Different Faces of Cathecolaminergic Polymorphic Ventricular Tachycardia on Epinephrine QT Stress Testing

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ABSTRACT

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is an inherited arrhythmic disorder characterized by exercise-induced ventricular arrhythmias and risk for sudden cardiac death. An exercise stress test is a standard method for diagnosis of CPVT, characterized by typical bidirectional polymorphic ventricular tachycardia. In patients who are not suitable for an exercise stress test, epinephrine stress testing (epinephrine challenge) can be used instead. There are a few reports in the literature discussing supraventricular tachycardias during exercise testing in pediatric CPVT patients. We present a 6-year-old boy who was admitted with syncopy. Epinehrine stress testing was performed according to the Ackerman protocol and revealed different types of tachycardias.

Key Words: Catecholaminergic polymorphic ventricular tachycardia; epinehrine stress test; supraventricular tachycardia

Adrenalin Stres Testi Sırasında Ortaya Çıkan Katekolaminerjik Polimorfik Ventriküler Taşikardinin Farklı Yüzleri

ÖZET

Katekolaminerjik polimorfik ventriküler taşikardi (CPVT), egzersize bağlı ventriküler aritmi ve ani kardiyak ölüm riski ile karakterize kalıtımsal bir aritmik hastalıktır. Egzersiz stres testi, tipik çift yönlü polimorfik ventriküler taşikardilerle sonuçlanan CPVT tanısında standart bir yöntemdir. Egzersiz stres testi için uygun olmayan hastalarda, adrenalin stres testi kullanılabilir. Literatürde pediatrik CPVT hastalarında egzersiz testi sırasında supraventriküler taşikardilerle ilgili az sayıda rapor vardır. Bu olgu sunumunda senkop şikayeti ile başvuran altı yaşında erkek çocuk hastada Ackerman protokolüne göre uygulanan adrenalin stres testiyle ortaya çıkan farklı taşikardi türlerini paylaştık.

Anahtar Kelimeler: Adrenalin stres testi; katekolaminerjik polimorfik ventriküler taşikardi; supraventriküler taşikardi

INTRODUCTION

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is an inherited arrhythmic disorder characterized by exercise-induced ventricular arrhythmias and risk for sudden cardiac death[(1)]. In CPVT, the heart is structurally normal and the resting electrocardiogram (ECG) is normal. A mutation in the ryanodine receptor 2 (RYR2) gene is found in the majority of patients with this disorder, causing excess calcium leak from the sarcoplasmic reticulum[(2)]. A maximal exercise stress test is a standard method of diagnosing CPVT, because arrhythmias can only be detected during exercise[(3)].

Pharmacological stress tests have been valuable in the diagnosis of several inherited arrhythmia disorders, such as Ajmaline infusion in Brugada syndrome or adrenalin infusion in the diagnosis of congenital long QT syndrome[(4,5)]. In CPVT, several studies have used intravenous epinephrine infusion in addition to an exercise stress test in diagnostic evaluation[(6,7)].

CASE REPORT

A 7-year-old boy was admitted with syncopy occuring for the past 3 years, especially when he was excited. A 12-lead surface ECG, echocardiography, and ambulatory ECG showed results in the normal range. Treadmill exercise stress testing was inconclusive for CPVT as a result of noncooperation during the test. Therefore, epinephrine stress testing

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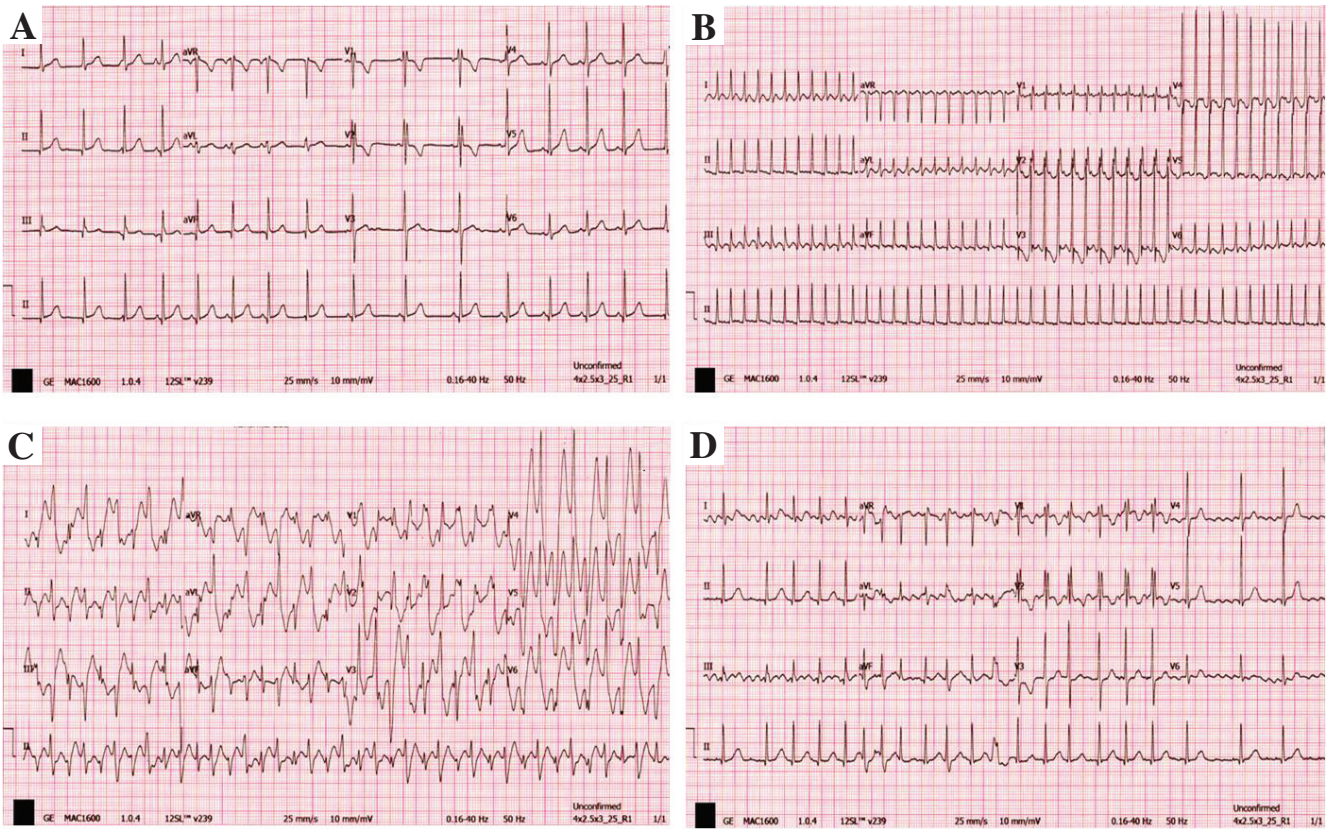

Figure 1. A. Basal electrocardiogram was consistent with normal sinus rhythm. **B.** Supraventricular tachycardia with narrow QRS complex. **C.** Bidirectional polymorphic ventricular tachycardia. **D.** Atrial flutter with varying degrees of atrioventricular block. This is likely the same narrow QRS tachycardia at the beginning of the test, but with varying degrees of atrioventricular block this time.

was undertaken. Basal ECG was consistent with normal sinus rhythm (Figure 1A). First, a supraventricular tachycardia with narrow QRS complex was induced during a 0.25 μg/kg/min infusion (Figure 1B). Next, a bidirectional polymorphic ventricular tachycardia developed (Figure 1C). Finally, atrial flutter with varying degrees of atrioventricular block was induced. Medical therapy with dideral and flecainid was initiated. The patient was under control on medical therapy without a need for an implantable cardioverter-defibrillator or symphatectomy.

CONCLUSION

Both bidirectional polymorphic ventricular tachycardia and several types of supraventricular tachycardias can be induced during epinephrine stress testing for the diagnosis of CPVT.

Informed Consent: Written informed consent was obtained from the patient's parents.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept/Design – HCK, YE; Analysis/Interpretation – HCK, EÖ; Data Collection – HCK; Writing – HCK; Critical Revision – YE; Final Approval – YE; Overall Responsibility – HCK, YE

Conflict of Interest: The authors have no conflict of interest to declare.

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