Case Report:
Successful Pregnancy Outcome in a Patient With Aortoarteritis
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Abstract:
Aortoarteritis, also known as Takayasu’s arteritis, aortic arch syndrome or pulseless disease is a rare vascular disease that cause progressive damage including inflammation, scarring, narrowing and abnormal ballooning inside the wall of aorta and major arteries. This case report of aortoarteritis in pregnancy was diagnosed during pregnancy and successful pregnancy outcome was achieved by combined multidisciplinary supervision of experts.

Key Words: Takayasu’s disease, Aortoarteritis, Intrauterine growth restriction

Introduction:
Aortoarteritis is a chronic inflammatory disease of unknown etiology of the aorta and its major branches. In 1908, Mikito Takayasu, a Japanese ophthalmologist, reported the association of retinal arteriovenous anastomoses and absent upper extremity pulses.[1] Takayasu arteritis(TM) is a large vessel vasculitis of unknown origin and is more frequently found in Asian populations. Japanese patients with TA have a higher incidence of aortic arch involvement. In contrast, different series from India report higher incidences of thoracic and abdominal segment of aortic involvement. It is more common in young females and thus it is not uncommon to encounter this disease during pregnancy. In some patients the diagnosis is made for the first time in pregnancy. Aortoarteritis becomes an important cause for severe hypertension detected during routine prenatal examination. Surgical repair before conception reduces the risk of aortic complications during pregnancy, and also decreases the incidence of fetal complications, such as intrauterine growth restriction (IUGR) or congenital heart disease. In uncomplicated coarctation, pregnancy and delivery are usually uneventful. Cesarean section is done mainly for obstetric indications.

Case Report:
A 24-year old woman, P0+0, married for 1year was admitted to our institution with chief complaints of headache, vertigo, dizziness and raised BP of 170/110 mm of Hg. She was carrying 26th week of pregnancy on admission. The patient was diagnosed to be hypertensive immediately after her marriage when she had severe headache and blurring of vision. She was taking antihypertensive drugs irregularly before she conceived. On admission she was prescribed 60mg nifedepine and in consultation with physician 2gm umethyl dopa and 200mg labetalol were added. But her BP remained elevated in the range of 160/110 mm Hg with occasional increase to 190/130mm Hg. Her radial and brachial pulse were normal but femoral, popliteal and dorsalis pedis pulsation were very low. The patient was short in height(4’8”) with puffy and congested face. Neck veins were neither engorged nor pulsatile. On auscultation of precordium, S1 and S2 were normal, and a soft systolic murmur was heard over the back in the inter-scapular region. The patient had mild anaemia, pitting oedema in the legs, and a 28week uterus with a cephalic presentation. Ophthalmoscopic evaluation aortic arch and thoracic aorta did not reveal any coarctation but aortoarteritis was suspected. USG with Doppler study showed blood flow in upper limb arteries was disproportionately more than in lower limb arteries. The blood flow in the renal and uterine vessels was normal. MRI of aorta confirmed aortoarteritis and showed multiple segmental narrowing at thoracic and abdominal aorta without much narrowing in renal and placental vessels. Complete blood count showed normal haemoglobin, WBC and platelets with an ESR value of 20mm/hr(Westergreen). Routine examination of urine revealed absence of proteinuria; serum urea, creatinine, bilirubin, ALT, AST and LDH were essentially normal. Urinary VMA excretion values were within normal limits and possibility of pheochromocytoma was ruled out. Pregnancy continued successfully and fetal profile was normal without any sign of intrauterine growth restriction. BP remained stable at around 150/100mm of Hg. In view of her short stature and small gynaecoid pelvis elective caesarean section was done at 38weeks under general anaesthesia under the supervision of cardiologists and physicians. Blood pressure lowering drugs like Inj. Esmolol, Inj. Fentanyl hydrochloride, Inj. lignocaine were used to control per- and postoperative hypertensive crises. BP was kept in and around 140/90mm of Hg throughout the operation. A female baby of 2.8 kg with a good Apgar score was delivered. The patient recovered uneventfully in the postoperative period and was dis-
Charged on the 10th postoperative day. Her BP was 140/90 mm Hg on discharge and remained stable with 60 mg nifedipine. The patient has been kept in follow-up in the cardiology outpatient department.

Discussion:

Takayasu arteritis (TA) is found in both sexes. Male-to-female ratio is about 1:1.58 in India, 1:1.58 in Japan, 1:2.9 in Thailand and 1:1.66 in Korea.[2] The major clinical signs on physical examination include a) blood pressure difference greater than 30 mm Hg between arms b) asymmetric pulses c) diminished or absent pulses d) poststenotic dilatations producing bounding pulses e) paroxysmal hypertension resulting from renovascular compromise g) bruits over subclavian arteries or aorta, and h) varying degrees of hypertensive retinopathy.

Diminished or absent pulse are found in 96%, bruits are observed in 94%, hypertension and heart failure are detected in 74% and 28% respectively.[3] Hypertension develops in this disease as a result of: a) Renal artery stenosis b) involvement of baroreceptors by aortitis, c) loss of elasticity of aorta, d) coarctation like lesion, e) aortic regurgitation. Treatment usually begins with prednisolone. If glucocorticoids alone are not effective, stronger cytotoxic drugs like methotrexate, azathioprine or cyclophosphamide are added. Blood pressure medications may also be necessary to control hypertension.

Angiography is the most important test for the diagnosis of TA. Drawbacks to arteriography, include morbidity from use of contrast dye in patients with renal disease and cumulative radiation exposure, and therefore can not be done in pregnancy. This can be avoided by using Magnetic Resonance Angiography which is safe in pregnancy. Arteriography demonstrates long, smooth, tapered narrowings or occlusions. Stenoses are seen in 90-100% of patients with TA and aneurysm formation is observed in 25%-47%. Magnetic resonance imaging are useful for early diagnosis of TA. Use of contrast reveal inflammatory lesions prior to the development of stenoses; these lesions are missed by angiography. Aortic lesions including stenosis, dilatation, wall thickening, and mural thrombi are well visualized on MRI. It is less adequate in visualizing distal lesions of the subclavian vessels and common carotids.

Following the acute phase, patients with fibrotic changes require surgical treatment of symptomatic stenotic or occlusive disease. This can be achieved by percutaneous angioplasty of aorta[6] or endovascular stenting for long segment renal artery stenosis or, in severe cases, by resection and placement of a manmade graft.

The outcome of pregnancy in this disease is usually favourable as in our case. In a study of 12 patients with aortooartitis who had 24 pregnancies, Sharma et al[7] noted a favourable outcome in 71% of patients while 19% had either an abortion or intrauterine death. The poor outcome are mainly related to adverse sequel of severe hypertension, abdominal aortic involvement and intrauterine growth restriction. Wong et al[8] showed that fetal weight appeared to be related to maternal abdominal aortic involvement. They devised a prognostic score based on four factors. These are a) involvement of abdominal aorta and renal arteries b) highest mean arterial pressure during 3rd trimester c) time when superimposed pre-eclampsia started d) time when proper treatment was started. For each factor a score of 0 to 2 is given with maximum score of 8. There appears to be a cut-off level of 4 or more, above which fetus is at high risk of IUGR. Medical treatment is almost same as done in nonpregnant state. BP should be monitored in all four limbs. Central aortic pressure monitoring has been recommended during intrapartum period to control the hypertensive crisis and convulsion. Vaginal delivery is possible in absence of any other obstetric contraindication. The 2nd stage should be curtailed by instrumental aids to avoid any fetal and maternal hypertensive complication. Caesarean section is justified in the presence of suspected maternal and fetal complications.

Conclusions

This is a rare type of chronic inflammatory vessel disease, commonly called as Takayasu’s disease. As it is common in females, the possibility of aortooartitis should be kept in mind in a pregnant woman when hypertension remains uncontrolled despite conventional antihypertensive therapy.

References:


