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【 CASE REPORT 】

Paroxysmal Atrial Fibrillation in Patients Successfully Treated by Radiofrequency Catheter Ablation with Severely Compression, Lateral Displacement, and Clockwise Rotation of Their Hearts due to Severe Pectus Excavatum

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Abstract:

Two cases with severe pectus excavatum and symptomatic atrial fibrillation (AF) underwent radiofrequency catheter ablation (RFCA). Their chest X-ray and computed tomography (CT) findings revealed lateral displacement and clockwise rotation of their hearts, and severe right atrial and mild right ventricular compression against the sternum, but no left atrium compression against the spinal column. The procedure was therefore carefully performed under guidance with CT, intra-cardiac echography, atriography, and a three-dimensions mapping system. Finally, the AF was successfully treated by RFCA without any complications. These findings underscore the importance of understanding cases of abnormal anatomy and carefully designing a strategy before performing any procedure.

Key words: atrial fibrillation, radiofrequency catheter ablation, pectus excavatum

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Introduction

Pulmonary vein antrum isolation (PVAI) has proven to be a useful strategy for radiofrequency catheter ablation (RFCA) of atrial fibrillation (AF) worldwide (1). We and other physicians have previously demonstrated that RFCA of AF utilizing a three-dimensional (3D) mapping system is feasible and safe for achieving a favorable outcome even in patients with an abnormal cardiac anatomy (2) and/or location (3).

We herein report two cases of paroxysmal AF successfully treated by RFCA in patients with severe compression, lateral displacement, and clockwise rotation of their hearts due to severe pectus excavatum.

Case Report

A 69-year-old woman and 54-year-old man were admitted to our hospital with a chief complaint of palpitations due to drug-refractory AF to undergo RFCA. Their $CHADS_2$ scores were both 0. They had a hollow in their thorax that was compressing their heart due to severe pectus excavatum (PE) (Fig. 1A and B). The pectus severity index (PSI) (4) was 6.13 in the woman and 4.90 in the man, indicating severe PE (PSI >3.25). Their other physical examinations and laboratory analyses yielded no abnormalities, including no abnormal heart murmurs. 12-lead electrocardiograms showed a normal sinus rhythm and AF in both patients without any tachycardia and no medications (Fig. 1C and D). Their echocardiograms yielded no evidence of clinically overt

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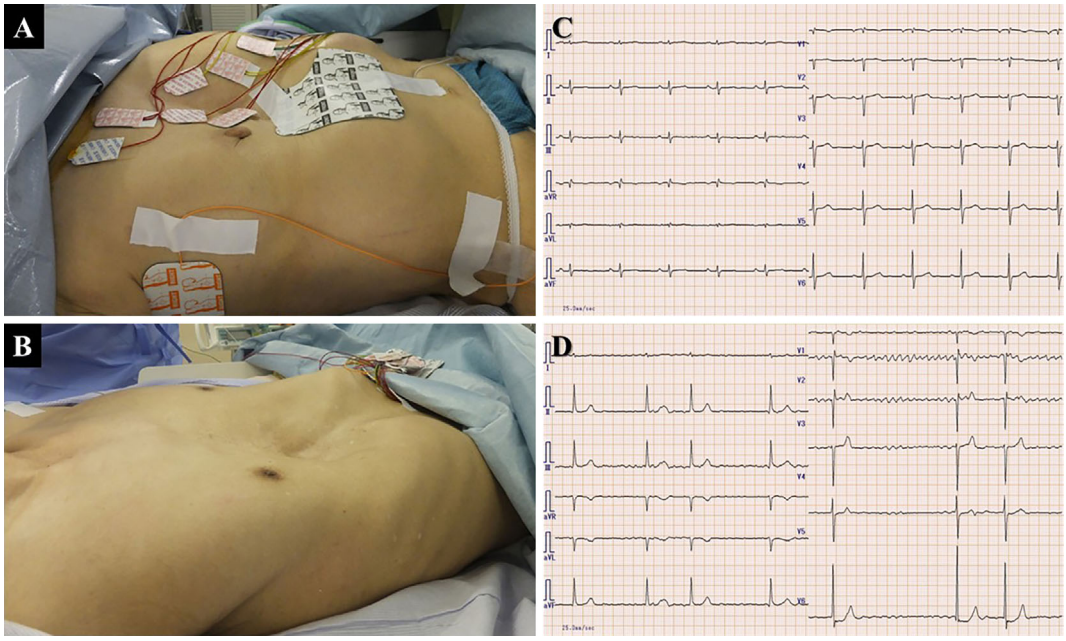

Figure 1. **The photos of the chest (A, B) and 12-lead electrocardiograms (C, D) of a 69-year-old woman and 54-year-old man.**

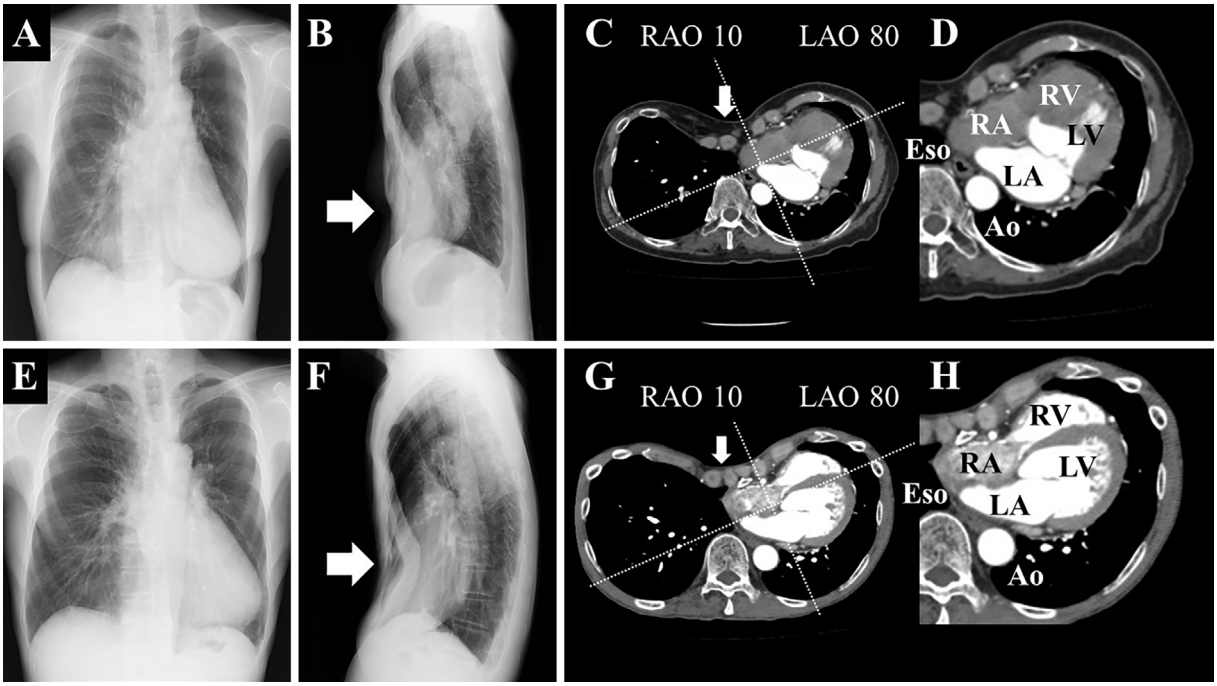

Figure 2. **Chest X-rays in the frontal (A, E) and lateral (B, F) views, and computed tomography images (C, D, and G, H) of the 69-year-old woman and 54-year-old man. The white arrows in panels B, C, F, and G indicate pectus excavatum. SVC: superior vena cava, IVC: inferior vena cava, Ao: aorta, RA: right atrium, LA: left atrium, RV: right ventricle, Eso: esophagus**

structural and/or organic heart disease, including mitral valve prolapse, tricuspid regurgitation, or pulmonary hypertension, and they had normal ejection fractions and left atrial (LA) dimensions of 38 mm in the woman and 45 mm in the man.

Chest X-rays revealed an enlarged cardio-thoracic ratio (Fig. 2A, B, E and F), and computed tomography (CT) (Fig. 2C, D, G and H) revealed lateral displacement and clockwise rotation of their hearts along with severely right atrial and mild right ventricular compression against the sternum; however, no LA compression was noted against the aorta or spinal column, probably due to the left-sided displacement of their hearts. CT also revealed that the area of the atrial septum was small, and the septum was perpendicu-

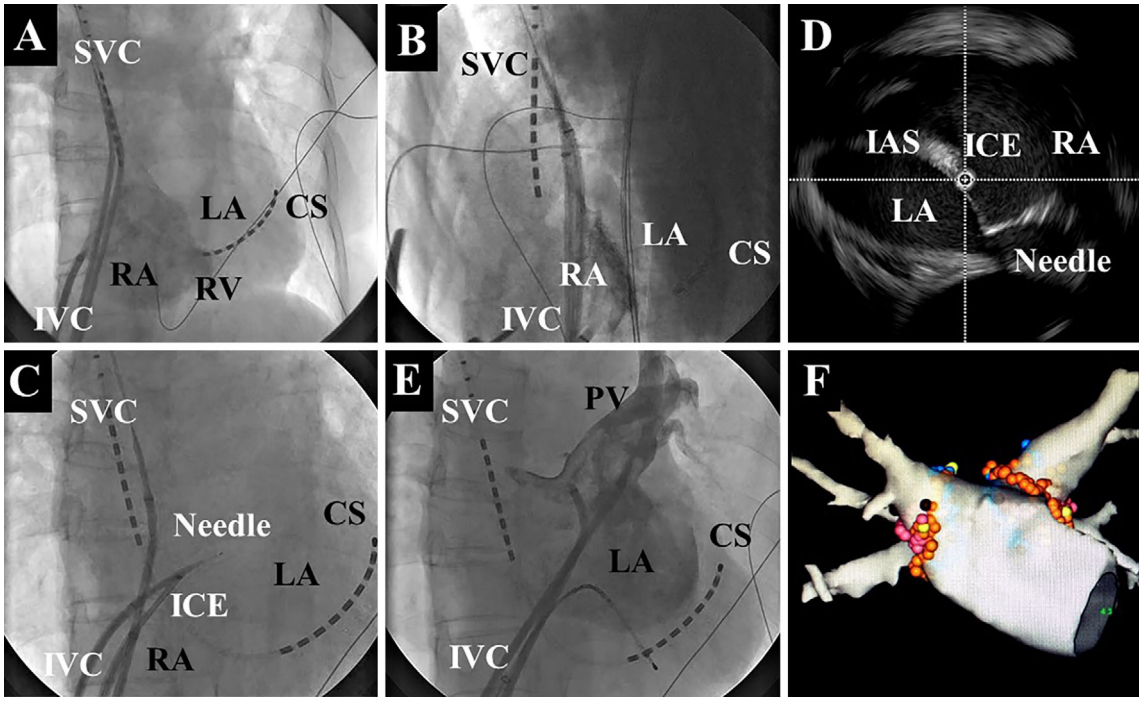

Figure 3. **Right atriography in the right anterior oblique (RAO) (A) and left anterior oblique (LAO) (B) views, intracardiac echography images (C), double transseptal puncture under guidance of an intracardiac echogram (D), left atriography in the RAO view (E), and an EnSite image in the RAO view (F) of the 69-year-old woman. The circles are the points of the pulmonary vein antrum ablation. SVC: superior vena cava, IVC: inferior vena cava, RA: right atrium, LA: left atrium, RV: right ventricle, CS: coronary sinus, IAS: interatrial septum, ICE: intra-cardiac echogram, PV: pulmonary vein**

lar in the 10° right anterior oblique (RAO) view and parallel in the 80° left anterior oblique (LAO) view (Fig. 2C and G). As such, right atriography was performed in those views before transseptal puncture (Fig. 3A and B).

A temperature probe (SensiTherm™; St. Jude Medical, St. Paul, USA) for monitoring the esophageal temperature was inserted and placed between the levels of the left superior and inferior PVs. Double transseptal puncture (Fig. 3C) was then carefully performed under guidance with intracardiac echography (Fig. 3D) (Ultra ICE catheter; EP Technologies, Boston Scientific Corporation, San Jose, USA). After left atriography was performed (Fig. 3E), an open irrigated 3.5-mm-tip ablation catheter (FlexAbility™; St. Jude Medical) and circular mapping catheter (Optima™; St. Jude Medical) were positioned in the LA. The LA was then reconstructed by a 3D mapping system (EnSite NavX/Velocity ™ Cardiac Mapping System; St. Jude Medical), and EnSite voltage maps revealed that the PVs had PV potentials, but there were no low voltage areas in LA in either patient. Circumferential PVAI was therefore performed under electroanatomic guidance with the 3D mapping system until the achievement of a bidirectional conduction block between the LA and PVs under the administration of isoproterenol.

Because the run of the phrenic nerves was unknown, pacing maneuvers to capture them were performed before each ablation. Furthermore, the procedures were carefully performed while confirming the diaphragmatic movement on fluoroscopy. Firing from both the left and right superior PVs was confirmed after PVAI. However, the firing from the non-PV foci was not induced under the intravenous administration of isoproterenol. Finally, programmed stimulation failed to induce any arrhythmias, including AF.

Both patients have remained well without any recurrence of AF and no medications for four years in the woman and two years in the man after the RFCA.

Discussion

AF is the most common clinical arrhythmia and is associated with significant clinical morbidity and increased mortality (4). PE is a well-known congenital skeletal abnormality occurring in 1:800 live births (4, 5) that may have a cardiac manifestation, resulting in right atrial, right ventricular, and right ventricular outflow tract compression against the sternum, LA compression against the spinal column, and lateral displacement of the heart to the left, with resultant cardiac distortion and an increased incidence of mitral valve prolapse (4, 5). It has been recently reported that a high incidence of non-valvular AF is associated with PE (4). Furthermore, the incidence of AF associated with severe PE is five-fold higher than in patients without PE (4). This association suggests a potential genetic or mechanical abnormality may be common to the two disorders.

The potential mechanisms underlying the occurrence of

AF associated with PE have been previously described (4, 5) and include the following: 1) mechanical stimulation of the LA and/or activation of vagal reflexes during swallowing, 2) mitral valve prolapse resulting from heart compression, 3) sympathetic nerve activation resulting from a low cardiac output due to heart compression, and 4) pulmonary hypertension associated with the PE. However, the AF in the two present patients was not induced by swallowing, no mitral valve prolapse or pulmonary hypertension were observed on echocardiography, and no abnormal heart murmurs or tachycardias resulting from sympathetic nerve activation were observed. Furthermore, there were no low-voltage areas in the LA, indicating no or little damage to the LA myocardium resulting from mechanical stress induced by PE in the present cases. We therefore suspected a probable mechanism of ectopic beats originating from the PVs and targeted those sites for RFCA. If the cause of the AF in those patients had been compression of the heart due to PE, they might have needed to consider surgery for the PE to prevent/treat the AF.

In addition, the present study underscored several points, as previously mentioned (3). First, the relationship between the position and torsion of each chamber of the heart was confirmed using preoperative CT. Second, in order to confirm the atrial septum, right atriography was performed before transseptal puncture with the atrial septum perpendicular in the 10° RAO view and parallel in the 80° LAO view (Fig. 3A-D). Third, because the run of the phrenic nerve was unknown, the procedure was performed while confirming the diaphragmatic movement using fluoroscopy during the RF energy delivery. Fourth, in order to confirm whether or not substernal pericardiocentesis could be performed, we preprocedurally confirmed the possible route by transthoracic echocardiography. Finally, AF was successfully treated by RFCA utilizing a 3D mapping system without any complications and obtained a good clinical course.

These points underscore the importance of understanding the anatomy in patients with an abnormal cardiac anatomy, considering the potential mechanisms of AF, and carefully planning a strategy before the procedure in patients with an abnormal cardiac anatomy and/or PE in an abnormal location. Furthermore, physicians should be aware that RFCA of AF can be steadily performed using newly developed modalities, including 3D mapping incorporating CT images and intracardiac echography-guided transseptal puncture, even in patients with an abnormal anatomy, such as those with PE, dextrocardia, and scoliosis. Finally, to our knowledge, this is the first report concerning PVI in patients suffering from PE.

The authors state that they have no Conflict of Interest (COI).

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