LONG SURVIVAL OF PATIENT WITH ROMANO WARD SYNDROME ON BETA BLOCKERS

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ABSTRACT

Romano Ward syndrome is an inherited condition characterized by prolongation of QT interval, evident on electrocardiogram, associated with syncope and life threatening arrhythmias and ultimately sudden cardiac death. Cardiac events occur most commonly in the middle age. We present here a case of 58 years old female who was diagnosed as a case of Romano Ward syndrome 30 years ago in 1985 and was on beta blockers since then with no episodes of syncope or ventricular tachycardia during this period.

KEY WORDS: Long QT interval; Ventricular tachyarrhythmia; Syncope; Beta blocker.


INTRODUCTION

Romano Ward syndrome (RWS) is an autosomal dominant disorder and one of the genetic variants of long QT syndrome. It is an inherited form of ions channelopathy resulting in prolonged cardiac repolarization and abnormal prolongation of QT interval on electrocardiogram (ECG). The patients are likely to develop ventricular arrhythmias causing Tor-sade de pointes, syncope and sudden death. Gene mutations in KCNQ1, KCNH2 and SCN5A account for 90% to 95% cases of long QT syndrome. Out of these KCNQ1 which is a potassium channel gene, its heterozygous mutation causes RWS while its homozygous mutation causes a much rarer condition Jervell Lange-Nielsen syndrome in which along with long QT interval deafness also occurs.1-9

CASE PRESENTATION

Thirty years ago, a 28 years old married non pregnant female was admitted from emergency with the complaint of palpitations and three episodes of syncope which lasted for 5 to 7 seconds. She had a history of loose motions and vomiting for five days which settled according to the patient.

On examination the patient was of average height and built, anxious looking, conscious and oriented. Her heart rate was regular with a rate of 110/min, blood pressure (BP) 110/60 mmHg, respiratory rate 18/min, and temperature 98°F. She had no evidence of anemia, jaundice, cyanosis, clubbing, koilonychia, lymphadenopathy, thyroid enlargement, or raised jugular venous pressure. Her circulatory, respiratory, gastrointestinal and nervous system examination was unremarkable. Her 12-lead ECG showed monomorphic ventricular tachycardia (VT). The patient was admitted in cardiac care unit and pharmacologically cardioverted. During her course of admission, she had multiple episodes of ventricular tachycardia which were successfully pharmacologically cardioverted.

Laboratory investigations revealed Hemoglobin 13.0 g/dl (11.5-15.4 g/dl), PCV 40 (35-47%), platelet count 186 (150-440), total leukocyte count 9.5 (4.0-10), serum Na 140 mmol/L (136-139 mmol/L), K 3.4 mmol/L (3.8-5.2 mmol/L), HCO3 18 mEq/L (22-29 mEq/L), Cl 100 mmol/L (98-107 mmol/L), urea 23 (10-50) mg/dl, creatinine 0.52 mg/dl (0.6-1.5 mg/dl), serum albumin 3.9 (3.63-4.92 g/dl), Ca 10.1 (8.1-10.4 mg/dl), Mg 1.8 mg/dl (1.58-2.55 mg/dl), and thyroid function tests were within normal limits. Repeated ECGs showed long QT interval and corrected QT was 560 mSec. Patient was tested for deafness and no hearing disorder was detected.

Her family history revealed that her mother was diagnosed as idiopathic long QT syndrome and had sudden cardiac death. Patient had five brothers. According to her, one had sudden cardiac death at the age of 20, one died of some other medical illness and the other one in road traffic accident and two are alive.
The patient was diagnosed as a case of RWS. She was prescribed beta blocker on discharge from hospital. She was advised to avoid physical exertion and emotional excitement and in case of palpitation or syncope she should report to the hospital.

During these 30 years she got two children both having ECG with normal QT interval and no deafness. Until now she is on follow-up, well-compliant to beta blocker and has not experienced any syncope or palpitation since then.

**DISCUSSION**

RWS is an inherited form of ion channelopathy resulting in prolonged cardiac repolarization and abnormal prolongation of QT interval on ECG. Other causes of prolongation of QT interval include electrolyte imbalance (hypokalemia, hypocalcaemia, hypomagnesaemia), hypothermia, drugs like anti-arrhythmic, tricyclic antidepressants, non-sedative antihistamines, antibiotics), antimalarial are also responsible for long QT syndrome. Cardiac events in patients with RWS occur from childhood through middle age and common in teenage years through 20s. The diagnosis of RWS is made by having prolongation of the QTc interval in the absence of other causes that can cause long QT syndrome like QT prolonging drugs. In our patient there was no other cause for the long QTc interval.

The mainstay of therapy for RWS is beta blocker. Propranolol and nadolol are the beta blockers commonly prescribed. Patients in whom drug treatment is not successful then surgical stellate ganglionectomy can be done as an imbalance between the right and left side of sympathetic nervous system plays a role in the etiology of this syndrome. In patients in which symptomatic bradycardia is associated with the beta blocker therapy then implantable cardioverter defibrillator (ICD) becomes the key treatment.

**CONCLUSION**

In conclusion regular follow-up is essential for all patients with long QT syndrome on drugs and regular periodic evaluation of ICD for inappropriate shocks and its complications. Patients should be counseled to avoid the drugs that cause further prolongation of QT interval or periodic Torsade de pointes. Intense physical activity, sports and emotional stress should be avoided. Genetic counseling should be done as each child of an individual with Romano Ward syndrome has a 50% risk of inheriting the disease.

**REFERENCES**


**CONFLICT OF INTEREST**

Authors declare no conflict of interest.

**GRANT SUPPORT AND FINANCIAL DISCLOSURE**

None declared.