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ABSTRACT

Upper venous system anatomic variations may cause difficulties during cardiac pacemaker implantation. Persistent left superior vena cava (PLSVC) and absent right superior vena cava could be an arrhythmogenic source of atrial arrhythmias and cardiac conduction disease. We represent dual-chamber pacemaker implantation in a patient with a very rare upper venous system anomaly, paroxysmal atrial fibrillation, sick sinus syndrome, that cause unusual fluoroscopic image.

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1. Introduction

Persistent left superior vena cava (PLSVC) is the most common anatomic variation of the upper venous system. Absent right superior vena cava associated with a PLSVC is a rare occurrence of 0.1% [1]. This rare combined venous anomaly is also associated with an increased incidence of atrial arrhythmias and cardiac conduction abnormalities [2,3]. This rare anatomic variation could be challengeable during device implantation. Herein, we describe dualchamber pacemaker implantation in a patient with paroxysmal atrial fibrillation, sick sinus syndrome, isolated PLSVC, and absent right superior vena cava.

2. Case presentation

69 years old male referred to our hospital for paroxysmal atrial fibrillation (PAF) ablation. The CHADSVasc Score was 1. Electrocardiogram revealed low atrial rhythm with a heart rate of 54 bpm and a 2.1-sec pause (Fig. 1). Ambulatory rhythm monitoring showed an average heart rate of 56 bpm and pauses detected after atrial fibrillation termination episodes. He was also diagnosed with sick sinus syndrome (SSS). Echocardiography revealed EF 60%, left atrial size 31 mm, and no remarkable sign. The treadmill exercise test was performed for evaluation of chronotropic incompetence. The

maximum heart rate was reached 125 bpm. On follow-up, the patient had a traumatic syncope episode.

In patients with PAF and SSS, rhythm or rate control strategies can be problematic because of anti-arrhythmic drugs (AADs). AADs can suppress sinus node function and bradycardia-related symptoms may become more prominent. AF ablation might be a good option to restore sinus rhythm but had no affect on sinus node dysfunction. PLSVC may be an arrhythmogenic source of atrial fibrillation and may require extensive ablation in addition to pulmonary vein isolation. Since patient had symptomatic bradycardia we decided to implant dual chamber pacemaker instead of AF ablation.

At the beginning of pacemaker implantation, the guidewire did not advance inferior vena cava via a left axillary vein. Venography performed (Video 1). PLSVC was observed. Right side venography showed right sided venous system was drained to PLSVC. Procedure canceled and computed tomography was planned to confirm the upper system venous anomaly. CT images confirmed PLSVC and agenesis of right superior vena cava (Fig. 2A and B). Both upper venous systems drained to PLSVC. In the second procedure, we implanted active fixation RV lead through the PLSVC to the basal septal proportion of RV with handshape 'C' stylet (Fig. 3). The active atrial lead was located to the posterior RA because of the unexpected dense fibrosis detected in the lateral RA. At the end of the procedure, an unusual course of both electrodes cause unexpected fluoroscopic images 'bow-tie' at the right anterior oblique (RAO) view (Fig. 4A and B- Video 2).

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Fig. 1. 12 leads electrocardiogram showed low atrial rhythm with 2.1-s pauses.



Fig. 2. Computer tomography: two and three dimensional consecutive coronal reformatted (A and B) images show agenesis of the right superior vena cava (black long arrow), bridging vein (black short arrow) drained to persistent left superior vena cava. Ao: Aortae.



Fig. 3. 'C' shaped handmade stylet led the electrode through the PLSVC to the basal septal proportion of RV.



Fig. 4A. Fluoroscopic images at the right anterior oblique view (30°) of both leads. The active atrial electrode was located to the posterior RA (short arrow) and the active RV electrode implanted through the PLSVC to the basal septal proportion of RV (long arrow).



Fig. 4B. Fluoroscopic images at the left anterior oblique view (45°) of both leads. Both electrodes come down from the PLSVC implanted. The active atrial electrode was located to the lateral RA (short arrow) and the active RV electrode to the septal proportion of RV (long arrow).

Supplementary video related to this article can be found at https://doi.org/10.1016/j.ipej.2021.10.007

3. Discussion

PLSVC could be associated with other congenital heart diseases, cardiac conduction diseases such as sick sinus syndrome, and a source of atrial arrhythmias. The presence of a dilated coronary sinus on echocardiography should alert the clinician towards the possibility of PLSVC. Computer tomography and/or venography will confirm the diagnosis [4].

In patients with PAF and SSS, rhythm or rate control strategies can be problematic because of anti-arrhythmic drugs (AADs). AADs can suppress sinus node function and bradycardia-related symptoms may become more prominent. Pacemaker implantation is a reasonable option to tolerate the AADs. Another choice is AF ablation. PLSVC may be source of the arrhythmogenic foci for AF initiation. PLSVC connection with left atrium may lead the AF episodes to continue even after pulmonary vein isolation. Electrical isolation of PLSVC seems to be necessary particularly in those patients [5]. However this procedure will require wider ablation.

Pacemaker implantation through a PLSVC and absence of right superior vena cava remains a difficult procedure. Regardless of the venous access side, the leads entered the right atrium through the coronary sinus. Ventricular electrode placement can be difficult relative to the atrium. Hand 'C' shaped stylets and active fixation electrodes are helpful to overcome difficulties of the right ventricular electrode placement. Implantation from the right pectoral side could make the procedure more difficult. Although it is applicable LV pacing through PLSVC with passive electrodes, complications such as coronary sinus thrombosis were reported [6,7]. And long-term electrode performance is also unknown. Transcatheter leadless pacemaker could be an alternative solution for the cases with failed upper venous system approach.

Fluoroscopic cardiac silhouette assessment should be done with both right anterior oblique 30° and left anterior oblique 45° views. In the RAO view, it can be assessed whether the electrode is located anterior vs posterior or basal vs apical. In the LAO view right or left chamber distinction can be made. In our case, the RAO view showed unexpected images because of both electrode implanted through the PLSVC. 'C' shaped curved posterior RA and basal septal RV leads created 'bow-tie' images (Fig. 4A). LAO view showed septal localization of RV and lateral localization of RA electrode (Fig. 4B).

In conclusion, we represent unusual dual-chamber pacemaker implantation in a patient with paroxysmal atrial fibrillation, sick sinus syndrome, isolated PLSVC, and absent right superior vena cava. This case showed the importance of CT imaging and venography in the evaluation of anatomic variation before pacemaker implantation and using different fluoroscopic projections to determine the correct pacing side.

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