient with TA reached successfully till term and had vaginal deliveries with fair fetal outcome. Those patients were mostly of types I and II TA [12, 13]. Incidences of secondary HT and IUGR were low in those patients. Poor perinatal outcome was usually seen in types III, IV, and V TA. Common complications seen were secondary HT, pre eclampsia, and IUGR. Operative interventions and NICU admissions were more common in those patients [14, 15]. Involvement of abdominal aorta and renal vessels could be the cause for higher incidences of IUGR and poor perinatal outcome. Course of pregnancy and fetal prognosis may not

correlate with severity of disease per se but are directly dependent on site of involvement of vessels. Therefore, a careful thought has to be given to the type of the disease while studying literature for patient management.

Another learning point from literature as well as this patient was that patient diagnosed for the first time during pregnancy may have unpredictable complications even if the disease appears to be stable. So, alertness at all times with preparation of timely intervention is prudent.

Conclusions

Delayed detection and secondary HT are poor prognostic factors for TA. So, attention to accurate BP measurement (preferably in legs) should be given. BP should be strictly controlled for favorable maternal and fetal outcome.

Patients with involvement of abdominal aorta and renal vessels usually show higher incidences of IUGR and poor perinatal outcome. So, all women should receive regular growth scans and fetal Doppler performed at 20 and 24 weeks of gestation. It will provide a predictor of utero placental function and help us identify high-risk pregnancies.

Type of TA should be specified in literature as course of disease, as fetal prognosis is mainly dependent on it.

Learning Points

- The importance of mindfully palpating the pulse cannot be overemphasized.
- IUGR and poor perinatal outcome usually seen with involvement of abdominal aorta and renal vessels.
- Regular visits, timely admission, close monitoring, and a multidisciplinary team approach are must.

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