Two Different Types of Atrial Arrhythmia in a Patient with Persistent Left Superior Vena Cava

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A 36-year-old male patient with a surgically repaired secundum-type atrial septal defect, persistent left superior vena cava (PLSVC), and a history of typical atrial flutter ablation, was referred to our division for an additional atrial flutter ablation. The 12-lead electrocardiography displayed a left bundle branch block-like QRS morphology and atrial flutter with a 1:1 atrioventricular conduction at a rate of 200 bpm. Three-dimensional electroanatomic mapping of the right atrium (RA) and PLSVC, using a high-density multipolar mapping catheter, identified two dense scar areas. These were located at the posterior wall of the RA, superior to the inferior vena cava, and at the interatrial septum near the ostium of the PLSVC (Figure 1 and Video 1). High-density activation with Coherent and propagation mappings revealed a figure-of-eight activation pattern, with a slow conduction zone located between the two dense scar areas (Figure 1 and Video 1). Intracardiac electrograms obtained via the high-density mapping catheter at that site showcased low-frequency multicomponent/fragmented long signals, indicative of the isthmus of the macro-reentrant scar-related flutter (Figure 1). Entrainment from the isthmus resulted in a similar coronary sinus atrial activation sequence and a post-pacing interval equal to the tachycardia cycle length (Figure 1). Ablation from the isthmus initially slowed, and then promptly halted the tachycardia within a few seconds. Ablation tags were additionally applied perpendicular to the isthmus to connect the two dense scar areas (Figure 1 and Video 2). Upon completion of the ablation points, isoproterenol infusion was administered to test for any inducibility of atrial arrhythmias, which subsequently triggered another atrial tachycardia with a cycle length of 415 msec. High-density activation and propagation mapping of both atria revealed a focal atrial tachycardia originating from the middle part of the PLSVC (Figure 2 and Video 3). Ablation at the earliest site immediately stopped the tachycardia, and a circular ablation at this middle part successfully electrically isolated the distal part of the PLSVC (Figure 2 and Video 4). Additionally, there was a small gap area close to the annular side of the cavo-tricuspid isthmus in both the voltage and activation maps of the first tachycardia (Video 1). This gap area was ablated following the ablation of the first tachycardia. The collision of two wavefronts at the cavo-tricuspid isthmus line was also observed in the mapping of the second tachycardia (Video 3). No tachycardias were inducible at the end.

The PLSVC is the most common congenital intrathoracic venous anomaly, presenting in various forms. Concomitant congenital heart anomalies such as atrial septal defects might be present, as in our case. Patients with repaired atrial congenital anomalies might experience scar-related micro- or macro-reentrant atrial arrhythmias, as well as focal ones. Particularly, the PLSVC might be a source of non-pulmonary foci for atrial fibrillation and focal atrial tachycardias, as demonstrated in our case.1

The selection of intracardiac stable reference(s) is crucial for atrial arrhythmia mapping, unlike ventricular arrhythmia mapping. Identifying optimal reference points using the software of electroanatomic mapping systems can be challenging when the signal quality is suboptimal. It is sometimes impossible to find a satisfactory signal-to-noise ratio in patients with an extremely enlarged PLSVC. In such cases, positioning the reference catheter more distally into the great cardiac vein can resolve the issue. In the current case, a stable position of the coronary sinus catheter and the use of...
the Advanced Reference Annotation (ARA) algorithm software of the electroanatomic mapping system provided a sufficient signal-to-noise ratio for adequate mapping of the arrhythmia. A limited ablation strategy seems appropriate for focal tachycardia in such cases. However, it is well-known that the remnant of the left superior cardinal vein, called the ligament of Marshall, and the persistence of the left superior cardinal vein, known as the PLSVC, can be highly arrhythmogenic, exhibiting triggered activity or increased automaticity and serving as non-pulmonary foci for both atrial tachycardia and fibrillation. Therefore, isolation, rather than focal ablation, may have a higher probability of preventing future arrhythmias.

Figure 1. The voltage map displays two dense scar areas and an intervening myocardial area between them in a posterior view (A). Activation maps depict the early-meets-late zone overlapping with the area shown on the voltage map (B,C). Local EGMs (indicated by a blue tag) at this site reveal long, fractionated, low-frequency signals (D), and the entrainment maneuver resulted in a post-pacing interval matching the tachycardia cycle length, alongside a similar CS activation pattern (E). Initiating ablation at this point led to an increased cycle length, eventually halting the tachycardia. A short ablation line (marked by red tags) was established between the two dense scars (F,G,H). The voltage color bar setting was adjusted to range between 0.05 mV and 0.50 mV.

ABL, ablation; CS, coronary sinus; EGM, electrogram; IVC, inferior vena cava; P, Pentaray; PLSVC, persistent left superior vena cava; SVC, superior vena cava.
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Video 1. Activation patterns with propagation (A) and coherent (B) maps, and synchronized voltage map (C) of the right atrium.

Video 2. Ablation tags on coherent (A) and voltage (B) maps.

Video 3. Bi-atrial activation (separate (A, B) and combined (C)) with propagation maps of the focal atrial tachycardia.

Video 4. Circumferential ablation tags in the PLSVC. PLSVC, persistent left superior vena cava.

References