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Electrocardiogram Quiz – Case 7

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A 69-year-old male presented to the emergency department of our hospital with a history consistent with several episodes of spontaneously resolving syncope worsening during the last two weeks. The patient's recent personal history included a pancreatic cancer with hepatic metastases under chemotherapy for the last two months. The patient was hemodynamically stable with normal vital signs. The initial ECG demonstrated ventricular tachycardia of the torsades de pointes type (fig. 1) that was automatically converted to normal sinus rhythm, followed by a new episode of ventricular tachycardia which needed external defibrillation at 200 Joules. The patient was admitted to the hospital for further investigation and treatment.

Questions

α. What could be the reason for the appearance of the torsades de pointes based on the depicted 12-lead ECG (fig. 2)?

β. How would you treat the patient having set the diagnosis?

Comment

Long QT-syndrome (LQTS) comprises a disorder of cardiac repolarization characterized by prolonged QT-interval on a surface ECG, syncope, T-wave abnormalities, ventricular tachycardia of the torsades de pointes type and an increased risk of sudden death. Classically, LQTS is divided into a congenital and an acquired

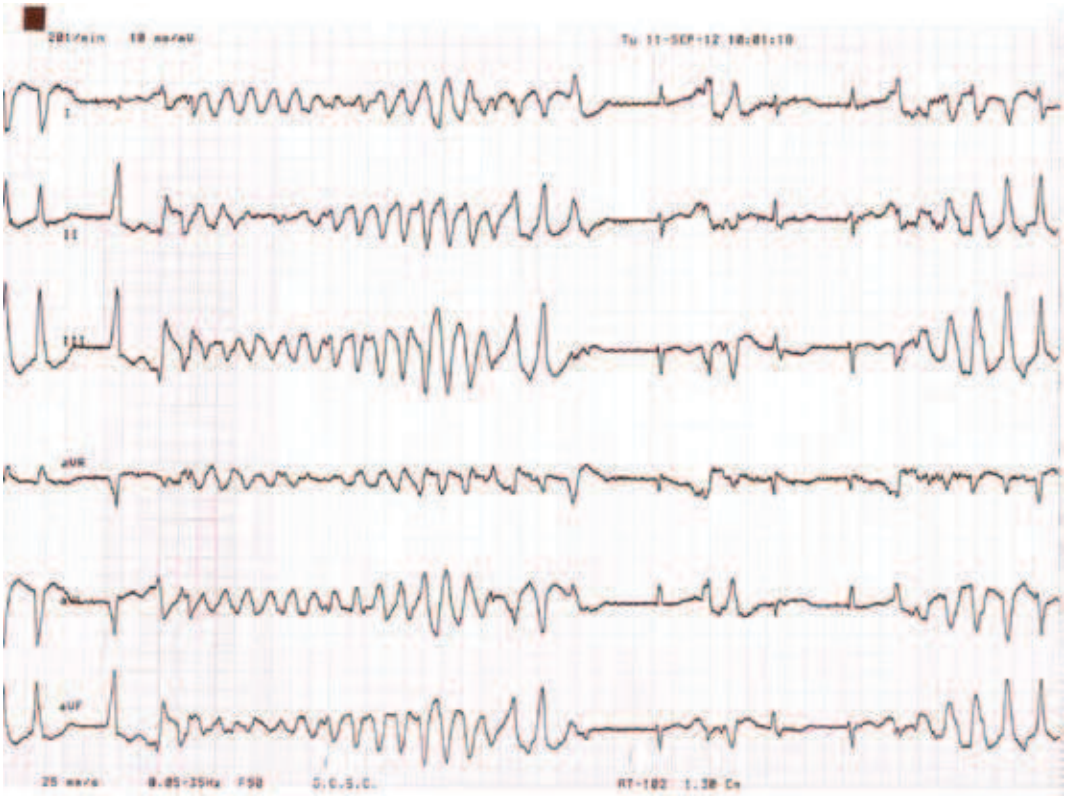

Figure 1

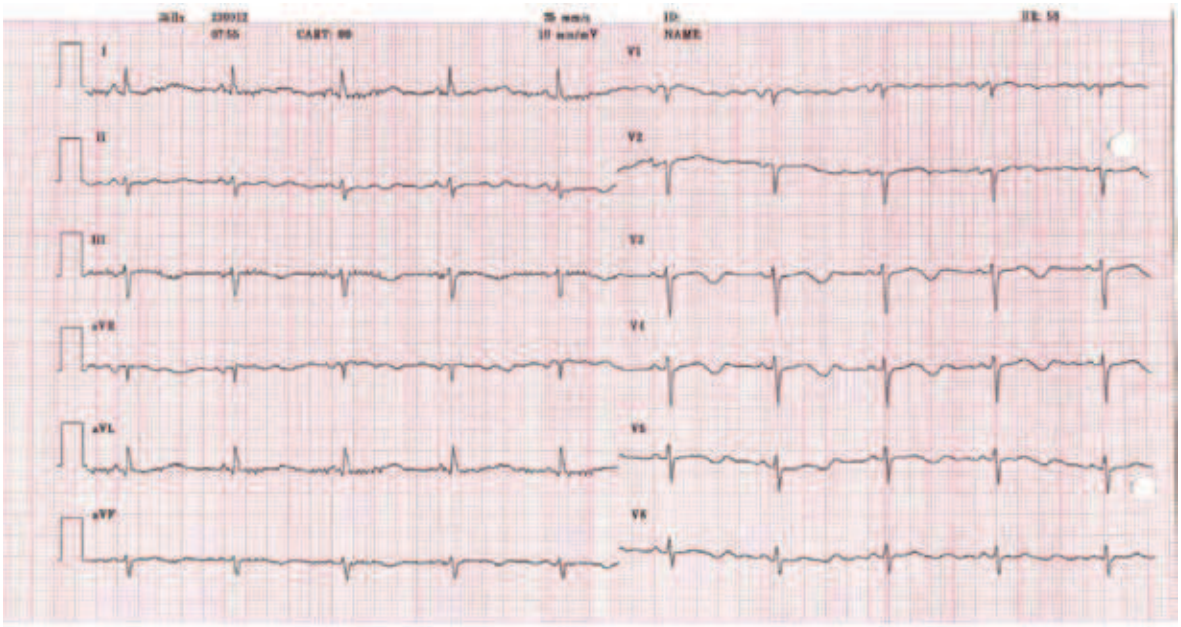

Figure 2

form. Four clinical types of congenital LQTS have been defined: the Romano-Ward syndrome, with autosomal dominant inheritance, the Jervell-Lange-Nielsen syndrome, wherein LQTS is associated with congenital deafness and the pattern of inheritance is autosomal recessive, the Andersen syndrome, where LQTS is variably present with other arrhythmias, periodic paralysis and malformations, and the very rare Timothy syndrome, characterized by a more malignant form of LQTS, cardiac and other somatic malformations, and autism. On the other hand, acquired LQTS is most often due to specific drugs, hypokalemia or hypomagnesemia. A number of specific gene loci have been identified that are associated with LQTS. Genetic testing is clinically available and may help to direct appropriate therapies. The most common causes of LQTS are mutations in the genes KCNQ1 (LQT1), KCNH2 (LQT2), and SCN5A (LQT3).

QT-interval on the surface ECG describes the manifestation of ventricular depolarization and repolarization. It is measured from the beginning of the QRS-complex to T-wave termination and averaged over 3 to 5 beats in a single lead. Prominent U-waves should be included in the measurement if they merge into the T-wave. QT-interval is influenced by heart rate. The RR-interval preceding the QT-interval should be measured for rate correction.

In our patient, the laboratory tests demonstrated no major electrolyte abnormality that could explain the initiation of ventricular tachycardia. Additionally, no drug in his out of hospital treatment could be associated with LQTS, and bedside echocardiography showed no structural or functional myocardial abnormality. However, a more detailed history revealed that the patient had experienced pre-syncope and syncope since the age of 8. The episodes were spontaneously resolving and hence he never sought for medical advice. The question of cardioverter defibrillator implantation arose, however the concomitant oncological history of the patient prevented us from such an intervention. The patient was finally transferred to an Internal Medicine Clinic for further support, and the first-degree relatives of the patient were scheduled for genetic and other investigations.

In conclusion, LQTS, both congenital and acquired, comprises a potentially life-threatening entity with a variable clinical presentation that when diagnosed requires immediate action.

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Diagnosis: *Long QT-syndrome*