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CASE REPORT

Management of Wolff-Parkinson-White Syndrome in a Patient with Post-partum Cardiomyopathy

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Abstract

Wolf-Parkinson-White (WPW) syndrome is a congenital heart condition in which the atrioventricular (AV) node is bypassed by an accessory pathway that connects the atria and ventricle directly. WPW syndrome in patients with a history of peripartum cardiomyopathy (pregnancy-related cardiomyopathy) is associated with a high risk of morbidity and mortality secondary to failure of the pump and the conduction system of the heart. Management of these cases deals with arrhythmia and systolic heart failure, which becomes more challenging in pregnant patients as it requires treatment methods that minimize risks to the fetus. We report a case of a young female patient with WPW syndrome and post-partum cardiomyopathy presenting with symptomatic arrhythmias (tachycardia).

Keywords: Wolf-Parkinson-White (WPW), Postpartum cardiomyopathy, Arrhythmias, Tachycardia

1. Introduction

Wolf-Parkinson-White (WPW) syndrome is a congenital heart disease in which an accessory pathway directly connects the atria and ventricle, bypassing the atrioventricular (AV) node. The characteristic WPW pattern EKG includes a short PR interval (0.12 s) and a delta wave with a wide (0.12 s) QRS complex. The delta wave results from rapid ventricular activation via the accessory pathway (preexcitation). In the general population, the prevalence of WPW pattern EKG is around 0.13 to 0.25 percent. In comparison prevalence of WPW syndrome is as low as 2 percent of patients with WPW patterns on surface EKG.

Diagnostic criteria involve pre-existing WPW patterns on EKG and arrhythmia involving the accessory pathway. Asymptomatic adults with WPW patterns are generally not treated. In contrast, those with symptomatic arrhythmias should be treated. We report a case of a young female patient with WPW syndrome and postpartum cardiomyopathy presenting with symptomatic arrhythmias (tachycardia).

2. Case presentation

29-year-old African American female with a past medical history of opiate abuse and currently two months into her post-partum period presented to ED complaining of constant, sharp left-sided chest pain for the past four days which was mildly relieved with ibuprofen. She described it to be exacerbated by deep breaths and associated with cough and blood-tinged sputum. She endorsed palpitations lasting all day long and two episodes of non-bloody, non-bilious emesis. Reported bilateral lower extremity edema with associated orthopnea and PND, for which she uses 2–3 pillows at night. Of note, she is bed-bound most of the time, secondary to her lower extremity swelling.

This was her fourth pregnancy and she reported having similar symptoms in her prior post-partum periods as well but was never formally evaluated.

In ED, the patient was found to have a BP of 76/50. EKG (EKG a) showed AVNRT with a nonspecific intraventricular block and a heart rate of 190 beats per minute, likely secondary to WPW syndrome. Also, a QTc of 533 ms on noted. Serum magnesium

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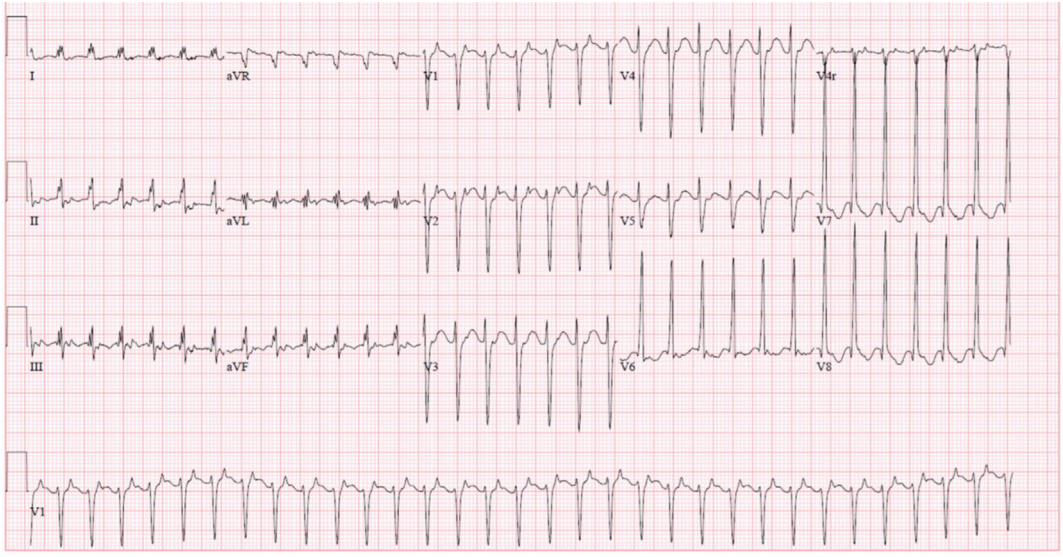
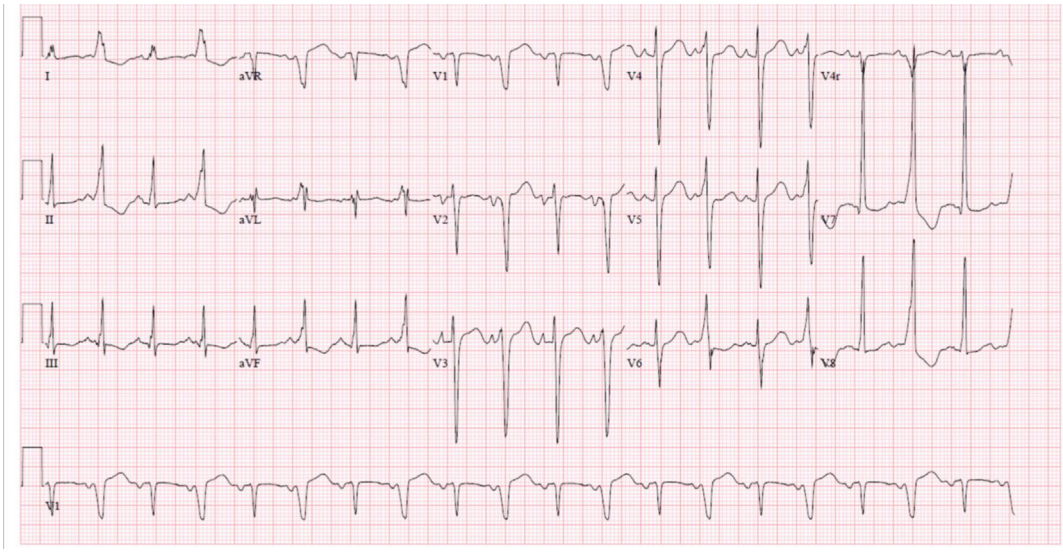
was 1.6 mg/dl, which was repleted. She was treated with two doses of 100 mg intravenous procainamide, which reverted the cardiac rhythm to sinus rhythm and raised the patients blood pressure to 110/70 mmHg. A white blood cell of 12.4/mm3 (Normal range 4–10) was noted. Brain natriuretic peptide levels were 916 pg/ml (Normal <100). Chest X-ray showed findings suggestive of a small left pleural effusion; CT angiography was negative for Pulmonary embolism but did show findings concerning left lower lobe bronchopneumonia with trace pericardial and bilateral pleural effusions. The patient was given ceftriaxone and doxycycline, was continued on procainamide infusion and admitted to the Intensive care unit for further management.

Cardiology was consulted, a transthoracic echocardiogram showed a left ventricular ejection fraction of less than twenty percent with moderate left ventricular hypertrophy and a moderate to severe tricuspid regurgitation. The following day, an electrocardiogram showed conversion to sinus rhythm with a heart rate of 100s–120s (EKG b). Procainamide was eventually discontinued, and she was started on intravenous furosemide infusion. Serum magnesium of greater than 2 mg/dl and potassium levels of greater than four mmol/l were maintained. The patient remained afebrile with improving leukocytosis; MRSA screen was negative, and blood cultures did not show any growth. She was continued on Ceftriaxone 2 g IV daily and doxycycline 100 mg PO BID given her QTC prolongation.



EKG a



EKG b

The patient was then transferred to a super-specialty center for further management, where she was placed on a life vest. Dobutamine and milrinone drip was started with visiting nurse as temporizing measures, and the patient is now being evaluated for left ventricular assist device implantation.

3. Discussion

Peripartum Cardiomyopathy (PPCM), also called pregnancy-associated cardiomyopathy, is an uncommon cause of heart failure during late pregnancy or early postpartum period. It is associated with significant morbidity and mortality related to heart failure and arrhythmias, which usually result from stretching of atrial and ventricular chambers.[1,2] It is estimated to account for less than 1% of pregnancy-associated cardiovascular complications, but the incidence has been increasing. In a large inpatient-based study on 9841 patients hospitalized for PPCM, 18.7% of the patients had an arrhythmia, of which 4.2% had ventricular arrhythmia, and 2.2% suffered cardiac arrest.[2,3] WPW syndrome is a pre-excitation/accessory pathway induced arrhythmia associated with pregnancy and can be seen in combination with PPCM, which involves simultaneous management of systolic dysfunction and can get challenging at times.

Management of arrhythmias during pregnancy is more or less similar to that of a non-pregnant state. However, care should be taken to avoid any detrimental effects on fetal well-being due to therapy or the underlying condition. Several AV nodal blocking agents are considered safe for supraventricular tachycardia during pregnancy; however, in patients with WPW syndrome, AV nodal blocking agents are contraindicated or are cautiously used due to the potential for life-threatening ventricular fibrillation.[4] Class IA antiarrhythmics like procainamide, quinidine, and Class IC antiarrhythmics like propafenone, flecainide are considered safe when there is no associated ischemic or structural heart disease. For these reasons, managing patients with WPW syndrome and simultaneous PPCM is a challenge and requires multidisciplinary team management as PPCM therapy involves using beta-blockers to manage the underlying heart failure.

In comparison, Class IC drugs are associated with a better benefit/risk ratio, whereas Class IA drugs have been noticed to be less potent than Class IC drugs and are associated with intolerable side effects.[5,6] Procainamide produces a lengthening of the refractory period of the accessory pathway in AVRT or of the fast pathway in AVNRT to terminate the re-entry tachycardia.[12] Beta blocking agents are occasionally used to suppress orthodromic AVRT in those with WPW syndrome with a low risk of ventricular tachyarrhythmias due to pre-excitation. Of note, AVRT occurs in two types: orthodromic and antidromic. Orthodromic AVRT re-entry impulse circulates in an antegrade direction through the AV node; in antidromic AVRT, the impulse travels in a retrograde direction through the AV node.

Amiodarone is effective but has significant side effects; thus, it can be employed in patients where other therapies failed or are not feasible. As per 2014 AHA guidelines, for patients with pre-excitation and rapid ventricular rate, management involves intravenous procainamide in hemodynamically stable patients and synchronized cardioversion in unstable ones. In a small, non-randomized trial containing a subset of patients with AVRT, ten patients treated with a combination of propafenone and beta-blocker therapy had no recurrence at >9 months after discharge.[7] Cardioversion can be performed during any week of pregnancy; although there is a theoretical risk of triggering an arrhythmia in the fetus, the risk is supposed to be small due to the small amount of energy directly reaching the fetus itself.[8,9] However, there have been cases reported of fetal arrhythmias requiring emergent C-section after cardioversion, and hence monitoring fetal heart rhythm is recommended.[10]

As per 2015 ACC/AHA/HRC guidelines, vagal maneuvers and IV adenosine are Class 1 A recommendations for orthodromic AVRT. In a hemodynamically stable patient where vagal maneuvers or adenosine is ineffective or not feasible, radiofrequency ablation can be performed as a class 1 recommendation irrespective of the patient's EKG demonstrating pre-excitation. If the patient is unstable, synchronized cardioversion is recommended (Class 1).

If the patient has pre-excitation and is unwilling to undergo ablation, medications including flecainide or propafenone (in the absence of structural heart disease (SHD))can be used as Class IIa recommendations. Amiodarone, beta-blockers, diltiazem, dofetilide, sotalol, or verapamil can be used as Class IIb recommendation. There is a class 1 recommendation for beta-blockers, diltiazem, or verapamil if there is no pre-excitation. If any of the aforementioned medications in the presence or absence of pre-excitation are ineffective, it is a class 1 indication for ablation.

After an acute event, radiofrequency ablation of the accessory pathway is recommended to prevent recurrent episodes of arrhythmia in patients with WPW syndrome. This procedure is generally avoided during pregnancy due to the risk of exposure of the fetus to ionizing radiation. However, as the amount of radiation is small and only involves the area of the

thorax of the mother for the procedure, patients with drug-resistant and severe arrhythmias during pregnancy can be considered for this procedure provided adequate precautionary measures to minimize fetal radiation exposure are undertaken.[11]

4. Conclusion

Although peripartum cardiomyopathy is rare, it is associated with significant morbidity and mortality from both systolic heart failure and arrhythmias' especially in cases of underlying WPW syndrome. Treatment involves using class Ia and III antiarrhythmic agents in the absence of structural or ischemic heart disease. Beta-blockers can also be used to manage heart failure in patients with orthodromic AVRT and low risk of ventricular arrhythmia.

Unstable patients need synchronized cardioversion which carries a risk of fetal arrhythmia, and fetal cardiac monitoring is recommended during the procedure.

Conflict of interest

No potential conflict of interest was reported by the authors.

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