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Case Report / Olgu Sunumu

Sympathetic denervation in the treatment of fatal arrhythmias in long QT syndrome with restrictive cardiomyopathy

Restriktif kardiyomiyopati birlikteliği olan uzun QT sendromunda ölümcül aritmilerin tedavisinde sempatik denervasyon

Serhat Koca,[1] Feyza Ayşenur Paç,[1] Alkın Yazıcıoğlu,[2] Dursun Aras,[3] Erdal Yekeler[2]

Departments of [1]Pediatric Cardiology, [2]Thoracic Surgery, [3]Cardiology,
Türkiye Yüksek İhtisas Training and Research Hospital, Ankara, Turkey

ABSTRACT

A 12-year-old female patient was diagnosed with long QT syndrome during screening performed as another member of her family had long QT syndrome. Beta-blocker therapy was initiated and an intra-cardiac defibrillator was implanted for syncopes. During follow-up, as defibrillator shocks suggesting ventricular tachycardia were detected, ablation was performed for the polymorphic ventricular extrasystoles, which were considered to induce ventricular tachycardia. Despite this intervention and ongoing proper defibrillator shocking, restrictive cardiomyopathy signs were added to the clinical presentation. The patient underwent left cardiac sympathetic denervation by the video-assisted thoracoscopy. During a three-month postoperative follow-up, no intra-cardiac defibrillator shocking occurred. Video-assisted thoracoscopic left cardiac sympathetic denervation can be safely and effectively performed in long QT patients with life-threatening ventricular arrhythmias.

Keywords: Long QT syndrome; sympathectomy; thoracoscopy.

ÖZ

On iki yaşında kız hastaya, başka bir aile ferdinde uzun QT sendromu olması nedeniyle yapılan tarama sırasında uzun QT sendromu tanısı kondu. Beta-bloker tedavisi başlandı ve senkoplar nedeniyle intrakardiyak defibrilatör implante edildi. İzlemde ventriküler taşikardiye uygun defibrilatör şokları gözlendiği için, ventriküler taşikardiyi tetiklediği düşünülen polimorfik ventriküler ekstrasistollere ablasyon yapıldı. Bu girişime ve uygun defibrilatör şoklamalarının devam etmesine rağmen, klinik tabloya restriktif kardiyomyopati bulguları eklendi. Hastaya video yardımlı torakoskopi ile sol kardiyak sempatik denervasyon uygulandı. Cerrahi sonrası üç aylık izlem süresince, intrakardiyak defibrillatör şoklaması görülmedi. Yaşamı tehdit eden ventriküler aritmiler ile birlikte uzun QT sendromu olan hastalarda, video yardımlı torakoskopik sol kardiyak sempatik denervasyon güvenli ve etkili bir şekilde uygulanabilir.

Anahtar sözcükler: Uzun QT sendromu; sempatektomi; torakoskopi.

Congenital long QT syndrome is a genetic disease, characterized by the prolongation of the QT interval and syncope episodes due to polymorphic ventricular tachycardia or ventricular fibrillation, which may cause sudden cardiac death.[1] An implantable cardioverter defibrillator (ICD), beta-blocker therapy, and left cardiac sympathetic denervation (LCSD) are the recommended treatment modalities to avoid sudden cardiac death events.[2]

Herein, we present a girl with long QT syndrome and restrictive cardiomyopathy who was successfully treated with video-assisted thoracoscopic LCSD in our clinic.

CASE REPORT

A 12-year-old girl underwent screening in our clinic, as her brother was diagnosed with long QT syndrome. She had long corrected QT interval on electrocardiography (ECG) and recurring syncope episodes; therefore, she was diagnosed with long QT syndrome. Her family history revealed that two uncles also had sudden cardiac death before the age of 35. The patient

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Correspondence: Serhat Koca, MD. Türkiye Yüksek İhtisas Eğitim ve Araştırma Hastanesi, Pediatrik Kardiyoloji Kliniği, 06230 Altındağ, Ankara, Turkey.
Tel: +90 506 - 581 82 48 e-mail: drserhatkoca@gmail.com

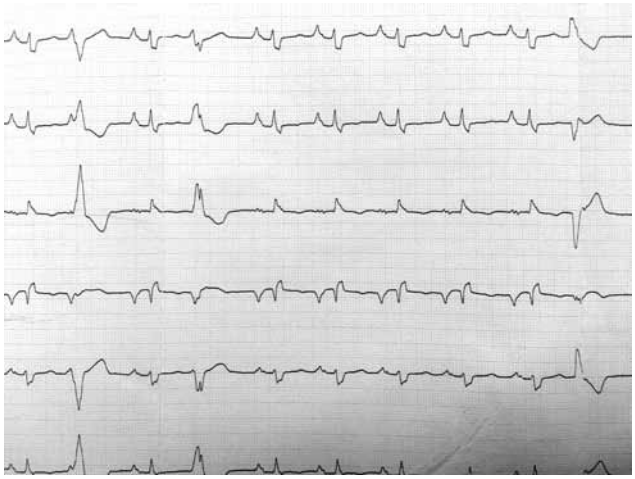

Figure 1. Ventricular extrasystoles with different morphologies shown in electrocardiography. Two different ventricular extrasystoles with two different morphologies are shown here.

was given high-dose (4 mg/kg/day propranolol) beta-blocker therapy. Unfortunately, syncope episodes continued, despite high-dose beta-blocker therapy. Subsequently, an ICD was implanted with continuous monitoring. During follow-up, defibrillator shocks suggesting ventricular tachycardia were detected. Once the cardiac Holter records of the patient revealed similar results, an electrophysiological study was carried out (Figure 1).

In the electrophysiological study, three different morphological ventricular extrasystoles, originating from the right ventricular outflow tract and apex were ablated. Ventricular extrasystoles decreased after this electrophysiological study, as assessed by the Holter examination. Ventricular tachycardia and ICD shocks disappeared after the procedure for 10 months. During follow-up, restrictive cardiomyopathy with pulmonary hypertension (sPAB: 45 mmHg) developed, as revealed by echocardiography (Figure 2).

Despite high-dose beta-blocker therapy, polymorphic ventricular tachycardia episodes with every attack resulting in appropriate ICD shocks were observed. As a result, a LCSD was planned to avoid tachycardia attacks and sudden cardiac death. The surgical procedure was planned using a single port, minimally invasive technique. After intubation with a left selectively, double-lumen endotracheobronchial tube, external defibrillators were deployed for any possible, but undesired cardiac arrhythmias. The thoracic incision for a single port was conducted through the fourth intercostal space and the exposure of the mediastinum pleura was perfected after the deflation of the left lung. Subsequently, the mediastinum pleura was opened and the sympathetic trunk from T_1 to T_4 was dissected and excised (lower half of the left stellate ganglion together with the thoracic ganglia T_2 to T_4). A 16F chest tube was deployed and surgery was completed without complication. Neural tissue was confirmed by pathology. Next day, the chest tube was removed and the patient was discharged on the postoperative fourth day. The patient has been under follow-up in our outpatient clinic for three months without any ICD shock.

DISCUSSION

Family history is of utmost importance for the diagnosis of congenital long QT syndrome.[1] This syndrome is characterized by rapid, chaotic heartbeats; these rapid heartbeats may induce a sudden fainting

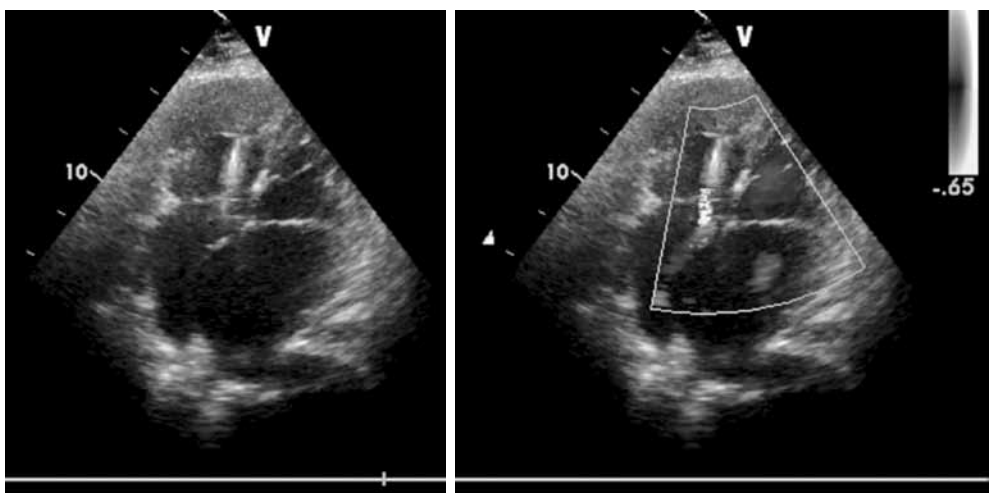

Figure 2. Restrictive cardiomyopathy during follow-up. Note significant extensions of both atria.

spell or seizure.[1] Long QT syndrome has an estimated prevalence of 1/2000, accounting for 20% of the autopsy negative sudden deaths in children and adolescents and for 10% of sudden infant death syndrome cases.[1] Our case did not visit a physician previously for ongoing syncope episodes; however, she was diagnosed during a family screening conducted following the detection of long QT in her sibling.

Following the application of medical treatment (high-dose beta-blocker therapy) and ICD implantation, polymorphic ventricular extrasystoles, which may induce fibrillation, were ablated to reduce the corresponding ICD shocks. A decrease in the corresponding ICD shocks was achieved through ablation, and, therefore, the life quality of the patient could have been improved. Although several studies have shown that ablation treatment is effective in ICD implanted patients, there is no study investigating the effect of ablation on the mortality risk in long QT syndrome.[3,4] In our case, although LCSD was in the first visit, it was unable to be applied due to the preference of the patient.

Despite the coexistence of several cardiomyopathies, including non-compaction, Takotsubo, dilated, and hypertrophic cardiomyopathy with long QT syndrome have been reported in the literature, the concomitance of restrictive cardiomyopathy with long QT syndrome has not been reported, yet.[5,6] The coexistence of long QT syndrome with restrictive cardiomyopathy is an extremely rare. Restrictive cardiomyopathy and pulmonary hypertension, which increase the mortality and morbidity, were detected in our case. Heart or heart/lung transplantation may be applicable in such patients at the last treatment of choice. Similarly, our case has been added in the transplantation/ventricular support device list, after she was assessed by the Heart Transplantation Council in our hospital.

Left cardiac sympathetic denervation is a long-standing intervention, being a safe and effective adjunctive therapy for patients with life-threatening ventricular arrhythmias.[7] In addition, LCSD is also recommended for reducing corresponding shocks in long QT syndrome in patients on beta-blocker therapy or implanted with an ICD.[8] The efficacy of this intervention is described as the reduction in QT intervals on the elimination of the sympathetic activation of the heart, the reduction of norepinephrine at a ventricular level, and the reduction of early post-depolarization.[8] Despite the statistically meaningful benefit of this intervention, its efficacy in reducing the cardiac deaths in the long-term has not yet been confirmed yet.[8] The video-assisted thoracoscopy in this intervention offers many advantages for such patients with comorbidities. Since it is a less invasive technique, LCSD is applied by video-assisted thoracoscopy with the main advantage being that the patient has a reduced risk for major comorbidities, including an increased risk for open surgery, pulmonary hypertension secondary to restrictive cardiomyopathy, and fatal ventricular arrhythmia secondary to long QT syndrome.[9] Left sympathetic ganglion denervation is a treatment method recommended following the ICD implantation, as it is more effective compared to the catheter ablation.[2]

In conclusion, long QT syndrome may present with a varying clinical features. Although such patients may be asymptomatic, they may also present with uncontrolled ventricular arrhythmia. In addition to beta-blocker therapy and implantable cardioverter defibrillator, left cardiac sympathetic denervation may be considered as a method of treatment in the management of life-threatening ventricular arrhythmias. The coexistence of restrictive cardiomyopathy and pulmonary hypertension, being developed concomitantly with long QT syndrome, has not been previously reported in the literature. Heart or heart/lung transplantation may be applicable as a treatment method for such patients as the last surgical option. However, before such major transplantation surgeries, patients should be given the chance of an effective and minimally invasive surgery with left cardiac sympathetic denervation which decreases the risk for fatal cardiac arrhythmias.

Declaration of conflicting interests

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