Left atrial access via an unroofed coronary sinus to eliminate fast/slow atypical AVNRT: A case report

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Introduction

Unroofed coronary sinus is a rare congenital anomaly that may alter the complex anatomy of the atrioventricular (AV) node. Arrhythmia circuits occurring among these patients may not be located in the usual position, especially those circuits involving the AV node. We report a case of the coexistence of this uncommon coronary sinus (CS) anomaly and the atypical form of atrioventricular nodal reentrant tachycardia (AVNRT) that could not be ablated by the conventional right-sided slow pathway modification but required left-sided septal ablation through an unroofed CS.

Case report

A 52-year-old female patient presented with recurrent intermittent palpitations and dyspnea for preoperative cardiovascular evaluation. A 2-dimensional echocardiogram revealed an enlarged right ventricle, and a dobutamine stress echocardiogram showed inducible wall motion abnormalities in the inferior wall. A cardiac computed tomography scan demonstrated no evidence of obstructive coronary artery disease but found, incidentally, a completely unroofed CS without persistence of left superior vena cava (Figure 1). Cardiac catheterization showed a small left-to-right shunt (pulmonary circulation to systemic circulation ratio [Qp/Qs] = 1.35) without pulmonary hypertension.

Subsequently, a 12-lead electrocardiogram captured a narrow-complex, long RP tachycardia that terminated with intravenous adenosine (Figure 2A). An electrophysiologic study (EPS) found evidence of dual AV nodal physiology, with a significant VA jump (retrograde slow pathway) during ventricular pacing. Right ventricular (RV) pacing induced the clinical tachycardia easily (Figure 2B). The tachycardia cycle length was 370 milliseconds, with long VA time (210 milliseconds) and concentric atrial activation. Attempts to entrain the arrhythmia with RV pacing continuously terminated the tachycardia without accelerating or advancing the atrial electrogram. A septal accessory pathway–mediated arrhythmia could not explain the arrhythmia, as it would be unusual to observe a long RP tachycardia in this situation. Furthermore, all the maneuvers performed in the study, such as PVC on His, as well as the mode of induction ruled out an accessory pathway–mediated arrhythmia. All these findings were consistent with the diagnosis of the fast/slow type of atypical AVNRT.

Radiofrequency (RF) energy applications with a 4-mm catheter at the anatomic slow pathway area in the right atrium between the CS and the tricuspid valve were unsuccessful despite the occurrence of slow junctional rhythm. A 3-dimernsional electroanatomic mapping system (NavX; St Jude Medical, St Paul, MN) was used to create a right and left atria activation map during tachycardia. Left atrial access was obtained by advancing the ablation catheter through the unroofed CS to map the left atrial septal area (Figure 3A). The earliest atrial activation was found at the lower left septal area (Figure 3B). RF ablation at the earliest atrial activation site rendered the tachycardia not inducible, without arrhythmia recurrence for more than 18 months.

Discussion

The prevalence of CS anomalies in patients with SVT has rarely been described. In a previous study including 408 patients with accessory pathways and AVNRT, significant CS abnormalities were found among only 12 patients (2.9%). In all, 6 patients had angulation of the CS, 4 patients had hypoplasia of the CS, 1 patient had narrowing of the proximal CS, and 1 patient had a fistula from a persistent left superior vena cava to the CS.[1] Another study evaluated the CS anatomy before EPS in 204 consecutive patients with SVT (43 with AVNRT). CS diverticula were found in 9 patients, with 5 patients having persistent left superior vena cava. An enlarged CS ostium (>25 mm) was seen in 9

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ABBREVIATIONS **ABL** = ablation catheter; **AV** = atrioventricular; **AVNRT** = atrioventricular nodal reentrant tachycardia; **CS** = coronary sinus; **EPS** = electrophysiologic study; **HIS** = His-bundle recording catheter; **HRA** = high right atrial recording catheter; **LA** = left atrium; **LAO** = left anterior oblique; **PVC** = premature ventricular contractions; **RA** = right atrium; **RAO** = right anterior oblique; **RF** = radiofrequency; **RV** = right ventricle; **RVA** = right ventricular apex; **SVC** = superior vena cava; **SVT** = supraventricular tachycardia (Heart Rhythm Case Reports 2015;1:457–460)

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KEY TEACHING POINTS

- Recognize the importance of anatomic variations of the heart and thoracic vessels in the treatment of supraventricular tachycardias.
- Recognize that some atypical forms of AVNRT cannot be ablated successfully in the conventional slow pathway area at the low right atrial septum.
- Recognize that activation mapping of the insertion of the retrograde limb of atypical slow/fast forms of AVNRT may help to achieve ablation success when the conventional anatomic-based ablation fails.

patients, of whom 4 had AVNRT.[2] None of these studies, however, have reported unroofing of the CS as a congenital anomaly present in cohorts of patients with SVT.

Unroofed CS is a rare anomaly[3] in which the common wall between the CS and the left atrium is absent. The anatomic defect may range from fenestrations in the roof of the proximal CS to the complete absence of the wall of the CS in the left atrium, where the coronary veins drain directly to the left atrium. A persistent left superior vena cava (SVC) is often but not invariably associated with the unroofing of the CS. Four anatomic variants are described: completely unroofed CS with or without persistent left SVC (types I and II respectively) and partially unroofed CS in the midportion (type III) or terminal portion (type IV).[4] The association between unroofed CS and arrhythmia substrate is limited to 1 AV accessory pathway,[5] 1 atrial tachycardia with unsuccessful endocardial ablation,[6] and 1 case of typical AVNRT successfully ablated in the classic right posterior paraseptal area.[7]

To the best of our knowledge, the coexistence of atypical AVNRT and unroofed CS has not been previously reported, and it is likely a coincidental association; the ablation in the left septum for atypical fast/slow AVNRT has not been previously reported either. During fast/slow AVNRT, the atrial activation is typically concentric (and is most commonly ablated at the low right septal area); nevertheless, an eccentric CS activation pattern has been observed in 18% of patients with fast/slow AVNRT requiring ablation up to 15 ± 4 mm inside the CS ostium to achieve success.[8] This finding, although controversial for some investigators is probably explained by the presence of a left atrial extension of the slow pathway to the human AV node,[9] which constitutes the retrograde limb of some atypical forms of AVNRT. This left atrial slow pathway extension of the AV node has been proposed as the ablation target for typical AVNRT when right-sided slow pathway ablation has failed (<5% of typical forms of AVNRT)[10]; however, ablation of the left septal area has not been reported as an effective treatment for atypical AVNRT.

The anatomy of the slow pathway in CS anomalies such as unroofed CS remains unclear. In our case, since conventional right-sided slow pathway ablation was not successful in preventing the arrhythmia, the next step would have been to map the earliest retrograde atrial activation at the CS ostium or a few millimeters inside the CS. Since the initial portion of the CS is absent among patients with unroofed CS, the left-sided slow pathway (retrograde limb of the fast/slow AVNRT) may have relocated to the left inferoseptal area during fetal development, as suggested in our case, therefore requiring ablation in the low left septal area at the earliest site of atrial activation to render the tachycardia noninducible.

Conclusion

We present an unusual case of the coexistence of a congenital anomaly of unroofing of the CS and atypical fast/slow AVNRT requiring RF slow pathway modification from the left septal area to achieve success.

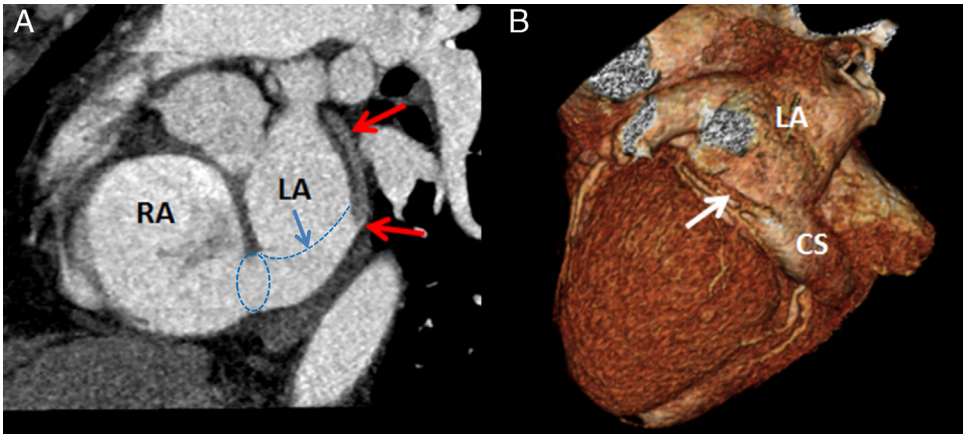

Figure 1 Cardiac computed tomography angiogram. **A:** Multiplanar reconstruction at the level of the atrial septal defect demonstrating completely unroofed coronary sinus, with the cardiac vein (red arrows) draining noncontrasted blood directly into the left atrium. Blue dotted lines represent the coronary sinus ostium and the area of the absent coronary sinus (CS) roof. **B:** A 3-dimernsional reconstruction demonstrating the cardiac vein (white arrow) entering into the coronary sinus. Notice that there is no definite separation between the mid CS and left atrium, confirming CS unroofing. LA = left atrium; RA = right atrium.