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Torsades de pointes in amiodarone-associated acquired long-QT syndrome

Amiodarone was developed in Belgium in 1961 and became popular in Europe for the treatment of angina. Based on Dr. Bramah Singh's investigation, (1) the Argentine physician Dr. Mauricio Rosenbaum started using amiodarone for the treatment of ventricular and supraventricular arrhythmias with good outcomes. (2, 3) This drug is a class III agent in the Vaughan Williams classification, with class I. II and IV antiarrhytmic effects. Amiodarone produces bradycardia, prolongs myocardial action potential and delays ventricular repolarization. Due to these three pharmacological properties amiodarone prolongs the QT-interval, predisposing to torsade de pointes (TDP), a polymorphic ventricular tachycardia (VT), in about 1% of patients under this treatment. (4)

We report the case of a 69-year-old woman with history of hypertension, coronary artery disease with previous myocardial infarction requiring stent implantation in the proximal left anterior descending coronary artery. Figure 1 shows the electrocardiogram (ECG) after the angiography. A carotid endarterectomy was performed 5 months before due to an atheroembolic stroke. During the postoperative period, she presented atrial fibrillation with rapid ventricular response and amiodarone was added to her normal treatment. Her current treatment is ASA 325 mg/day, atenolol 50 mg bid, enalapril 20 mg bid and amiodarone 200 mg bid. One month before the event she attended the outpatient clinic and an echocardiogram was performed showing: normal left ventricular dimensions, mildly increased wall thickness, normal left atrium and aorta, mild left ventricular dysfunction with an estimated ejection fraction of 50%, hypokinetic basal inferior and mid inferior segments and mitral inflow filling pattern of delayed relaxation (according to her age). Right chamber dimensions and right ventricular function were normal (TAPSE of 20 mmHg). A fibrocalcific trileaflet aortic valve with normal leaflet excursion was observed, with normal gradients and no regurgitation. The mitral valve was normal, without regurgitation, and the tricuspid and pulmonary valves were also normal. There was absence of pericadial effusion and both septae were intact.

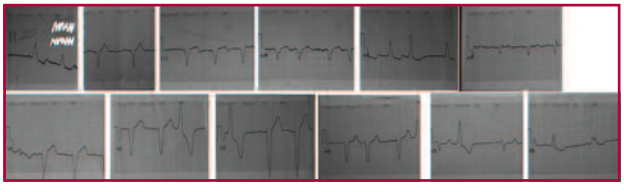

Fig. 1. Electrocardiogram before atrial fibrillation.

She was attended at home by an emergency care system due to several episodes of syncope at rest. When the emergency system arrived, the patient was in cardiac arrest (CA) due to ventricular fibrillation (VF); advanced cardiopulmonary resuscitation was initiated (ACPR) and was defibrillated with 300 J with return of spontaneous circulation (ROSC). Another episode of CA with VF occurred while she was being transferred, requiring two 300 J shocks and 300 mg of intravenous amiodarone, with ROSC.

She was admitted to the emergency department, with a Glasgow score of 9 and breathing room air. Her blood pressure was 90/70, presenting slow capillary refill time and regular heart rhythm at 50 bpm. Another episode of CA in VF occurred at the emergency department. ACPR was initiated and required two 300 J shocks, with ROSC in sinus bradycardia at a heart rate of 40 bpm. Inotropic support was started with dobutamine and a loading dose of 300 mg of amiodarone was administered intravenously. The patient was sedated, intubated, received mechanical ventilation assistance and was transferred to the intensive care unit. The laboratory tests showed: BUN 1.37 g/L, creatinine levels 3.13 mg/dl, K+ 4.8 mEq/L, Na+ 138 mEq/L, Ca++ 10.8 mg/dl, Mg++ 1.9 mEq/L.

Coronary angiography was performed due to a diagnosis of electrical storm secondary to myocardial ischemia.

The ECG showed sinus bradycardia at a heart rate of 35 bpm, normal P-wave and PR interval, old inferior myocardial infarction and poor R wave progression from V1-V6; the QTc was 509 ms. In Figure 2, lead II shows the episode of self-limiting ventricular tachycardia (VT) of 16 beats with change in QRS polarity at 250 bpm and one ventricular triplet in V3.

A diagnosis of polymorphic VT in a patient with long QT-interval (TDP) was made. Amiodarone and inotropic agents were stopped and 2 g magnesium sulphate was administered intravenously. A Furman temporary pacemaker (Figure 3) was implanted and a unipolar lead stimulated the atria at 80 bpm with adequate capture and blood pressure of 120/80 mm Hg.

During the next 48 hours she did not present further episodes of TDP and pacing was stopped. The QT

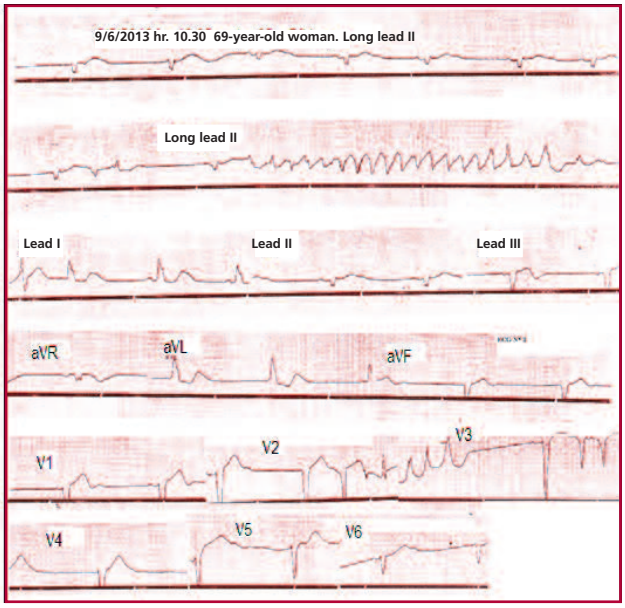

Fig. 2. Torsade de pointes.

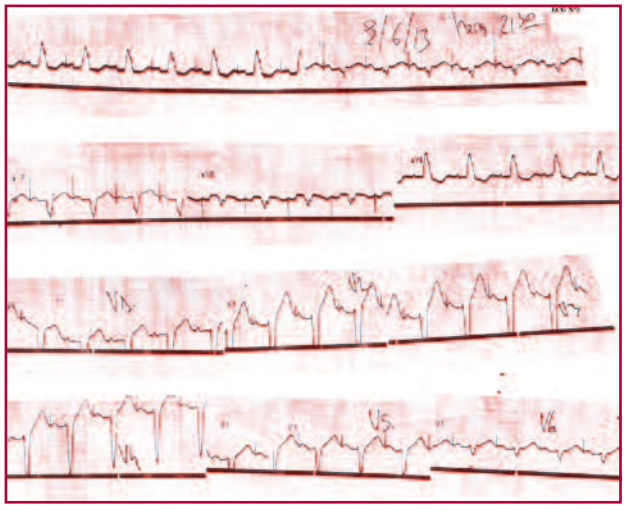

Fig. 3. Electrocardiogram after implanting a Furman pacemaker, showing atrial paced rhythm (unipolar pacing) with a heart rate of 80 bpm and 100% capture.

interval gradually returned to normal values. After 7 days of hospitalization in an internal medicine ward, the patient was discharged with sinus rhythm at 70 bpm, QTc of 440 ms and was followed-up in the outpatient clinic with favorable outcome.

Long QT syndrome (LQTS) refers to a group of channelopathies mainly affecting sodium and potassium channels and characterized by a dispersion of ventricular repolarization expressed in the electrocardiogram by a prolonged QT interval. It may be congenital or acquired. Several mutations have been identified distributed in 10 genes. The severe form of the disease is sporadic, but there are a number of common polymorphisms that may confer susceptibility to the development of the disease when specific drugs are being administered. (5) Patients with long QT syndrome are predisposed to sudden death due to polymorphic VT (TDP). Torsade de pointes is a polymorphic VT associated with LQTS which may be abolished by increasing the heart rate. (6)

The name torsade de pointes describes a sequence of consecutive QRS complexes that then changes its polarity, like twisting around the isoelectric line. (6) Abnormal QTc values are ≥ 440 ms in men and ≥ 460 ms in women. (5) The susceptibility of suffering TDP-VF is higher if the QTc exceeds 500 ms. (4) Torsade de pointes with heart rates >220 bpm are more prone to degenerate into VF.

The factors contributing to prolong the QT interval are: drugs, electrolyte disorders (hypokalemia, hypomagnesemia, hypocalcemia), ischemia, myocarditis, severe bradycardia, T wave alternans, R-on-T phenomenon, female sex, intoxications (as cocaine or organophosphates) neurological disorders (stroke or subarachnoid hemorrhage.) and endocrine diseases (hypothyroidism or pheochromocytoma). (6)

In our patient, long QT was due to the administration of amiodarone after an episode of postoperative AF. Amiodarone is a Vaughan Williams class III antiarrhythmic drug which blocks potassium channels prolonging repolarization. Amiodarone produces bradycardia, prolongs myocardial action potential and delays ventricular repolarization. Due to these three pharmacological properties, amiodarone prolongs the QT-interval, predisposing to TDP, a polymorphic VT in about 1% of patients under this treatment. (4) Initially, our patient presented an episode of syncope evolving with cardiac arrest with shockable rhythm, probably due to TDP. As the patient had a history of coronary artery disease with stent implant to the left anterior descending coronary artery and repetitive episodes of cardiac arrest with shockable rhythm considered VF refractory to treatment, a diagnosis of electrical storm secondary to ischemia was made. After ROSC, a correct ECG analysis showed a QTc interval of 509 ms and a run of polymorphic VT at 250 bpm (both variables increase the probability of degenerating into VF); the diagnosis was then changed to TDP due to drug-induced LQTS. Kay et al. described that TDP is preceded by a sequence of long RR interval of the dominant cycle followed by a short extrasystolic interval with R-on-T phenomenon. This sequence was observed in our patient (Figure 2). Amiodarone was immediately withdrawn, magnesium sulphate was administered and a temporary pacemaker was implanted, with ventricular capture at 80 bpm. This treatment increased heart rate and shortened the QTc to 480 ms, with favorable outcome.

During the next 48 hours the patient did not present further episodes of TDP and pacing was stopped. The QT interval gradually returned to normal values. After 7 days of hospitalization in an internal medicine ward, the patient was discharged with sinus rhythm

at 70 bpm, QTc of 440 ms and was followed-up in the outpatient clinic with favorable outcome. In conclusion, TDP amiodarone-induced LQTS is an infrequent but severe condition requiring timely electrocardiographic diagnosis and specific treatment.

Carlos Ferrando[1], Federico Rivero[1], Federico Machado[2]
[1] Emergency Department, Hospital Maciel. Montevideo Uruguay
[2] Cardiologist. Sanatorio SEMM-Mautone - Sanatorio Cantegril. Punta del Este, Uruguay
e-mail: sanocuore@gmail.com

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Non-compaction peripartum cardiomyopathy: just a coincidence?

The prevalence of non-compaction cardiomyopathy (NCC) is low; yet, its diagnosis is progressively increasing as a consequence of advances and use of the different diagnostic imaging techniques. It is characterized by a thin, compacted external layer and an extensive non-compacted internal layer, with prominent trabeculation. Although its pathogenesis is still controversial, the American Heart Association includes this condition as a primary genetic cardiomyopathy. The interruption of the myocardial compaction process which normally occurs during intrauterine development may play a role in its pathogenesis. The clinical presentation of NCC is variable; subjects may be completely free of symptoms and the diagnosis is incidentally made or may present heart failure, arrhythmias, thromboembolism or sudden death. (1)

Peripartum caripomyopathy (PPCM) is defined as the development of heart failure in the last month of pregnancy or within 5 months of delivery in the absence of another etiology and in the absence of demonstrable previous heart disease. Its prevalence is very low and varies in different regions from 1 per 100 live births in Zambia and Nigeria to 1 per 3000 live births in the United States. Although the etiology of PPCM is unknown, many potential causes have been proposed, including malnutrition, hormonal abnormalities, infections, abnormal immune response and abnormal response to increased hemodynamic burden of pregnancy. It is more frequent in black women, and in those with hypertension, diabetes, multifetal pregnancies and >30 years. (2)

We report the case of a patient who developed signs and symptoms of heart failure in the immediate peripartum period with complementary tests and morphologic criteria of NCC.

A 30-year-old woman complained of progressive dyspnea four months after delivery, with edema of the lower extremities, fever and bloody sputum. She reported a family history of sudden death in a 15-year-old brother and a 3-year old nephew. She had had five pregnancies and five uneventful deliveries, the last one in September 2011. Two months before consultation (on the second month after delivery) she began presenting edema of the lower extremities and progressive dyspnea in FC III. A few days before seeking medical assistance, she complained of fever and bloody sputum. At physical examination, she looked severely ill and presented tachypnea, tachycardia and fever. Blood pressure was 120/80; 2+ pitting edema in the lower extremities and 2/3 jugular engorgement were observed. The apex beat was laterally displaced and was palpable in early diastole. The first heart sound and the second heart sound were normal and a gallop third sound was heard. Crackles were heard at both lung bases; vesicular breath sounds were decreased and percussion produced a dull tone at the right lung base. The electrocardiogram showed sinus tachycardia and signs of left ventricular hypertrophy. The hematocrit was 32%. A right pulmonary effusion was observed in contrast-enhanced computed tomography scan, with areas of consolidation of the right middle lobe and anterior segment of the right lower lobe, and filling defects in the segmental branches of the pulmonary artery. A diagnosis of heart failure and pulmonary embolism was made. Oxygen therapy was administered, anticoagulant drugs were started and negative fluid balance was indicated, with favorable response. Transthoracic echocardiography showed spheroid left ventricular dilatation with areas of non-compacted myocadium in the mid inferior, apical inferior, mid lateral and apical lateral segments (end-systolic noncompacted-to-compacted ratio: 2.8) with severe left ventricular dysfunction (ejection fraction = 29%). The right ventricle was moderately dilated but with normal systolic function. Diastolic filling had a restrictive pattern and moderate mitral regurgitation and tricuspid regurgitation were observed (Figure 1). Gadolinium-enhanced cardiac magnetic resonance imaging confirmed the echocardiographic diagnosis of NCC, with absence of myocardial edema and late enhancement (a patchy pattern at the level of the septum suggestive of non-ischemic fibrosis). Therapy