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Case report

Iatrogenic life-threating condition in a patient with multiple rare disorders

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ABSTRACT

Andersen–Tawil syndrome (long QT syndrome 7) is a rare inherited disorder, characterized by periodic paralysis, long QT, ventricular arrhythmias and skeletal abnormalities.

A 52-year-old female with a history of long QT-syndrome, ICD implantation (secondary prevention of cardiac arrest) and systemic vasculitis was admitted due to an electrical storm caused by endocarditis. She was admitted again short after discharge due to multi-organ failure, which was caused probably by withdrawal of steroids and VKA. Characteristic dysmorphic features resulted in Andersen–Tawil syndrome suspicion.

If patients have one rare disorder they should not be excluded from further diagnostics, and very detailed outpatient care.

Introduction

Congenital long QT syndrome (LQTS) is a genetic disorder characterized by QT interval prolongation due to gene mutations encoding cardiac ion channel proteins. It predisposes to sudden cardiac death in the mechanism of ventricular tachycardia (VT, mostly *torsade de pointes*) or ventricular fibrillation (VF) [1]. Diagnosis of LQTS is established based on medical history, clinical outcomes and electrocardiography (ECG). Also, genetic tests can confirm the right diagnosis [1].

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One of the LQTS is LQTS7, named also Andersen–Tawil syndrome (ATS). This syndrome is caused by KCNJ2-gene mutation, coding the potassium (K) channel Ik1. Features characteristic for ATS include periodic paralysis, dysmorphic features and heart rhythm disorders (prolonged QT-interval and ventricular arrhythmias) [2].

Case report

A fifty-two-year old white female patient, diagnosed with congenital LQTS at the age of 47 years, just after successfully resuscitated pre-hospital cardiac arrest followed by cardioverter defibrillator (ICD) implantation, with medical history of superficial left arm and brachiocephalic vein thrombosis, epilepsy and anti-PR3 ANCA antibodies vasculitis, was admitted to our Department of Cardiology due to electrical storm (with the most probable cause – electrode damage during the course of endocarditis). Ventricular electrode dysfunction requiring immediate replacement was revealed during ICD control. Implanted cardioverter defibrillator, with all electrodes and the device itself, was removed, a smear from the tip of the ventricular electrode was performed and the bacterial cause of the electrical storm was proven. Additional microbiology results showed the occurrence of methicillin-resistant *Staphylococcus epidermidis*, gentamicin and vancomycin sensitive. Additionally, three blood cultures were collected; each of them proved positive. Transesophageal echocardiography (TOE) demonstrated infective endocarditis with vegetation on both: the atrial electrode (Image 1B) and mitral valve (MV) (Image 1A) leading to severe regurgitation. Antibiogram guided therapy with gentamicin and vancomycin was implemented and conducted for 6 weeks. Due to systemic vasculitis, concomitant therapy with azathioprine and corticosteroids was required as well as antithrombotic treatment due to autoimmune disease, history of thrombosis and temporal immobilization. Therapy with enoxaparin resulted in thrombocytopenia with platelets count 20,000/μL 12 h after low-molecular heparin administration followed by diagnosis of heparin-induced thrombocytopenia (HIT). After the withdrawal of enoxaparin and azathioprine and elevation of steroids doses, the platelet level increased to 50,000/μL.

The patient was transferred to the Department of Cardiothoracic Surgery for MV replacement and tricuspid valve (TV) repair. During cardiopulmonary bypass surgery, biological prosthesis and tricuspid annulus were implanted. During the post-operative period TTE was performed showing that the TV was tight and no dysfunction of MV was observed. Twenty days after surgery the patient was discharged. The patient required long-term antithrombotic treatment due to biological prosthesis, with the following risk factors: history of venous thromboembolism and hypercoagulation – systemic vasculitis. Vitamin K antagonist (VKA) therapy was initiated with the INR ranging between 2.5 and 3.0.

Twenty-six days after discharge the patient was admitted again to the Department of Cardiology in very severe general condition, close to circulation and respiratory collapse. Clinical condition suggested severe, most likely infective, exacerbation of congestive heart failure, with multi-organ failure features, such as acute renal and hepatic failure. The patient reported nausea and emesis for the past several days, in addition to an increasing cough for approximately 3 weeks, persistent elevated body temperature, dyspnea (NYHA class IV), as well as pain in the right calf (including its contusion and edema). Laboratory data included increased alanine and aspartate aminotransferases (3291 IU/L and 7186 IU/L respectively), decreased estimated glomerular filtration rate (30 mL/min/1.73 m^2) as well as increased creatinine and C-reactive protein (CRP) levels (1.8 mg/dL and 44.6 mg/L respectively). Additionally, D-dimer was increased up to 52,605 pg/mL, platelets were below normal range (100×10^3/μL), NT-proBNP value was elevated up to 23,265 μg/L and INR was 1.92. After administration of enoxaparin in the ER the platelet count at admission was 55,000/μL.

Intensive i.v. infusion of furosemide, followed by fractionated doses and oral therapy was introduced. Gradual therapy with metoprolol, which due to congenital LQTS, the patient received permanently, short-acting angiotensin-converting enzyme inhibitor and spironolactone were implemented. Medications doses were adjusted to the blood pressure and heart rate. Increased CRP level was observed; therefore, blood cultures were collected and empirical therapy with piperacillin with tazobactam was initiated. We began with 4.5 g i.v. every 12 h. Then the antibiotic dosage was adjusted to patient's renal

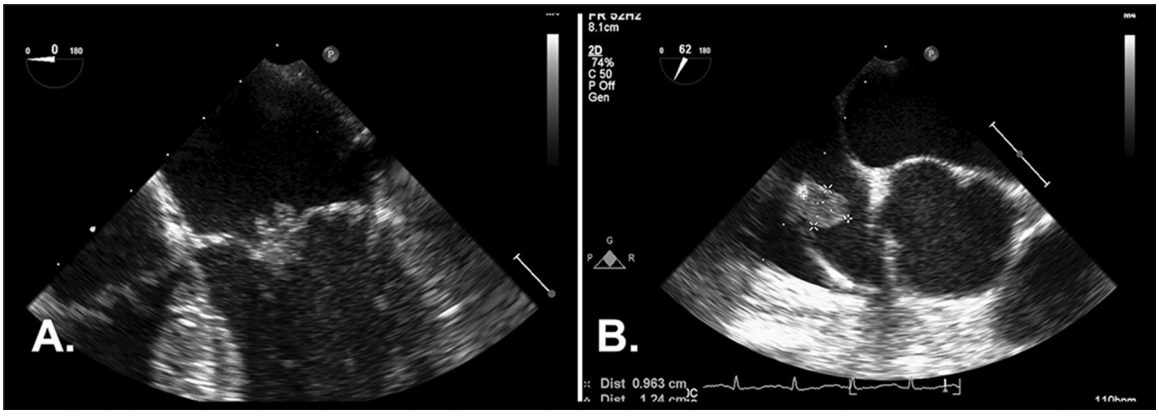

Image 1 – (A) Vegetation on mitral valve; (B) vegetation on atrial electrode's tip.

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