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EXAMINATION AND TREATMENT OF A FEMALE PATIENT WITH SYMPTOMATIC MANIFESTING WPW PHENOMENON: CASE REPORT

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We describe a clinical case of 37 y.o. woman with anteroseptal accessory pathway associated with left ventricular dyssynchrony and ejection fraction reduction. Wolff-Parkinson-White syndrome and phenomenon diagnostic criteria are discussed.

Key words: WPW phenomenon; WPW syndrome; accessory pathways; left ventricular dyssynchrony; ventricular premature beats; atrial fibrillation; paroxysmal atrioventricular re-entry tachycardia; radiofrequency catheter ablation

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The examination of patients with manifestation phenomenon and Wolff-Parkinson-White syndrome (WPW) does not usually cause any difficulties. The differences between WPW phenomenon and WPW syndrome are defined by the current Recommendations of the Ministry of Health of the Russian Federation and the European Society of Cardiology (ESC) [1, 2]. The guidelines of the Ministry of Health of the Russian Federation state that for a diagnosis of WPW syndrome, the following are required: PR interval shorter than 120 ms; QRS complex longer than 120 ms (in adult patients); registration of a so-called delta wave in the beginning of the R wave and recurrent supraventricular tachycardias (SVT). Importantly, SVT in this context includes paroxysmal atrioventricular reciprocal tachycardias, both orthodromic and antidromic, as well as atrial fibrillation (AF) and atrial flutter with anterograde conduction of excitation via an accessory pathway (AP). The guidelines of the Russian Federation Ministry of

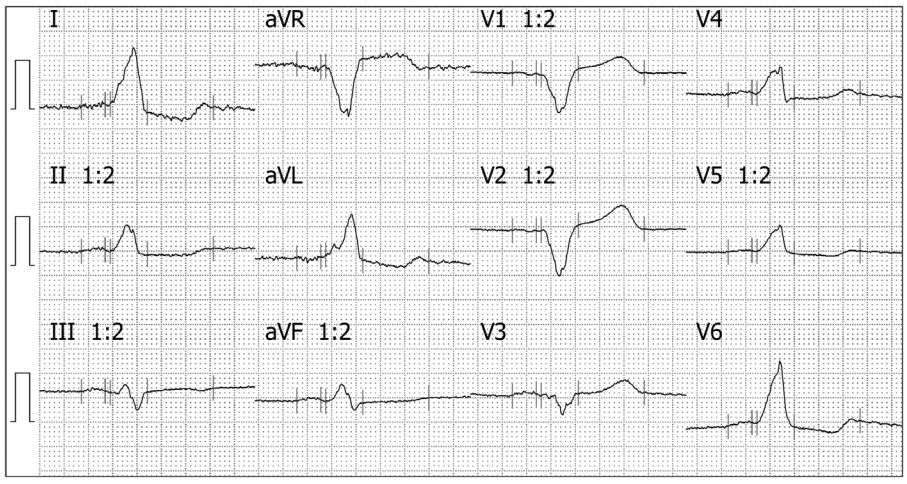

Fig. 1. ECG of patient: signs of pre-excitation with anterior septal location of an additional conduction pathway are recorded.

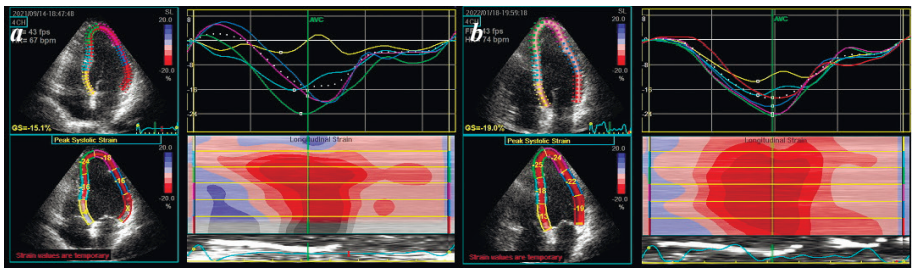

Fig. 2. Results of echocardiographic study of patient: a - baseline, b - after radiofrequency ablation. Explanations in the text.

Health further clearly state: “In the absence of SVT in patients with signs of ventricular pre-excitation, the electrocardiographic phenomenon of WPW is noted”.

In the ESC guidelines you will find important additions to these definitions of WPW syndrome and WPW phenomenon, we believe. It is noted that the delta wave can manifest as a smoothing of both the ascending and descending parts of the QRS complex. It has been observed that WPW phenomenon is found in most cases in patients with a structurally normal heart, except for rare familial (hereditary, genetically determined) forms of ventricular pre-excitation associated with left ventricular (LV) hypertrophy against a background of multisystem disease.

It should be noted that other definitions, such as asymptomatic patients with Wolff-Parkinson-White pattern, can be found in the international literature, in addition to the terms WPW phenomenon and WPW syndrome, which can be divided into high and low risk patients based on electrophysiological examination (EPE), and asymptomatic Wolff-Parkinson-White syndrome [3-5]. We have allowed the unusual term “symptomatic manifest WPW phenomenon” to appear in the title of this case because, on the one hand, the patient did not have SVT at the time of presentation (which prevented the diagnosis of manifest WPW syndrome) and, on the other hand, she had symptoms due to AP functioning with antegrade conduction (which has been proven by the treatment).

Patient G., 37, consulted in the Family Doctor medical center on April 19, 2021. She complained of pains in the left thorax, dyspnea on physical activity (sixth floor stairs) and palpitations. The patient denied having attacks of rhythmic or nonrhythmic heartbeat. The medical history shows that the patient had previously been examined at another health center. She had repeatedly undergone echocardiography (Echo), Holter monitoring (HM) of the electrocardiogram (ECG). Initially, the diagnosis was: Main: WPW Syndrome. Postmyocardotic cardiosclerosis? Complications: Chronic heart failure, functional class I, ventricular extrasystole, grade 5 according to Ryan. Paroxysmal supraventricular tachycardia. Unstable ventricular tachycardia. Six months before coming to our clinic, the patient had undergone transesophageal EPI. Conclusion of the examination: “No evidence of AP, impaired atrioventricular conduction of excitation and sinus node automatism function. Complete block of the left bundle branch. Shortened PQ phenomenon (no signs of ventricular pre-excitation registered during stimulation, St-R 138 ms). Single, paired, paced, frequent, programmed pacing with 1 and 2 stimuli failed to provoke paroxysmal tachycardia.” After a transesophageal EPE was performed, the patient’s diagnosis was changed to “Dilated cardiomyopathy. The phenomenon of shortened PQ. Complete blocka of the left bundle branch.

As there was no doubt about the presence of WPW phenomenon on the ECG presented by the patient (despite the previous transesophageal EPE finding), an examination including ECG recording, HM ECG, Echo (with dynamic assessment) and repeated transesophageal EPE was recommended to determine the further treatment tactics. A characteristic picture of WPW phenomenon was recorded on the registered ECG (Fig. 1). The use of the St. George’s algorithm [6] allowed us to estimate the localization of AP as anteroseptal. Noteworthy is the unusual presentation of a number of leads with 0.5 mV/cm amplification, as well as the results of the automatic QT interval estimation with WPW phenomenon, indicating its “proper” value. As far as we know, there are no generally accepted standards for QT interval estimation in WPW phenomenon and syndrome.

Echo study was performed on the patient on April 22, 2021. There was a marked decrease in LV contractili-

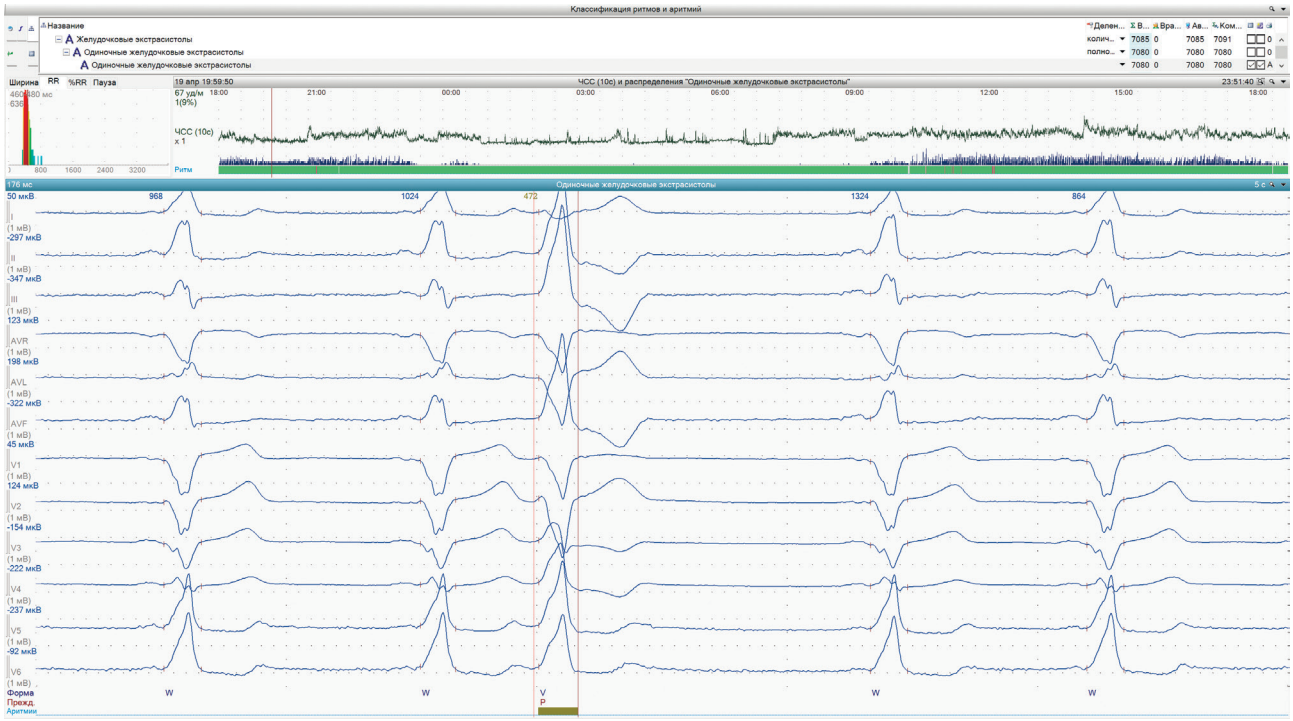

Fig. 3. Data of Holter monitoring of patient: signs of pre-excitation were registered, more than 7000 monomorphic ventricular extrasystoles from the right ventricle outflow tract were detected.

ty - Simpson ejection fraction was 49%, end-diastolic and systolic LV dimensions were 5.2 and 4.2 cm respectively, volumes were 105 and 53 ml respectively, interventricular septum was 0.9 cm, LV posterior wall was 0.7 cm, LV mass index was 94 g/m2. Markers of dyssynchrony were assessed: interventricular septal paradoxical motion was observed, aortic presystolic interval was 152 ms, interventricular mechanical delay was 60 ms, septal-lateral delay was 70 ms (Fig. 2a). Thus, it was concluded that there were signs of LV dyssynchrony. Zones of local contractility abnormality, as well as data for significant valve pathology were not detected.

During an ECG in 12 standard leads (Fig. 3), the ECG of WPW phenomenon was observed throughout the day, more than 7000 single monomorphic symptomatic ventricular extrasystoles (VE) were recorded, which were mainly observed during daytime hours. The configuration of VE as consistent with complete blockade of the left

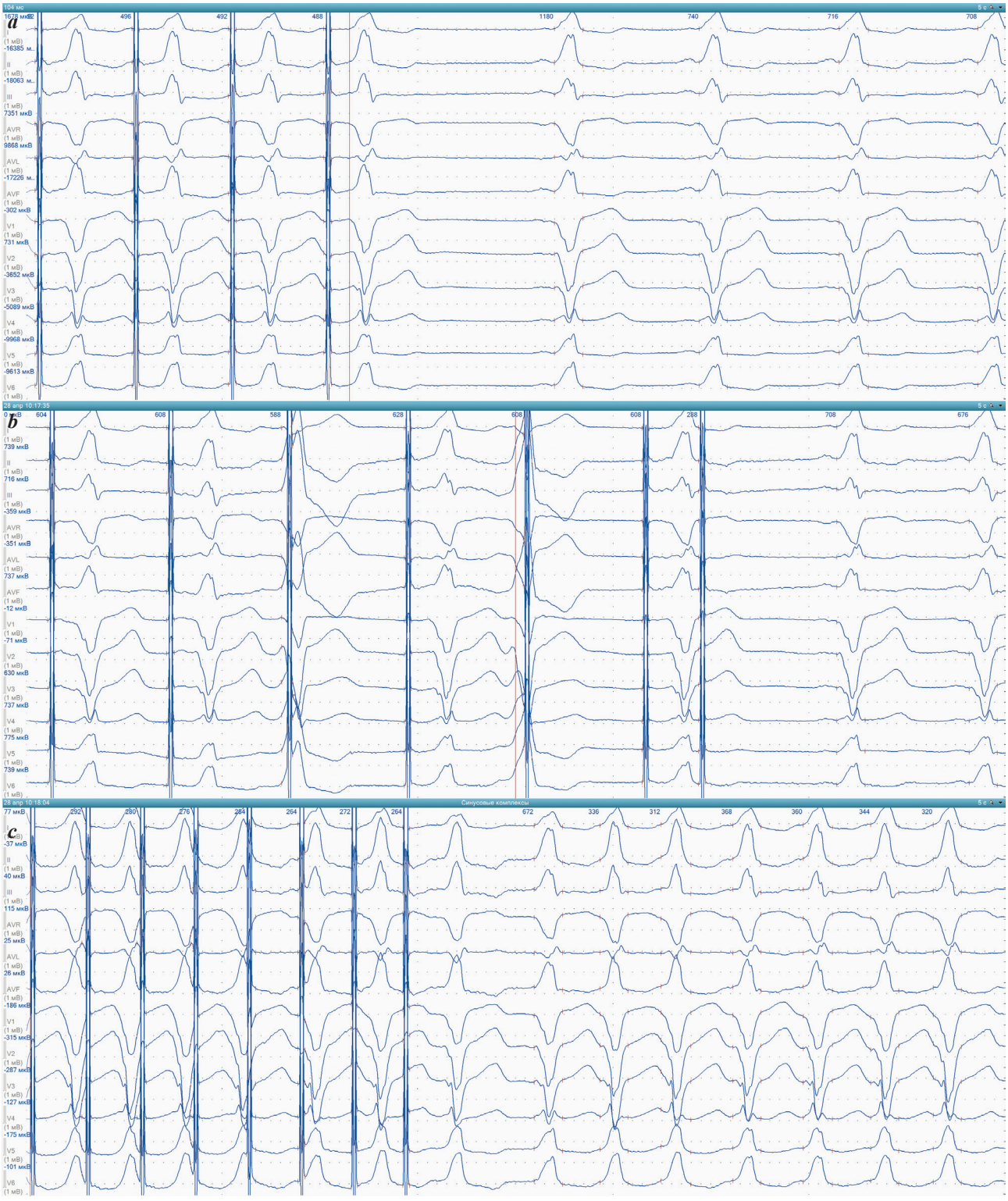

Fig. 4. Fragments of transesophageal electrophysiological study of patient: a - determination of time of sinus node function recovery, b - programmed pacing, c - induction of atrial fibrillation when determining Wenckebach point. Explanations in the text.

bundle branch and its posterior branch (alpha QRS angle greater than 90°), suggesting that the source of VE was in the right ventricular outflow tract (RVOT).

The patient underwent a second transesophageal EPE 6 months after the first one. In orthorhythmic pacing, the St-R interval did not exceed 104 ms (Fig. 4a), and the recovery time of sinus node function was within normal limits. In programmed pacing, the determination of the effective AP refractory period was difficult due to VE. A programmed pacing could not be performed in the absence of VE, so the effective AP refractory period was estimated to be 290 ms (Fig. 4b), which was approximately the same as the value obtained during the first EPE (despite a different interpretation of the results). When determining the Wenkebach point (its value was 220 ppm - Fig. 4c), a AF was induced. According to the patient, she had never had such palpitations before.

In AF (Fig. 5a), the maximum ventricular rate at 10-second intervals was 250 bpm, with "wide" QRS complexes predominating and alternating with single, smaller QRS complexes, which was associated with minimal AP involvement in excitation conduction. Interestingly, in these relatively narrow QRS complexes, there was a marked shift of the ST segment. We tend to regard it as a sign of myocardial ischaemia associated not with coronary artery constriction but with a sudden shortening of diastole duration (in which blood supply to the myocardium occurs) due to high tachycardia. As there was no spontaneous recovery of sinus rhythm within a few minutes, the decision was made to medicate cardioversion.

The patient was given 5 ml of 1% novocainamide solution intravenously slowly (over 15 minutes) to stop AF. A sinus rhythm was restored (Fig. 5b), with a quadrigeminia VE recorded in the background. Thus, a repeat EPE confirmed the presence of WPW phenomenon in the patient and induction of AF, which was not self-limited and required medication-assisted cardioversion. This made it possible to diagnose WPW syndrome and put the patient in

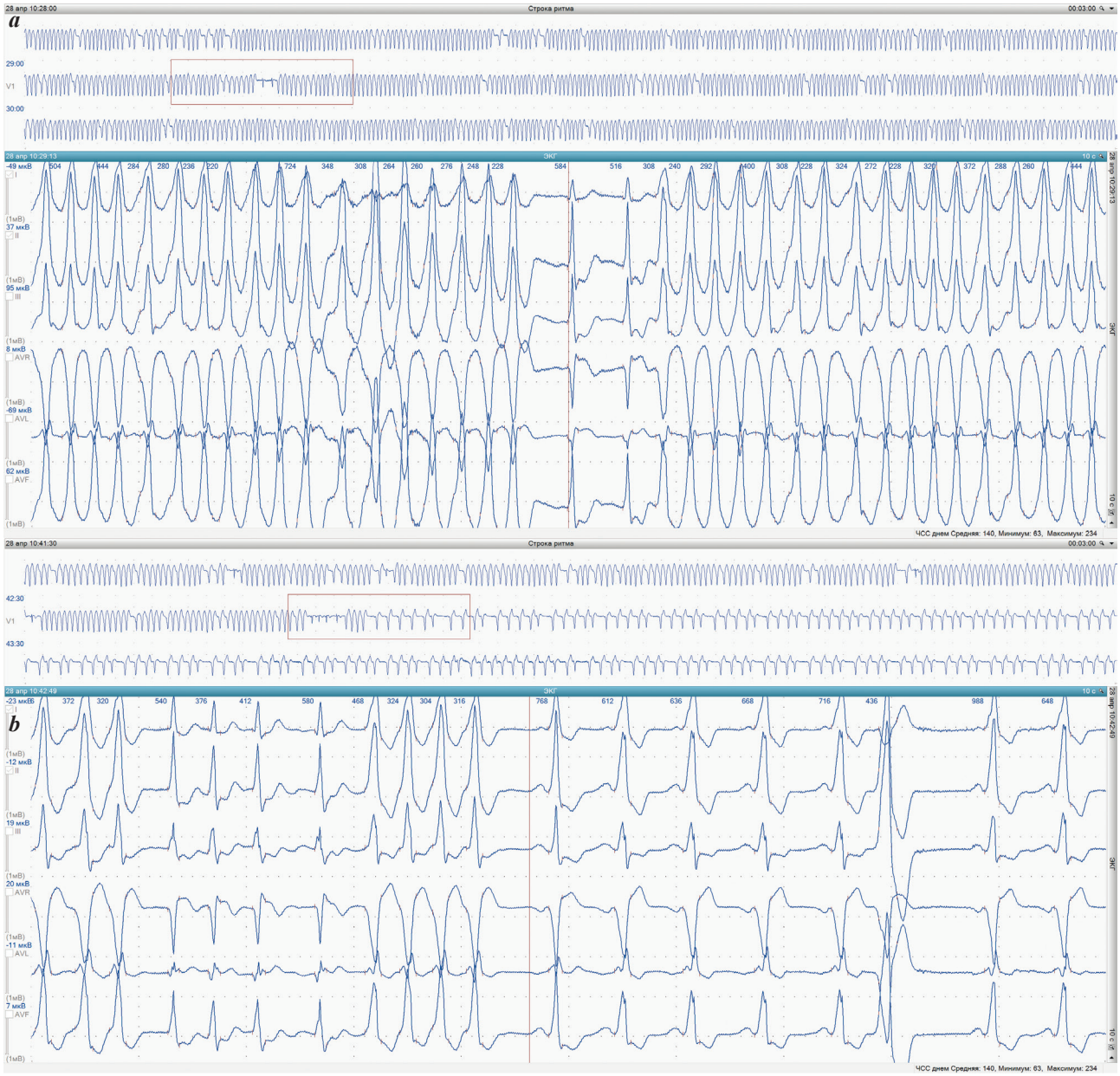

Fig. 5. Atrial fibrillation in patient: a - tachycardia with HR up to 250 per minute, b - recovery of sinus rhythm against the background of procainamide administration. Explanations in the text.

the high-risk category. The patient therefore had absolute indication for AP radiofrequency catheter ablation (RFA), which was performed after 5 months (taking into account the epidemic situation).

During endocardial EPE under local anaesthesia with 30 ml of 0.5% novocaine solution, the right femoral vein was punctured three times. The following electrodes are placed in the right heart chambers: a ten-pole diagnostic unguided coronary sinus catheter (Webster Decapolar 5F, 2-8-2 mm electrode spacing, 110 cm, Biosense Webster, USA) is positioned in the coronary sinus. A quadripolar electrophysiological catheter (diagnostic unguided, Avail Quadrapolar 6F, 10 mm electrode spacing, 115 cm, Biosense Webster, USA) is positioned in the apex of the right ventricle. A Celsius 7F, D curve (Biosense Webster, USA) non-irrigation controlled electrode was chosen as the treatment electrode.

Electrophysiological parameters of the heart, absence of antegrade and retrograde decrementation were determined. The test with intravenous bolus injection of 30 mg of adenosine triphosphate recorded a 'wide' QRS complex, short PQ interval. Repeatedly, tachycardia with retrograde conduction across the AP (Fig. 6a) was induced, relieved by frequent stimulation, and AF paroxysms with conduction across the AP were induced and relieved either by self or by atrial stimulation. Mapping was performed to determine the localization of the AP, which revealed a significant advance in the anterior septal region (Fig. 6b).

As a result of low power testing (20W for 5-7 seconds) in interest, when conduction along the AP disappeared, the PQ interval remained within normal limits, allowing the full effect (60 seconds with 35W) to be applied. According to the repeated EPE results, no evidence of AP function was seen during the 30-minute observation period and the bolus administration of adenosine triphosphate at a dose of 30 mg IV (Fig. 6c).

Fig. 6. Fragments of endocardial electrophysiological study and radiofrequency ablation of patient: a - induced paroxysmal reciprocal orthodromic atrioventricular tachycardia with cycle length of 416 ms and RP' interval of 118 ms, b - ventricular electrogram advance by 40 ms on distal pair of ablation catheter compared with QRS complex, c - ECG and electrogram after effective radiofrequency exposure. Explanations in the text.

Against a background of sinus rhythm without pre-excitation, frequent monomorphic VE was recorded on the ECG. The ECG showed that the VE foci were located in the RVOT. The treatment electrode was replaced with a Celsius ThermoCool 7F, D curve irrigated controlled electrode (Biosense Webster, USA). The electrode was inserted into the RVOT, and activation mapping revealed an excitation advance on the lateral wall of the RVOT of -30 ms, and stimulation mapping obtained a complete match on the 12-channel ECG. Performed RFA for 60 seconds, 35W at an irrigation rate of 30 ml/min, eliminating the VE.

Nine-month follow-up showed no evidence of pre-excitation on repeat ECGs, no rhythmic or irregular heartbeat, no palpitations or dyspnoea on daily physical activity. At 4 months after RFA, the HM ECG showed no signs of pre-excitation, with a single monomorphic VE from the RVOT. A repeat Echo study showed an improvement in LV contractile function with an increase in ejection fraction to 54%. There was no evidence of significant intraventricular dyssynchrony: aortic presystolic interval was 107 ms, interventricular mechanical delay was 8 ms, septal-lateral delay was 15 ms. In addition, there was an in-

crease in global systolic strain (from baseline) from 15 to 19%, combined with synchronisation of segmental strain curves and the absence of segments with postsystolic shortening (Fig. 2b).

DISCUSSION

This case study is interesting, in our view, both because of the difficulty in identifying AP in the patient and because of the difficulty in formulating a diagnosis. On the one hand, the patient had no complaints of rhythmic or irregular heartbeat and no evidence of recurrent atriventricular tachycardia or AF/atrial flutter prior to the repeat EPE. This, despite the presence of many symptomatic VEs, in accordance with current guidelines, prevented her from being diagnosed with WPW syndrome. On the other hand, the patient complained of dyspnea on moderate exertion, the Echo showed a decreased ejection fraction and LV a dyssynchrony, which diagnosed chronic heart failure, which in our opinion did not allow us to speak of an "asymptomatic syndrome or WPW phenomenon".

Unfortunately, RFA has not been able to dot the i's and cross the t's. The procedure eliminated both AP and the source of VEs in the RVOT, resulting in normalization of the LV ejection fraction and elimination of intraventricular dyssynchrony. This does not suggest that the decrease in LV contractility was due to the presence of AP alone. However, a significant role of around 7000 VEs per day in chronic heart failure in a young patient is highly questionable.

In several clinical cases the association between the presence of AP and dilatation and decreased LV contractility has been demonstrated. LV dysfunction has long been thought to develop because of recurrent episodes of tachycardia, but it has now been demonstrated to occur in patients with signs of ventricular pre-excitation and without a history of tachycardia [5, 7]. The true incidence of this phenomenon remains underestimated at present: in a number of such cases patients are diagnosed with idiopathic dilatated cardiomyopathy. On the other hand, LV dysfunction has been shown to be prone to progression over time. Therefore, it is possible to assume that in some patients RFA is performed before clinically significant LV systolic dysfunction has formed [5].

The key pathogenetic factor responsible for impaired LV function in WPW is impaired synchrony of LV myocardial contraction [8]. The group of M.Tomaske et al (2008) studied electrocardiographic and echocardiographic parameters in 34 patients with right septal and posterior septal AP location initially and 1 month after RFA [9]. In this group of patients there was a decrease of QRS complex duration, increase of LV ejection fraction, decrease of delay time between septum and posterior wall after effective RFA.

It has been shown that an important predictor of LV dysfunction in WPW is AP localization. In the described cases of LV dysfunction in WPW, right-sided septal or posterior septal location of the accessory pathway was fixed [9-11]. In support of this, Y.Nakatani et al. (2017) demonstrated higher levels of atrial natriuretic peptide in patients with right-sided and septal location of AP compared to its left-sided location, which probably indicates a greater risk of chronic heart failure in these AP localizations [12].

A recent paper by S. Akimoto et al. (2021) described the characteristics of longitudinal and circular deformation in patients with left- and right-sided AP compared with healthy subjects [13]. More pronounced functional abnormalities were demonstrated in the group with right-sided AP. According to the authors' conclusion, the earliest manifestation of functional impairment is the reduction of circular deformation in the endocardial layer of myocardium, which was observed in both groups of patients with WPW. At the same time, a decrease in the longitudinal deformity indices was recorded only in the group with right-sided AP.

CONCLUSION

Thus, our clinical case illustrates the significance of LV dyssynchrony manifestations associated with AP in a patient without complaints characteristic for paroxysmal tachycardia. RFA resulted in improvement of LV myocardial mechanics, which is quite consistent with the available literature data regarding similar clinical situations. In this regard, it seems timely to initiate a discussion on the revision of the boundaries of "WPW syndrome" and "WPW phenomenon", as well as the possibility to consider LV dysfunction associated with the presence of AP and typical ECG changes as a variant of WPW syndrome, given the obvious clinical significance of such a combination.

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