Case report

Persistence of left superior vena cava and focal right atrial tachycardia: Challenges and interventional treatment

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ABSTRACT

We describe the case of a patient with persistence of left superior vena cava and right atrial tachycardia originating between right superior vena cava and crista terminalis. We present the initial suspect of anatomical anomaly, the diagnosis and the interventional treatment of the arrhythmia. Potential challenges and the role of anomalous thoracic veins in promoting rhythm disturbances are also discussed.

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Introduction

The role of pulmonary veins in initiating arrhythmias was widely described [1]; a similar role was also described for superior vena cava, coronary sinus, Marshall vein, inferior vena cava and even for anomalous veins such as persistent left superior vena cava [2–7].

Clinical history

A 58-year-old man with history of previous mitral valvuloplasty (posterior annuloplasty with Sorin hemi-ring and posterior leaflet quadrangular resection 10 years before) and two previous episodes of persistent atrial fibrillation (treated with electrical cardioversion in the last year) underwent evaluation for relapsing palpitation. 12-lead ECG showed a regular atrial tachycardia with 5:1 atrioventricular conduction, with P-wave morphology and P-P interval suggesting clockwise right atrial flutter (Fig. 1). A previous echocardiogram showed mild mitral regurgitation, normal left ventricular function and a marked enlargement of the coronary sinus (CS). To rule out a thoracic veins anomaly, a cardiac magnetic resonance (MRI) was performed, demonstrating the persistence of left superior vena cava (LSVC) with associated “normal” right superior branch (Fig. 2a).

Electrophysiologic evaluation

We performed an electroanatomical mapping of the arrhythmia using Ensite NavX technology (St Jude Medical Inc) with Fusion algorithm (map merged with cardiac MRI). A left
subclavian vein approach allowed the stable placement of a hexapolar deflectable mapping catheter in the CS, recording a right-side originating atrial arrhythmia. The catheter was advanced through the LSVC to the CS, with the tip facing the CS ostium and the proximal dipole in the CS distal part (Fig. 2b). A slight bend of the deflectable tip allowed sufficient stability to use this catheter as the geometric reference for the electroanatomical mapping system.

The recording from a circular mapping catheter placed in the right atrium excluded an isthmus-dependent flutter, suggesting a focal arrhythmia originating from the crista terminalis (Fig. 3), with eccentric excitation from high right atrium propagating toward septum and lateral atrial wall. The electroanatomical mapping with a quadripolar 4 mm irrigated ablation catheter confirmed the presence of earliest electrical activation from a focus at the junction between right superior vena cava and crista terminalis. Radiofrequency ablation at this site interrupted the arrhythmia, obtaining stable sinus rhythm (Fig. 4).

Discussion

Persistent LSVC is the most common congenital anomaly of thoracic veins (0.3–0.5% of general population and 3–5% of congenital heart disease patients) [8-11]. During normal fetal life, cardinal veins coming from the upper district of the body merge into the sinus venosus; the left cardinal vein then disappears, leaving the vestigial Marshall ligament and the coronary sinus. If the left system persists, then a LSVC is observed; LSVC drains in the right atrium through the coronary sinus in 50% of the cases, in the other 10% LSVC joins directly the right atrium or the pulmonary veins [12]. LSVC is usually an occasional finding, especially during placement of pacemakers/defibrillator leads or central venous catheters or during thoracic open surgery [13-15]; it can also be suspected on the basis of thoracic radiogram or echocardiography [16,17]. The diagnosis can be confirmed using contrast echocardiography [18]. Thoracic CT scans or MRI can obviously better define anatomy and are recommended before proceeding to intervention [19].

The demonstration of LSCV deserves attention in performing arrhythmia ablation, especially for localizing the starting point of the arrhythmic circuit and for placement of the catheters and lesion sets: in our case, the major challenge was to achieve a stable position for the CS mapping catheter, used as reference for the electroanatomical mapping system; we obtained the goal using a steerable catheter, slightly flexed in the vein. LSVC was an “innocent bystander” for the arrhythmia demonstrated in our patient, but this is not always the case [6,7]: frequently it has an active role in promoting reentrant circuit, or contains automatic focal activity that can trigger arrhythmias.
Fig. 2 – (a) MRI clarified the anatomy: a left persistent superior vena cava was documented (marked with *, artefactually interrupted due to scale). DICOM data were then merged with Ensite NavX maps. Left anterior oblique view on the left, right anterior oblique view on the right. (b) The placement of the CS catheter is showed. We advanced the catheter through the left superior vena cava (red dotted line), placing the tip at the ostium of the coronary sinus and the proximal dipolo into the CS distal part. Bending the deflectable part of the catheter was needed to obtain adequate mechanical stability inside the markedly enlarged CS. Ao, aorta; PA, pulmonary artery; RV, right ventricle; LV, left ventricle.
Fig. 3 – A tachycardia originating from the upper part of the right atrium was documented with a 20-polar catheter and coronary sinus activation. The earliest potential is on 17–18 (facing the roof-high lateral wall of the right atrium); the activation then proceeds to both 19–20 (medial roof of right atrium) and 15–16 (lateral wall of right atrium). One-two (lower lateral wall-floor of right atrium) comes slightly later than CS signals. This activation is not compatible with a common typical atrial flutter (isthmus-dependent macroreentry).

Fig. 4 – Interruption of the arrhythmia with radiofrequency ablation at the junction between right superior vena cava and right atrial roof.
and the electrophysiologist should pay attention to search for these findings when the anatomical anomaly is suspected.

**Conflict of interest**

No conflict of interest.

**Ethical statement**

I declare, on behalf of all authors that the research was conducted according to Declaration of Helsinki.

**Informed consent**

The authors declare that informed consent requirements do not apply to this manuscript.

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**Appendix A. Supplementary data**

Supplementary material related to this article can be found in the online version, at doi:10.1016/j.crvasa.2015.05.008.

**REFERENCES**


