Management of scar-related atrial flutter in a patient with dextrocardia, inferior vena cava interruption, and azygos continuation

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Introduction

Dextrocardia is detected in approximately 1 in 12,000 live births, and one-third of these have complete situs inversus (1). Dextrocardia has been reported to be associated with inferior vena cava (IVC) stenosis or interruption in 8%–18% of cases, and the anomalies of IVC can coexist with azygos continuation in approximately 0.6% of cases (2). We report a case of catheter ablation of scar-related atrial flutter (AFL) in a patient with dextrocardia and complex venous anomaly.

Case Report

The patient was a 44-year-old male with dextrocardia, situs inversus, IVC interruption, and azygos continuation and an 8-year history of highly symptomatic chronic AFL. In 1975, when he was 3 year of age, he had undergone a surgical correction for two ostium secundum atrial septal defects (ASD). A schematic diagram of the anatomy of the heart is shown in Figure 1a. He underwent several electrical cardioversion because of symptomatic AFL episodes after 1999. He had EHRA Class III when he was referred to our clinic. Baseline 12-lead electrocardiography (ECG) showed a macroreentrant atrial tachycardia and dextrocardia (Fig. 1b). After local anesthesia, three long sheaths were placed at the SVC–RA junction via femoral veins to stabilize the catheters and control them (Fig. 1c). Then, a decapolar coronary sinus catheter and a duodecapolar halo catheter were placed in the coronary sinus and RA, respectively (Fig. 1d). An activation and voltage map of RA were obtained using Carto-3 system with an irrigated RF ablation catheter. Pacing entrainment was performed at the hepatic vein-tricuspid valve, which revealed a PPI−TCL of >50 ms excluding a peri-tricuspid typical AFL. Two scar areas were detected on the interatrial septum (Fig. 2a). The pacing entrainment between the two scars demonstrated a short PPI−TCL value (254–242=12 ms, Fig. 2b), and the pacing site was demonstrated in Figure 2c with a “white dot.” Activation mapping suggested that the tachycardia spread between the two scars. When a linear ablation was created between the two scars (Fig. 2c), tachycardia stopped (Fig. 2d). No tachycardia occurred with rapid or programmed extrastimulus pacing with isoproterenol infusion. The patient was discharged the following day, and his clinical status improved to EHRA Class I. He had no recurrence of arrhythmias at the 1-year follow-up.

Discussion

Radiofrequency (RF) catheter ablation of supraventricular tachycardia (SVT) has rarely been reported in patients with...
dextrocardia, and only few cases, who have typical AVNRT or accessory pathway, have previously been described in patients with dextrocardia, IVC interruption, and azygos continuation (3, 4). To the best of our knowledge, the present case may be the first case of RF ablation of scar-related AFL due to surgical repair of ASDs in a patient with dextrocardia and complex venous anomaly.

Dextrocardia or complex cardiac anatomy may be very challenging to electrophysiologists during catheter ablation procedures. An interrupted IVC with azygos continuation to SVC may complicate the femoral venous approach typically used for diagnostic or interventional cardiac catheterization because of the abrupt 180° turn at the level of the superior azygos arch, and ablation of left atrial arrhythmias in such cases is more difficult. Therefore, we used three long sheaths to stabilize the catheters and control them. Femoral venous approach is not feasible in left atrial arrhythmias, which requires atrial septal puncture in an interrupted IVC, which will eventually require a superior approach.

Atrial tachycardias are common after repair of many types of complex congenital heart disease (5). The most common late-onset atrial arrhythmias in these patients are cavotricuspid isthmus-dependent AFL, incisional atrial reentrant tachycardia, and atrial fibrillation and less commonly focal atrial tachycardia (6). Arrhythmia mechanisms are related to surgical incisions, atrial enlargement, and structural remodeling with slow conduction creating the substrate for macroreentry (7). The efficacy of antiarrhythmic drugs in this type of arrhythmias has been unsatisfactory, and these tachycardias are difficult to medically manage and frequently recur after electrical cardioversion. In patients with surgically corrected ASD, electroanatomic mapping-guided RF ablation of late-onset macroreentrant atrial arrhythmias demonstrated a high success rate in a very long-term follow-up (8).

**Conclusion**

This case demonstrated a complex venous anomaly with dextrocardia and successful management of scar-related AFL due to surgical repair of ASD. The use of RF ablation with electroanatomic mapping system is effective and safe in such patients.

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