Long QT syndrome and left ventricular non-compaction

Niescalenie miokardium i zespół wydłużonego odstępu QT

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The coincidence of left ventricular (LV) non-compaction (NC) and long QT syndrome (LQTS) is rarely described. We present the case of a 22-year-old woman transferred to the intensive care unit after ventricular fibrillation, which she experienced under stress while giving a lecture. Her 45-year-old mother had experienced sudden cardiac death (SCD). She was unconscious (GCS6). Echocardiography revealed severe systolic dysfunction (LVEF 20%) with apical lateral and posterior segments non-compacted. The criteria of Jenni et al. (Heart, 2001; 86: 661–671) (non-compacted/compacted ratio at end-systole > 2) and Chin et al. (Circulation, 1990; 82: 507–513) (compacted/non-compacted+compacted end-diastolic ratio ≥ 0.33 with communication between the recesses and LV) were fulfilled. Electrocardiograms (ECG) confirmed prolonged QT interval (Fig. 1) — QT interval in lead II is 760 ms. The overt T-wave alternans is seen best in limb leads. The presence of QT interval dispersion above 80 ms confirms the LQTS diagnosis. In the course of intensive care, many ventricular tachycardias — torsade de pointes (TdP) were observed (Fig. 2) and treated (amiodarone, then lignocaine). On the 5th day brain death was observed. The autopsy revealed biventricular dilatation with mild endocardial fibrosis with persistent sinusoid vessels — pathomorphological synonym of NC. In the reported patient, one should suspect a congenital origin: the family history of SCD, acute onset, no history of disease or medication intake and another genetic disorder — LVNC. LQTS includes also the T shape changes: widening, reversing and notching and macroscopic T-wave alternans. In the presented ECG, on the one hand, T-wave is slowly generated with a wide base that may resemble the LQT1 pattern; but on the other hand, T wave is inscribed after a very long ST segment as in LQT3. A clinical history of stress is characteristic for LQT1. A recently released document states that LQTS should be diagnosed in the presence of a QT interval corrected for heart rate using Bazett’s formula (QTc) ≥ 500 ms in repeated 12-lead ECG, and in the absence of a secondary cause for QT prolongation. TdP episodes start with a ‘short–long–short’ sequence of RR cycles including premature ventricular beat with short coupling (Fig. 2). The pathomechanism of QT prolongation is transmural myocardial repolarisation dispersion with action potential prolongation in the medial layer. The interlayer differentiation is also well recognised in LVNC. We may search for the pathogenesis of QT prolongation not solely in channelopathy, but also in the layer alterations in LVNC. Arrhythmic potential might have been increased by amiodarone. In summary, the coincidence of LVNC with systolic dysfunction and LQTS with TdP is rarely described. In our case, it proved fatal, despite adequate therapy.

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