Guidelines recommend using VKAs for at least 3 months in patients with LVT or those who are at risk for LVT development. However, the duration of triple antithrombotic therapy should be minimized because of an increased bleeding risk (2, 8). We stopped warfarin treatment because of gastric bleeding with INR of 7 and patient non-compliance. Luckily, we could detect LVT on echocardiography in controls. However, the "safe anticoagulation problem" arose in this patient, and we thought that any NOAC would be better and safer than VKAs, which could not be monitored. Because the incidence of gastrointestinal bleeding is lower in AF patients, we chose apixaban as an anticoagulant in this patient (9). Despite the absence of trials in literature regarding the use of NOACs in case of LVT, some reports encourage the use of these agents (6, 7). Investigations that specifically address late LVT formation in STEMI patients are absent in literature. Regarding stroke and mortality, any superiority of warfarin over acetylsalicylate has not been shown in patients with systolic heart failure with sinus rhythm. Hence, decisions regarding the use of anticoagulants in this population should be individualized (10). Antithrombotic effectiveness and low bleeding risks of apixaban may alter our knowledge and change the practices in the future with randomized controlled trials.

Conclusion

LVT can be seen even years after acute STEMI, and long-term anticoagulation decisions must be individualized. We demonstrated the success of apixaban in the resolution of LVT. Randomized clinical trials in the future are necessary to determine the clinical benefits of apixaban in patients with LVT.

Video 1. Mobile apical thrombus located on the septal apical aneurysmal segment.

Video 2. Suspected thrombus image can be seen in the septal apical aneurysm.

Video 3. Apical thrombus could not be detected.

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A rare association with suffered cardiac arrest, long QT interval, and syndactyly: Timothy syndrome (LQT-8)

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Introduction

Timothy syndrome (TS), also referred to as syndactyly-associated long QT syndrome (LQTS) or LQT8, is a multi-system disorder characterized by developmental defects causing dysmorphic facial features, congenital heart abnormalities, neurocognitive impairment, and webbing of the toes and fingers (syndactyly) (1). TS is caused by mutations of the CACNA1C gene, which encodes L-type calcium channel Ca (V) 1.2. Two types of TS have been defined according to the mutation sites: G406R in exon 8A (TS1) and G402S/G406R in exon 8 (TS2). These gain-of-function mutations result in an impaired open-state and voltage-dependent inactivation of the L-type calcium channel, ultimately

leading to a markedly prolonged myocardial action potential (delayed ventricular repolarization) (1-4).

This paper presents a two-and-a-half-year-old female patient who had cardiac arrest (CA) during an operation for inguinal hernia (IH) and syndactyly upon which short-term resuscitation was performed. The diagnosis of TS was established on the basis of the electrocardiography (ECG)-Holter findings [long QTc (corrected QT) (>600 ms), 2:1 AV block (AVB), and T-wave alternans (TWA)] and typical clinical features, and it was confirmed by molecular genetic analysis (MGA). An epicardial dual-chamber implantable cardioverter defibrillator (EDCICD) was implanted in the patient, and treatment with propranolol was initiated. To the best of our knowledge, this is the first report from Turkey about TS.

Case Report

The female patient at the age of two-and-a-half years was referred to us because of the development of CA and the performance of short-term resuscitation during surgery for IH and syndactyly. According to the physical examination, her weight was 10 kg (<3.p) and height was 80 cm (<3.p). She had physical and neuro-motor development retardation. The patient had a slightly dysmorphic-looking face and syndactyly between the 4th and 5th fingers and 2nd and 3rd toes (Fig. 1). According to her cardiovascular examination, she had bradycardia; her peak heart rate was 45–50/min and blood pressure 105/60 mm Hg. She had a 2/6 systolic murmur over the mesocardiac, and her femoral pulses were bilaterally palpable. All other system examinations were unremarkable was normal, and she had an LQTc interval (QTc: 696 ms) and a 2:1 AVB, and T wave alter-

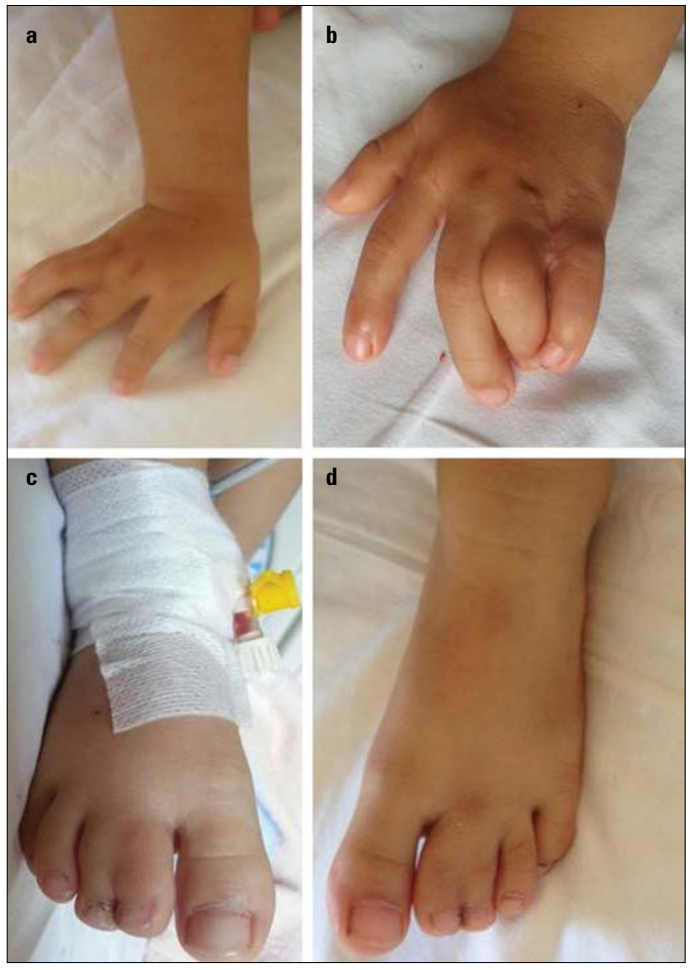

Figure 1. a-d The two-and-a-half-year-old female patient was observed to have syndactyly between her 4th and 5th fingers and 2nd and 3rd toes

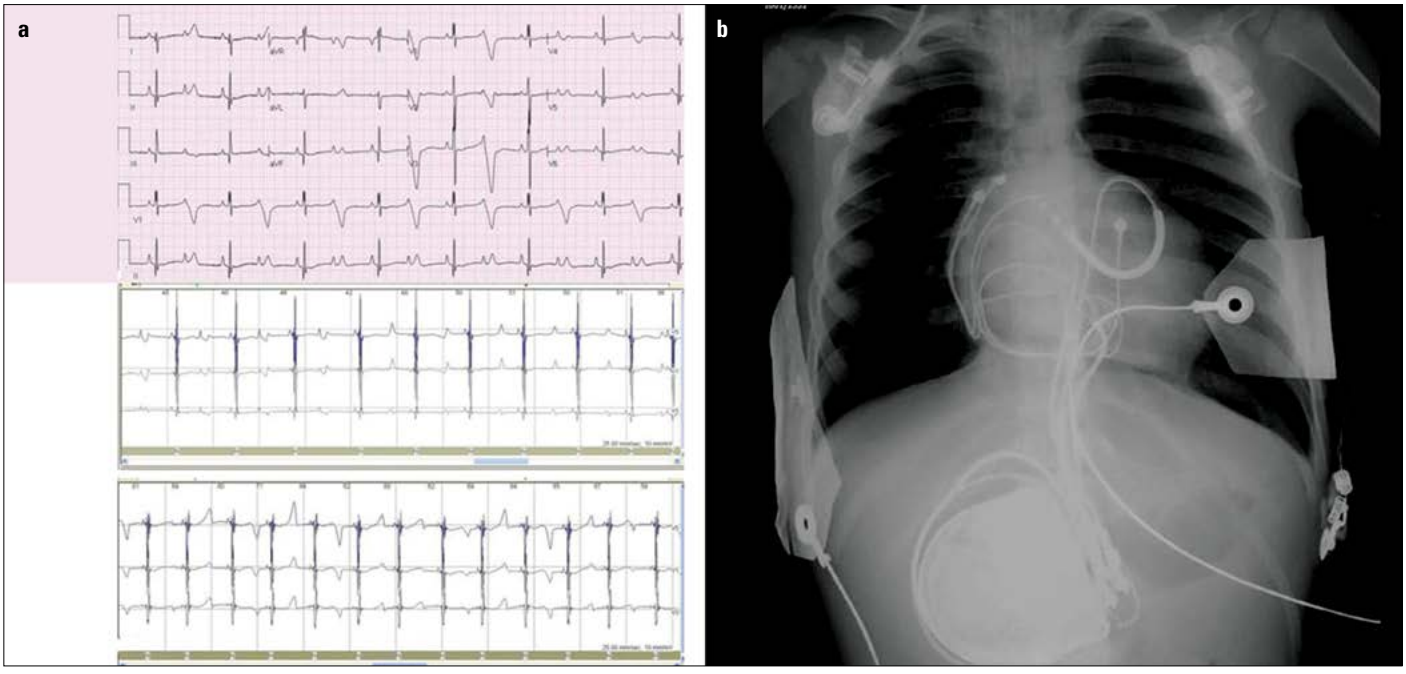

Figure 2. a, b. In the 12-lead-ECG trace of the patient, a long QTc interval (QTc 696 ms) and 2:1 AV block were observed, and Holter ECG monitoring revealed long QT and 2:1 AV block as well as T-wave alternans (a). In the X-ray image, an implanted epicardial dual-chamber ICD (b) can be seen

nans (TWA) as confirmed by 12-lead ECG and 24-h Holter ECG monitoring results (Fig. 2a). Two small mid-muscular ventricular septal defects and a small, patent ductus arteriosus in the echocardiograph were observed. The diagnosis of TS was established on the basis of the ECG and Holter findings and typical clinical features, and it was confirmed by MGA, which showed the typical G406R mutation of exon 8 of the CACNA1C (c.1216G>A) gene. Treatment with a beta-blocker (propranolol, 3 mg/kg/day) was started and EDCICD was implanted in the patient (Fig. 2b).

Discussion

Most children with TS have potentially fatal arrhythmias including 2:1 AVB, torsade de pointes, and ventricular fibrillation (1, 2, 5, 6). The number of TS cases reported in literature in English to date is below 35 (4), and our case is the first genetically diagnosed TS case reported in Turkey.

TS is one of the most severe types of LQTS, and its high mortality at a very young age is likely caused by cardiac arrhythmias precipitated by infections, severe illnesses, or anesthesia (1, 4). In our case, because of CA after anesthesia, LQTS should be considered in such cases. Because ventricular tachyarrhythmia is the leading cause of death in patients with TS, effective antiarrhythmic medications and implantable cardioverter defibrillators (ICDs) are the mainstay of therapy. Because of the small number of patients, no validated drug therapies have been established so far. For other forms of LQTS, beta-blockers are reported to be a treatment option. The calcium channel blockers ranolazine and mexiletine have been used to prevent ventricular tachyarrhythmia (4, 5, 7, 8). Gao et al. (4) showed that mexiletine shortens QTc, attenuates QT–RR slope, and abolishes 2:1 AVB and TWA in a TS patient and TS model via the inhibition of late I_{Na} channel. TS should be considered in every patient with a confirmed diagnosis as soon as the body weight allows the procedure for primary prophylaxis of sudden cardiac death even in patients without documented ventricular tachycardia (1-5). Because the age and weight of our patient were appropriate, an epicardial ICD was implanted in addition to beta-blocker treatment.

Conclusion

The presence of bradycardia, LQT-interval, and 2:1 AVB in combination with multi-systemic signs during the newborn stage and infancy period should suggest TS. Because of the nature and poor progress of the disease, ICD implantation should be performed along with medical treatment for patients at the appropriate age and weight.

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