A patient with long QT, sinus bradycardia, and ventricular ectopy: part II

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For figures, please see Part I, on page 1346

Answers to question 1

The 12-lead ECG demonstrated atrioventricular (AV) junctional rhythm with ventricular bigeminy. The premature ventricular complexes (PVCs) were an ‘R-on-T’ pattern with right bundle branch block morphology. Cardiac repolarization was prolonged with a QTc of 490 ms (Bazett’s formula). The T-waves were merged with the U-waves (Figure 1A).

During exercise (Figure 1B), AV junctional rhythm was replaced by sinus rhythm. Initially, no PVCs could be seen, but later the frequency of PVCs increased, with periods of ventricular bigeminy. There was no supraventricular tachycardia during the exercise test. The maximum heart rate was 126 bpm. The QT interval shortened during exercise to 320 ms at heart rate 120 bpm, resulting in a QTc interval of 450 ms. During recovery, AV junctional rhythm returned.

Ambulatory Holter recording (Figure 2) showed periods of sinus rhythm, sinus bradycardia, ectopic atrial rhythm, and AV junctional rhythm with a mean heart rate of 52 bpm and a maximum of 123 bpm. There were approximately 1800 PVCs, often in bigeminy. There were periods of QTc prolongation which were not correlated with heart rate or the time of the day. However, there seemed to be a correlation between the periods of QTc prolongation and ventricular bigeminy. There was a salvo of ventricular tachycardia of three complexes at a rate of 178 bpm, starting with a PVC of same morphology as the solitary PVCs. Furthermore, there were 11 short episodes of supraventricular tachycardia.

Answers to question 2

These electrocardiographic findings in the absence of structural cardiac abnormalities are suggestive of a cardiac ion channel or protein abnormality. In this patient, there are three prominent electrocardiographic abnormalities. First, there is evidence of chronotropic incompetence, expressed by periods of AV junctional rhythm and low heart rate during sinus rhythm. Second, there is intermittent prolongation of the QT interval. Third, there is frequent ventricular ectopy and bursts of ventricular tachycardia.

Discussion

The combination of ventricular ectopy and QT interval prolongation may be suggestive of long QT syndrome type 7, also known as Andersen Tawil syndrome (ATS). This condition is caused by a mutation in the KCNJ2 gene resulting in a loss of function in Kir2.1. Andersen Tawil syndrome is characterized by ventricular ectopy, periodic muscle weakness, and a dysmorphic features.

Exercise-related ventricular arrhythmia may also be caused by mutations in the ryanodine receptor gene (RyR2) which are responsible for catecholaminergic polymorphic ventricular tachycardia. Mutations in the gene encoding for RyR2 channels may also cause sinus bradycardia. However, in patients with RyR2 mutations, the QT interval is usually normal.

Finally, a combination of the long QT interval and sinus bradycardia with periods of the AV junctional rhythm may be suggestive of long QT syndrome type 4 (LQT4). This condition is caused by a ‘loss of function’ mutation in the ankyrin-B gene (ANK2). Ankyrin-B is a plasma membrane protein that binds the lipid bilayer to the cytoskeleton and is involved in the coordination of cardiac ion channels. In LQT4, however, atrial fibrillation very often is present.

There were no neurological symptoms, such as muscle weakness, and no morphological features typical for ATS. Genetic testing showed no mutations in KCNJ2 and RyR2 genes; the results of genetic testing for ANK2-relation mutations were not available at the time of publication. Therefore, at present, the underlying cause of the arrhythmias in this patient remains unknown.

The patient was treated with a dual chamber pacemaker for symptomatic bradycardia, which helped to resolve her symptoms. As the patient has never experienced syncope.

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or sustained ventricular tachycardia and her family history was unremarkable for sudden cardiac death, an ICD was not implanted.

References


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Pacemaker-mediated dermatitis

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Pacemaker insertion causing contact dermatitis is a rare but recognized complication. We report a 60-year-old gentleman, who presented 2 years after his pacemaker implantation with erythema and itching at site of pacemaker insertion. He was initially treated with antihistamines; however, the erythema persisted over the pacemaker site as shown in Figure 1.

He underwent patch testing that showed him to be reactive to the various components of the pacemaker such as nickel, chromium, and titanium. He is therefore under regular follow-up.

Pacemaker-mediated dermatitis is thought to be a delayed hypersensitivity type 3 or 4 mediated reaction. Titanium and nickel are the most common allergens. There is a role for topical corticosteroid that can reduce skin symptoms but recurrence is common. The only complete treatment is the removal of all the allergens. It is recommended that the pacemaker system be coated with non-allergenic materials. Recently, it has been reported in Japan that 0.2 mm thick PTFE (polyte-trafluoroethylene) sheet coating is an effective method, but there are still possibilities that PTFE allergy exists. The best treatment for pacemaker dermatitis is the removal of the device and replacement with another model not containing documented allergens for the patient. Other coatings believed to be hypoallergenic are silicone, parylene, and gold.

With the ever widespread use of various devices, such as biventricular pacemakers and implantable defibrillators, we would expect to see this more often and therefore it is important to recognize this potential problem and its effective management.

Conflicts of interest: none declared.