

An Unusual Case of Pregnancy with Severe Takayasu's Arteritis

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Abstract

Takayasu arteritis, though a life threatening disease, successful management of such high risk pregnancies is possible by the joint efforts of obstetrician, cardiologist and neonatologist. Needless to add, it requires the presence of a good infrastructure with proper monitoring facilities.

Introduction

Takayasu's arteritis is a chronic inflammatory arteriopathy of uncertain origin involving large and medium sized arteries with predominant incidence in young women (M:F 1:8), especially of Oriental origin. It was first described in 1908 by two Japanese ophthalmologists, Takayasu and Onishi. The onset of symptoms is often in the form of nonspecific complaints such as low-grade fever, muscle ache, joint pain, weight loss, night sweats, suggestive of severe inflammatory reaction. This makes the diagnosis almost impossible in its early stage. In the severe phase of the disease more specific symptoms suggestive of circulatory insufficiency such as vision disturbances, myocardial ischaemic chest pain and stroke appear.

In patients with Takayasu's arteritis, pregnancy acts as a burden to the already compromised circulatory system. Uncontrolled hypertension, cerebrovascular ischaemia, near complete occlusion of systemic and pulmonary arteries and congestive cardiac failure are known life threatening complications during late pregnancy and labour. Our report describes successful management of a 27 year old

primigravida with severe Takayasu's arteritis.

Case Report

A 27 year old primigravida came to the antenatal OPD at 24 weeks for registration. She was referred from Vapi (Gujarat), as a diagnosed case of Takayasu's arteritis. Patient was advised termination in first trimester which she refused with due risk.

Past History

Diagnosis of Takayasu's arteritis was confirmed three years ago, though she had been symptomatic for the last ten years. Three years back when she had developed Bell's palsy, the attending physician discovered absent pulses in the upper extremities. The diagnosis of Takayasu's arteritis was confirmed after whole body angiographic studies and was then treated with oral prednisolone and ticlopidine. Later she developed hypertension associated with continuous headache, dyspnoea on exertion and chest pain. Oral nicardipine and atenolol were added and coronary angiography was done. Left ventricle ejection fraction was 40%, left main coronary artery showed 100% ostial block and anterolateral wall motion abnormality. This was associated with intermittent 'black out' episodes followed by blurring of vision. A month later, she developed paroxysmal nocturnal dyspnoea and had to undergo two vessel stent angioplasty along with bilateral renal artery stenting for severe stenosis.

Antenatal Management

At her first visit patient was well aware of her medical status, counselling regarding the complications and prognosis was done.

On examination we found that left upper extremity pulses were absent, right radial and brachial pulses were sluggish, lower extremity pulses were well felt,

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so blood pressure was monitored on thigh using popliteal pulsations.

Investigations done were:

Routine investigations (CBC, LFT, RFT) were normal.

ECG showed left axis deviation. 2D-Echo, obstetric ultrasonography was normal.

Repeat angiography revealed 100% left subclavian block and bilateral complete internal mammary artery block. Bilaterally stented renal arteries were patent and functioning well.

She was followed up in the antenatal OPD every fortnightly, along with regular follow up with cardiologist and rheumatologist. For hypertension nicardipine and atenolol were continued, dosage of prednisolone was reduced.

Pregnancy progressed well under strict supervision and monitoring (especially of hypertension and cardiac status) until 32 weeks of gestation, when clinical assessment and ultrasonography showed intrauterine growth retardation.

Weekly foetal monitoring was done with NST. Ultrasonography was repeated at 34 weeks which showed IUGR along with oligohydramnios. Colour Doppler study was done and was found to be normal. Thereafter biweekly NSTs were done.

Keeping in view severity of the disease and associated IUGR and oligohydramnios, a decision of elective caesarean section was taken at 36 weeks of gestation. Caesarean was done under epidural anaesthesia.

Outcome was a healthy female baby weighing 2.2 Kg, apgar scores were 8 and 9 at 1 and 5 minutes respectively. Twenty units of pitocin infusion was given slowly after baby's delivery without significant haemodynamic changes. Postoperatively ICU monitoring of the patient was done as complications are more common in the postpartum period.

Baby was observed in NICU for 24 hours, no congenital malformations were detected. Postpartum period was uneventful. Patient was discharged on seventh postoperative day. No wound complications were encountered inspite of patient being on steroids for long term.

Discussion

Takayasu's arteritis is a progressive and frequently fatal nonspecific obstructive arteritis, probably of autoimmune aetiology.

Ueno *et al* classified Takayasu's arteritis into four types:

Types	Anatomical Location
I	Aortic arch and its branches,
II	Thoracic and abdominal aorta,
III	Combination of I and II,
IV	Pulmonary artery.

Ishikawa severity grading based on presence of complications (retinopathy, HTN, aneurysm and AR).

I	no complication,
IIa	one complication (mild),
IIb	one complication (severe),
III	two or more complications.

Our patient was in group III i.e. having more than one complication- hypertension and retinopathy. Overall maternal mortality rate of women with Takayasu's arteritis is 4-8 % and further pregnancy is not advised in patients with group IIb and III disease.

The main maternal risks of Takayasu is due to :

- arterial HTN with superimposed preeclampsia (60%),
- congestive cardiac failure,
- cerebrovascular events (5%).

Risk is greatest during III trimester and perinatal period.

Blood pressure changes during labour are greater in patients with Takayasu arteritis than in healthy patients, hence reasons for caesarean section depend on obstetric indications as well on severity of the disease.

Major foetal risks are intrauterine death (2-5%), IUGR (18%) and prematurity.

State of Takayasu in early pregnancy and magnitude of blood pressure elevation in late gestation are the most critical factors in determining the management and main aim of treatment is suppression of inflammation and preservation of vascular competence.