

SUCCESSFUL OUTCOME OF 2ND TRIMESTER MEDICAL TERMINATION OF PREGNANCY IN A CASE OF AORTOARTERITIS WITH DILATED CARDIOMYOPATHY

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ABSTRACT

BACKGROUND

Aortoarteritis is also known as Takayasu Arteritis, Aortic Arch Syndrome and Pulseless Disease. The 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 with formation of aneurysms. Thus, the disease carries high morbidity and mortality. The occurrence of aortoarteritis during pregnancy is a very rare occurrence. The complications are fatal with the continuation of pregnancy. We are reporting a case of aortoarteritis in pregnancy diagnosed first at 19 weeks of gestation for Medical Termination of Pregnancy and successful outcome was achieved by combined multidisciplinary approach.

KEYWORDS

Aortoarteritis, Medical Termination of Pregnancy, Blood Pressure, Tablet Mifepristone, Arteriography.

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BACKGROUND

CASE REPORT

A 24-year-old, unmarried primigravida has come with 5 months of amenorrhoea for MTP. Patient had no specific complaints, no significant illness in the past. On examination, patient was conscious, oriented, afebrile, not anaemic, no icterus, no cyanosis, no clubbing, no lymphadenopathy, no pedal oedema, PR: 96/min. (right radial artery), regular, BP: 200/100 mmHg (right upper limb). Pulse and blood pressure was not recordable in the left upper limb. Dorsalis pedis pulse absent on both sides.

Cardiovascular Examination: S1, S2 present, ejection systolic murmur present. Respiratory System: Bilateral air entry equal on both sides, no added sounds. Per abdomen: Uterus enlarged to 20 wks. size, foetal parts felt. Per Vaginal Examination: Cervix uneffaced, os closed. Central Nervous System: No abnormalities detected.

On evaluation, echocardiogram shows dilated cardiomyopathy/left ventricle and right ventricle systolic dysfunction present. LVEF: 37%. Arterial Doppler ultrasonogram shows evidence of severe arterial wall thickening with severe luminal narrowing noted in descending thoracic aorta. Severe arterial wall thickening with complete occlusion involving left subclavian artery. Poststenotic flow pattern seen in both renal arteries-features suggestive of aortoarteritis. Rheumatologist and vascular surgeon opinion obtained. Ophthalmologist opinion

obtained regarding fundal changes. No evidence of hypertensive retinopathy. Patient was treated with T. Lasix 40 mg half twice a day, T. Aldactone 25 mg once a day, T. Enalapril 2.5 mg twice a day. Her blood pressure was controlled.

Even though, heart disease is a contraindication for 2nd trimester MTP in view of DCM and very low ejection fraction, patient being an unwed in view of social reasons, MTP was planned. T. Mifepristone 200 mg given on day 1 followed by T. Misoprostol 400 micrograms after 48 hours. Patient was closely monitored. She delivered a dead born male baby. Patient was haemodynamically stable. Her postnatal period was uneventful. Inj. Anti-D 300 micrograms IM given. (Rh negative mother-AB negative).

DISCUSSION

Aortoarteritis is a chronic inflammatory disease of the aorta and its major branches. Incidence is rare one in 2 lakhs. It predominantly affects young females.⁽¹⁾ Takayasu arteritis was first described in 1908 by two Japanese ophthalmologists, Takayasu and Onishi, who observed retinopathy in the absence of peripheral pulses.⁽²⁾ The disease is characterised by nonspecific granulomatous inflammation of blood vessels. Wall thickening, fibrosis, stenosis and thrombus formation cause end-organ ischaemia. More acute inflammation destroys the arterial media leading to aneurysm formation. Arterial media leading to aneurysm formation. Stenotic lesions predominate and have been reported in 90% of cases while aneurysms are only reported in approximately 25%.⁽³⁾ Patients with aortoarteritis may present with a variety of clinical manifestations, but arterial hypertension is the most common feature of this disease. The rarity of the disorder and the heterogeneous nature of its clinical manifestation predispose to a late diagnosis and delayed treatment.

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Clinical suspicion and proper imaging are crucial for the correct diagnosis and management of patients with TA.⁽⁴⁾

Aortoarteritis is found in both the genders. Female-to-male ratio is about 1.58:1 in India and Japan. Diagnosis is made by physical examination including difference in blood pressure in both arms of about 30 mmHg, asymmetrical pulse, absent pulse and presence of bruit (94%) bounding pulse as a result of poststenotic dilation, hypertension (74%) resulting in hypertensive retinopathy, claudication, carotidynia, visual loss, stroke and heart failure (28%).⁽⁵⁾ The deaths occurred mostly from failure to treat systolic hypertension.⁽⁶⁾ The various problems arising during pregnancy are related to pressure elevation and heart failure.⁽⁷⁾

The disease is classified clinically into stages depending on the presence of complications such as hypertension, retinopathy, aneurysms and aortic insufficiency.⁽⁸⁾ Group I - Uncomplicated disease. Group IIa - Single complication with uncomplicated disease. Group IIb - Severe single complication with uncomplicated disease. Group III- 2 or more complications. Causes of hypertension in aortoarteritis include renal artery stenosis, coarctation of aorta and involvement of baroreceptors and loss of elasticity of aorta. Mainstay treatment is glucocorticoids (prednisolone).

If the condition remains uncontrolled, then cyclophosphamide, methotrexate, azathioprine can also be used.

Arteriography is commonly used to diagnose this condition, but because need of contrast and use of radiation it is not safe in pregnancy and renal disease patients. MRI is preferred in such condition as it can detect the inflammation and diagnose the disease earlier, but it is not able to detect lesions in distal subclavian artery and common carotid artery. High resolution Doppler ultrasonography is useful in such condition, but has a limitation of less visualisation of aorta. Contrast-enhanced MR imaging provides information about the disease activity, which may be useful in diagnosis and treatment.⁽⁹⁾

The course of the disease remains unaffected during pregnancy inflammatory activity and the haemodynamic state improve with pregnancy in patients with Takayasu arteritis. The physiologic aspects which cause this improvement should be maintained even after pregnancy.⁽¹⁰⁾ However, there is increased chances of developing preeclampsia, eclampsia, placental abruption, IUGR and death. Vaginal delivery is preferred at term. Change in systolic blood pressure during the second stage of labour should be kept in mind and use of instrumental delivery should be considered to cut second stage labour. Poor outcome of disease occurs mainly in severe hypertension with involvement of abdominal aorta showed that abdominal aorta involvement is directly related to foetal weight.

There are no reported direct maternal death in these cases. Almost, 83.9% women deliver a healthy, mature newborn, an incidence of 8.2% of stillbirth and 19.7% of growth restricted neonates have been reported in the literature. BP should be monitored in all four limbs with central aortic pressure monitoring during intrapartum period

to prevent hypertensive complications and convulsions. Caesarean section is done only for obstetric indications. Takayasu's arteritis should be regarded as a risk factor for aortic dissection. It is important to treat systemic hypertension in Takayasu's arteritis patients and suspect the diagnosis of aortic dissection in any pregnant patient complaining of chest pain as dissection is a leading cause of maternal mortality in the developed world.⁽¹¹⁾

Patients should be nursed in a high care unit postoperatively/post-delivery to detect complications early. After delivery, maternal peripheral resistance and left ventricular workload increases. This physiological change may lead to the development of pulmonary oedema, heart failure, renal dysfunction or cerebral haemorrhage. Use of immunosuppressive treatment may also increase the risk of puerperal infection.

CONCLUSION

Aortoarteritis, a rare vascular disorder if diagnosed by appropriate clinical physical examination and confirmed by MRI and arteriography is amenable to successful outcome with close cardiovascular monitoring. Although, pregnancy was complicated by hypertension and its sequelae, successful outcome could be achieved with timely admission, judicious medication and multidisciplinary approach.⁽¹²⁾

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