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Peripartum cardiomyopathy: report of a catastrophic case of a poorly understood disease

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Topic(s):
Echocardiography: Masses and Sources of Emboli

Citation:
European Heart Journal - Cardiovascular Imaging (2019) 20 (Supplement 1), i284

Peripartum cardiomyopathy (PPCM) is an idiopathic form of heart failure that occurs from the last months of pregnancy to the first 5 months of the postpartum period. Its pathophysiology is poorly understood and is associated to significant morbidity and mortality, with a high risk of systemic and pulmonary arterial embolic events.

A 37 years-old female presented to the emergency room (ER) after 5 weeks of delivery, with a 5 days history of progressive dyspnea. She was previously healthy, and her prenatal follow-up was entirely normal. Physical exam revealed regular pulse and in auscultation a +/6+ systolic murmur over the mitral area, with a marked S3 sound. Of note, the patient had jugular distension, was tachypneic at rest, and had late inspiratory fine crackles in the lung bases. Electrocardiogram showed sinus rhythm, with signs of left ventricular overload and left atrial enlargement. Transthoracic echocardiogram (TTE) revealed 4 chamber enlargement and severe right (RV) and left ventricular (LV) systolic dysfunction with diffuse hypokinetic walls. There were signs of elevated left atrial pressure and moderate mitral regurgitation (functional). A large mass was seen occupying almost the entire LV apex, multilobulated and highly mobile, with heterogeneous echogenicity, better depicted with intravenous contrast, suggesting thrombus with different ages. Another elongated mass was seen extending from the apex through the anterior wall, very close to the LV outflow tract, with great mobility, depicted in details by three-dimensional echocardiography. Cardiac magnetic resonance confirmed the aspect of an apical thrombus, with no significant late-enhancement on the myocardium. Non-fractionated heparin was started immediately, and after a few days with oral warfarin the patient was discharged, after clinical improvement and total disappearance of the masses.

After 5 days the patient evolved with abdominal pain, returning to the ER for evaluation. Abdominal computed tomography (CT) revealed ischemic lesions in the liver and kidneys. A new TTE showed the recurrence of thrombus in the LV and new thrombus in the RV, with high mobility, and low molecular weight heparin was initiated. After a few hours the patient suddenly developed torpor, deviated gaze and aphasia. A cerebral CT angiography revealed acute occlusion of the right middle cerebral artery and the patient was submitted to intra-arterial thrombolysis, with complete angiographic and clinical resolution. The patient was discharged after 30 days of hospital stay, with partial improvement of biventricular dysfunction and complete resolution of the intraventricular thrombus.

As a conclusion our case underscore a rare and catastrophic presentation of PPCM, and highlights the use of multimodality imaging for the assessment of the disease and its complications. TTE is a first line tool, and provides important information for the prognosis and to elaborate the correct therapeutic strategy for these patients.
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