Case Report

Atrial Standstill: a Rare Case

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Abstract

We introduce a 32-year-old man who was evaluated for a dizziness and headache of unknown origin for at least two months and was referred to our center after ECG findings. He was finally diagnosed as a case of idiopathic, familial, diffuse, persistent atrial standstill, which is a rare arrhythmogenic condition characterized by the absence of electrical and mechanical activity in the atria. He successfully received a single-chamber permanent pacemaker.

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Introduction

Atrial standstill or silent atrium is a rare condition presenting with the absence of electrical and mechanical activity in the atria. It presents with the absence of P waves, bradycardia, and wide QRS complex in the electrocardiogram (ECG) and has several substantially different etiologies, ranging from hereditary mutational causes to secondary conditions such as amyloidosis.[1, 2] This case report features a patient with familial atrial standstill diagnosed accidentally.

Case Report

Our patient was a 32-year-old man referred to the electrophysiology clinic with episodes of headache and dizziness since the previous month and an unusual rhythm in the ECG (Figure 1). With normal clinical work up for headaches, he had been treated symptomatically but felt no change. He had no experience of any cardiac symptoms such as palpitation, chest pain, dyspnea, or syncope and could easily perform such hard physical activities as climbing and playing football. His past medical history and physical examination was unremarkable. Interestingly, his mother had a cerebrovascular accident at the age of 35 years and received a permanent pacemaker (PPM) due to atrial standstill. Also, his brother underwent PPM implantation at the age of 13 following several episodes of seizure with similar diagnosis.

In echocardiography, both atria were mildly enlarged and no A wave was observed in the tricuspid and mitral inflow pulse in Doppler recording. Tissue Doppler imaging also confirmed no visible atrial contracture (Figure 2). In a 24-hour electrocardiogram Holter monitoring, the patient's minimum, maximum, and mean heart rates were 44, 45, and 59 bpm, respectively.

The patient's electrophysiological study revealed a wide complex QRS bradycardia (40 - 45 bpm) without any discernable P wave. Also, no potential for the right atrium could be recorded, and electrical stimulation failed to provoke atrial depolarization. There was a lack of atrial

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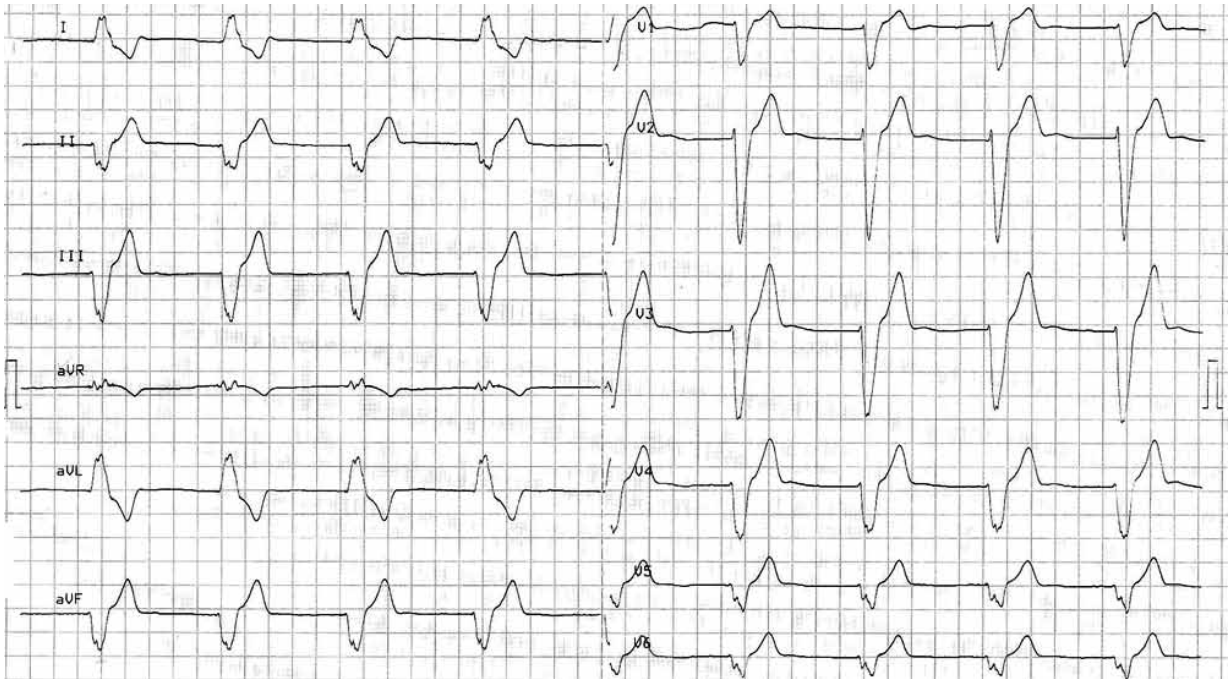

Figure 1. Twelve-lead electrocardiography of the patient. Which shows absence of P wave, bradycardia and wide QRS complexes

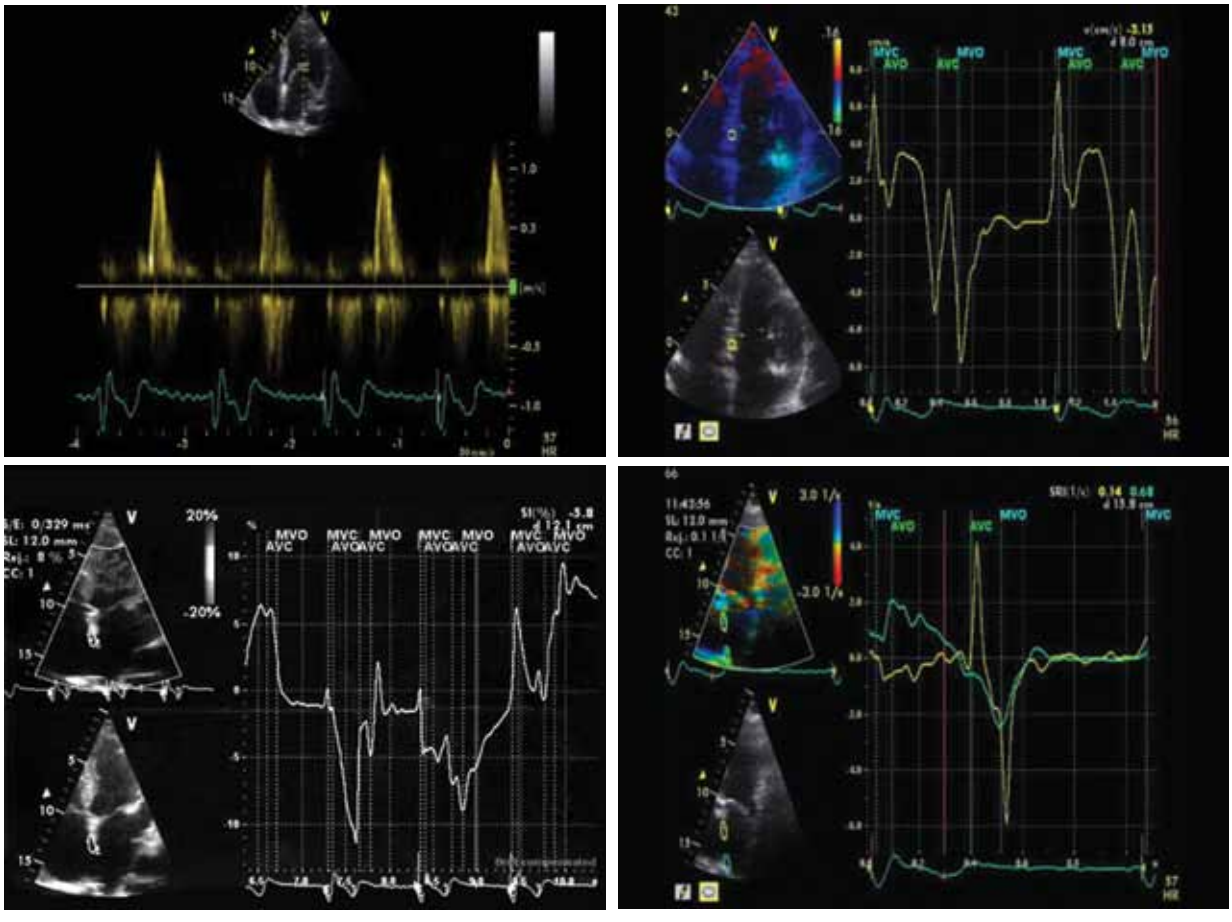

Figure 2. Conventional Doppler echocardiography of mitral valve with no visible A wave (A). Tissue Doppler imaging, base septal view (B). Strain imaging of left atrial (LA) wall (septal side), with no visible atrial contracture (C). Strain rate imaging of LA wall with no visible atrial contraction (D)

capture during high output, right atrium and coronary sinus pacing, without His potential around the His bundle. No ventricular arrhythmia could be induced during programmed ventricular stimulation. After excluding the secondary causes of atrial standstill, a single-chamber PPM in the VVIR mode was implanted with its active lead positioned in the right ventricular apex. The patient has no more symptoms after 11 months of follow-up.

Discussion

Our findings pointed toward a diagnosis of atrial standstill, which is a rare condition. Familial atrial standstill is usually diagnosed in the third to fifth decade of life, with low cardiac output symptoms, syncopal attacks, peripheral embolism, and neurological symptoms. It is generally classified into two types: transient and persistent. The former has been observed in drug intoxication (e.g. digitalis or quinidine), hyperkalemia, and acute myocardial infarction or following open cardiac surgery. The latter is uncommon and manifests with syncope or peripheral embolism. A digenic inheritance of polymorphism has been reported in patients with congenital form, linked to SCN5A (a cardiac sodium channel) and Connexin 40 (an atrial-specific gap junction Connexin), and the coexistence of both genes is necessary for atrial standstill.[1, 2]

Our patient had an idiopathic, familial, diffuse persistent atrial standstill. There was no evidence of other cardiac disease or any specific finding. However, the main etiology remained unknown as our patient did not give consent for a genetic evaluation. In such patients, implanting a PPM is thought to confer a better life style and symptom reduction.[3]

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