A Diagnosis that Escapes Our Attention: Short QT Syndrome

Dikkatimizden Kaçan Bir Tanı: Kısa QT Sendromu

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ABSTRACT

Short QT sydrome (SQT) is one of the rarely seen arythmia causes related to sudden cardiac death. An important part of the patients are diagnosed during the examination due to cardiac arrest. Therefore, diagnosing asymptomatic patients is lifesaving. In this case report, we presented a five-year old asymptomatic female patient how to put the SQT diagnosis. There was no complaint of the patient who had been directed to pediatric cardiology polyclinic due to murmur. In her electrocardiography (EKG), QTc duration was determined in lower limits. However, when the patient's family history was examined in a detailed way, suspicious deaths were recognized. In the patient's 24-hour holter EKG; SQT was diagnosed by establishing average QTc duration as 340 ms together with sharp, narrow T waves. Satolol treatment was initiated to the patient. To diagnose asymptomatic SQT patients, a decent family history must be obtained. Spot EKG is insufficient to rule out the diagnosis. All patients with suspicious death histories in their familieis must be directed to pediatric cardiologists.

Key Words: Asymptomatic short QT, family history, diagnosis

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ÖZET

Kısa QT sendromu (SQT) oldukça nadir görülen, ani kardiyak ölüm ile ilşkili aritmi nedenlerinden biridir. Hastaların önemli bir kısmı kardiyak arrest ile tanı almaktadır. Bu nedenle asemptomatik hastaların tanısını koymak yaşam kurtarıcıdır. Biz bu olgu sunumunda asemptomatik, 5 yaşında kısa QT tanısı koyduğumuz bir kız hastayı sunduk. Üfürüm nedeniyle pediatrik kardiyoloji polikliniğine yönlendirilmiş olan hastanın herhangi bir yakınması yoktu. Elektrokardiyografisinde(EKG) QTc süresi şüpheli alt sınırlarda saptandı. Ancak hastanın aile öyküsü detaylı sorgulandığında şüpheli ölümler olduğu farkedildi. Hastanın 24 saat holter EKG'sinde sivri, dar T dalgaları ile birlikte ortalama QTc süresi 340 ms saptanarak SQT tanısı konuldu. Hastaya sotalol tedavisi başlandı. Asemptomatik SQT hastalarına tanı koyabilmek için mutlaka iyi bir aile öyküsü alınmalıdır. Spot EKG tanıyı dışlamak için yeterli değildir. Ailesinde şüpheli ölüm öyküsü olan tüm çocukların ileri inceleme için pediatrik kardiyologlara yönlendirilmesi gerekir.

Anahtar Sözcükler: Asemptomatik kısa QT, aile öyküsü, tanı

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INTRODUCTION

Short QT syndrome (SQT) is one of the arythmia causes regarding sudden cardiac death. Gussak et al. were the first ones describing the relationship between short QT duration and arythmia (1). Later, it was understood that the disease was associated with the acceleration of ventricular repolarization owing to potassium (K) channel mutation (2,3). SQT syndrome is regarded as one of the rarely seen arythmia causes. Nevertheless, it is observed that the number of patients has increased when people are more aware of the disease.

QTc duration calculated by Bazzet formula was <340 ms (4) in the electrocaardiographies of the patients with SQT syndrome. QTc duration of SQT patients may be closer to lower limits of normal (4), on the other hand, QTc duration can be determined as <340 ms in a very few healthy individuals, too (5). Therefore, it is not appropriate to use only QTc value for SQT diagnosis. Diagnosis criteria are utilized recommended by Gallob et al. for SQT diagnosis (6). We presented a 5-year-old female patient diagnosed with asymptomatic SQT sydrome in this study. We also wanted to discuss how to diagnose asymptomatic SQT patients over our patient.

CASE REPORT

A five-year-old female patient. Her weight is 17 kg (25-50 percentile), height is 124 cm (50-75 percentile). Our patient was directed to pediatric cardiology polyclinic due to murmur hearing during physical examination by her pediatrician for whom she had been taken for upper airway infection. When our patient referred to pediatric cardiology polyclinic, no pathological finding was determined other than 1/6 pansystolic murmur in mesocardiac focus during physical examination. There was no syncope history in the background story. In the family history, though, it was found out that the mother of our patient died of heart attack at the age of 21, however, no autopsy was carried out for her. In the electrocardiography (EKG) of our patient, QTc duration calculated by Bazzet formula was discovered as 388 ms. Nonetheless, it was noticed that QT duration did not change although heart rate changed (respiratory arythmia) in EKG. When QTc was calculated for every QRS complex, it was observed that QTc value was 340 ms and all QTc durations changed between 340-388 ms (Figure 1).

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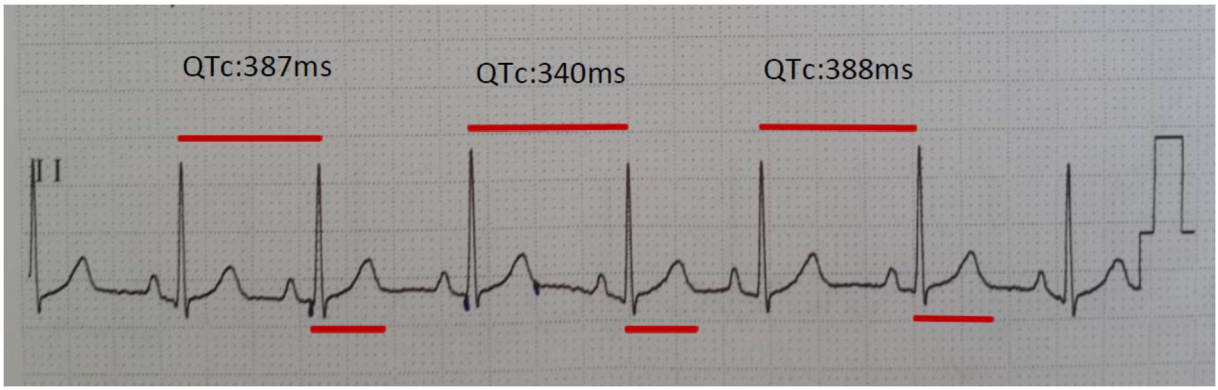

Figure 1. During the initial referral of the clinic, QT durations at the DII leads in the ECG appear to be at the lower limit.

Thereupon, some data related to the death of mother in question were detailed. Other people from the family were communicated and it was revealed that "death due to heart attack" definition was not based on a medical evidence; moreover, 4 siblings of mother including 3 males and a female had also died.

It was found out that the youngest one of all was a 5-year-old male and the oldest one was a 35-year-old male and no autopsy procedure had been applied to none of them. We evaluated our patient with holter EKG for 24 hours. We observed typical narrow and sharp T waves for SQT in Holter records (Figure 2). We also observed that QT duration did not change even though heart rate changed in holter records (Figure 3).

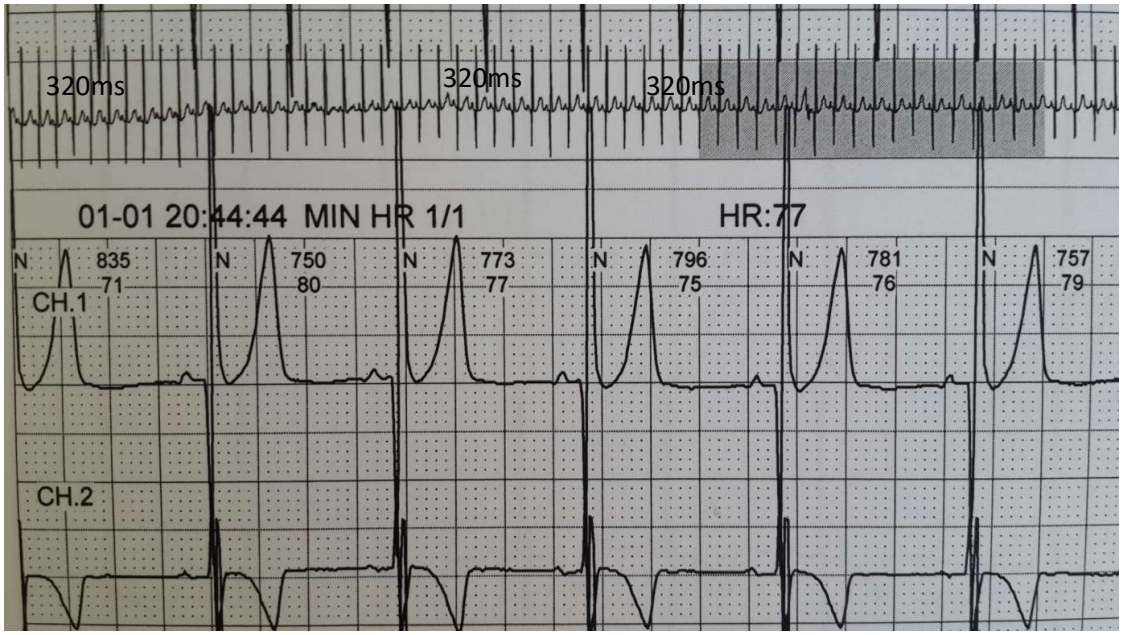

Figure 2. Narrow and sharp T waves are seen in 24 hour holter ECG record of the patient.

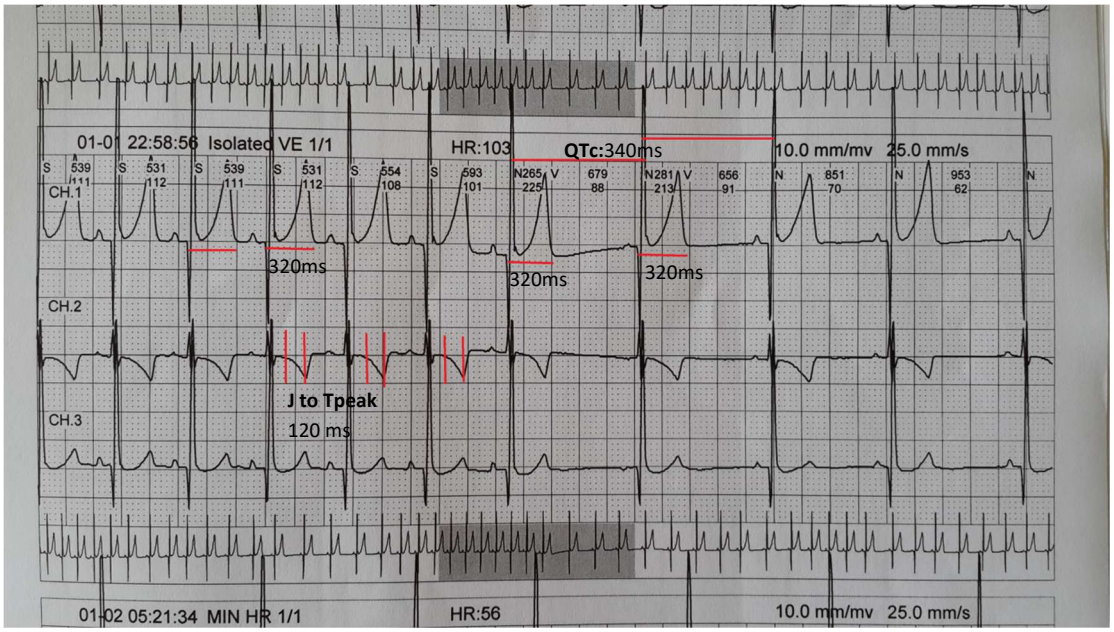

Figure 3. The 24 hour holter ECG record shows that the QT interval did not change with heart rate and the QT interval was 340 ms.

Average QTc duration was established as 340 ms in Holter EKG, however, no arythmia pattern was seen. Sotalol treatment from 90 mg/m^2 was initiated for the patient.

DISCUSSION

Our patient was diagnosed with SQT thanks to family history, holter and EKG findings. We performed a genetic examination for SQT. Known genetic mutations were not determined in the patient for SQT. With Gollob scoring our patient got 4 points in total including 2 points for being QTc<350 ms and 2 points due to family history (6).

She did not get any points from J to Tpeak duration (since 120 ms was determined). Sotalol treatment was commenced for our patient as there is no hydrokinidin in our country. Following sotalol treatment, QTc duration was observed to be as 450 ms (Figure 4). In a multicenter study in which Giustetto et al. presented long term follow up results of SQT patients, it was reported that arythmias such as atrial fibrillation (AF), ventricular fibrillation (VF) and ventricular tachycardia (VT) were more commonly determined in holter records of patients with syncope or cardiac arrest history (7). We followed up our patient without attaching defibrillator as the patient was very young, she had no symptom, there was no arythmia pattern in the repeated holter records and she tolerated sotalol treatment well. We decided to follow up our patient with frequent holter controls.

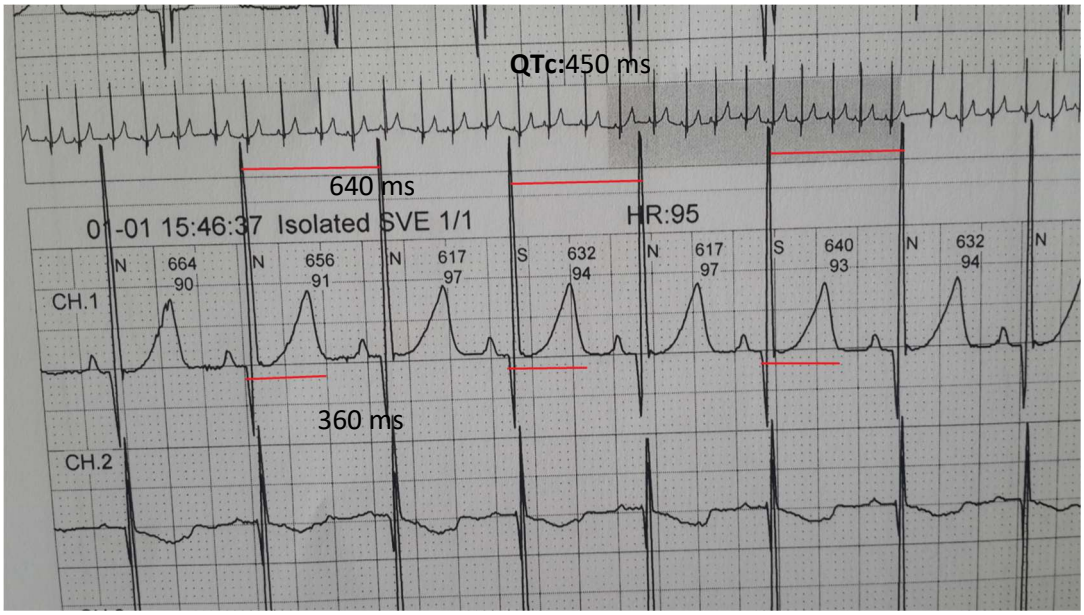

Figure 4. The 24-hour holter ECG record after sotalol treatment shows a QTc duration of 450 ms

The QTc duration in the spot ECG for SQT diagnosis is significant, but for some subtypes of SQT, the QTc duration can be borderline. Spot ECG alone may not suffice to allow the diagnosis of SQT in these patients. In these patients, a good family story, holter ECG and genetic screening are needed to be able to recognize. We diagnosed a little girl with SQT who had no symptom and no specific finding in her spot EKG. We owe our accurate diagnosis to only getting a detailed family history. If we had confined ourselves to first mentioned information “her mother died of heart attack” by the family members during the anamnesis, we could not have diagnosed the patient. We can say that; a good family history should be obtained to diagnose asymptomatic SQT patients. Every patient whose family history looks suspicious should be evaluated with EKG and holter EKG for 24 hours.

CONCLUSION

Firstly paediatricians should be made aware that SQT is one of the causes of sudden cardiac death. Pediatricians should direct every patient with suspicious death history in the family to pediatric cardiologists for further examination. Spot EKG is insufficient to rule out SQT syndrome.

Conflict of interest

No conflict of interest was declared by the authors.

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