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ATRIAL FLUTTER AS THE FIRST MANIFESTATION OF PROGRESSIVE CARDIAC CONDUCTION DISEASE IN A YOUNG APPARENTLY HEALTHY PATIENT: A CASE REPORT

I. Stepanenko[1], D. Frolov[1], V. Salukhov[1], M. Didenko[2], V. Kitsyshin[1], Z. Khalaeva[1], S. Kushnarev[3]

[1]Molchanov First Department of Internal Medicine Postgraduate Training, Kirov Military Medical Academy, Saint Petersburg, Russian Federation
[2]Kupriyanov First Department of Postgraduate Surgical Training, Kirov Military Medical Academy, Saint Petersburg, Russian Federation
[3]Radiology Department, Kirov Military Medical Academy, Saint Petersburg, Russian Federation

Abstract

We reported a case of a twenty-one-year-old man with an atrial flutter as the first manifestation of progressive cardiac conduction disease. The patient was admitted to the cardiology department due to complaints of shortness of breath and a decrease in exercise tolerance, which had happened after physical exercises (running). During ambulatory ECG monitoring persistent AFL was observed with atrial rate 262-297 bpm and ventricular rate 26-136 bpm (average 56 bpm). AV conduction was very variable – 4:1-14:1. The results of ambulatory ECG monitoring during the whole period of recording indicated signs of atrioventricular conduction disturbances. After cardioversion sinus rhythm was restored additional rhythm and conduction disorders were revealed. Ambulatory ECG monitoring was performed two weeks after the initial one, and throughout this recording were registered sinus rhythm on the background of first-degree AV block; transient Mobitz I AV block; and type 2 second-degree sinoatrial block. Trans-esophageal electrophysiology study was performed. During pharmacological denervation of the heart, signs of slowing of the atrioventricular conduction and sinus node recovery time persisted. These changes along with right bundle branch block were regarded as a progressive cardiac conduction disease with an apparently hereditary cause.

Keywords

progressive cardiac conduction disorder • atrial flutter • Lev-Lenegre disease • young patient

Introduction

Progressive cardiac conduction disease (PCCD) is a hereditary, potentially life-threatening [1] heart disease that is often primarily a genetic disease, but may also be associated with structural heart disease [2–4]. In this article, we report PCCD with atrial flutter (AFL) as a manifestation. To our knowledge, there is only one similar case report, which has been presented in the available literature [5].

Case report

A young, apparently healthy twenty-one-year-old man was admitted to the cardiology department due to complaints of shortness of breath and a decrease in exercise tolerance, which had happened after physical exercises (running). The patient's father died due to sudden cardiac death at the age of 47 years. The physical examination revealed accelerated heart rate (102 bpm) and normal blood pressure. Examination of other organs and systems were without apparent pathology.

On admission to the cardiology department, the electrocardiogram (ECG) showed AFL with pretty regular ventricular rate, (98 bpm), right axis deviation, complete right bundle branch block (RBBB), left posterior fascicular block (LPFB). (Figure 1). Complete blood count showed an increase in the hemoglobin level to 167 g/L, in the biochemical blood analysis, the CPK level exceeded the upper limit of reference values (221.02 U/L), while the CPKMB level was normal. Other biochemical blood parameters were normal. Thyroid function was not impaired.

The echocardiography showed mild mitral regurgitation. The sizes and function of ventricles as well as thickness

Corresponding author: I. Stepanenko
E-mail: icetp@rambler.ru
ORCID: 0000-0001-6436-5656

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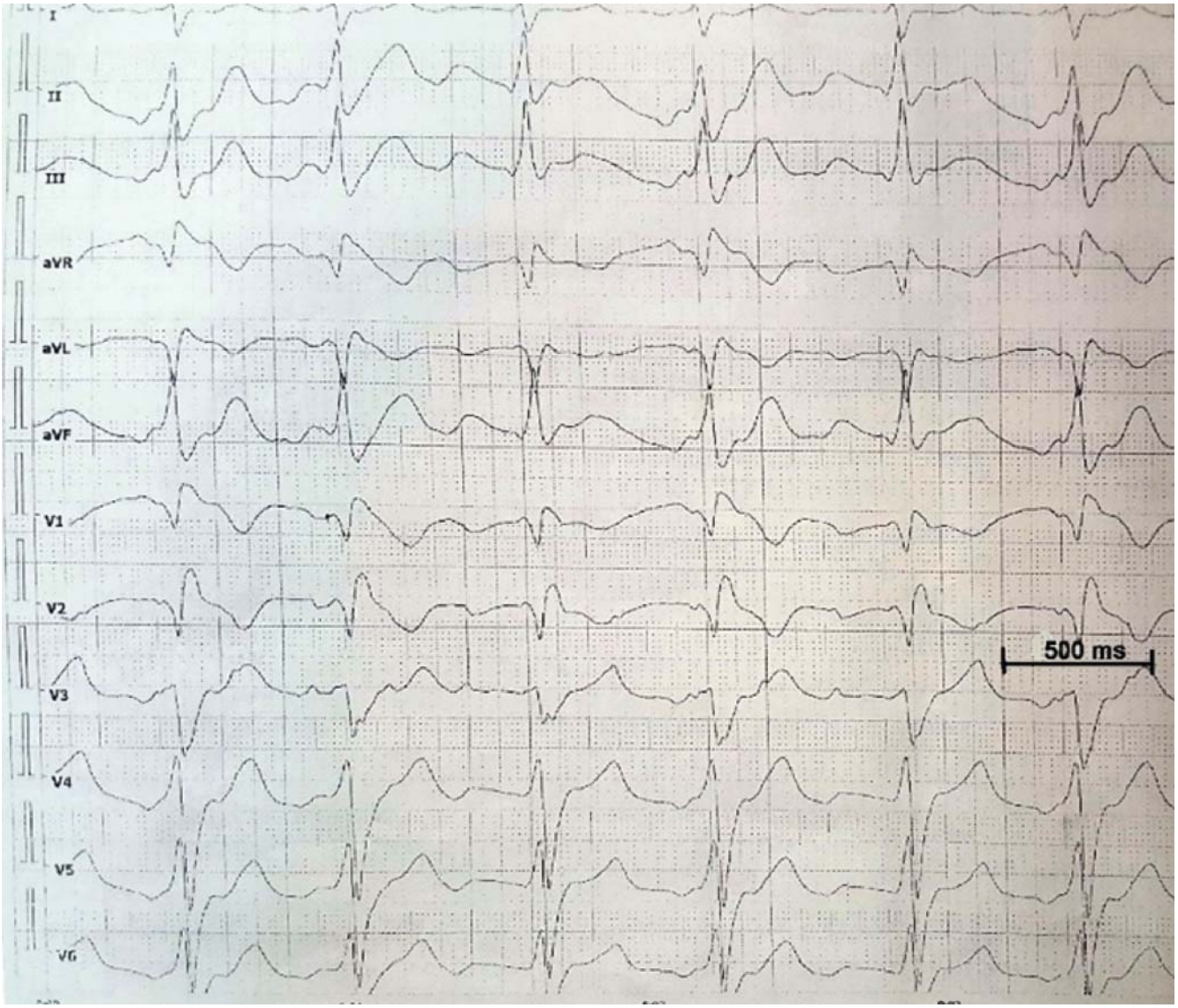

Figure 1. 12-Lead ECG on admission (paper speed 50 mm/s) with AFL with pretty regular ventricular rate (98 bpm), right axis deviation, complete RBBB, LPFB. AFL, atrial flutter; RBBB, right bundle branch block; LPFB, left posterior fascicular block.

of myocardium were normal. Late gadolinium-enhanced cardiac magnetic resonance revealed no evidence of any specific pathologic abnormality including diagnostic features associated with cardiomyopathy or/and inflammation, such as myocardial edema, global hypermedia, and focal necrosis, fibrosis, scar, etc. Similarly, chest computed tomography revealed no abnormality.

During ambulatory ECG monitoring persistent AFL was observed with atrial rate 262-297 bpm and ventricular rate 26-136 bpm (average 56 bpm). AV conduction was very variable – 4:1-14:1. (Figure 2A). 74 pauses (more than 2500 msec) were recorded with maximum duration – 3107 msec (without clinical symptoms). The results of ambulatory ECG monitoring during the whole period of recording indicated signs of atrioventricular conduction disturbances. Abdominal and thyroid ultrasound revealed no pathology.

The patient was successfully cardioverted by 50 J biphasic electrical cardioversion. After a short period of bradyarrhythmia (junctional rhythm 45 bpm, then accelerated junctional rhythm with atrioventricular (AV) dissociation 55-60 bpm) which required an additional 0.1% atropine (1.0 ml) administration, 76 bpm sinus rhythm was restored. The patient received anticoagulant therapy with Rivaroxaban 20 mg once daily before cardioversion (11 days) and 6 weeks afterwards.

According to the ambulatory ECG monitoring repeated two weeks after the initial one, throughout this recording were

registered sinus rhythm on the background of first-degree AV block (Figure 2B); transient Mobitz I AV block (Figures 2C); and type 2 second-degree sinoatrial block (Figure 2D).

A trans-esophageal electrophysiology study was performed. Sinoatrial conduction time (SACT) was 316 msec; maximal SNRT 1587 msec; rate-corrected sinus node recovery time (CSNRT) 712 msec; Wenckebach point was 120 impulses per minute. The effective refractory period (ERP) of the AV node was 400 msec (normal range 280–320 msec). After 3 minutes on the background of pharmacological denervation of the heart, a sinus rhythm with a heart rate of 78 bpm (<80% of the proper value) was recorded; maximum SNRT was 1370 msec; CSNRT 594 msec; Wenckebach point was 130 impulses per minute; ERP of the AV node 360 msec. It was not possible to induce any tachyarrhythmias during programmed stimulation. Conclusion: during pharmacological denervation of the heart, signs of slowing of the atrioventricular conduction and sinus node recovery time persisted.

Genetic testing was not available at the time of patient hospitalization.

Thus, the following diagnosis was made: primary progressive cardiac conduction disease (PCCD) with sick sinus syndrome in the form of transient second-degree sinoatrial block type 2 and paroxysmal AFL; conduction disturbance in the form of a permanent first-degree AV block; transient second-degree AV block type 1; complete right bundle branch block and left posterior fascicular block; without heart failure.

The patient was discharged without health complaints, and with the recommendations to repeat ambulatory ECG monitoring and to be under cardiologist observation.

Discussion

Along with the progressive delay of impulse conduction through the His-Purkinje system, atrioventricular block, right or left bundle branch block (RBBB or LBBB), and development of syncopal states, PCCD can lead to such fatal consequences as sudden cardiac death [6]. In this regard, especially if the

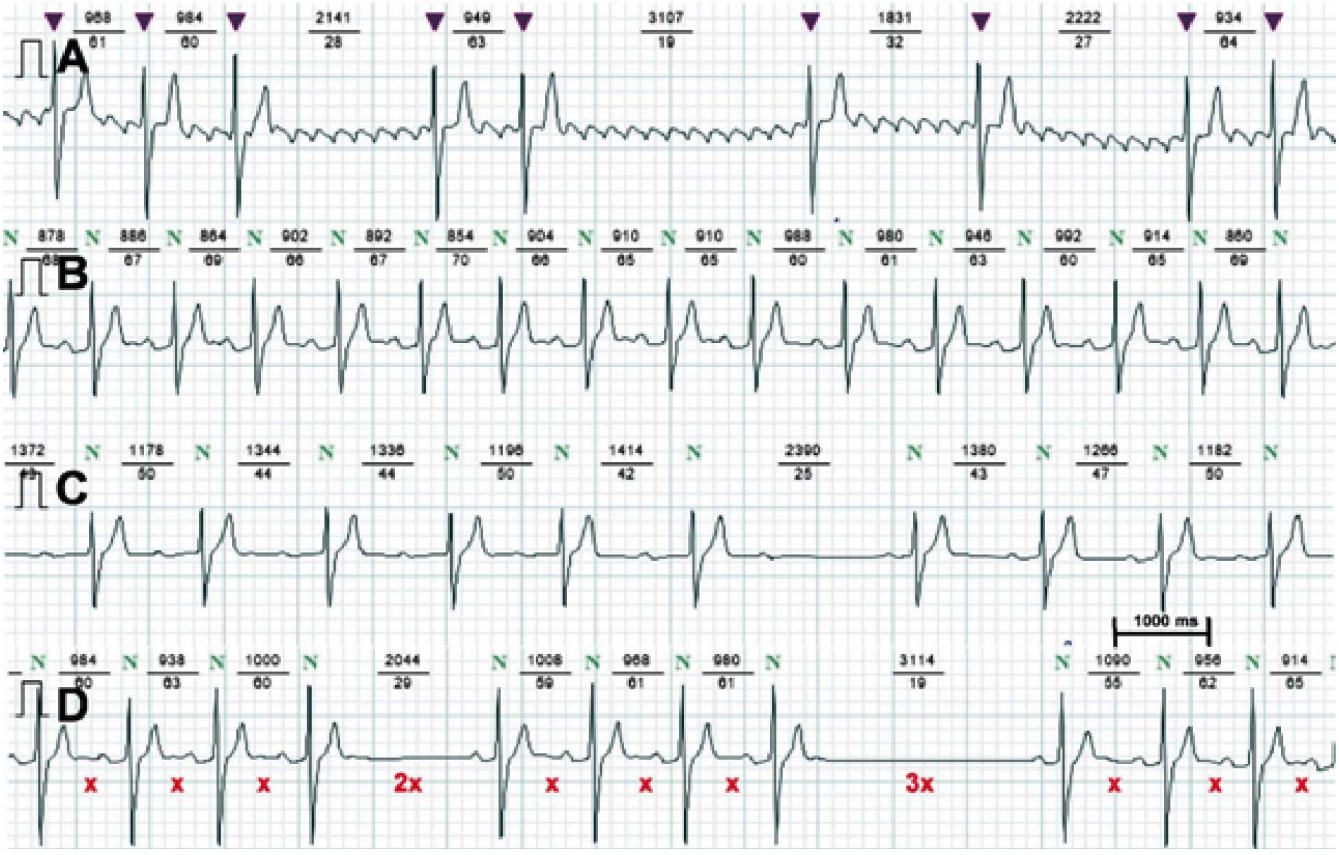

Figure 2. Ambulatory ECG monitoring before (A) and after (B,C,D) cardioversion (speed of recordings 12.5 mm/s). (A) AFL with 4:1-14:1 AV conduction. (B) First-degree AV block. C. Second-degree AV block type 1. (D) Type 2 second-degree sinoatrial block. AFL, atrial flutter; AV, atrioventricular.

patient has a family history of sudden death of first-degree relatives, the presence of signs of PCCD dictates the need to assess risks for the patient and determine the tactics of observation and treatment, including the consideration of pacemaker (PM) implantation [7].

AFL, which was the direct cause of the patient's admission to the hospital, is rare at a young age, and has the most common association with structural heart diseases, including rheumatic heart disease, inherited pathology, and various forms of cardiomyopathy. Despite the fact that according to a number of studies, the risk of thromboembolism in AFL is relatively low [8], the recovery of sinus rhythm in young patients is preferred [9]. Successful cardioversion in this clinical case also allowed us to study in detail the status of the cardiac conduction system of the patient.

Anamnesis (the father died from sudden cardiac death) suggests that the pathology seems to be hereditary. In this regard, the question of the need for genetic testing was raised, but according to the current HRS/EHRA Expert Consensus Statement on the State of Genetic Testing for the Channelopathies and Cardiomyopathies, genetic testing has the recommendation class IIb (may be considered) [10]. The prerequisite for conducting a genetic examination in this case would be the presence of concomitant hereditary heart disease, which was not confirmed during a comprehensive examination of the cardiovascular system.

The rare case presented here demonstrated that PCCD can manifest with supraventricular arrhythmia, which has become an independent factor in the deterioration of the patient's well-being. Given that the disease has a progressive development, such patients should be closely monitored by a cardiologist, so that when the indications appear, the implantation of a pacemaker and the catheter ablation of the atrial flutter may be done in a timely manner.

Conflict of interest

Authors declare no conflict of interests for this article

ORCID

Ivan A. Stepanenko https://orcid.org/0000-0001-6436-5656

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