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PERIPARTUM CARDIOMYOPATHY ASSOCIATED WITH LONG QT SYNDROME AND POLYMORPHIC VENTRICULAR TACHYCARDIA SYNCOPE: A CASE REPORT

CARDIOMIOPATIA PERIPARTO ASSOCIADA À SÍNDROME DO QT LONGO E SÍNCOPE POR TAQUICARDIA VENTRICULAR POLIMÓRFICA: RELATO DE CASO

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ABSTRACT

Introduction: Peripartum cardiomyopathy is a rare cause of heart failure during the period between the last month of pregnancy and five months after delivery. Long QT syndrome is characterized by a delay in ventricular repolarization and may manifest with syncope and sudden death due to a type of polymorphic ventricular tachycardia known as torsades de pointes. Case description: J.S., 26-years-old, female, born and residing in São Paulo, Puerperal - 40th day (G3C3A0), went to the emergency room complaining of syncope during breastfeeding and dyspnea on moderate exertion. During evaluation in the ER, the patient developed thoracic discomfort and psychomotor agitation, with non-sustained ventricular tachycardia on the cardiac monitor (torsades de pointes), which was controlled with electrical cardioversion and intravenous magnesium sulfate. The electrocardiogram showed sinus rhythm, diffuse alteration of ventricular repolarization and QTc interval of 580 ms. The echocardiogram showed moderate systolic dysfunction, with a left ventricular ejection fraction of 43% influenced by diffuse hypokinesia. After evaluation by the arrhythmology team, the diagnosis of peripartum cardiomyopathy associated with long QT syndrome was made. Optimized treatment for heart failure was initiated and a cardioverter-defibrillator was implanted due to recurrent episodes of arrhythmia during hospitalization. Discussion: Peripartum cardiomyopathy is a rare disease, but it has a high mortality rate, between 18% and 56%. The patient described met the 4 diagnostic criteria: symptoms of heart failure in the first 5 months after delivery, absence of prior cardiomyopathy, unknown etiology, and systolic dysfunction with LVEF<45%. Long QT syndrome is a genetic disease of varying presentations. The factors that trigger the tachyarrhythmias are situations of electrical instability due to sympathetic system hyperactivity and rare situations, such as peripartum cardiomyopathy. In cases of severe ventricular arrhythmias, the treatment is a cardioverter-defibrillator implant. Conclusion: The association of peripartum cardiomyopathy with long QT syndrome is rare. The severity associated with these conditions points out early diagnosis and immediate treatment important because of the potential risk of death associated with both clinical conditions.

Keywords: Peripartium Period; Tachycardia, Ventricular; Torsades de Pointes

RESUMO

Introdução: A cardiomiopatia periparto é uma causa rara de insuficiência cardíaca no período entre o último mês de gestação e os cinco meses após o parto. A síndrome do QT longo caracteriza-se pelo atraso da repolarização ventricular e pode se manifestar com síncope e morte súbita devido a um tipo de taquicardia ventricular polimórfica conhecida como torsades de pointes. Descrição do caso: J.S., 26 anos, sexo feminino, natural e procedente de São Paulo. Paciente puérpera - 40º dia (G3P3A0), procurou o pronto-socorro com queixa de síncope durante amamentação e dispneia em moderados esforços. Durante a avaliação no PS, evoluiu para desconforto torácico e agitação psicomotora, sendo notada taquicardia ventricular não sustentada no monitor cardíaco (torsades de pointes), que foi controlada com cardioversão elétrica e sulfato de magnésio intravenoso. O eletrocardiograma mostrou ritmo sinusal, alteração difusa da repolarização ventricular e intervalo QTc de 580 ms. O ecocardiograma mostrou disfunção sistólica moderada, com fração de ejeção do ventrículo esquerdo de 43% à custa de hipocinesia difusa.

Após avaliação da equipe de arritmologia chegou-se ao diagnóstico de cardiomiopatia periparto associado à síndrome do QT longo. Foi iniciado tratamento otimizado para insuficiência cardíaca e implantado cardiodesfibrilador por causa de episódios recorrentes de arritmia durante a internação. Discussão: A cardiomiopatia periparto é uma doença rara, porém, tem taxa de mortalidade elevada, entre 18% e 56%. A paciente descrita satisfez os quatro critérios para o diagnóstico: sintomas de insuficiência cardíaca nos primeiros 5 meses depois do parto, ausência de cardiomiopatia prévia, etiologia desconhecida e disfunção sistólica com FEVE < 45%. A síndrome do QT longo é uma doença genética de apresentações variáveis. Os fatores que desencadeiam as taquiarritmias são situações de instabilidade elétrica por hiperatividade do sistema simpático e também situações raras, como a cardiomiopatia periparto. Em casos de arritmias ventriculares graves, o tratamento é o implante de cardiodesfibrilador. Conclusão: A associação da cardiomiopatia periparto com a síndrome do QT longo é rara. A gravidade associada a essas condições torna importante o diagnóstico precoce e tratamento imediato pelo potencial risco de morte associado a ambas as condições clínicas.

Descritores: *Cardiomiopatia Período Periparto; Síndrome do QT Longo; Taquicardia Ventricular Polimórfica; Torsades de Pointes.*

INTRODUCTION

Peripartum Cardiomyopathy (PPCM) is a rare clinical condition whose diagnosis is based on three clinical criteria according to the European Society of Cardiology (ESC) Working Group on Peripartum Cardiology: a) onset of symptoms of heart failure (HF) between the last month of gestation and the fifth month of puerperium; b) left ventricular systolic dysfunction (ejection fraction <45%); c) absence of other apparent etiology for HF. Cardiac arrhythmias occur in about 19% of patients with CMP patients. A registry that included 9,841 hospitalized women diagnosed with CMP demonstrated a prevalence of ventricular tachycardia of 4.2%. In-hospital morbidity and mortality, length of stay, costs and number of procedures were significantly higher in the group of patients with arrhythmia. In rare cases, it may be associated with diseases such as Long QT Syndrome (LQTS), whose delayed ventricular repolarization may determine predisposition to malignant ventricular arrhythmias, with highlight to polymorphic ventricular tachycardia, especially torsades de pointes.[4]

CASE DESCRIPTION

A 26-year-old female patient admitted to the emergency department on the 40th postpartum day, currently in her third pregnancy. She reported recent onset of dyspnea on moderate exertion (not previously present) and two episodes of loss of consciousness during breastfeeding, without prodrome and without apparent cause. Denied cardiovascular or obstetric pathological history. Denied alcoholism and smoking. She was in good general condition, eupneic, BP: 128/70 mmHg, HR: 72 bpm, without peripheral edema, remaining with normal physical examination. The initial electrocardiogram (ECG) showed sinus rhythm, diffuse repolarization alteration and QT interval widening (QTc 580ms). Figure 1 Initial laboratory tests were: Hb: 12.5 g/dl; Leukocytes: 8,200/mm3; Platelets 205,000/mm3; Cr: 0.83 mg/dL; U: 11 mg/dL; Na: 143 mmol/l; K: 3.3 mmol/L; Mg: 2.1 meq/l. The patient suddenly developed thoracic discomfort, without hemodynamic instability and without alteration of the level of consciousness, showing non-sustained ventricular tachycardia and polymorphic ventricular tachycardia (torsades de pointes). (Figure 2) Considering the hemodynamic stability, it was decided to rapidly administer Magnesium Sulfate (2g) IV, obtaining a reversal to sinus rhythm. The patient underwent transthoracic echocardiography showing diffuse left ventricular hypokinesia - ejection fraction: 0.43 (Simpson). The patient was hospitalized and evaluated by Arrhythmology team. Measurements for HF were initiated, and cardiodesfibrillator (ICD) implantation was indicated, assuming that the two episodes of syncope without prodromes were probably related to the presence of complex ventricular arrhythmia. The patient developed well, with improvement of symptoms and was discharged two days

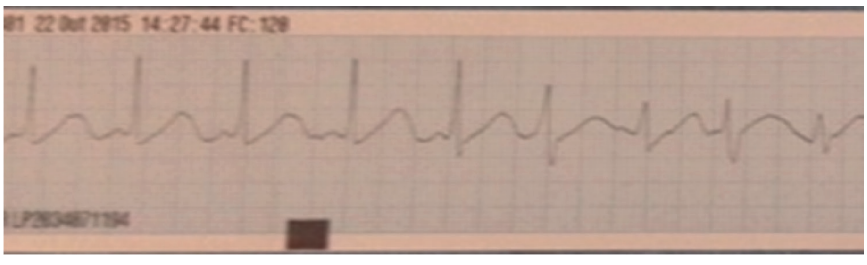

Figure 1. Electrocardiogram. DII derivation: sinus rhythm, HR: 101 bpm.

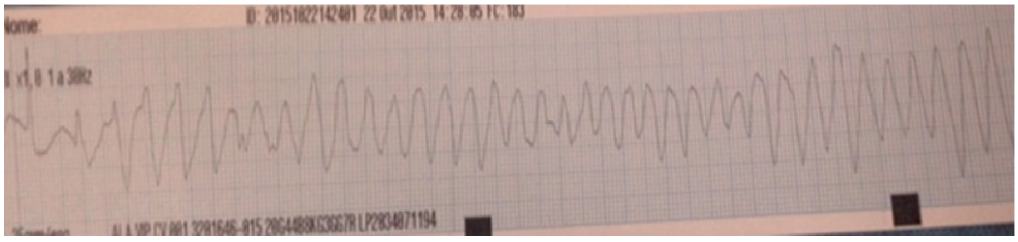

Figure 2. Electrocardiogram. IBD derivation: polymorphic ventricular tachycardia (torsades de pointes).

after ICD implantation, maintaining the use of beta-blocker. It was reevaluated on an outpatient basis and showed no further episodes of syncope.

DISCUSSION

Among the clinical criteria for diagnosis of CMP recommended by the European Society of Cardiology (ESC) Working Group on Peripartum Cardiology the patient described presented the following: a) development of cardiomyopathy in the first five months of the postpartum period; b) absence of previous cardiomyopathy; and c) left ventricular systolic dysfunction. The incidence of CMP varies according to geographic location, with variation in different countries: Japan (1: 20,000), Denmark (1: 10,149), the United States (1: 4,000), and South Africa (1: 1,000). In Brazil, we do not have data on estimated incidence of CMP. Among the risk factors described for its development are: black race, multiparity, advanced maternal age, hypertension and preeclampsia. During pregnancy, some symptoms such as dyspnea, fatigue and peripheral edema may not be recognized as related to HF and are misinterpreted as part of the cardiac adaptations of pregnancy. The pharmacological treatment to be instituted during pregnancy should carefully consider the contraindications, risks and benefits of the established therapy.[6] In the postpartum period, the treatment should be similar to that used for HF by other etiologies, such as angiotensin-converting enzyme inhibitors, angiotensin II receptor blocker, beta blockers and spironolactone. Up to 32% of patients may have complete or almost complete recovery of ventricular function within the first six months of evolution. It is a disease with high mortality, with rates that can reach up to 32% depending on the clinical presentation and the repercussion on ventricular function.

Long QT syndrome (LQTS) is a cardiac canalopathy characterized by delayed myocardial repolarization with QT interval prolongation (> 480 msec) and can be divided into congenital and acquired forms, with an estimated incidence of 1:2,500. In this pathological condition severe cardiac arrhythmias such as ventricular tachycardia and ventricular fibrillation are described. The presence of these arrhythmias aggravates the prognosis, with potential evolution to syncope and sudden death. Among the factors that trigger arrhythmias are conditions that can trigger electrical instability due to sympathetic system hyperactivity (physical and emotional stress) and even less frequent situations, such as CMP. Some genetic associations and phenotypic profiles are described, such as LQTS1 (cardiac events triggered by physical exercise and swimming), LQTS2 (cardiac events related to the postpartum period), LQTS3 (sleep related events). The treatment is based on the use of beta blockers and the efficiency depends on the genotype presented, being LQTS1 and LQTS2 the types with better response. For LQTS patients without a history of syncope, ventricular arrhythmias, family history of sudden death, or QT interval not exceeding 500 milliseconds, beta-blockers may not be required, but for patients with syncope without other apparent cause or reversed sudden death, ICD implantation is recommended in addition to beta--blocker therapy. In mothers with LQTS, a beta-blocker should be started and maintained during pregnancy, including in the puerperium, even in breastfeeding patients (recommendation class I, level of evidence B)., In patients with acquired forms that progress to torsades de pointes, intravenous magnesium sulfate and atrial/ventricular pacemaker may be options for cases presenting without instability. Beta-blocker-associated ICD implantation may be indicated with a view to reducing the risk of sudden death in patients with LQTS who have documented ventricular syncope and/or tachycardia and have a life expectancy of more than 1 year (recommendation class II, level of evidence B). As the patient presented syncope without prodromes and without apparent cause during breastfeeding, it was admitted as probable cause the polymorphic ventricular tachycardia (torsades de pointes) associated with LQTS, and ICD with secondary prevention of sudden death was indicated.

CONCLUSION

The association of peripartum cardiomyopathy with long QT syndrome is rare. The severity associated with these conditions makes early diagnosis and prompt treatment important because of the potential risk of death associated with both clinical conditions.

CONFLICTS OF INTEREST

The author declares that he has no conflicts of interest in this work.

AUTHORS' CONTRIBUTIONS: Each author contributed individually and significantly to the development of the manuscript. MGNB, FQA and TAM contributed to the writing of the manuscript. MGNB, FQA, LOJ, TAM, PGMB and MS followed the patient and reviewed the clinical and medical records. MGNB, TAM, FQA, LOJ, PGMBS, MCS, JCGT, VF conducted bibliographic research, revised the manuscript, and contributed to the intellectual concept of the study.

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