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Case Report

A rare case of peripartum cardiomyopathy with takayasu arteritis

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ABSTRACT

29 years old primigravida 38.3 weeks came with complaints of bleeding per vaginum and sudden onset breathlessness. Major finding on examination was that there was no pulse in the left hand along with raised BP, low SPO2 and B/L crepitations. Emergency management was given and patient stabilized temporarily. D/D were discussed with acute LVF due to PPCM most likely and patient was taken up for emergency LSCS. Post section 2D ECHO was performed showing decreased EF. Peripartum cardiomyopathy is an uncommon but potential life-threatening cardiac failure of unknown etiology, encountered late in pregnancy or in the postpartum period. Diagnosis of PPCM should essentially include echocardiographic substantiation of left ventricular dysfunction. TA is a vasculitis that mainly affects women of childbearing age, so it is possible to find pregnant patients with the disease. Women with Takayasu arteritis require multidisciplinary management prior to and during pregnancy. Monitoring for and reducing risk of preeclampsia, FGR and thromboembolic disease are helpful in achieving favorable outcomes.

Keywords: Peripartum cardiomyopathy, Takayasu's arteritis, Vasculitis, Aortic arch syndrome

INTRODUCTION

Peripartum cardiomyopathy is a relatively rare disease, which can have devastating consequences and should be promptly identified and correctly treated. Overall prognosis is a good majority of the cases, although some patients may progress to irreversible heart failure. Early diagnosis is important and effective treatment reduces mortality rates and increases the chance of complete recovery of ventricular systolic function. Mortality of these women is quite high. The use of inotropic drugs and mechanical circulatory support devices may be necessary in the initial phase of severe forms of acute PPCM. Many patients, after initial stabilization, recover left ventricular dysfunction.[1-3]

Unfortunately, some patients need further mechanical circulatory support or urgent heart transplantation despite maximal therapy. In addition, the time frame and extent of recovery is unpredictable, and patients may suffer from cardiac arrest due to ventricular fibrillation in the first months after diagnosis.[4] The clinical course may be further aggravated by atrial and/or ventricular thrombus formation with subsequent cardio-embolic complications.[5]

We report on an interesting case with a favorable outcome and discuss the clinical presentation, therapy and outcome of this condition with undiagnosed Takayasu's arteritis. Takayasu's arteritis is a large vessel vasculitis affecting younger to middle aged women. The importance of physical examination especially of peripheral pulses is important especially in cases of peripartum cardiomyopathy with Takayasu's arteritis.

CASE REPORT

A 29 years old primigravida 38.3 weeks came to casualty. She presented with cough 3-4 days. Leaking per vaginum since 1 hour. And sudden onset breathlessness for 1 hour.

On general examination: patient was agitated and unable lie in supine position due to tachypnoea. Pulse-140 b/min in right arm. pulse was not recordable in left arm. RR-28/min, BP-130/80 mmhg. SpO_2 was found to be 89 to 90% on RA, RS-bilateral crepitations +, CVS S1S2+, no murmur.

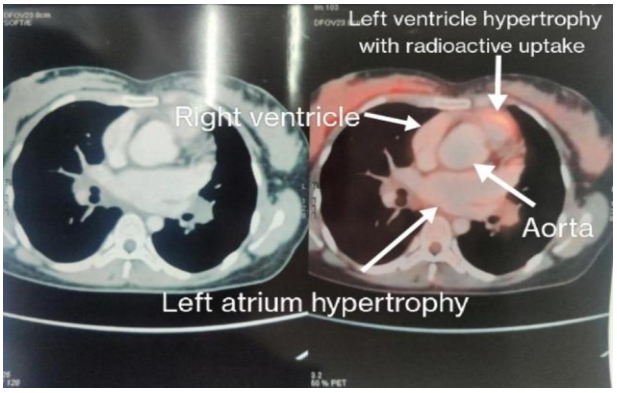

Figure 1: PET scan suggesting LV and LA.

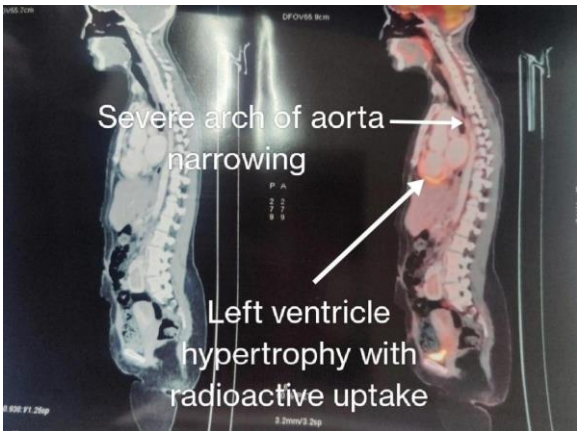

Figure 2: Severe narrowing of arch of aorta.

Per abdominal examination revealed full term uterus with cephalic presentation with fetal heart rate of 140 beats per minute and uterine activity of 1/10/15.

Per vaginal examination showed cervical dilatation of 2 cm with 40% effacement and absent membranes and frank leak present with station at -2.

Emergency management followed and effort was made to stabilize to patient. Propped up position with 100% oxygen supplementation was started by face mask with that saturation became 94 to 95%. Bilateral IV line inserted and foley catheter put under aap.an ECG was done immediately which tachycardia with left bundle branch block s/o LVF. Injection Lasix 40 mg IV stat given with sublingual nitrate 0.2 mg stat. Continuous ctg monitoring done and Investigations were sent.

Case discussed with ICU intensivist and physician anesthesiologist and T neonatologist informed. Differential diagnosis discussed were-1) LVF mostly due to PPCM 2) pulmonary /amniotic fluid embolism 3) acute lvf due to preeclampsia.

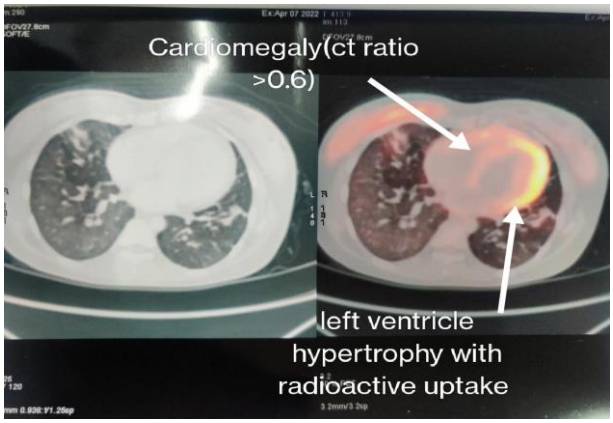

Figure 3: Cardiomegaly (increased CT ratio).

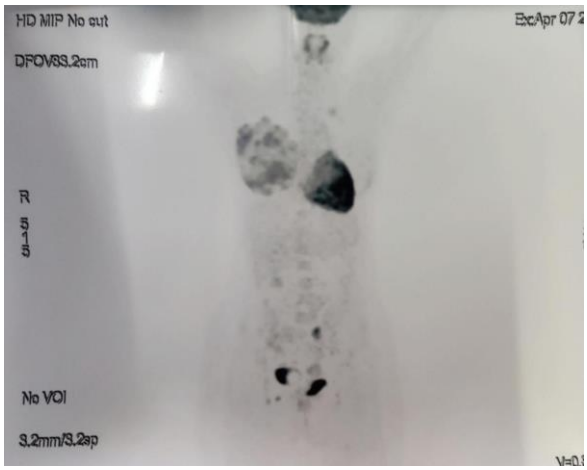

Figure 4: Radioactive uptake in various tissue and severe vascular stenosis.

Probable diagnosis of acute LVF due to PPCM was made and continuous ctg monitoring showed late decelerations. pv examination done and patient was assessed. Patient was posted for emergency LSCS i/v/o? left ventricular failure with fetal distress. Post section patient shifted to ICU. 2D ECHO done s/o EF 20-25% with deranged LFT. Patient was on mechanical ventilatory support and weaned off subsequently, DIURETICS started (T digoxin, T ivabrad, T ecosprin 75), IV Antibiotics were continued with INJ CLEXANE 0. 4MG. Patient was off O_2 and SpO_2 of 98% on RA and shifted to ward on day 3 of LSCS. 2D ECHO was done within 10 hours' s/o EF 20 TO 25% and diagnosis confirmed. LMWH started 0.4 mg sc started 12 hours after surgery. Patient required ventilatory support for 48 hours and weaned off subsequently. Patient was shifted to ward on day 5 repeat 2D ECHO on day 8 s/o EF 35 to 40%. patient was stabilized by day 5 so expressed breast milk was given to baby. Serial monitoring and follow-up advised to diagnose residual LV dysfunction.

PECT SCAN was done on follow up. S/o findings confirming Takayasu's arteritis. And showed diffuse hypermetabolism of the arch of aorta with left atrium and ventricle affection.

After 10 days when EF improved. Patient started breastfeeding the baby. Patient was discharged. And explained about recurrence of PPCM in next pregnancy and contraception was advised.

DISCUSSION

This case report highlights the diagnostic and therapeutic dilemma that physicians face when encountering patients with the rare, but interesting condition of peripartum cardiomyopathy with Takayasu's arteritis which warrants prompt identification and treatment.

PPCM usually presents with classical symptoms and sign of systolic heart failure with ventricular enlargement and dysfunction seen on echocardiography. Often there is significant mitral and tricuspid regurgitation.[6]

Unusual presentations include thromboembolism or hepatic failure secondary to heart failure. The development of heart failure and the usual time of diagnosis are during the post-partum period in more than 90% of the cases.[7]

PPCM can occur at any age with a higher incidence in women older than 30 years.[8-9]

The diagnosis of PPCM is challenging because most women in the last months of a normal pregnancy or soon after the delivery experience dyspnea, fatigue and pedal oedema. Symptoms and signs which should raise the suspicion of heart failure and could help the clinicians in the diagnosis, are the presence of paroxysmal nocturnal dyspnea, nocturnal cough, new regurgitant murmurs, pulmonary crackles, jugular venous distention and hepatomegaly.[10]

The etiology and pathogenesis seem to be multifactorial and poorly understood with the available literature rather conflicting.

Gestational hypertension, tocolytic therapy and twin pregnancy have been proposed as possible risk factors because they were commonly associated with PPCM. These observations were not confirmed from other authors who did not find any association between PPCM and history of hypertension during pregnancy or use of tocolytic agents.[11] The association between PPCM and twin pregnancy could support the theory of autoimmunity as a possible mechanism. This could depend on an excessive traffic of hematopoietic lineage cells from the fetus to the mother as manifest in twin pregnancy.[12]

Usually, the lower concentrations of these foreign proteins could contribute to tolerance of the fetus while increased levels could theoretically lead to the initiation of autoimmune disease.[8]

The weak immunogenicity of the paternal haplotype of the chimeric cells or the naturally immunosuppressive state of the mother or both could avoid rejection of fetal cells during pregnancy. Following postpartum, the recovery of immune competence could trigger a pathologic autoimmune response against cardiac cells where hematopoietic cells have taken up residence during pregnancy and therefore myocardial cells are recognized as nonself.[13]

Multiparity could be another risk factor for the development of PPCM10 and again, this observation has not been confirmed by other authors. In fact, more than 50% of the patients are at their first or second pregnancy.[9]

Molecular markers of an inflammatory process are found in most of the patients. 90% of the patients show high levels of plasma C-reactive protein that correlated positively with LV end-diastolic and end-systolic dimensions and inversely with LV ejection fractions.[11] This could indicate the chronic inflammatory state at baseline, which is higher in more unstable patients. The presence of a low-grade chronic inflammatory process could be due to the release of endotoxins and subsequent release of pro-inflammatory cytokines.[14]

Interestingly, the possibility of developing PPCM in a surrogate mother whose embryo was received from the commissioning couple of whom the mother was diagnosed with PPCM following the delivery of her first child, points out the possible role of the presence of a transmissible agent, either infectious or non-infectious as well as genetic susceptibility.[15] A hereditary predisposition is also suggested by familial reports of PPCM and strong consideration should be given to screening family members because PPCM may be of a genetic predisposition to cardiomyopathy.[13]

Management

Management of acute PPCM The management of heart failure around pregnancy is challenging and in the absence of evidence-based data, the initial management of patients with PPCM is similar to the treatment of AHF or other etiologies. Interdisciplinary approaches of cardiologists, intensivists, obstetricians, neonatologists, anesthetists and cardiac surgeons are necessary in cases of severe AHF. Prespecified protocols of interdisciplinary work-up of these patients are helpful. Timely diagnosis and treatment delivery are crucial, the initial treatment of patients with severe forms of acute PPCM is significantly different to those of stable patients.

Prognosis

Overall prognosis of PCM is good in the majority of the cases, although some patients may progress to irreversible

heart failure. Progression of the condition requiring heart transplantation is described in 4% and death in 9% at two years follow up.[14]

Other studies showed a much higher mortality rate such as 15% or 32% at 6 months.[13-15] Patients who eventually die tend to have worse NYHA functional class, LVEF and larger LV dimensions at diagnosis.[13] There seems to be an initial high-risk period with 25-50% of the women dying within the first 3 months postpartum.

Sudden cardiac death has been reported to account for up to 50% of the mortality and therefore attention should be paid to identify those patients who are likely to experience a late recovery of systolic function from those who should be considered for implantation of a cardioverter-defibrillator. Mortality rates have decreased over the past 10 years due to advances in medical therapy for heart failure and use of implantable defibrillators. Normalization of left ventricular systolic function occurs in 23% and in 54% of the patients respectively at six months 13 and at two years and is more likely if EF at diagnosis is more than 30%.[9]

Higher left ventricular end diastolic dimension and lower fractional shortening at diagnosis seems to be associated with a worse prognosis. A fractional shortening of less than 20% and a left ventricular end diastolic dimension of 6 cm or greater was associated with a more than 3-fold higher risk for persistent left ventricular dysfunction. 75% of the patients, who recover, have an EF of more than 45% at two months after diagnosis.

Complete recovery of systolic function occurs usually in the first 6 months after delivery 9 although the recovery phase need not be limited to the first 12 months. Continuing improvement was observed in the second and third year after diagnosis.[17] Persistence of the disease after 6 months portends worse survival.

To get an assessment of prognosis at the time of diagnosis, a dobutamine stress echocardiography study could be performed in non-critically ill patients. Inotropic contractile reserve accurately correlated with subsequent recovery of left ventricular function and usually associated with a benign prognosis. MRI could be a useful alternative to dobutamine stress echocardiography in predicting outcome. Delayed gadolinium enhancement is more likely to be present in patients less responding to conventional therapy but could resolve over time in the recovering patients.

Even if left ventricular function recovers completely, exercise tolerance may remain abnormal and this could be more objectively assessed by an abnormal response to dobutamine stress echocardiography.

Management involves conventional therapy for heart failure with diuretics, ACE-inhibitors, beta-blockers and aldosterone antagonists. Angiotensin-receptor blockers should be added in case of ACE-inhibitors intolerance. Anticoagulant therapy should be considered in view of the low left ventricular EF, which predisposes to thrombus formation, especially in the peripartum period when a hypercoagulable state exists. In patients not improving on conventional therapy or in patients with critical hemodynamic state with cardiogenic shock, hemodynamic support with pressors should be considered. There have been some reports about the use of levosimendan in non-responsive patients.

Non-responsive patients should be considered for heart transplantation even if there are some reports about effective use of extracorporeal membrane oxygenation, intra-aortic balloon pump or mechanical assist devices.

Among patients who eventually recover, the withdrawal of heart failure medications was not associated with decompensation over a follow-up of 29 months. The duration of heart failure treatment is determined by the patient's heart performance at rest and with exertion. Patients with normal EF at rest and during dobutamine could taper off medical therapy in 6-12 months; patients with normal EF at rest and abnormal EF during dobutamine should be treated for a longer period with ACE-inhibitors and beta blockers. Patients who continue to have a depressed ventricular function at rest have a poorer prognosis and should receive medical therapy indefinitely. In any case, it seems reasonable to continue at least for a year with ACE-inhibitors and beta blockers even in case of complete recovery.

It is important to note that the use of ACE-inhibitors should be limited to after delivery since they have teratogenic effects. Other drugs like immunosuppressive drugs are still under evaluation.

A subsequent pregnancy carries a high risk of relapse, significant decrease of left ventricular function and mortality. Mortality rate is described to be approximately 55% during subsequent pregnancy even though it seems associated more with patients who entered the subsequent pregnancy with abnormal systolic function i.e., without making a complete recovery. Complete recovery from a relapse is very rare. There is no consensus regarding recommendations for future pregnancy after PCM but patients whose left ventricular size or function does not return to normal should be counseled strongly to avoid subsequent pregnancy.[13]

Takayasu's arteritis is a LVV with aortic inflammation leading to proximal occlusion and/or aneurysms of carotid, subclavian, pulmonary, iliac, and renal arteries. Mean age is typically reported between the second and third decade of life. Its etiology remains primarily idiopathic. Autoimmunity, sex hormones, and genetic (predisposition of the human leukocyte antigen, HLA BW52) factors have often been hypothesized as plausible factors causing it. These factors were conspicuously absent in the case described. Various types of TA have been acknowledged

in the past: type I (disease embroiling aortic arch and its branches), type II (lesions constrained to descending thoracic aorta and abdominal aorta), type III (patients with characteristics of types I and II), type IV (involvement of pulmonary artery), and type V (combined features of types IIb and IV).

Hypertension is fairly common due to reduction in elasticity and narrowing of the arteries, besides abnormalities in functioning of aortic and carotid baroreceptor's function, it should be prudently contained during pregnancy, as severe renovascular hypertension, cardiac involvement, or pulmonary hypertension is associated with poor fetomaternal prognosis like abortion, preterm labor, and low birth weight babies. Blood pressure in such patients should be also measured in the lower extremity to pick up blood pressure discrepancies; like in our case BP recordings in lower extremity were higher than upper extremity.

Besides, pulselessness of unilateral or bilateral radial arteries and vascular bruit should be seen in all cases of hypertension. Involvement of abdominal aorta is associated with adverse pregnancy outcomes, which was fortunately absent in the present case. Arterial ultrasound Doppler, quantifying the flow in the uterine arteries, is beneficial in evaluation of fetal well-being and growth in women with TA. Management of TA entails an interdisciplinary approach with involvement of obstetricians, anesthesiologists, cardiologists, rheumatologists, and neonatologist in a tertiary care center. The aims are control of inflammation, prevention and treatment of complications like hypertension and revascularization by percutaneous angioplasty, use of endoprosthesis, or surgical correction for occlusive and stenotic lesions.

CONCLUSION

PPCM is a relatively rare disease, which can have devastating consequences and should be promptly identified and correctly treated. Early diagnosis is important and therefore women who develop symptoms of heart failure during pregnancy or shortly after should be investigated for this condition. Effective treatment reduces mortality rates and increases the chance of complete recovery of ventricular systolic function. Management of TA entails an interdisciplinary approach with involvement of obstetricians, anesthesiologists, cardiologists, rheumatologists, and neonatologist in a tertiary care center. The aims are control of inflammation, prevention and treatment complications like hypertension and revascularization by percutaneous angioplasty, use of endoprosthesis, or surgical correction for occlusive and stenotic lesions.

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