Int Cardiovasc Res J.2020;**14**(4):153-156.icrj.105732

International Cardiovascular Research Journal

www.ircrj.com

Atrial Standstill as a Probable Cause of Stroke in A Young Female Patient; A Case Report

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ARTICLE INFO

Article Type:
Case Report

Article History:
Received: 23 May 2020
Revised: 7 Jul 2020
Accepted: 14 Jul 2020

Keywords:
Arrhythmia
Stroke
Atrial Standstill

ABSTRACT

Introduction: Atrial standstill is a rare cardiac arrhythmia, which may present with different clinical symptoms, including longstanding dyspnea, congestive heart failure, syncope, cerebrovascular accidents, and even sudden cardiac death. According to such various presentations, diagnosis of atrial standstill may be complicated despite a high level of suspicion.

Case Presentation: The present report demonstrated a case of bilateral idiopathic atrial standstill in a young female patient. The initial presentation of the disorder was the prolonged history of dyspnea and reduced effort tolerance, which was complicated by an episode of ischemic stroke two years ago. The patient was discharged with oral anticoagulants and anti-convulsants, but she had discontinued medication arbitrarily and was diagnosed in an electrophysiological study prior to pacemaker placement because of the junctional rhythm and premature ventricular beats. She was finally diagnosed as a case of biatrial standstill, which was characterized by the absence of electrical and mechanical activity in both atrias. Single-chamber permanent pacemaker was successfully implanted and she was discharged with oral anticoagulants and was recommended for further evaluation for idiopathic or familial cardiomyopathy.

Conclusions: Atrial standstill could present in a silent manner as longstanding dyspnea and reduced effort tolerance, and remain misdiagnosed even after the development of cerebral ischemia in the absence of a high level of suspicion. These patients are eligible to receive oral anticoagulants life-long, and implanting PPM is almost indicated.

1. Introduction

Absence of atrial mechanical and electrical activity is a rare condition named atrial standstill. This serious condition may manifest through stroke, syncope, and congestive heart failure (1, 2). The true nature of atrial standstill is not completely understood, but according the possible etiologies, it can be categorized into inherited, idiopathic, and secondary forms (3, 4). The inherited forms of this condition may be diagnosed according to the positive family history in most cases, but other forms may be diagnosed incidentally alongside various manifestations (1, 2, 5). Thromboembolic events are among uncommon complications in patient with atrial standstill and may provide the possible diagnostic clue (5). However, despite the development of such complications, some patients may be misdiagnosed. The present report aims to discuss a delayed diagnosis of atrial standstill in a young female patient presented with a previous history of longstanding dyspnea, reduced effort tolerance, an episode of stroke, and arrhythmias.

2. Case Presentation

A 29-year-old female patient admitted to our hospital with dyspnea on effort and fatigability since few months ago, which had aggravated during the last few days. Her pulse rate was 48 bpm and her blood pressure was 120/80 mmHg. On physical examination, her lungs were clear with a holosystolic murmur in the left sternal border. The right-sided hemiparesis and dysphasia were remarkable. Electrocardiogram (ECG) showed a narrow QRS bradycardia without discernible P waves with the rate of 48 bpm (Figure 1). Transthoracic echocardiography revealed reduced left ventricular ejection fraction (40%), abnormal septal motion, severe bilateral enlargement, severe

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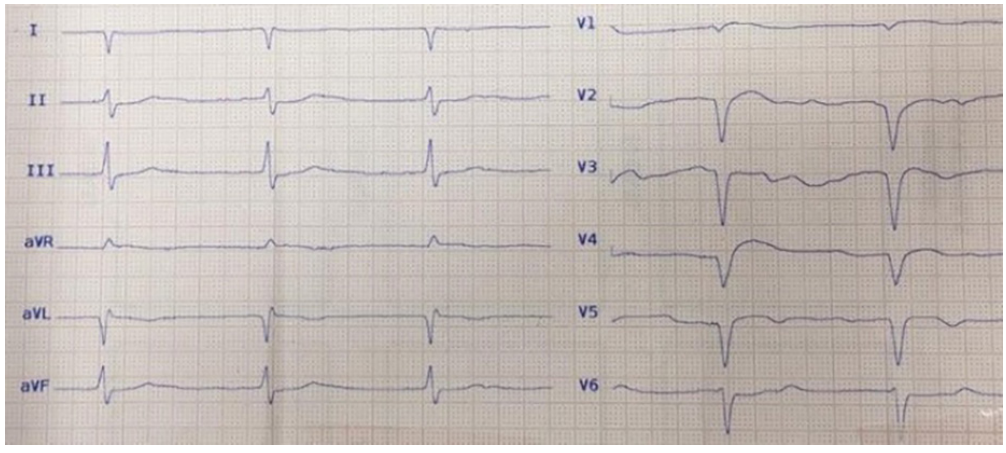

Figure 1. ECG Upon Arrival

tricuspid valve regurgitation, and mild pulmonary artery hypertension (pulmonary artery pressure estimated as 40 mmHg). She had a history of a probably thromboembolic stroke in 2018, with right sided hemiparesis and aphasia, and the same ECG as the recent presentation (bradycardia, junctional rhythm, and no P waves).

Her brain CT scan showed an ischemic cerebrovascular accident in the fronto-temporo-parietal region as well as ischemia of the basal ganglia territory. Moreover, the brain CT angiography revealed the occlusion of the internal carotid and middle cerebral arteries. Based on the hospital discharge note, no further workup was done to rule out cardiogenic emboli. The patient had been discharged with anticoagulant medication and had undergone rehabilitation. She denied further follow-up for her symptoms after the stroke event and she had discontinued her medications.

A Permanent Pacemaker (PPM) was decided to be implanted for her. Before PPM implantation, a quadri polar and a decapolar steerable diagnostic electrophysiology catheter were inserted to the heart to back up temporary pacing and evaluate the electrical activity of the heart (Figure 2). There was no atrial electrical activity in the right atrium (Figure 2-A), inter-atrial septum, and coronary sinus, and atrial capture could not be induced at maximum stimulation output during the electrophysiological study. HIS to ventricle activation time was 50 ms, which demonstrated that the atrioventricular block was above the HIS bundle (Figure 2). After failure to implant the HIS bundle pacing, a VVIR pace-maker was implanted in the right ventricle and the patient was discharged with standard heart failure treatments and Rivaroxaban 20 mg daily.

3. Discussion

The present report demonstrated that atrial standstill without previous positive family history required high levels of suspicion in order not to be misdiagnosed. This type of cardiac arrhythmias is a rare disorder, which may be manifested in different ages from pediatrics to geriatrics (1, 2). The familial forms are mostly diagnosed according to the positive family history, while other forms may be misdiagnosed (1). In the present study, the patient's initial symptom was an episode of stroke. Stroke, as the first presentation of atrial standstill, has been even reported in pediatric populations (1, 2). The primary clinical

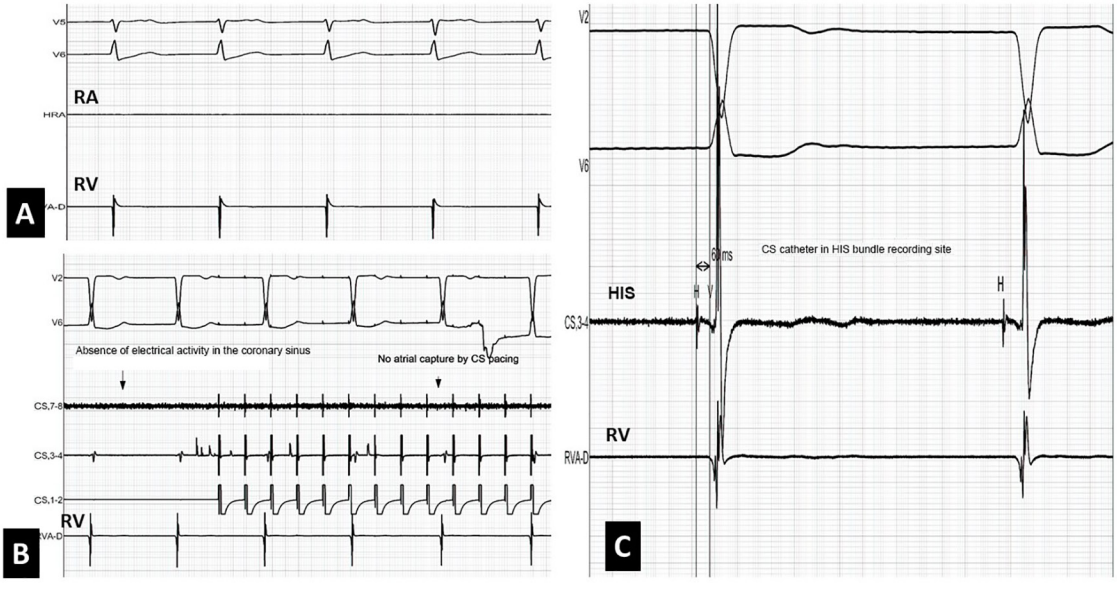

Figure 2. A-B: No Intrinsic Electrical Activity and Capture Failure of Both Atria, **C:** Normal HIS Bundle Conduction

manifestations of this disease are widely varied and a high level of suspicion is mandatory for diagnosis of most patients. These patients may present with a longstanding episodic dizziness, fatigue, or reduced effort tolerance as the primary manifestations. More acute and life-threatening manifestations may override these chronic symptoms, including stroke, heart failure, syncope, and sudden cardiac death (6). Abnormal or absence of P wave is also one of the main electrocardiographic findings in atrial standstill (6, 7). Chavez et al. proposed several diagnostic criteria for atrial standstill, which were mainly based on absent or abnormal P waves, presence of atrial asystole, supraventricular QRS morphology, and inability to stimulate the atria by temporary pacing during the electrophysiological study (2, 7). Some authors have related the term 'pseudo' atrial fibrillation to their patients presented with atrial standstill. Turkoglu et al. reported a case of atrial standstill presented with palpitation and dyspnea. Similar to the present case, their patient had no visible P wave on her ECG (8). Their patient received direct current cardioversion and cardiac asystole, necessitating cardiopulmonary resuscitation (8). According to the bilateral dilation of atriums and severe tricuspid regurgitation, an electrophysiological study was performed and the diagnosis of atrial standstill and irregular junctional ectopic rhythm was made (8). In the present case, despite discernable P waves in the ECG, the co-presentation of an ischemic stroke misled the diagnostic workup for the possible impression of atrial standstill. The first acute presentation of atrial standstill as a stroke is not quite common. The latest report of stroke was seen in a seven-year-old girl who developed cardiogenic cerebral embolism because of atrial standstill (2). According to that report, despite the lack of established treatments for atrial standstill, the authors recommended their successful management by oral anticoagulants and permanent cardiac pacemaker implantation in order to prevent further strokes as well as cardiac events (2). In another report on atrial standstill in a pediatric population, a 14-year-old boy presented with stroke. The authors stated that their patient had flutter episodes and bradycardia, as well (9). In contrast to the present case and those mentioned above, atrial standstill is accompanied by other cardiac manifestations in some patients. For instance, Lee et al. reported a 29-year-old male patient who had cerebral infarction and isolated left ventricular non-compaction without any cardiac symptoms. According to their report, the patient was the first case of a rare form of cardiomyopathies with spontaneous left-sided atrial standstill (10). Moreover, some patients with rare genetic disorders, including emery Dreifuss muscular dystrophy, may develop both atrial standstill and stroke (11).

Regardless of the different clinical presentations of atrial standstill syndrome, treatment of this rare type of arrhythmias is another challenging issue (3). Among the patients presented with stroke, long-term administration of anticoagulants should be considered in order to prevent further cardiovascular adverse events. Moreover, some authors have suggested that atrial standstill is a progressive disorder (12). For instance, Cappelli et al. reported a case of pediatric atrial standstill who received a VVI pacemaker (12). After 13 years of follow-up, their patient showed a reduced excitable atrial myocardium and need for increasing the pacing threshold (12). Such findings demonstrate the need for close follow-up of such patients in order to reduce the possible complications of incorrect pacing thresholds.

3.1. Conclusion

The present report demonstrated a case of bilateral atrial standstill syndrome, which was misdiagnosed. The findings indicated that atrial standstill could present in a silent manner as longstanding dyspnea and reduced effort tolerance, and remain misdiagnosed even after the development of cerebral ischemia in the absence of a high level of suspicion. These patients are eligible to receive oral anticoagulants lifelong, and implanting PPM is almost indicated. For the patient under the present investigation, probable idiopathic or familial cardiomyopathies had to be considered, but she and her family discontinued further evaluations.

3.1. Ethical Approval

Not applicable.

3.2. Informed Consent

The patient was called for signing a written informed consent form, but unfortunately, she had expired due to aspiration and long standing hypoxiemia resulting from a seizure attack.

Acknowledgements

There is no acknowledgment.

Authors' Contribution

SHBR developed the original idea and the protocol, abstracted and analyzed the data, wrote the manuscript, and was a guarantor. AHB contributed to the development of the protocol, abstracted the data, and prepared the manuscript.

Funding/Support

This study was not supported by any grants.

Financial Disclosure

The authors have no financial interests related to the material in the manuscript.

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