

Permanent atrial standstill with irregular junctional ectopic rhythm mimicking atrial fibrillation

Atriyal fibrilasyonu taklit eden kalıcı atriyal duraklama ve düzensiz ektopik kavşak ritmi

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Summary – We present an interesting case of “pseudo” atrial fibrillation which was further diagnosed as atrial standstill with irregular junctional ectopic rhythm during electrophysiologic study. A 56-year-old woman presented to a health facility with symptoms of palpitation, lightheadedness, and shortness of breath. Upon detection of irregular rhythm with narrow QRS complexes and no visible P waves on the electrocardiogram, newly developed atrial fibrillation was considered and a direct current cardioversion was performed, during which cardiac asystole developed necessitating cardiopulmonary resuscitation. The patient was then transferred to our institution. Echocardiographic examination showed biatrial dilatation, normal left ventricular systolic function, marked left ventricular hypertrophy, severe aortic stenosis, moderate mitral regurgitation, and severe tricuspid regurgitation. The electrocardiogram showed an irregular rhythm with narrow QRS complexes without any fibrillatory f waves and 24-hour Holter monitoring revealed three episodes of ventricular asystole lasting for more than 3.5 seconds. During the electrophysiologic study, no electrical activity was observed at the high and low levels of the right atrial lateral free wall and septal wall. The final diagnosis was established as atrial standstill and irregular junctional ectopic rhythm. The patient refused aortic valve replacement and died due to progression of the underlying disease one year following permanent pacemaker implantation.

Özet – Bu yazıda, ilginç bir “yalancı” atriyal fibrilasyon olgusu sunuldu; daha sonra yapılan elektrofizyolojik çalışma sayesinde hastaya atriyal duraklama ve düzensiz ektopik kavşak ritmi tanısı kondu. Elli altı yaşında kadın hasta bir sağlık kuruluşuna çarpıntı, bayılma hissi ve nefes darlığı yakınmalarıyla başvurmuş. Elektrokardiyogramında dar QRS kompleksinin eşlik ettiği düzensiz ritim izlenmesi ve P dalgalarının görülmemesi üzerine, yeni gelişen atriyal fibrilasyon düşünülerek hastaya doğru akımlı kardiyoversiyon uygulanmış ve bu sırada kardiyopulmoner canlandırma girişimi gerektiren kardiyak asistol gelişmiş. Hasta daha sonra kurumumuza yönlendirildi. Ekokardiyografik incelemede iki atriyumda da genişleme, belirgin sol ventrikül hipertrofisi, ciddi aort darlığı, orta derecede mitral ve ciddi triküspit yetersizlik izlenirken, sol ventrikül sistolik fonksiyonu normal bulundu. Elektrokardiyografide dar QRS kompleksinin eşlik ettiği düzensiz ritim ve fibrillatuvar f dalga yokluğu, 24 saatlik Holter izleminde ise 3.5 saniyeden uzun süren üç adet ventrikül asistol atağı izlendi. Elektrofizyolojik çalışmada, sağ atriyum lateral serbest duvarının ve septal duvarının yukarı ve aşağısında elektriksel aktivite izlenmedi. Bu bulgularla tanı atriyal duraklama ve düzensiz ektopik kavşak ritmi olarak kondu. Hasta aort kapağı değişimini kabul etmedi ve kalıcı kalp pili yerleştirildikten bir yıl sonra altta yatan hastalığının ilerlemesi nedeniyle kaybedildi.

Atrial standstill is rare cardiac disorder characterized by the absence of electrical and mechanical atrial activity. It was first reported by Chavez et al. in 1946 and was later classified by Levy et al. into two forms: partial and total.^[1] To distinguish these

forms, careful atrial mapping should be performed. From pathological point of view, fibroelastosis, fatty infiltration, amyloid deposits, myocarditis,

Abbreviations:

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| AF | Atrial fibrillation |
| AS | Atrial standstill |
| DC | Direct current |

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and signs of secondary heart disease have been reported on examination of atrial biopsy materials in patients with AS.^[2-6]

We present an interesting case of “pseudo” atrial fibrillation which was diagnosed as atrial standstill with irregular junctional ectopic rhythm during electrophysiologic study.

CASE REPORT

A 56-year-old female patient presented to a health facility with symptoms of palpitation, lightheadedness, and shortness of breath. Electrocardiographic examination showed irregular rhythm with narrow QRS complexes and no visible P waves. The patient was considered to have newly developed atrial fibrillation and a direct current cardioversion was planned. Following three weeks of anticoagulation therapy, DC cardioversion was performed, during which cardiac asystole developed necessitating cardiopulmonary resuscitation. The patient was then transferred to our institution for further diagnostic and therapeutic considerations.

On physical examination, she had an irregular pulse with a heart rate of 95 beats per minute, blood pressure was 90/60 mmHg, and an apparent systolic ejection murmur was heard over the right second intercostal space radiating to the apex. Echocardiographic examination showed biatrial dilatation, normal left ventricular systolic function, marked left ventricular hypertrophy, severe aortic stenosis (aortic valve area 0.8 cm²), moderate mitral regurgitation, and severe tricuspid regurgitation with a pulmonary artery systolic pressure of approximately 70 mmHg. Electrocardiographic evaluation performed at our institution also showed an irregular rhythm with narrow QRS complexes without any fibrillatory f waves. Because of the history of lightheadedness and palpitations, 24-hour Holter rhythm monitoring was performed, which revealed three episodes of ventricular asystole lasting for more than 3.5 seconds. No clinically significant tachycardia episodes were observed. Then, an electrophysiologic study was planned. Diagnostic catheters were placed at the high right atrium, right ventricular apex, and His position. There was no electrical activity at the high and low lateral right atrial free wall or high or low right atrial septal wall. It was impossible to engage the coronary sinus or enter the left atrium through the patent foramen ovale to record left atrial activity. Transseptal catheterization was not performed

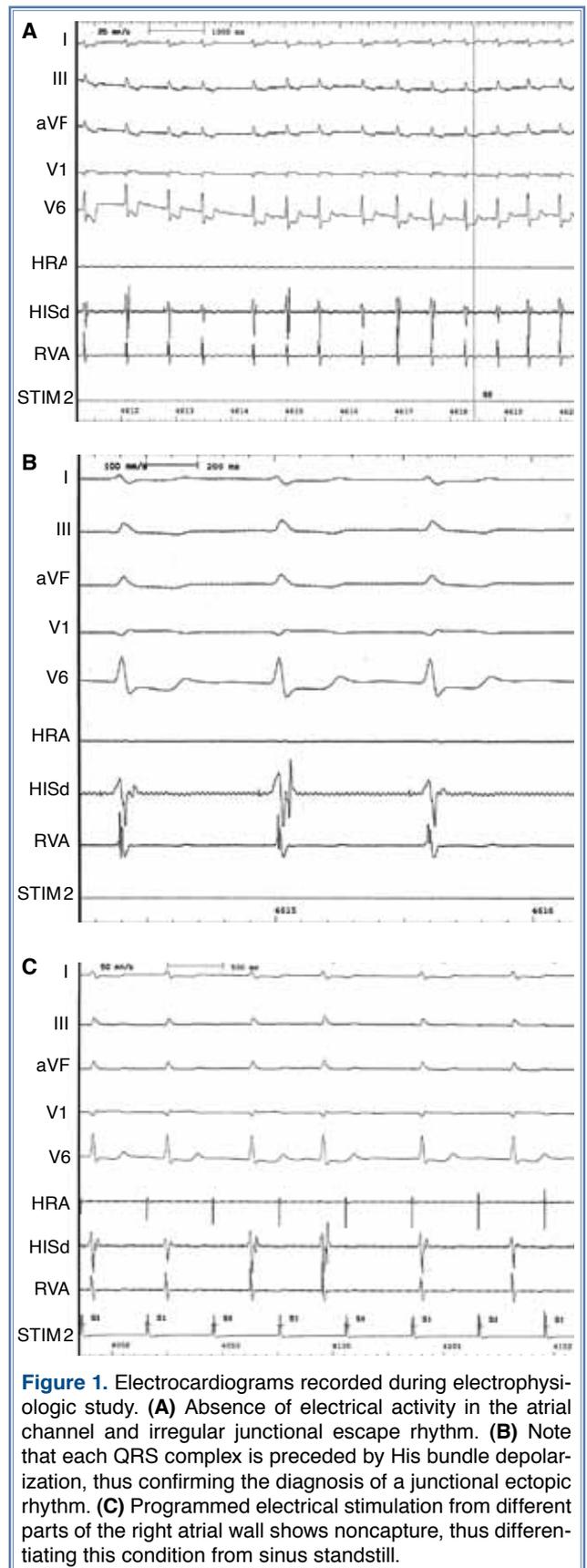


Figure 1. Electrocardiograms recorded during electrophysiologic study. **(A)** Absence of electrical activity in the atrial channel and irregular junctional escape rhythm. **(B)** Note that each QRS complex is preceded by His bundle depolarization, thus confirming the diagnosis of a junctional ectopic rhythm. **(C)** Programmed electrical stimulation from different parts of the right atrial wall shows noncapture, thus differentiating this condition from sinus standstill.

because of its invasive nature. The final diagnosis was established as AS and irregular junctional ectopic rhythm based on the findings of the electrophysiologic study (Fig. 1). The patient refused aortic valve replacement and died due to progression of the underlying disease one year following permanent pacemaker (VVIR mode) implantation.

DISCUSSION

Atrial standstill is generally associated with an underlying primary cardiac disease or arises from secondary involvement of the heart. Familial forms of AS have also been reported.^[4,7-9]

The presentation of our case was challenging and out of the routine daily practice. This case is important because of its electrocardiographic presentation mimicking AF. This kind of electrocardiogram can be easily misinterpreted as AF, which can lead to delivery of inappropriate therapy, as it was in our case. Direct current cardioversion should be avoided in patients with absence of fibrillatory activity on the surface electrocardiogram due to the high risk for postprocedural asystole or severe bradycardia. If one considers DC cardioversion in a patient in whom fibrillatory activity is absent on the surface electrocardiogram, a temporary pacing wire should be placed and connected to a battery prior to the procedure. This approach is further supported by the fact that approximately 50% of patients with AS suffer from syncope.^[10] Due to the progressive nature of the disease, at least 24–48 hours of cardiac rhythm monitoring must be performed periodically in every patient with AS irrespective of the presence or absence of symptoms related to bradyarrhythmias.

Although the use of magnetocardiograms has been reported to be useful in the diagnosis of partial AS, it is difficult to establish diagnosis of AS with noninvasive diagnostic tools.^[11] Electrocardiographic demonstration of prominent fibrillatory f waves can rule out AS, but it is not always possible. In patients with chronic AF, voltage of f waves sometimes decreases and it may be difficult to distinguish AF from AS in the presence of a regular ventricular rate (patients taking digitalis and/or other therapies for rate control, or patients with digitalis intoxication). Decreased f wave voltage generally indicates progression of atrial dilatation leading to progression of atrial dysfunction, which itself can degenerate into AS.

Echocardiographic assessment is important in diagnosing morphological cardiac abnormality and

should be performed in all patients with AS. Two-dimensional echocardiography generally shows dilatation of both atria together with signs of an underlying cardiac pathology or presence of thrombus. Doppler assessment of mitral inflow pattern can reveal the absence of A wave, which points to the absence of mechanical atrial activity, but this is not enough to make a differentiation between AS and AF.

We are of the opinion that junctional ectopic rhythm observed in our patient has the same features with junctional ectopic tachycardia. This is a narrow QRS complex tachycardia characterized by an irregular cycle length, sinus capture beats, periods of variable atrioventricular and/or ventriculoatrial relationships, and ventriculoatrial dissociation. Electrophysiologic hallmark of tachycardia is that each QRS complex is preceded by His bundle depolarization. The His-ventricular interval is always normal except in the setting of underlying conduction system disease.^[12] Another possible cause of irregular ventricular rate during the course AS is electrical activity originating from atrial myocytes in close vicinity of the tricuspid annulus. Indeed, in most cases of AS, the disease first involves the high lateral right atrium, shows slow progression toward the lower right atrium, and finally atrial electrocardiograms are recorded only around the tricuspid annulus and interatrial septum.^[10] However, careful annular mapping revealed absence of such an electrical activity in our patient, thus confirming the advanced stage of the disease in this case. We cannot rule out the possibility of AF localized to the left atrial free wall, but even so it cannot be conducted to the ventricles because of the presence of exit block in the area surrounding the localized AF. If this electrical activity could be conducted to the ventricles, then we would record this activity at the vicinity of the atrioventricular junction. However, it should be noted that we cannot absolutely exclude the possibility of AF localized to a small portion of the left atrium that might have conducted down to the ventricles via electrically active fibers of the coronary sinus. Nonetheless, this kind of conduction is very rare.

In conclusion, electrophysiologic study is an essential tool in the diagnosis of AS. The absence of electrical activity and failure of pacing at a maximum output at multiple sites within the left and right atria can establish the diagnosis of complete AS.

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Key words: Atrial fibrillation; diagnosis, differential; electrocardiography; electrophysiology; heart atria; heart conduction system.

Anahtar sözcükler: Atriyum fibrilasyonu; tanı, ayırıcı; elektrokardi-yografi; elektrofizyoloji; kalp atriyumu; kalp iletim sistemi.