Peripheral Vascular Disease: An Unusual Presentation of Peripartum Cardiomyopathy

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Peripartum cardiomyopathy (PPCM) usually presents with the heart failure in the last month of pregnancy or up to 5 months post-partum period, without any underlying cause of cardiac failure. This is a hypercoagulable state associated with left ventricular (LV) systolic dysfunction, thereby causing thromboembolic complications. We report a case of 2 months post-partum female who presented with left lower limb arterial thrombotic occlusion which turned out to be secondary to embolization from a large LV thrombus resulting from PPCM. The present case is an unusual presentation of PPCM that usually presents with a clinical feature of cardiac failure.

Keywords: Cardiac failure, Peripartum cardiomyopathy, Peripheral vascular disease, Thromboembolism

INTRODUCTION

Peripartum cardiomyopathy (PPCM) is a disease which occurs between the last month of pregnancy and the first 5 months post-partum and presents with left ventricular (LV) systolic dysfunction.¹ The etiology of the disease is unknown, and it is a diagnosis of exclusion, wherein patients have no prior history of heart disease and there are no other known possible causes of heart failure. The incidence of PPCM varies from one case per 4000 live births in the US to one case per 1000 live births in South Africa or one case per 299 live births in Haiti.² PPCM is associated with dilatation of both the ventricles with reduced ejection fraction (EF). In these women, contraction abnormalities with stagnant blood flow can lead to LV thrombus formation. A high initial end-systolic volume, as well as a LV EF <35% are found to be major predictors of LV thrombus formation. There is a consistent, graded relationship between increasing LV size (LV end diastolic dimension [LVED] diameter > 60 mm) and the occurrence of LV thrombus. LV thrombus is detected in up to 17% patients of PPCM, with a potential risk of embolization, manifesting as either stroke or peripheral arterial disease. The characteristics of thrombus associated with highest risk for embolization include large size, increased friability, mobility, protrusion into the LV cavity, diffuse LV dilatation, and impaired systolic function. A peripheral embolic event is associated with threefold higher mortality compared to patients with severe systolic dysfunction without an embolic event. The highest sensitivity and specificity for LV thrombus detection is reported for contrast-enhanced magnetic resonance imaging (MRI) compared with transthoracic echocardiography and transesophageal echocardiography. However, two-dimensional (2D) echocardiography provides a non-invasive means of imaging the LV and its apex with precise spatial orientation and is a valuable tool for the detection and follow-up of a LV thrombus. Despite the high risk for systemic embolization, there is a lack of standardized therapeutical regimens for the treatment of LV thrombi. Anticoagulation, antiplatelet therapy, surgical thrombectomy, and thrombolytic therapy are the treatment options. Anticoagulant therapy has had variable success, with thrombus resolution rates ranging from 13% to 59%. Thrombolysis with urokinase, streptokinase, and tissue plasminogen activator has been tried, but the risks of hemorrhage may be unacceptable. Surgical thrombectomy is also advocated, but generally patients with PPCM are at an increased risk for perioperative morbidity and mortality. Most studies suggest that when LV thrombus is diagnosed in these patients, anticoagulation may be started and surgical treatment may be delayed. Family planning counseling is an important aspect after diagnosis of PPCM.
A subsequent pregnancy after a diagnosis of PPCM carries a higher risk of relapse and patient should be advised an appropriate method of family planning. Since combined oral contraceptives increase the risk of thromboembolism, their use should be discouraged. We present a case report of a 2 months post-partum female with PPCM with LV thrombus, presenting with left lower limb thrombotic arterial occlusion secondary to embolization.

CASE REPORT

A 25-year-old, 2 months post-partum, non-diabetic, non-hypertensive, none smoker female presented to the emergency department of Guru Gobind Singh Medical College and Hospital, Faridkot (Punjab) in February 2015 with complaints of pain and swelling of left leg for 1-month. She had a normal vaginal delivery at home 2 months prior to admission. The patient denied any history of fever, trauma, prolonged immobilization or usage of oral contraceptive pills prior to the onset of symptoms. There was no history of easy fatiguability, chest pain, exertional dyspnea, and similar episodes in the past in the family.

On physical examination, patient had a regular rapid pulse rate of 118/min, blood pressure of 110/80 mm Hg, respiratory rate of 18/min, and temperature of 37°C. Cardiovascular examination revealed an apex beat that was shifted down and out with normal heart sounds and no murmur. Chest examination revealed bilateral vesicular breathing. Patient’s left leg was swollen, tender, and had decreased surface temperature with black discoloration of toes (Figure 1). Peripheral pulses including dorsalis pedis, anterior and posterior tibial were diminished.

Chest X-ray showed moderate cardiomegaly (Figure 2), and electrocardiogram showed sinus tachycardia. Laboratory investigations included complete blood count with a hemoglobin of 7.0 g/dL, a total white cell count of 6,000/mm³ and platelet count of 306,000/mm³. The patient had normal prothrombin time (15 s), normal liver and renal function tests. Arterial Doppler of the left leg showed thrombus at the junction of left external iliac and common femoral arteries with a poor and patchy flow in the superficial femoral artery.

A 2D Transthoracic Echocardiogram showed enlarged LV with a large intracardiac thrombus of size 2.5 cm × 2.5 cm located at the LV apex protruding into the LV cavity (Figures 3 and 4). Global LV systolic dysfunction was also present with LV EF of 34%, mild mitral regurgitation, mild tricuspid regurgitation and minimal pericardial effusion. Hematologic tests for hypercoagulable states including protein C and S deficiency, antithrombin III deficiency, Factor V Leiden, lupus anticoagulant, antiphospholipid antibody and homocystenemia were also done to rule out confounding causes of peripheral vascular disease, and they were within normal limits. With all the above parameters, the patient was diagnosed to have PPCM with LV thrombus with peripheral thromboembolism.

Cardiac MR (CMR) was also done and revealed a hypointense thrombus in the LV (Figures 5 and 6).
was adjusted to 4 mg daily to maintain an international normalized ratio between 2 and 3 without any bleeding complications.

On the 21st day, she revisited the hospital for follow-up and repeat echocardiogram, which showed no visible intracardiac thrombus, although the LV systolic function remained depressed.

**DISCUSSION**

A disease of unknown etiology, PPCM presents with LV systolic dysfunction and symptoms of heart failure between the last month of pregnancy and the first 5 months post-partum. Increased age, gravidity or parity, toxemia or hypertension of pregnancy, use of tocolytics, twin pregnancy, obesity, and low socioeconomic status are some of the risk factors known to cause the disease. Some potential etiologies like myocarditis, abnormal immune response to pregnancy, maladaptive response to the hemodynamic stresses of pregnancy and stress-activated cytokines are known to be associated with the disease. Signs and symptoms of PPCM resemble systolic heart failure and it is a diagnosis of exclusion. Diagnosis is not considered until other causes of heart failure are ruled out. Echocardiography is central to the diagnosis and it reveals an EF of $<45\%$ and/or fractional shortening of $<30\%$, along with a LVED $>2.7$ cm/m$^2$ of body surface area.

In women with PPCM, LV thrombus is detected in up to 17% of patients in whom the risk of systemic embolization is high. Ventricular thrombus formation occurs more frequently in PPCM patients with an EF of $<35\%$. Post-partum period is a phase of procoagulant activity due to the elevation of factors VII, X, VIII, fibrinogen, and von Willebrand factor and therefore PPCM carries a high risk of thromboembolism.

The presence of an LV thrombus carries a high risk of peripheral embolization. An increased risk for embolization is reported for cases in which the thrombus protrudes into the ventricular cavity or shows increased mobility with an annual risk of $1.4-12\%$. An embolic peripheral event is associated with a significantly higher mortality ($38.9\%$) compared to patients with severe systolic dysfunction without an embolic event ($10.3\%$).

Although 2D trans-thoracic echocardiography is the most common used technique for non-invasive identification and follow-up of LV thrombus, the recent introduction of CMR may improve thrombus detection.

Patients with PPCM have a varied natural course, with 50-60% of patients showing complete or near-complete recovery within the first 6 months post-partum; the
remaining may show further clinical deterioration, leading to cardiac transplantation or premature death, or persistent LV dysfunction and chronic heart failure. A baseline EF of <30% carries higher chances of improvement in LV systolic function. Patients with low EF have a poor long-term outcome and a higher-risk of thromboembolic complications. So, a minimum of 6 months of anticoagulation is recommended to prevent life-threatening embolization in women with PPCM.

A subsequent pregnancy after a diagnosis of PPCM is associated with a higher relapse rate if LV systolic function is not fully recovered previously, and even full recovery carries higher chances of relapse.

**CONCLUSION**

This case of peripheral vascular disease secondary to embolization from a large intracardiac thrombus, without clinical evidence of heart failure, is extremely unusual in PPCM. Hence, intracardiac thrombus with peripheral embolization is a potentially dangerous complication of PPCM. So, every peripartum female presenting with a newly oncoming peripheral vascular disease should be evaluated for PPCM so that early intervention can reduce further morbidity and mortality.

**REFERENCES**


How to cite this article: Garg R, Aggarwal S, Kaur S, Chawla SP, Samberia Y. Peripheral Vascular Disease: An Unusual Presentation of Peripartum Cardiomyopathy. IJSS Case Reports & Reviews 2015;2(3):16-19.

Source of Support: Nil, Conflict of Interest: None declared.