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Persistent Isolated Right Atrial Standstill Associated With Left Atrial Tachycardia

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Introduction: Atrial standstill is a rare condition, characterized by absence of atrial electrical and mechanical activity evident in surface electrocardiography echocardiography, or fluoroscopy, which is associated with unresponsiveness of atria to maximal output electrical stimulation. This condition can be present with thromboembolic complication, low cardiac output, and sometimes palpitation.

Case Presentation: Here we presented a woman with right atrial stand still and left atrial tachycardia. It was confirmed by electrocardiogram, echocardiography, and intracardiac electrogram in basal state and during maximal output electrical stimulation. We treated her by implanting pacemaker to control bradycardia, oral calcium channel blocker to control palpitation episodes, and anticoagulation.

Conclusions: Atrial standstill can be present partially that can be localized in one atrium and is associated with tachycardia in the other atrium.

Keywords: Atrial stand-still; Atrial Tachycardia; Electrocardiogram

1. Introduction

Persistent atrial standstill is a rare disorder, first described by Chavez et a1. (1). The current diagnostic criteria include: (a) absence of P waves in surface and intracavitary electrocardiograms; (b) absence of A waves in jugular venous pulse and right atrrial atrial pressure tracings; (c) supraventricular type QRS complex; (d) immobility of atrium on fluoroscopy; and (e) inability to stimulate the atrium electrically (2). In present case report, we explained a case with isolated right atrial standstill and associated atrial tachycardia (AT) in left atrium (LA).

2. Case Presentation

A 32-year-old woman, who presented by chronic fatigue and dyspnea on exertion, was referred to the arrhythmia clinic of Shiraz University of Medical Sciences for investigation of an irregular rhythm on electrocardiography (ECG). She had not any previous disorder and there was no history of cardiac disease in her family. On physical examination, the patient was a slender woman with a resting pulse rate of 75 beats per minute and blood pressure of 135/80 mmHg. The "a" waves were absent in jugular venous pulse. The apical impulse was normal. First heart sound was variable, the second heart sound was normal, and a systolic ejection murmur of grade 2/6 with maximal intensity in apex was heard. ECG and Holter monitoring showed fine P waves resembling f waves with group beating of narrow QRS complexes and without pre-excitation (Figures 1 and 2). Exercise tolerance test with Bruce protocol was performed. During exercise, the narrow QRS complexes rate increased regularly due to increased atrioventricular (AV) node conduction. Transesophageal echocardiography revealed left ventricular ejection fraction of 50%, enlarged atria without clot in LA, left atrial auricle or RA. There was no intracardiac shunt such as atrial septal defect, ventricular septal defect, or patent foramen ovale. By impression of atrial flutter with variable block, due to fine P waves in ECG, after obtaining informed consent, the patient was transferred to the electrophysiology (EP) laboratory in fasting state and EP study was performed. EP study revealed variable cycle length (CL) of 834 to 1538, QRS of 100, and QT of 376 and HV of 44 millisecods with variable AV conduction. AH was not recordable. During EP study, no electrical activation was recorded from RA (Figure 3). Moreover, maximal output pacing failed to capture the RA (Figure 4), which was confirmed electrically and fluoroscopically. Coronary sinus (CS) activation by decapolar catheter showed AT with CL of 365 milliseconds, with atrial activation from proximal to distal part. No electrical activity was detected from different poles of duodecapolar catheter in RA. AT was stopped with overdrive pacing from proximal of CS. Slow escape rhythm from distal of CS with CL of 850 milliseconds was started (Figure 5). EP study confirmed right atrial standstill with AT from in LA. A single chamber permanent pace maker (VVIR) was implanted and oral anticoagulation and calcium channel blocker started.

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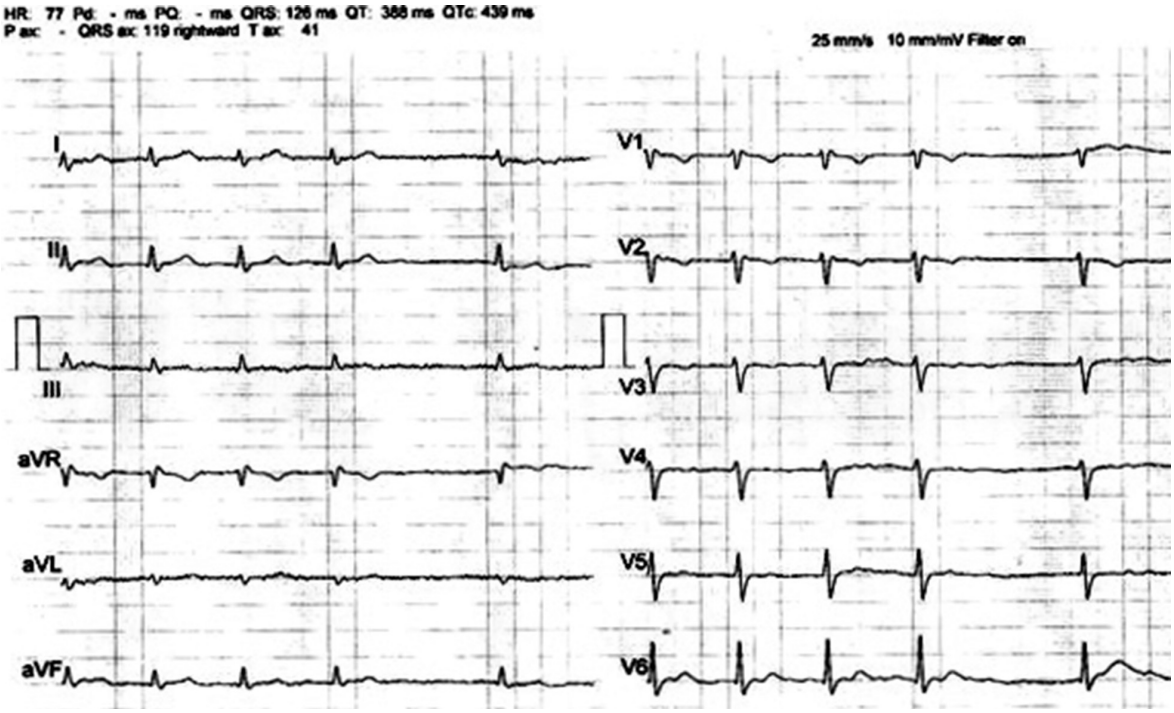
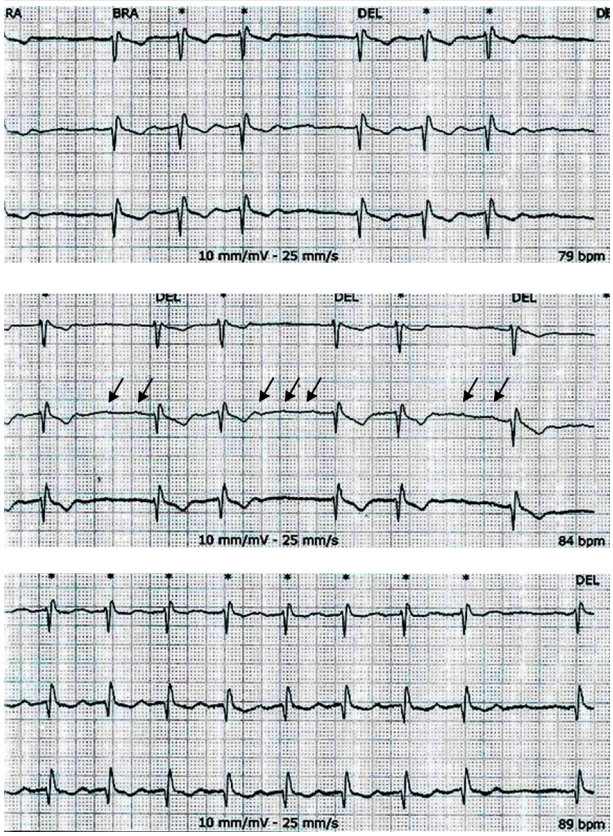
Figure 1. Electrocardiogram at Presentation



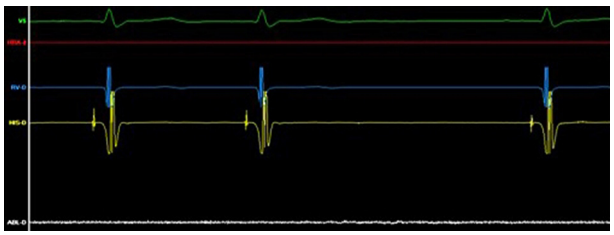
There are fine "P" waves resembling "f" waves with group beating of QRS.

Figure 2. Holter Monitoring of the Patient Showed Fine "P" Waves With Group Beating of Narrow Complex QRS



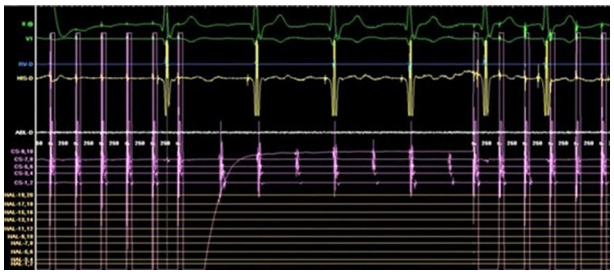
Black arrows indicate fine "P" waves.

Figure 3. Surface Electrocardiography and Intracardiac Electrogram



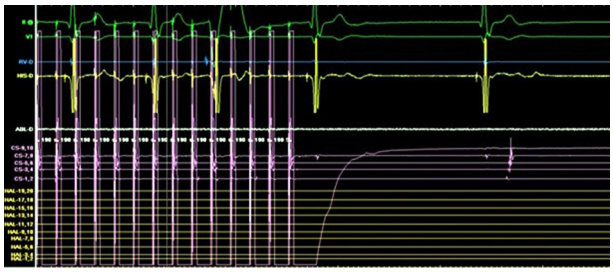
There was no electrical activation in right atrium.

Figure 4. Surface Electrocardiography and Intracardiac Electrogram



Electrical activation recorded from right atrium by duodecapolar catheter and from left atrium with coronary sinus decapolar catheter; no electrical activity is visible on Halo catheter during left atrium capture by proximal coronary sinus pacing.

Figure 5. Surface Electrocardiography and Intracardiac Electrogram



Electrical activation recorded from right atrium by duodecapolar catheter and from left atrium with coronary sinus decapolar catheter. Termination of atrial tachycardia with overdrive pacing from proximal of coronary sinus and escape rhythm from distal of coronary sinus is illustrated.

3. Discussion

Atrial standstill is a rare disease, described by absence of atrial mechanical and electrical activity. This abnormality can involve both atria or only one of them. In the later condition, standstill can be seen in one atrium and is associated with tachycardia in the all part or partial segment of the other atrium. It can be persistent or temporary and can be associated with neuromuscular dystrophy or other cardiac conditions such as valvular heart disease, myocardial infarction, Brugada syndrome, or amyloidosis. Association between Ebstein's anomaly and atrial standstill was reported previously (3). There is also a report about this disorder and idioventricular rhythm in a patient with thalassemia intermedia (4). This arrhythmia has also been reported in familial forms. In our case, atrial standstill was confirmed by the absence of electrical activity in the RA and failure of capture of the RA electrically by maximal output pacing from different parts of it. Nevertheless, LA showed AT that was stopped by overdrive pacing from proximal CS, followed by escape rhythm from distal part of CS. Isolated atrial standstill was reported for the first time by Harley (5). Atrial standstill in association with dilated cardiomyopathy and ventricular tachycardia has been reported too (6). The cause of abnormality in this patient is unknown. *SCN5A* mutation seems to underlie familial atrial standstill. She had no history of muscular dystrophy or progressive cardiac disease as mentioned previously. Her siblings and her parents were not affected. Atrial standstill might lead to syncope, stroke, or heart failure. Treatment is directed toward underlying cause and if it failed, permanent pacemaker implantation and anticoagulation would be recommended for reducing the risk of complications, as we did for this patient.

Authors' Contributions

Mohammad Vahid Jorat and Mohammad Hosein Nikoo: gathering data, studying patient, final decision about the patient, and revision of final manuscript. Aida Yousefi: data gathering and preparing the first draft of manuscript.

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