Syncope Due To Social Phobia May Be A Sign Of Serious Ventricular Arrhythmia

Sosyal Fobi Nedeni İle Meydana Gelen Senkop Ciddi Ventriküler Aritminin Bir Göstergesi Olabilir

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ABSTRACT

Catecholaminergic polymorphic ventricular tachycardia is a rare inheritable cardiac channelopathy characterized by malignant polymorphic ventricular tachycardias that are triggered by catecholaminergic stress. During physical exercise or emotional stress, patients typically encounter syncope or sudden cardiac death within the first two decades of life. Here, we report the case of a nine-year-old female patient suffered from syncope due to feeling fear or anxiety to enter crowded environments. She was examined and followed up by several other departments, however no significant improvement of her symptoms was observed and then referred to our clinic. Catecholaminergic polymorphic ventricular tachycardia was detected at the end of cardiac examinations and successfully treated with propranolol. In patients with recurrent syncope attacks and dizziness, rhythm disturbances should be kept in mind that if it occurs especially after effort or emotional stress.

Keywords: *Dizziness, Emotional stress, Syncope, Ventricular tachycardia*

ÖZET

Katekolaminerjik polimorfik ventriküler taşikardi, katekolaminerjik stres ile tetiklenen malign polimorfik ventriküler taşikardiler ile karakterize ve nadir görülen kalıtsal bir kardiyak kanalopatidir. Hastalar tipik olarak 20 yaşından önce fiziksel egzersiz sırasında veya duygusal stres ile senkop veya kardiyak arrest ile karşılaşırlar. Burada kalabalık ortamlara girmede endişe veya korku hissetmesinden dolayı senkop geçiren dokuz yaşında kız bir olgu sunulmuştur. Diğer bölümlerde incelemeleri ve takipleri yapılan, ancak semptomlarında belirgin bir iyileşme gözlenmeyen olgu kliniğimize yönlendirilmiştir. Kalp incelemeleri sonucunda katekolaminerjik polimorfik ventriküler taşikardi tanısı konulmuş ve propranolol ile başarılı bir şekilde tedavi edilmiştir. Özellikle duygusal stres veya efor sonrasında meydana gelen tekrarlayan senkop atakları veya baş dönmesi olan hastalarda ritim bozukluklarının akılda tutulması gereklidir.

Anahtar Kelimeler: Baş dönmesi, Duygusal stres, Senkop, Ventriküler taşikardi

INTRODUCTION

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rhythm disorder that causes syncope and sudden death following exercise or emotional stress without underlying structural heart disease.(1) It was first described in the late 1970s. Its prevalence is estimated to be about 1/10000. Ventricular tachycardia, ventricular fibrillation, cardiac arrest and sudden death may be triggered by the causes that increase sympathetic stimulation.(2) However, as the cause of the complaints is attributed to other diseases, these cases are usually diagnosed late. It is often inherited through the RyR2 gene encoding the ryanodine receptor in the heart. It can also occur less frequently with a CASQ2 mutation encoding the calsequestrin gene, or sporadically.(3,4) In this article, we present a nine-year-old patient who was diagnosed with CPVT and has been followed-up with propranolol treatment.

A nine-year-old female patient presented with complaints of dizziness, palpitations, and syncope that had developed after excitement and effort for approximately five months. In her anamnesis, she was afraid of fainting and also was afraid of playing games with her friends and going out to crowded environments and was not even going to school. Firstly, the patient applied to a psychiatrist because of social phobia. Sertraline and lorazepam treatment were started. At the same time, she was followed up by a pediatric neurologist, thinking that she had an epileptic attack. The patient was referred to us with no significant improvement in her complaints during follow-up. On physical examination, there was no pathological sound or murmur in her heart examination. Blood pressure was 110/70 mmHg and heart rate was 110 beats/min (bpm). Body weight was consistent with three percentiles. The other system examinations were all normal. There was no specific family history of syncope or sudden cardiac death. Baseline electrocardiographic (ECG) evaluation was normal; corrected QT interval was found to be normal and also there was no Brugada pattern. Echocardiographic examination revealed no structural or functional abnormalities. During the 24-hour holter ECG follow-up, catecholaminergic polymorphic ventricular tachycardia (CPVT) was considered primarily because bidirectional ventricular extrasystoles appeared and non-sustained bidirectional polymorphic ventricular tachycardia attacks were seen when the heart rate increased above 140 bpm. Syncope attacks after exertion were thought to be caused by possible cardiac arrhythmias due to catecholamine release. So CPVT was considered primarily and also the diagnosis was confirmed with exercise test. The test was terminated because of dizziness and ventricular ectopic beats were observed immediately after heart rate increased above 125 bpm during exercise test. Then, effective dosage of oral propranolol treatment was prescribed to her. During the follow-up it was learned that the symptoms and signs of patient were improved dramatically. She started to go to school, went out of the house more frequently, and had no dizziness or syncope attacks. Moreover, there was no more need for the drugs that she was taking before. Control holter ECG examination showed significant improvement and no significant arrhythmia was observed. The patient has been followed up with successful propranolol treatment for approximately six months.

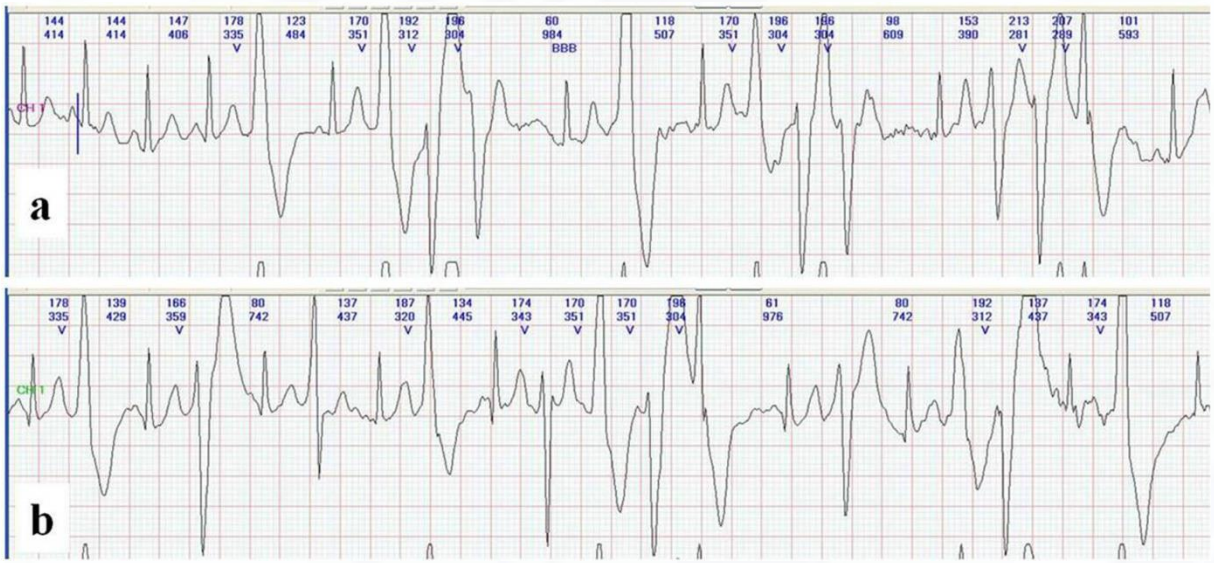

Figure 1. Bidirectional triplet (a) and quadruplet (b) ventricular extrasystoles (non-sustained polymorphic ventricular tachycardia)

DISCUSSION

Since it was first discovered in 1970's, catecholaminergic polymorphic ventricular tachycardia (CPVT) has been reported as a cause of syncope, ventricular arrhythmias and sudden cardiac death. Catecholaminergic polymorphic ventricular tachycardia typically manifests as syncope between 7 and 9 years of age, but sudden death may be the first presentation.(5) In 30% of CPVT patients, there is family history of sudden death before the age of forty.(6) The difficulty to recognize CPVT patients was reported by Roston et al (7), who found in a study on 226 CPVT patients that the establishment of diagnosis was approximately two years after the first symptomatic episode. In addition, more than 60% of patients received a missed diagnosis at the initial evaluation.(7) It has been shown that approximately 60% of patients with CPVT have RyR2 (Ryanodine type 2 receptor) gene mutations. Patients with RyR2 mutation become symptomatic earlier.(8) Catecholaminergic polymorphic ventricular tachycardia is associated with two genetic mutations; RyR2 and CASQ2. RyR2 is inherited in an autosomal dominant pattern and mediates the release of calcium from the sarcoplasmic reticulum that is required for myocardial contraction. The RyR2 mutation increases calcium release and can trigger life threatening ventricular arrhythmias. A second genetic form of CPVT, with an autosomal recessive inheritance, involves *mutations* in the *gene encoding* cardiac *calsequestrin* (*CASQ2*). The CASQ2 protein, which serves as the major calcium reservoir within the sarcoplasmic reticulum, has an ability to bind extremely large amounts of calcium. Mutations have also been reported in genes such as calmodulin1 (CALM1), triadine (TRDN) and SCN5A.(4) Catecholaminergic polymorphic ventricular tachycardia can be difficult to diagnose, as ECG is normal in the absence of symptoms and echocardiography shows no specific findings. A typical finding on ECG is ventricular tachycardia with 180-degree alteration of the QRS axis (bidirectional tachycardia). Catecholaminergic polymorphic ventricular tachycardia is not inducible by programmed electrical stimulation. In patients suspected to have this disease, the arrhythmia must be recorded by holter monitoring or induced by exercise treadmill testing.(5) Lifestyle change should be recommended in the follow-up of the disease. Patients should avoid competitive sports, heavy exercise and stressful environments. When catecholaminergic polymorphic ventricular tachycardia is diagnosed, treatment should be planned on the basis of the patient's hemodynamic condition.(8) The focus of treatment is to suppress the adrenergic activity, therefore, beta-blockers are the most important drugs in the treatment of CPVT. Beta-blockers are effective for acute phase and maintenance treatment. Hayashi et al. (9) found that only five out of 81 patients with CPVT who had a mean follow-up of 7.9 years with beta blocker therapy had only fatal/non-fatal ventricular arrhythmias, and beta blocker treatment completely prevented recurrent arrhythmia attacks in the majority of patients. Implantable cardioverter defifrillator implantation and/or left cardiac sympathetic denervation is recommended if symptoms persist under medical treatment or if they have a polymorphic ventricular tachycardia attack. Flecainide is also an effective choice for arrhythmias developing under beta blocker treatment.(10)

Syncope or sudden cardiac death in childhood might occur due to other arrhythmogenic entities. These include arrhythmogenic right ventricular cardiomyopathy, Brugada syndrome, long QT syndrome, pre-excitation syndrome, commotio cordis, and Andersen-Tawil syndrome.(11,12) Any of these were not observed in our case. Our patient received an effective doseage of oral beta-blocker (propronalol) treatment and then no significant symptom or arrhythmia were detected during the follow-up.

In conclusion, rhythm disturbances should be considered in patients with recurrent syncope attacks. In the history, it should be questioned whether syncope attacks start with effort or emotional stress. Patients diagnosed with epilepsy or vasovagal syncope due to misdiagnosis may present with syncope as in our case or with sudden cardiac arrest as a result of adrenergic stimuli. Exercise stress test and holter ECG can be used in the diagnosis of these patients.

Although catecholaminergic polymorphic ventricular tachycardia can be rarely seen, it should be considered in the differential diagnosis of recurrent syncope attacks.

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