

Mortality in a second gravida following dissecting aortic aneurysm due to Takayasu's Arteritis

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Abstract: Takayasu's arteritis is a vasculitis involving the aorta and its branches. This is a case of undiagnosed Takayasu's arteritis in a 32 weeks pregnant lady, who presented to the emergency department with complaints of severe chest and epigastric pain and signs consistent with aortic aneurysm. The woman died of aortic dissection two days later.

Key words: Pregnancy, Takayasu's arteritis, aortic aneurysm, dissection

Introduction

Takayasu's arteritis is a chronic inflammatory disease of unknown origin that usually affects the aorta, its primary branches and occasionally the pulmonary and coronary arteries. The disease is most common in young reproductive women and more prevalent in eastern Asia compared to western countries.¹ The estimated incidence in the western world is 2.6 cases per million persons per year.² Aortic aneurysm associated with Takayasu's arteritis is not rare. The aorta with little calcification has a greater possibility of aneurysm formation in patients with Takayasu's arteritis. Aortic dissection following aneurysm is rare but life threatening and catastrophic. The most common predisposing cause of dissection is chronic hypertension, pregnancy, connective tissue disorder, and congenital abnormality of the aortic valve, coarctation of aorta and in patients with Marfan's disease. There is increase risk for dissection in pregnant than in non-pregnant women.

Case Report

Twenty four years old G₂ P₁₊₀, a housewife from Sindhupalchok at 32 weeks of pregnancy presented to labour room with the chief complaints of sudden onset of central and right sided chest and epigastric pain for about 2 hours which radiated to the back. The pain aggravated on lying flat. This was accompanied by shortness of breath,

profuse sweating and an episode of vomiting. However, there were no complaints of headache, blurring of vision, palpitation or paroxysmal nocturnal dyspnoea.

The index pregnancy was a supervised one and during her first four antenatal checks up, the 1st being at 16 weeks and the last at 29 weeks, she was normotensive, without albuminuria or edema.

There was history of hypertension in her previous pregnancy 2 years back which was recordable only in the intrapartum period. The pregnancy had ended in preterm delivery at 8th gestational months preceded by decrease in fetal movement and resulting in an intrauterine fetal death (IUFD). Nevertheless, she was not prescribed any antihypertensive or special medications during pregnancy or after the delivery.

At the time of admission in labour room, the patient was in distress due to severe epigastric pain, the radial pulse on the right was 76 beats/ min with good volume and blood pressure was 180/120 mm Hg. The pulse and blood pressure on the left side were unrecordable. Pallor and edema were absent, chest was bilaterally clear and flow murmur was heard in the apical region. Per abdomen examination revealed epigastric tenderness, uterus of 30 weeks size, single fetus, longitudinal lie, cephalic, not engaged and the fetal heart sound was 154/ beats minute with no uterine contraction

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or tenderness. On inspection of genitalia there was no swelling or oedema. Per speculum and vaginal examinations were withheld, as she was not in labour. Urinary protein in the dipstick showed 1+.

At the time of admission, medical consultation was done that showed a repeat record of unequal BP of 170/100 mm Hg on the right and 110/70 mm Hg on the left. The pulse on the right was 80 beats/minute. The left radial and the brachial pulses were weak while the rest of the peripheral pulses were palpable and had normal volume. There was no radio-radial or radio femoral delay, chest was clear and no obvious murmur was appreciated by the medical team. So with the differential diagnosis of arteritis, aortic aneurysm, dissection of the aorta she was admitted in the Intensive Coronary Care Unit (ICCU) and treated with intravenous fluids, parenteral glyceryl trinitrate, H₂ blockers, antacids, metachlorpromide, metoprolol and morphine. Investigations revealed normal electrocardiography (ECG), x-ray chest lateral view and serum amylase. Abdominal ultrasonography (USG) done in the following morning showed normal caliber of the abdominal aorta (2.2 cm), with separation of the intimal flap at the level of superior mesenteric artery; however the extent of which could not be well assessed. The flow in the distal segment was normal including the iliac bifurcation. So, the impression was dissection of abdominal aorta. Obstetric scan showed single live fetus of 31-32 weeks pregnancy. She was further advised to undergo magnetic resonance imaging (MRI) to see the extent of the dissection. Echocardiography showed minimal pericardial and left pleural effusion with no evidence of dilatation of the ascending aorta.

Electrocardiography (ECG) done twice was normal but the third ECG showed sinus tachycardia with short QT interval.

Vascular surgery team was also consulted and they advised to continue with the same antihypertensive to keep the blood pressure on the lower side and to repeat an echocardiography to assess the need of surgical intervention in case there was involvement of the ascending aorta.

On the second day of admission the patient was transferred to Coronary Care Unit (CCU), where she started to complain of sudden onset of shortness of breath. At that time her heart rate was 120-130/minute. She looked extremely pale. Her peripheries were cold and clammy. The blood pressure was not recordable. Oxygen was supplemented, wide bore intravenous cannula was secured and intravenous fluids were started. With the development of gradual bradycardia, cardiopulmonary resuscitation (CPR) was initiated with the observation of sinus rhythm. USG repeated in the CCU could

not localize the aorta. Repeat echocardiography revealed pericardial effusion which on tapping showed frank blood.

Despite all the measures she went into asystole and was declared dead. The probable cause of death was right haemothorax with pericardial tamponade with rupture aorta leading to cardiogenic and hypovolumic shock.

Discussion

In 1990, the American College of Rheumatology suggested a set of criteria for the diagnosis of Takayasu's arteritis. The criteria consisted of

- a) Age less than 40 years
- b) Claudication of an extremity
- c) Decreased brachial artery pulse
- d) More than 10 mmHg difference in the systolic pressure between the left and the right arm
- e) A bruit over the subclavian arteries or the aorta
- f) Angiographic evidence of narrowing or occlusion of the aorta or its primary or proximal branches.²

Presence of three of the six criteria is required for the diagnosis. This patient fulfilled three out of six criteria, so this could be a case of Takayasu's arteritis.

Though rare, aortic dissection should be thought of whenever a pregnant women presents with preeclampsia superimposed on chronic hypertension and with intractable chest pain and normal ECG. Usually patients with history of Marfan's syndrome and Ehlers-Danlos-syndrome are the ones who have aneurysm and dissection of the aorta.³ However it has also been seen that the rupture of the dissecting aneurysm of aorta occur more frequently in pregnant than in non pregnant women.

This patient was 24 years old and studies have shown that the mean age of the presentation was usually 23.6 ± 3.6 years.⁴ Half of the aortic dissection in women below 40 years has been seen to occur in association with pregnancy. The presenting symptoms during pregnancy are usually severe hypertension and unequal pulse which this patient had.⁴ Absent or decrease deficit of pulse is usually present in 30% of the patients.⁵

The risk in Takayasu's arteritis is mainly during the third trimester. Many reports have stated about its detection and fatal consequences at 32-38 weeks period of gestation which is related to the expansion of aneurysm more during labour and delivery with or without Marfan's syndrome.⁶

Most of the patients do present like the preeclamptics. It is one of the presentations with poor fetal prognosis. The

patient also had a past history of increase blood pressure and probably she was preeclamptic then, that lead to the loss of her baby, but the diagnosis of Takayasu's arteritis was missed then as well as in this pregnancy.

This patient had normal and palpable pulse as well as blood pressure in her right hand but the pulse and the blood pressure in the left hand was difficult to appreciate. Her blood pressure throughout the antenatal period was normal, till she came to labour room with the hypertensive crisis of 180/120 mm Hg only in her right arm; probably the blood pressure in the left hand was never ever taken during the ANC. Thus, the habit of taking the blood pressure bilaterally in the entire patients coming for the antenatal check up is advisable, when there is a suggestive history of fetal loss or if at all hypertension is detected, even if it might have been just transient record.

Studies depicted a large number of cases coming to emergency room with the history of sudden increase in blood pressure, headache, haemoptysis, chest and epigastric pain, pain in neck, throat, in abdomen and lower back, syncope, paresis, dyspnoea etc. These signs and symptoms are actually boon in disguise which may help making the right diagnosis of the condition before it may have fatal consequences like in this case. A lady with Takayasu's arteritis with aortic dissection and haemopericardium as this case had initial presentation of maxillary pain which later radiated to the chest with fatality occurring in less than 12 hours.⁷

Dissection of the ascending aorta is infrequent during pregnancy. Given this low prevalence, diagnosis is often delayed or even unrecognized. The delay in the diagnosis can have a highly jeopardizing effect both on mother and fetus including ours.

The prognosis of the mother with dissecting aortic aneurysm is extremely fatal and frightful with 94% of cases dying, most of them within first 48 hours as did this patient.⁷ Fourty eight percent of the women under the age of 40 who have acute dissecting aortic aneurysm are pregnant.⁸ Aortic dissections seldom affect young women. Nevertheless the structural changes that take place in the arterial wall, which occurs during pregnancy, may predispose to this complication, particularly in the third trimester and the postpartum period.

The diagnosis is usually delayed or missed as there is predilection for the patient with Takayasu's arteritis to present with the features of preeclampsia leading to aneurysm and then to dissection.⁹ Obstetricians are therefore forced with the dilemma of suboptimal management in these patients, as we too initially thought it to be a case of

impending eclampsia because she had features like rise in the blood pressure of 180/120 mmHg on the right side, severe epigastric pain and intractable chest pain, vomiting, sweating, but when the pulse and the blood pressure on the left side was found to be very feeble and almost unrecordable, medical consultation was then sought for, changing the diagnosis from impending eclampsia to probably Takayasu's arteritis

Aortic dissection requires a prompt diagnosis and must be treated without delay in order to reduce the mortality. This patient died of sudden and intractable cardiovascular shock and probably we could have been able to save the patient if there was an early and prompt diagnosis, immediate caesarean section, followed by the surgical repair of the dissection which might have saved her is our speculation. So, delay in diagnosis was fatal for both the mother and the baby.

Conclusion

In any patients presenting with a features of preeclampsia, a rare possibility of Takayasu's arteritis with aneurysm should be borne in mind. Thus, keeping a high level of suspicion in such cases; early diagnosis and prompt treatment are very crucial to achieve a successful outcome, as these could terminate with fatal consequences from aortic aneurysm and dissection ultimately leading to maternal and fetal mortality as in this patient.

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