Successful Implantation of a Dual-Chamber Permanent Pacemaker in a Patient with Persistent Left Superior Vena Cava and Absence of Right Superior Vena Cava: Tips and Tricks

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SOUHRN

Ageneze pravostranné horní duté žíly při perzistující levostranné horní duté žíle (persistent left superior vena cava, PLSVC) představuje vzácnou a obecně asymptomatickou vrozenou malformaci. Obvykle se zjistí náhodně během implantace kardiostimulátoru. V této kazuistice popisujeme naše zkušenosti s implantací dvoudutinového kardiostimulátoru u pacienta s tak složitými anatomickými poměry a vyzdvihujeme klinický význam venografie pro jednoznačný popis anatomie konkrétního žilního systému na operačním sále. Zdůrazňujeme zvláště metody použité pro zajištění správného umístění elektrody.

ABSTRACT

An absent right superior vena cava associated with persistent left superior vena cava (PLSVC) is a rare and generally asymptomatic congenital malformation. It is usually discovered incidentally during pacemaker (PM) implantation. In this report, we describe our experience with implanting a dual-chamber pacemaker in a patient with such complex anatomy and the clinical value of peripheral phlebography for clearly describing venous anatomy in the surgical room. In particular, we highlight the methods used to ensure correct lead positioning.

Introduction

Persistent left superior vena cava (PLSVC) is a rare vascular anomaly that is usually identified incidentally during cardiovascular imaging or procedures. This anomaly arises because of the failure of the left anterior cardinal vein to regress during embryonic development, which occurs in 0.3–0.5% of healthy individuals.[1,2] Generally, the PLSVC drains into a dilated coronary sinus (CS); however, in rare instances, it can drain into the left atrium via an unroofed CS or, less commonly, directly. Almost 90% of cases present with bilateral superior venae cavae; conversely, the absence of the right superior vena cava (ARSVC) is rare, with a prevalence of 0.09% to 0.13% in the population.[3]

PLSVC might be associated with abnormal electrophysiological function resulting from anatomical and structural heart abnormalities, presenting clinically as either tachycardia or bradyarrhythmia.[4,5] Although PLSVC is often benign, it can complicate invasive procedures, such as pacemaker (PM)/Implantable Cardioverter-Defibrillator (ICD) implantation, central venous access, and cardiothoracic surgery. We report successful and uncomplicated implantation of a dual-chamber PM in a patient with PLSVC and ARSVC who experienced a complete heart block.

Case presentation

A previously healthy 46-year-old male patient was referred to our hospital with complaints of shortness of breath, dizziness, and weakness. He had no significant medi-

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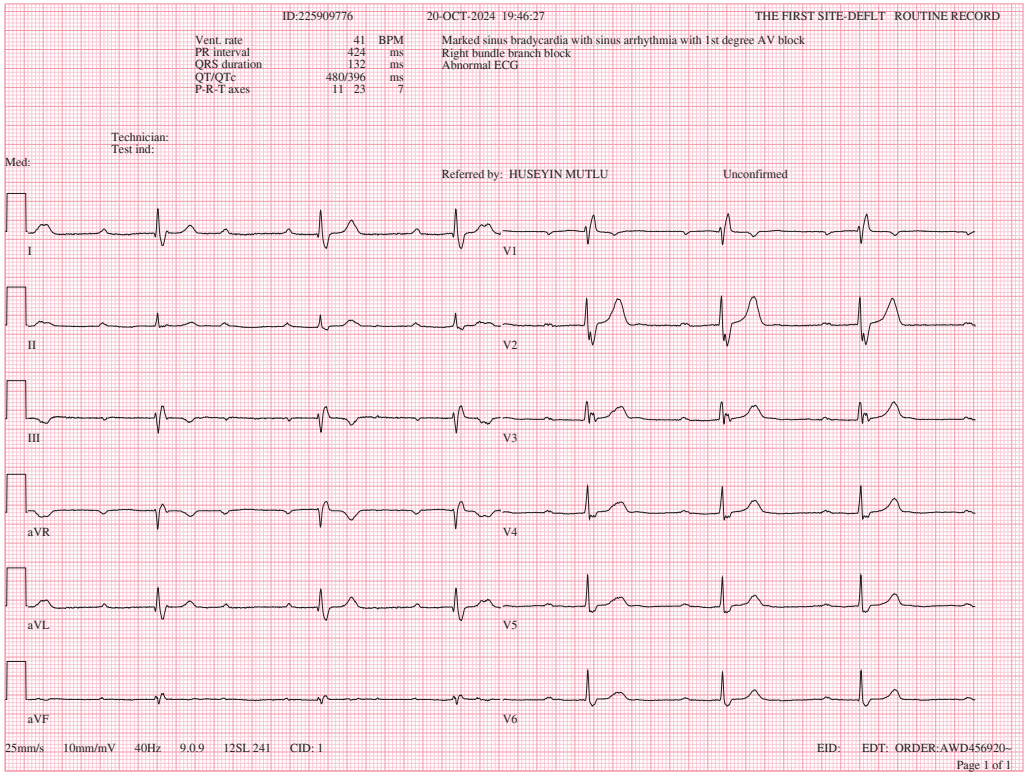

Fig. 1 – A 12-lead ECG showed a ventricular rate of 4 1/min with atrioventricular dissociation.

cal history and denied experiencing angina, palpitations, syncope, or any related symptoms. On examination, his blood pressure was 148/66 mmHg with a regular pulse of 38 beats per minute, with no clinical evidence of heart failure. His examination was otherwise unremarkable. A 12-lead electrocardiogram revealed a complete atrioventricular block with a ventricular rate of 34 beats per minute and right bundle branch block morphology (**Fig. 1**). Transthoracic echocardiography revealed normal left ventricular size and function. There was no laboratory evidence of either a metabolic or ischemic cause of the conduction disease. Coronary angiography revealed normal coronary arteries, and dual-chamber PM implantation was scheduled.

The procedure was initially performed using the left subclavian approach with a left pectoral incision. Following the left subclavian puncture, resistance was encountered while attempting to advance the lead, leading to its retraction. A venogram revealed a PLSVC draining into the right atrium (RA) via the CS and its acute angle of emergence (**Video 1**). Therefore, we decided to place the lead contralaterally via the right subclavian vein. The right subclavian vein was cannulated under fluoroscopy. Unexpectedly, the venogram revealed the ARSVC, with the right brachiocephalic vein draining directly into the PLSVC.

The placement of the right atrial pacing lead into the RA cavity was not particularly challenging; however, positioning it within the right atrial appendage required careful manipulation. After entering the RA cavity, the straight stylet of the RA pacing lead was replaced with the curved "J" stylet, and clockwise torque was required to advance the lead anteriorly and place it in the adjacent atrial appendage location.

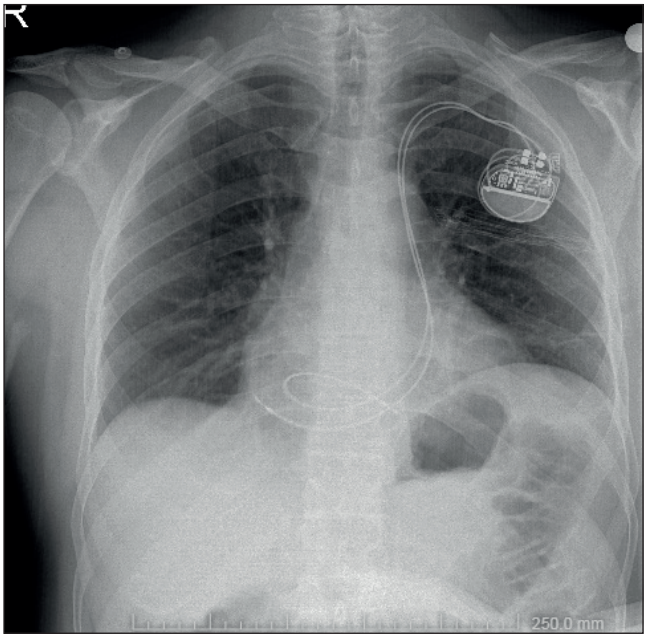

Fig. 2 – Radiographic view of the final placement of the right atrial and ventricular leads via persistent left superior vena cava and coronary sinus.

Positioning the right ventricular (RV) pacing lead was more challenging since the pacing lead gained atrial access through the CS ostium, which was very close to the RV inlet, but the CS ostium directed the lead away from

the RV inlet toward the lateral free wall of the RA. The stylet was shaped into a semicircle to facilitate entry into the RV and then pushed into the RV by making a loop against the lateral wall of the RA. Retracting the stylet a few centimeters was crucial to direct the tip of the RV pacing lead from the RA-free wall into the RV and ensure that the lead tip was no longer stiff, thus minimizing the risk of atrial perforation. The lead was positioned near the RV apex, which was confirmed by fluoroscopy (**Fig. 2, Video 2**). Care was taken to form an adequate loop in the RA to avoid dislodgement in the future. Both leads exhibited excellent parameters, with thresholds of 0.9 mV for each and resistances of less than 750 ohm. A dual-chamber PM was connected to the leads and implanted in a deep pocket. The wound was then closed in layers. The recovery was uneventful, and the patient was discharged after 2 days.

Discussion

In the current case, we reported our experience with PM implantation in a patient with a rare venous anomaly consisting of an absent right and a PLSVC.

PLSVC is the most common benign thoracic venous anomaly and is usually discovered incidentally. Nevertheless, the presence of PLSVC affects the structure of the heart and vessels. The PLSVC drains approximately 20% of the total venous return and significantly dilates the CS. Draining directly into the left atrium is often associated with complex cardiac pathologies, such as atrial septal defect and aortic coarctation.[1] Therefore, timely identification of PLSVC, which is associated with conduction abnormalities and congenital heart disease, is crucial for patient prognosis.

The PLSVC can be identified before PM implantation. This anomaly may be suspected during a routine chest X-ray performed after central venous access is established. Moreover, echocardiography that shows a dilated CS should also be considered. These findings should encourage clinicians to use preoperative venous imaging. Understanding the anatomy facilitates the procedure and allows for more complex interventions because it is associated with the use of techniques to overcome pathological angulations.

The potential for a PLSVC should be considered during device implantation if the guiding wire moves in a left downward direction or if resistance is felt while trying to push it forward. In this situation, performing venography before the procedure is an effective and simple method. Injecting 50 cc of contrast agent into each subclavian vein allows for precise mapping of the thoracic venous system, and the ARSVC can be easily documented.

The PM implantation with PLSVC remains challenging. The large size and thin wall of the CS increase the risk of injury during lead implantation, raising safety concerns. In addition, accessing through the PLSVC is technically complex, resulting in a higher likelihood of failed lead implantation and difficulties in achieving the optimal lead position. Hand-shaped styles and active fixation leads might help to overcome technical difficulties and achieve successful long-term results.

Conclusion

The ARSVC and PLSVC is an extremely rare congenital anomaly encountered unexpectedly during routine PM implantation. Intraprocedural venography is highly effective in verifying the ARSVC and in accurately determining the CS route. An experienced operator can overcome anatomical challenges by using shaped styles and active fixation leads.

Acknowledgements
None.

Conflict of interest
None.

Ethical statement
We declare that the case report was conducted following the applicable ethical standards and guidelines, as outlined in the Declaration of Helsinki.

Informed consent
Appropriate permissions, including written informed consent, were obtained.

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