CASE REPORT

Long –QT Syndrome Secondary to Hypocalcemia

JUAN JIMÉNEZ-JÁIMEZ, RICARDO SEBASTIÁN GALDEANO, ISABEL ALMANSA VALENCIA, MIGUEL ÁLVAREZ LÓPEZ, LUIS TERCEDOR SÁNCHEZ

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Address for reprints:
Dr. Juan Jiménez-Jáimez
Department of Cardiology,
Hospital Universitario Virgen de las Nieves
Avda. de las Fuerzas Armadas 2
18014 Granada
Tel. 667 242021- 34 958020563
E-mail:
jimenez.jaimez@gmail.com

SUMMARY

Long-QT syndrome is a congenital or acquired disorder that produces sudden death due to ventricular arrhythmias. Electrolyte disturbances and medications are the most common causes of acquired long-QT syndrome. We describe the case of a patient with long-QT syndrome secondary to hypocalcemia caused by primary hypoparathyroidism. The secondary causes of long-QT syndrome should be thoroughly examined as they are more common than the genetic causes. Also, as they are reversible with adequate etiological treatment, their correct identification avoids unnecessary diagnostic and therapeutic measures.

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Key words > Long-QT syndrome - Hypocalcemia

Abbreviations > **LQTS:** Long QT syndrome

INTRODUCTION

Long QT syndrome (LQTS) is a disease characterized by the prolongation of the ventricular repolarization time and the predisposition to fatal ventricular arrhythmias. The aetiology of LQTS may be congenital or acquired. Congenital LQTS is often of genetic origin. Acquired LQTS is caused mainly by the action of drugs which prolong the time of the action at the expense of the inhibition of sodium and/or potassium channels, as hypopotassemia, hypomagnessemia, hypocalcemia, which produce high predisposition to present torsade de pointes. Hypocalcemia extends phase 2 of the potential of action and, therefore, the time of repolarization. It is a reversible cause of LQTS, as the correct supplement with calcium shortens QT interval and decreases the probability of malign ventricular arrhythmias. (1, 2)

In this presentation the case of a patient with secondary long QT syndrome to hypocalcemia due to primary hypoparathyroidism is described.

CLINICAL CASE

Female patient, aged 18 years, smoker and asthmatic, who consulted for syncopes. She suffered three syncopal episodes during the night rest, seen by her father, of abrupt establishment and spontaneous quickly recovery, with a duration of few minutes. In some syncope movements of the upper limbs were associated. Never before had she suffered a similar episode. Se was referred to Cardiology as a QT corrected interval of 530 msec was detected in the electrocardiogram (Figure 1). The morphology of the QT interval consisting of the prolongation of the flat ST segment with final T wave led us to consider the diagnosis of type 3 congenital LQTS, (3) due to the affectation of alpha subunit of the cardiac sodium channel. An echocardiogram and a holter were performed, both of them normal. The patient started a treatment with beta-blockers and as she had high risk (corrected QT interval > 500 msec, LQTS phenotype of type 3 and syncope), the necessity of inserting an automatic implantable defibrillator was considered. (4) Likewise, a genetic study with complete sequencing of the three genes responsible for congenital LQTS (KCNQ1, KCNH2, and SCN5A) was performed. In this study associated polymorphisms with LQTS in all the genes, but any missence mutation that can explain a hypothetic congenital LQTS, were found. The patient suffered a new syncope, this time with neurological profile, tonic-clonic movements and postcritical confusion; it was considered as a convulsive episode. A cranial computerized axial tomography showing calcifications in the ganglia of the base and the bilateral frontal cortex was performed. A treatment with antiepileptic drugs was initiated and a level of serum calcium of 4 mg/dl and serum phosphorus of 6.3 mg/dl was observed in the assays. The levels of parathormona were undetectable in serum and finally the diagnosis of primary hypoparathyroidism with serious hypocalcemia was reached. The patient started a treatment with calcium and after some months, with the calcemia already corrected (9.2 mg/dl); a new electrocardiogram was performed showing a corrected QT interval of 436 msec (Figure 2), with no syncopal episodes.

DISCUSSION

The distinction between the genetic and acquired cause of LQTS is not always simple. In fact, in many

Arrhythmia Unit. Department of Cardiology, Hospital Virgen de las Nieves. Granada

occasions there is a genetic mutation and some exogenous factor, as electrolytic disorders that act as a precipitant factor of ventricular arrhythmias. (5) Even some mutations and polymorphisms responsible for LQTS that are sensitive to exogenous causes such as those drugs that prolong the QT interval were described. (6) Among the most frequent causes we find the hypocalcemia; some cases of prolongation of QT interval secondary to hypocalcemia have been reported, (7) many of them in the context of polyglandular autoimmune syndrome.

The identification of reversible causes is important in the management of LQTS. This may help to make an etiologic treatment reducing the QT interval and decreasing the incidence of malign ventricular arrhythmias. For this reason it is necessary to investigate the antecedent of drugs that can prolong QT interval and perform a blood test including calcium, magnesium and potassium.

In our case, taking into account the criteria of risk stratification published by Priori et al., (4) LQTS type 3 was diagnosed. A genetic study was performed and the necessity of an automatic implantable defibrillator was considered. As no genetic findings were observed and according to the results of the complementary tests we reach to the right diagnosis of LQTS acquired through secondary hypocalcemia to primary hypoparathyroidism.

RESUMEN

Síndrome de QT largo secundario a hipocalcemia

Introducción

El síndrome de QT largo es causa de muerte súbita por arritmias ventriculares y puede ser de origen congénito o adquirido. Entre las causas adquiridas, las más frecuentes son los trastornos iónicos y los fármacos. En esta presentación se describe el caso de una paciente con síndrome de QT largo secundario a hipocalcemia por hipoparatiroidismo primario. Es indispensable la detección de posibles causas secundarias y reversibles de síndrome de QT largo, que son más frecuentes que el origen genético, dado que tienen tratamiento etiológico eficaz y se evitan medidas diagnósticas y terapéuticas innecesarias.

Palabras clave > Síndrome de QT prolongado - Hipocalcemia

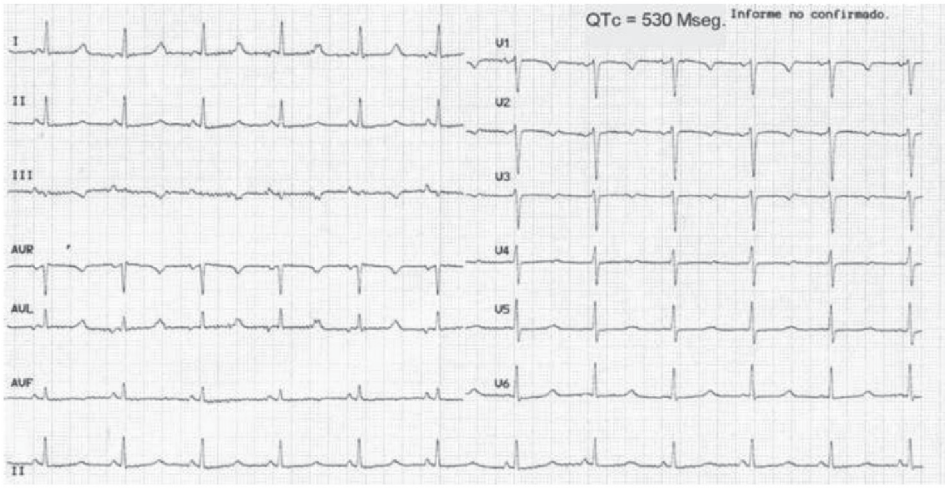

Fig. 1. 12-lead electrocardiogram of the patient before correcting the ionic disorder; the marked prolongation of QTc interval can be seen and its morphology is similar to congenital type 3 LQTS.

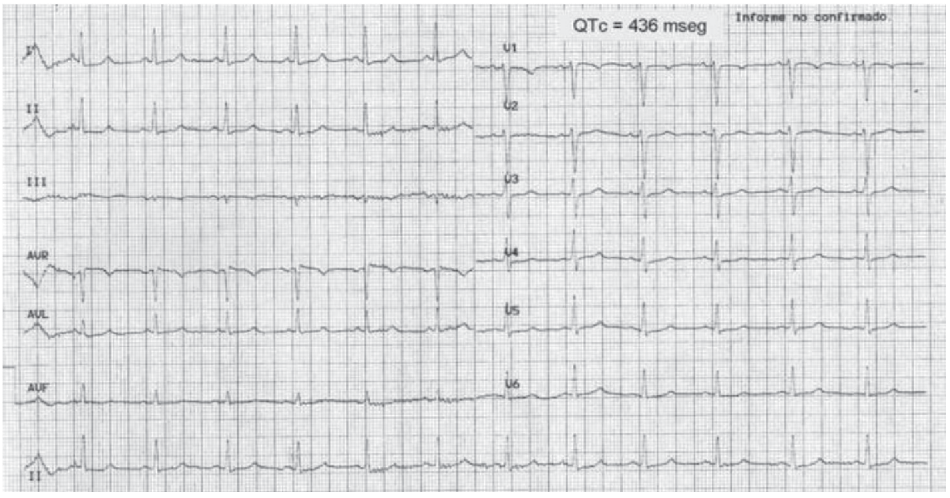

Fig. 2. 12-lead electrocardiogram after the correction of the ionic disorder (hypocalcemia); the normalization of QTc interval can be seen.

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