Clinical Case Reports

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CASE REPORT

Association of a congenital long QT syndrome type 1 with Takotsubo cardiomyopathy

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Key Clinical Message

The occurrence of takotsubo cardiomyopathy in a patient with congenital long QT syndrome has rarely been described. This case report discusses the occurrence of a clinically overt takotsubo cardiomyopathy accompanied by congenital long QT syndrome type 1 in a female patient.

Keywords

Apical ballooning, Congenital long QT syndrome, Takotsubo cardiomyopathy.

Case Report

A 72 year-old woman was admitted for an elective nasal septoplasty to our hospital. A day after surgery, she complained of symptoms of unstable angina pectoris, corresponding to a Canadian Cardiovascular Society (CCS) score of Grade IV. The Troponin I level was markedly elevated (1.973 μg/L) and a 12-lead electrocardiogram (ECG) recorded new T-wave inversions in leads I, aVL, V1-6 (Fig. 1). An immediate coronary angiography revealed insignificant coronary artery disease. However, a subsequent laevo-cardiography (Video S1 and Fig. 2 top) and a transthoracic echocardiography (Fig. 2 bottom) showed signs of typical "apical ballooning" with a concomitant highly reduced left ventricular ejection fraction (EF 33%) consistent with Takotsubo cardiomyopathy (TC). The patient was transferred to the intensive care unit and interestingly, the ECG now recorded a relevant QTc prolongation of 661 ms. A follow-up echocardiography at day 3 revealed an improved EF of 48% accompanied by insignificant regional wall motion abnormalities. In sharp contrast, the daily ECG recordings continually demonstrated a prolonged QTc interval (629 ms) despite optimal beta-blocker-therapy. To rule out a congenital type of long QT syndrome (LQTS), the most common LQTS genes were screened by polymerase chain reaction (PCR). Genetic screening tests proved a novel heterozygous LQTS type 1 mutation in KCNQ1 (3 bp-Deletion c. 1084_1086delAAG). There was no history of ventricular arrhythmias documented in the past. Five weeks after hospital discharge, the patient presented for follow-up in a clinically stable condition with a normal heart function (EF 58%) suggesting good recovery. The QTc interval was 487 ms in the follow-up ECG (Fig. 3).

Discussion

Takotsubo Cardiomyopathy (TC) is typically characterized by a transient and reversible dysfunction of the myocardium unrelated to obstructive coronary artery disease. The pathogenetic mechanisms underlying TC are poorly understood. Among others, high levels of catecholamines have been discussed as one potential mechanism causing

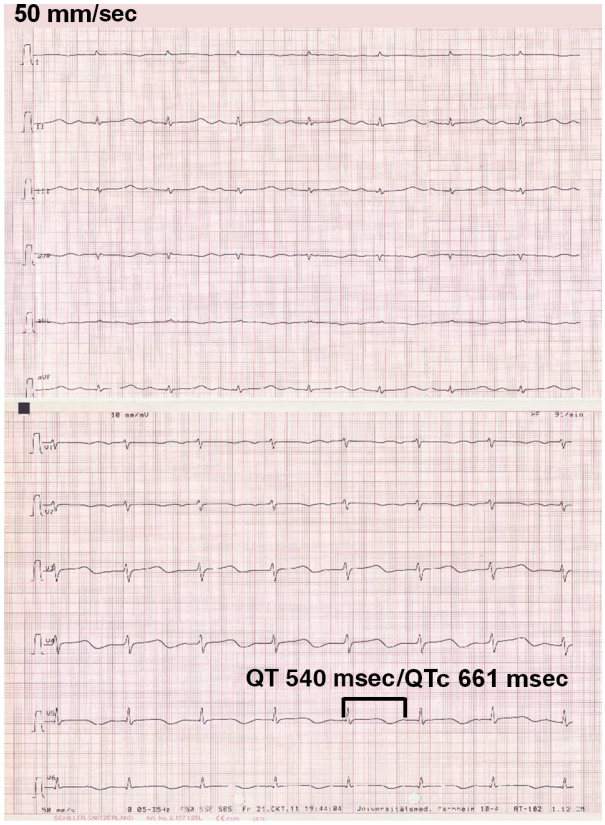

Figure 1. 12-lead electrocardiogram on admission shows new T-wave inversions in leads I, aVL, V1-V6 as well as a QTc prolongation of 661 ms.

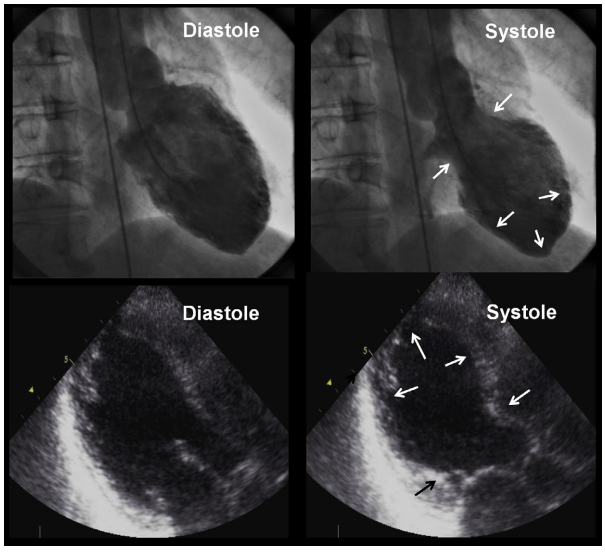

Figure 2. Laevo-cardiography (top) revealed severe wall motion abnormalities of the mid and apical portions of the left ventricle. Transthoracic echocardiography (bottom) demonstrated typical signs of an "apical ballooning" corresponding to TC.

the development of TC. On the other hand, it was demonstrated that electrical disturbances may be caused by toxic catecholamine excess [1]. However, the simultaneous occurrence of both TC and congenital electrical disease has to this date been reported in only two cases [2, 3].

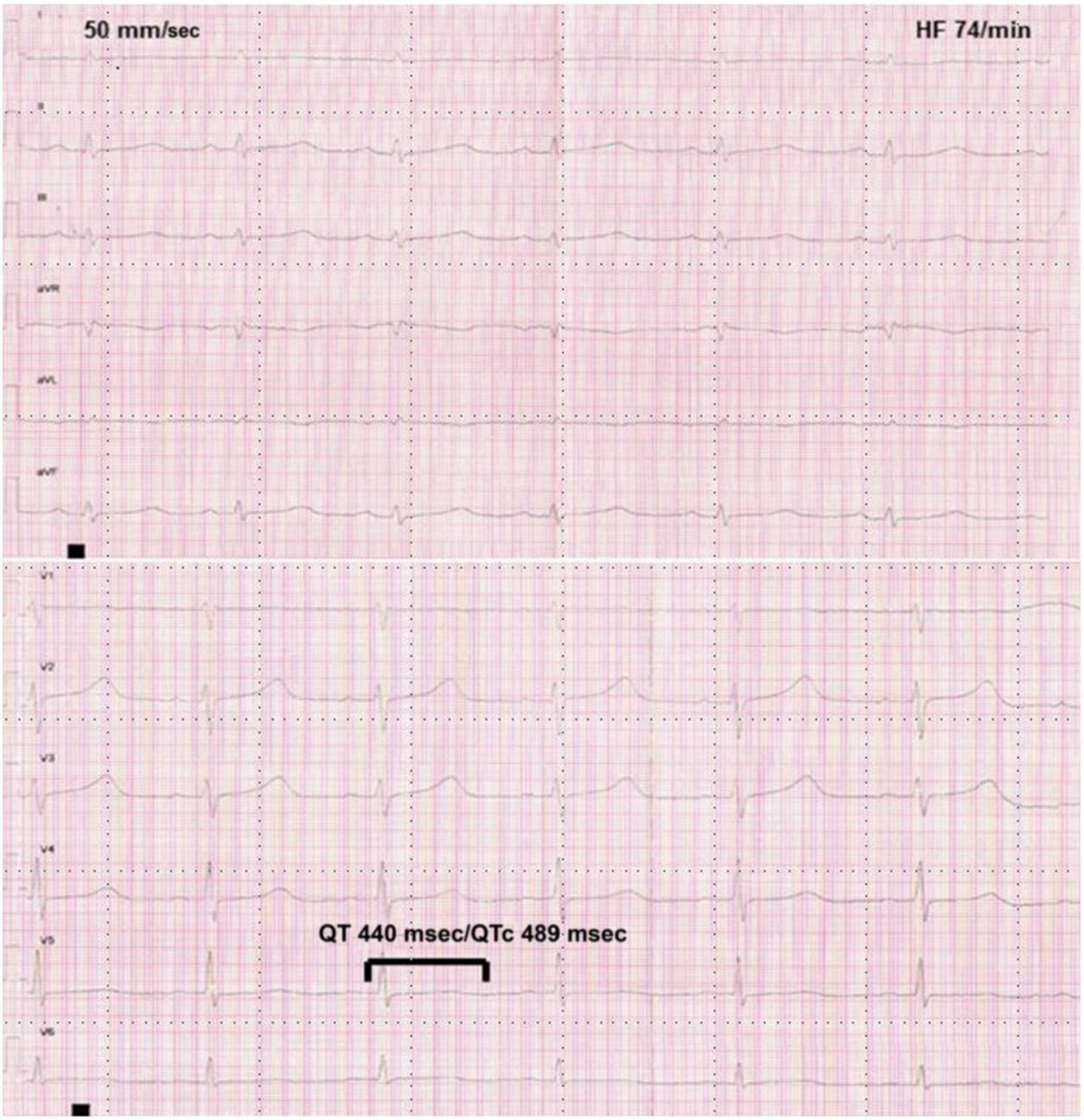

Figure 3. 12-lead electrocardiogram 5 weeks after hospital discharge shows absence of T-waves inversions as well as reducing the QTc duration to 487 ms.

To the best of our knowledge, this case is the first description of a clinically overt TC coincident with LQTS type 1. Both diseases may delay cardiac depolarization and TC has been shown to be associated with altered intracellular Ca++-handling [4]. Prolonged action potential duration, which determines how much calcium enters the myocytes during each contraction-relaxation cycle in LQTS type 1 patients, may have also exacerbated the catecholamine-dependent intracellular calcium overload. This could have increased the susceptibly to TC.

Drugs, electrolyte imbalance (hypokalemia, or hypomagnesemia), and bradycardia are causes which may provoke an acquired LQTS [5–7]. The impact of these causes have been excluded in our case report.

In summary, it could be speculated that the novel mutation of KCNQ1, as in this case, or congenital

electrical diseases in general, play a direct causative mechanical role for the development of TC.

Conflict of interest

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Supporting Information

Additional Supporting Information may be found online in the supporting information tab for this article:

Video S1. Laevo-cardiography proves hypo-/akinesia of the mid and apical left ventricular portions corresponding to TC.