Case Reports

A Histopathologic Study of the Conduction System in an Elderly Patient with Partial Atrial Standstill

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SUMMARY

Electrophysiologic and histopathologic studies were performed on an 83-year-old man with partial atrial standstill. In the electrophysiologic study, no atrial electrical activity was recorded in the right atrium, but f waves were seen in the coronary sinus electrogram. Although the high right atrium and the atrial septum could not be activated by electrical stimuli of up to 10 V, the low atrium could be stimulated. The pathologic examination showed that there was extensive fatty infiltration into the sinus node and fibrosis in the atrial muscle. In the low right atrium, the musculature was partially preserved.

Additional Indexing Words:

Partial atrial standstill Electrophysiologic study Histopathologic study Atrial fibrillation

ATRIAL standstill is rare,^{1),2)} and few histopathologic studies of the conduction system have been undertaken in autopsy cases with this condition. In this case report, we describe electrophysiologic and histopathologic studies in an 83-year-old man with partial atrial standstill.

CASE REPORT

This man had been well until he was admitted to another hospital because of a stroke when he was 71 years old. Hypertension and arrhythmia were noted on that occasion. He visited our hospital for the first time at the age of 74. His physical examination revealed a blood pressure of 182/86 mmHg and an irregular pulse of 66 beats/min. His chest X-ray film showed moderate cardiomegaly, and the cardiothoracic ratio was 60%. The elec-

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Fig. 1. A: Electrocardiogram recorded at the age of 74, demonstrating atrial fibrillation with approximately 90 beats/min, but no apparent f wave. The R-R intervals are less than 1,160 msec, and the mean electrical axis is -30° . B: Electrogram at the age of 82, demonstrating no atrial activity. A regular and narrow QRS complex of 34 beats/min suggesting complete atrioventricular block with junctional escape rhythm.

trocardiogram demonstrated atrial fibrillation and a rate-dependent right bundle branch block (Fig. 1-A). However, f waves were not clearly identified in any lead. Subsequently, he suffered from heart failure and was admitted to our hospital several times. At the age of 80, he was readmitted because of recurrent heart failure, and bradycardia (approximately 31 beats/min) was first noted at night. Fig. 2-A shows the esophageal electrogram at this time; f waves could be recognized, and f-f intervals were approximately 220 msec. As bradycardia became persistent throughout the day (Fig. 1-B) and heart failure worsened, an electrophysiologic study was performed, and a permanent pacemaker was implanted when he was 82 years old. However, the patient died of sepsis complicating disseminated intravascular coagulation at the age of 83.

Electrophysiologic study:

The enlarged surface electrocardiogram recorded at the time of the electrophysiologic study revealed f waves, and f-f intervals were approximately

Fig. 2. A: Precordial and esophageal electrocardiograms show f waves (arrow) on the esophageal lead and f-f intervals are approximately 220 msec. B: Enlarged surface electrocardiograms show f waves (arrow, left), and then f waves disappear in the same lead (right). C: Simultaneous recordings of leads II , aV_F , V_1 , high right atrium and coronary sinus electrograms show almost regular f waves on the coronary sinus electrogram, but no atrial electrical activity on the high right atrial electrogram. Abbreviations: CS=coronary sinus electrogram; E=esophageal electrocardiogram; HRA=high right atrial electrogram.

180 msec. Subsequently, f waves could not be found in the same leads. Thus, f waves were thought to appear intermittently (Fig. 2-A). Almost regular f waves (f-f intervals 280 msec) were also recorded in the coronary sinus, but not in the high right atrium (Fig. 2-C). These findings suggested that these f waves could have originated from the left atrium. The His bundle electrogram could not be obtained in this study. No atrial electrical activity was recorded in the surface electrocardiogram or intra-atrial electrograms

Fig. 3. Surface electrocardiograms, intracavitary electrograms and intra-atrial pressure tracings when paced at the high right atrium (A), atrial septum (B), low right atrium (C) and right ventricle (D). Arrows and "a" indicate f wave and atrial contraction wave, respectively. Abbreviations: E=esophageal electrocardiogram; HRA=high right atrial electrogram; LRA=low right atrial electrogram; P_{RA}=right atrial pressure tracing; RV=right ventricle; SEPTUM=atrial septum.

when the high right atrium was paced by electrical stimuli of up to 10 V. Furthermore, no atrial contraction wave (a wave) was seen in the right atrial pressure tracing (Fig. 3-A, B). However, the low right atrium could be activated by electrical stimuli of 10 V, and a waves were seen simultaneously in the right atrial pressure tracing (Fig. 3-C). During right ventricular pacing, retrograde A waves were recorded in the low right atrial electrogram, and V-A intervals were 560 msec, but retrograde A waves were not seen in the high right atrial electrogram. When the electrode at the high right atrium was placed at the low right atrium, retrograde A waves could be recorded. In the right atrial pressure tracing, no a wave could be recorded. It was suggested that this phenomenon might indicate electro-mechanical uncoupling (Fig. 3-D).

Pathologic findings:

The heart weight was 500 g, and both atria were markedly dilated. Although each of the three coronary arteries had severe stenosis (greater than 75%), no myocardial infarction was identified. Histopathologic examination of the conduction system was performed by serial sections according to the method of Lev et al.^{3),4)}

There were marked fatty infiltrations into the sinus node, and only a few conduction cells of the sinus node were seen (Fig. 4). There was marked and diffuse fibrosis in the high right atrium near the sinus node and in the left

Fig. 4. Photomicrographs (Elastica-van Gieson stain) showing the sinus node (A; arrows, $\times 10$), the high right atrium near the sinus node (B; $\times 10$), the low right atrium (C; $\times 25$) and the second portion of the right bundle branch (D; arrows, $\times 50$), respectively. See further explanations in the text. Abbreviations: At=atrium; RB=right bundle branch; SN=sinus node.

atrium, but the severity of the latter was milder than the former. There was also severe fibrosis in the low right atrium, but the musculature was partially preserved in this area. The atrioventricular node and the bundle of His were almost intact. Although the anterior fascicles of the left bundle branch showed moderate fibrosis, the posterior fascicles of the left bundle branch revealed severe fibrosis and disruption. In the right bundle branch, there was mild to moderate fibrosis in the first and third portions and severe fibrosis in the second portion.

DISCUSSION

Atrial standstill is a rare condition, and the diagnostic criteria^{1),2)} include 1) absence of p waves; 2) absence of A waves in the jugular pulse and atrial pressure tracing; 3) a supraventricular-type QRS complex; 4) immobility of the atria on fluoroscopy; and 5) inability of the atria to be electrically stimulated. Atrial standstill has been subdivided into temporary and persistent forms¹⁾ and may also be classified into total and partial forms.⁵⁾ Partial atrial standstill is defined as the absence of electrical activity in a discrete area of the atrium.⁵⁾

According to the electrophysiologic study, this case was thought to present partial atrial standstill, which intermittently revealed atrial flutter-fibrillation in the left atrium. However, neither electrical activity nor mechanical contraction in the right atrium were present, except in the low right atrium. Since this patient had shown rate-dependent right bundle branch block, complicating pathologic changes in the atrioventricular conduction system were suggested.

There have been a few case reports of atrial standstill. Zipes et al⁶⁾ reported one that was similar to ours. In their patient, no right atrial activity could be recorded, but the esophageal electrogram showed irregular left atrial electrical activity. Atrial pacing was accomplished only when the catheter tip was positioned in the area of the low atrial septum. Effendy et al⁷⁾ described a patient with an alternation of partial and total atrial standstill. In their case, standstill was present only in the high right atrium.

Histopathologically, both marked fatty infiltrations into the sinus node and diffuse fibrosis in the high right atrial muscle may have been responsible for partial atrial standstill in our case. Severe fibrosis of the second portion of the right bundle branch also may have contributed to the rate-dependent right bundle branch block. Rosen et al⁸⁾ performed an electrophysiologic study in a 73-year-old woman with persistent atrial standstill, and postmortem examinations with serial sections of the conduction system were obtained. Ac-

According to their observations, the musculature showed marked fibrosis, fibroelastosis, fatty infiltration and marked vacuolization throughout both the atria and atrial septum. Tanaka et al⁹⁾ reported a 69-year-old man with persistent atrial standstill complicated with idiopathic cardiomyopathy. The postmortem examination disclosed marked fibrosis of the right atrium and fibrous strands in the right ventricle. Patton et al¹⁰⁾ also described diffuse fibrosis of the atrium in a 54-year-old man with an electrically silent right atrium. We believe that atrial standstill may have been histopathologically characterized by marked fibrosis of the atrial musculature, and fatty infiltrations into the sinus node may have been specific in our case.

Atrial standstill has usually appeared to be associated with organic heart disease,⁵⁾ but, in some cases, the underlying disease is unknown.⁸⁾ In our patient, amyloid deposits in the heart were clinically thought to be an underlying disorder because of recurring attacks of heart failure and increasing low voltage on the surface electrocardiogram. Allensworth et al¹¹⁾ described three siblings with persistent atrial standstill accompanied by familial amyloidosis. A biopsy specimen of the right atrium in a 36-year-old man demonstrated hypertrophy of the atrial cells, and interstitial and perivascular deposits of amyloid. But postmortem examination of our patient revealed no amyloid deposits in either the atrial or ventricular musculature. Thus, the underlying disease of our patient is still unknown, except for atherosclerotic heart disease.

Impairment of the sinus node and atria in our case was more severe than those observed in 22 elderly patients with sick sinus syndrome previously reported.¹²⁾ Although the surface electrocardiogram (Fig. 1-B) suggested a junctional escape rhythm with a complete atrioventricular block, histopathologic study revealed that the atrioventricular node and the bundle of His were almost intact and that there was no trifascicular lesion. Lévy et al⁵⁾ reported that the atrioventricular junctional escape rhythm constituted a major electrocardiographic feature and might be associated with both total and partial atrial standstill. This suggested that complete atrioventricular block is not induced by an anatomical disturbance of the atrioventricular conduction system. We believed that electrical activity in the atrium was so weak that it could not activate the atrioventricular node, or that electrical activity could not pass through the atrioventricular node because of the severe paranodal fibrosis in the right atrium. Zipes et al¹³⁾ demonstrated summation in antegrade atrioventricular nodal conduction in an experimental model, and Ohkawa et al¹⁴⁾ suggested this possibility in a patient who showed complete atrioventricular block with a junctional escape rhythm and an intact atrioventricular conduction system.

According to a histopathologic study¹⁵⁾ that correlated the lesion of the right bundle branch block with the configuration of the QRS complex in 11 autopsy cases of complete right bundle branch block, lesions in the second or third portion frequently showed either an rsR, an rSR or an RSR configuration of the QRS complex in the V₁ and V₂ leads. Our case revealed the rsR pattern in the V₁ lead, and it corresponded to the lesion of the right bundle branch.

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