Implantable Cardioverter-Defibrillator Implantation in a Patient with Atrial Standstill

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We report a 55-year-old female patient who presented with no P waves but with a wide QRS complex escape rhythm at 44 beats/min and prolonged QTc of 0.55 seconds on ECG. The patient had recurrence of ventricular fibrillations and loss of consciousness, and underwent defibrillation and cardiopulmonary resuscitation (CPR) several times because of cardiac arrest. The transthoracic echocardiography showed dilated cardiomyopathy and enlargement of both atria. The Doppler echocardiography documented the absence of A wave in the tricuspid and mitral valve flow. An electrophysiologic study demonstrated electrical inactivity in the right and left atria. Atrial pacing with maximum output did not capture the atria. These findings together with her electrocardiographic finding indicated atrial standstill. Sudden cardiac death was her first clinical manifestation of ventricular arrhythmia. The patient remained asymptomatic after receiving a single chamber implantable cardioverter-defibrillator (ICD) with VVI pacemaker function.

Key Words: Atrial standstill, dilated cardiomyopathy, cardiopulmonary resuscitation, sudden cardiac death

INTRODUCTION

Atrial standstill is a rare condition characterized by the absence of electrical and mechanical activity in the atria. On surface ECG, atrial standstill is distinguished by bradycardia, junctional (usually narrow complex) or wide complex escape rhythm, and absence of the P wave. Atrial standstill can be persistent or transient, and diffuse or partial. Nearly 50% of patients suffer from Adams-Stokes attacks. According to most reports, atrial standstill may be secondary to other clinical disorders such as Ebstein's anomaly, muscular dystrophy, diabetes mellitus, amyloidosis, or myocarditis, and it has also been reported in rare cases of digitalis or quinidine toxicity, and ischemic heart disease. Only a few cases of familial clustering of primary atrial standstill have been reported. Some cases are associated with a cardiac sodium channel gene SCN5A mutation and connexin 40, and a mutation or deletion of the ryanodine receptor 2 gene (RYR2) has been found in patients with catecholaminergic polymorphic ventricular tachycardia (CPVT). Herein, we present a patient with atrial standstill, dilated cardiomyopathy, and sudden cardiac arrest.

CASE REPORT

A 55-year-old female patient was referred to our hospital for evaluation of her bradycardia and dyspnea on exertion (NYHA class II-III). Physical examination revealed chronically ill appearance, grade 2/6 systolic murmur at the mid right and left sternal borders, decreased breathing sounds with both basal rales, 2 finger-breath tender hepatomegaly, and weakness of right extremities. Laboratory findings revealed elevated BNP level (471 pg/mL), however, other parameters including CBC, electrolytes, cardiac enzymes, liver function test, etc. were within normal limits.

Twelve-lead electrocardiogram (ECG) showed an escape distal rhythm at 44 beats/min with a QRS duration of 0.12 seconds, but no atrial activity. T wave inversion in V1 to V6 was prominent, and
corrected QT interval (QTc) was prolonged to 0.55 seconds (Fig. 1A). Chest X-ray revealed marked cardiomegaly with right and left chamber enlargement. Echocardiography showed massively dilated right and left atria along with mild tricuspid and mitral regurgitation. Right and left ventricles were also enlarged, and left ventricular ejection fraction (LVEF) was 34%, which was compatible with the findings of dilated cardiomyopathy. A Doppler study did not demonstrate any atrial activity (A wave) in the mitral and tricuspid inflow regions.

Three hours after admission, she developed sudden onset ventricular fibrillation (VF) and loss of consciousness, and needed immediate defibrillation. There were several episodes of recurrent VF and cardiac arrest after defibrillation, and the patient needed immediate cardiopulmonary resuscitation. After she was stabilized from these events, we decided to implant ICD with pacemaker function. The patient underwent an electrophysiologic (EP) study which revealed total right and left atrial standstill (no recordable atrial electrogram plus lack of atrial capture during high output, right atrium and coronary sinus pacing), and HV interval of 56 ms (Fig. 1B). During programmed

Fig. 1. Twelve-lead ECG and intracardiac electrogram. (A) Twelve-lead ECG on the 1st day of admission showed an escape distal rhythm at 44 beats/min with wide QRS complex, of 0.12 sec. No P wave was found in ECG. QTc interval was prolonged to 0.55 sec. (B) Recording obtained during EPS after implantation of ICD with VVI pacemaker function. There was no electrical activity on atrial and CS electrograms. ECG, electrocardiogram; ICD, implantable cardioverter-defibrillator; HRA, high right atrium, CS, coronary sinus.

Fig. 2. (A) Chest PA of a single chamber ICD with VVI pacemaker function implanted to the patient. (B) DFT test showed induction of VF, and safe and successful defibrillation at 14.8 J. PA, postero-anterior; ICD, implantable cardioverter-defibrillator; DFT, defibrillation threshold test; VF, ventricular fibrillation.
ventricular stimulation, ventricular fibrillation was easily induced. Therefore, we implanted a single chamber ICD with VVI pacemaker function (Fig. 2). After the procedure, her surface ECG showed regular pacemaker rhythm, and her QTc interval progressively shortened to 0.44 seconds. Her cardiac performance improved progressively, and the BNP level returned to normal (7 pg/mL). She received conventional managements for heart failure and anticoagulation. The biopsy report of chest wall fat pad showed no evidence of amyloidosis. The gene study for SCN5A and connexin 40 revealed negative result.

DISCUSSION

Atrial standstill is a rare arrhythmogenic condition, and its diagnosis relies on ECG demonstration of bradycardia, the absence of P waves, and junctional narrow or wide complex escape rhythm. Intracavitary recordings also fail to demonstrate atrial depolarization. Typically, the atria are scarred and have little or no working myocardia. Few autopsy reports have described the pathologic changes in atrial standstill, showing fibroelastosis and fatty infiltration in atria. The high lateral RA is usually involved first in a progression toward the lower RA site. Finally, atrial electrograms can be recorded only in the vicinity of the tricuspid valve or interatrial septum. This phenomenon was observed in most cases, despite of different underlying diseases. Nakazato et al. observed the absence of P wave in 6 of 11 patients with atrial standstill, and observed the loss of P waves in the remaining 5 patients as the underlying disease progressed. Atrial standstill may be secondary to several clinical disorders, however, she had no evidence of muscular disease, ischemic heart disease, drug intoxication, and myocarditis. In the primary persistent form, atrial paralysis is paralleled by atrial dilatation, and mitral valve incompetence. Clinically, atrial standstill causes weakness and lethargy, and about 50% of patients suffer from syncope. Atropine does not increase the heart rate. Symptomatic cases require ventricular pacing and treatment for congestive heart failure if present, however, the prognosis is poor. Fazelifar et al. reported a family with atrial standstill associated with syncope, dilated cardiomyopathy, and sudden cardiac death, and agreed with the postulation that the atrial standstill is a disease primarily associated with SCN5A mutation and rare connexin 40 genotype. Bhuiyan et al. presented two families with typical CPVT in conjunction with additional features of progressive AV block, sinus node dysfunction, atrial fibrillation, atrial standstill, and dilated cardiomyopathy those related with the genetic defect in the N-terminus of RYR2. Although we did not investigate possible genetic defect such as deletion of ryanodine receptor 2 gene, cardiac evaluation through available hospital records of the patient’s relatives did not reveal any cardiac abnormalities, and the clinical features were different from those of CPVT. Dilated cardiomyopathy can cause fatal ventricular arrhythmias via any kind of arrhythmogenic mechanism.

Our case had atrial standstill which was persistent and advanced, and dilated cardiomyopathy accompanying bradycardia and long QT interval as well as the electrical and mechanical activity of both atria were not detected by intracardiac electrogram, Doppler recordings, and fluoroscopy. She developed recurrent ventricular fibrillations and cardiac arrest, therefore, we performed a single chamber ICD implantation with VVI pacemaker function to prevent sudden cardiac death. With the procedure and anticoagulation, she remained free of ventricular arrhythmias and syncope.

REFERENCES

1. Fazelifar AF, Arya A, Haghjoo M, Sadr-Ameli MA. Familial atrial standstill in association with dilated cardiomyopathy. Pacing Clin Electrophysiol 2005;28:1005-8.
2. Bensaid J. Persistent atrial standstill documented over a 22-year period. Am Heart J 1996;131:404-7.
3. Marini M, Arbustini E, Disertori M. Atrial standstill: a paralysis of cardiological relevance. Ital Heart J Suppl 2004;5:681-6.
4. Bloomfield DA, Sinclair-Smith BC. Persistent atrial standstill. Am J Med 1965;39:335-40.
5. Rosenbaum FF, Levine SA. Auricular standstill. Its occurrence and significance. Am J Med Sci 1939;198:774.
6. Koshimizu TA, Komori S, Ishihara T, Kohno I, Umetani...
K, Sawanobori T, et al. Restored atrial excitability after late recanalization in a patient with atrial standstill and acute myocardial infarction. Pacing Clin Electrophysiol 2002;25:217-9.

7. Groenewegen WA, Firouzi M, Bezzina CR, Vliex S, van Langen IM, Sandkuijl L, et al. A cardiac sodium channel mutation cosegregates with a rare connexin 40 genotype in familial atrial standstill. Circ Res 2003;92:14-22.

8. Shaw TR, Northridge DB, Francis CM. Left atrial standstill in a patient with mitral stenosis and sinus rhythm: a risk of thrombus hidden by left and right atrial electrical dissociation. Heart 2003;89:1173.

9. Bhuiyan ZA, van den Berg MP, van Tintelen JP, Bink-Boelkens MT, Wiesfeld AC, Alders M, et al. Expanding spectrum of human RYR2-related disease: new electrocardiographic, structural, and genetic features. Circulation 2007;116:1569-76.

10. Rosen KM, Rahimtoola SH, Gunnar RM, Lev M. Transient and persistent atrial standstill with His bundle lesions. Electrophysiologic and pathologic correlations. Circulation 1971;44:220-36.

11. Nakazato Y, Nakata Y, Hisaoka T, Sumiyoshi M, Ogura S, Yamaguchi H. Clinical and electrophysiological characteristics of atrial standstill. Pacing Clin Electrophysiol 1995;18:1244-54.

12. Williams DO, Jones EL, Nagle RE, Smith BS. Familial atrial cardiomyopathy with heart block. Q J Med 1972;41:491-508.

13. Amat-y-Leon F, Racki AJ, Denes P, Ten Eick RE, Singer DH, Bharati S, et al. Familial atrial dysrhythmia with A-V block: Intracellular microelectrode, clinical electrophysiologic, and morphologic observations. Circulation 1974;50:1097-104.

14. Jeong MH, Ahn YK, Gill GC, Park JH, Cho JG, Park JC, et al. Tsutsugamushi myocarditis with congestive heart failure and persistent atrial standstill. Jpn Circ J 1996;60:382-8.

15. Talwar KK, Radhakrishnan S, Chopra P. Myocarditis manifesting as persistent atrial standstill. Int J Cardiol 1988;20:283-6.