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CASE REPORT

Persistent Left Superior Vena Cava With Absent Right Superior Vena Cava

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ABSTRACT

A 52-year-old patient was referred to our hospital with the indication for a pacemaker implantation. The attempt to implant the pacemaker lead through the right subclavian vein revealed an unusual course of the lead. Phlebography through the right subclavian vein revealed the presence of a persistent left superior vena cava and the absence of a right superior vena cava.

INTRODUCTION

During the procedure of a pacemaker implantation, one may come across various venous anomalies. The most common congenital thoracic venous anomaly is a persistent left superior vena cava (PLSVC),[1] which poses a technical challenge while attempting to implant the pacing leads. A case of a PLSVC is herein presented and the challenges and alternative techniques and route(s) of lead implantation are discussed.

ABBREVIATIONS
PLSVC = persistent left superior vena cava
SVC = superior vena cava

CASE REPORT

A 52-year-old left-handed patient with a history of hypertension and diabetes was referred to our hospital for symptomatic (exertional dyspnea) 2:1 second degree atrioventricular block, prompting a decision for a pacemaker implantation.

An attempt to implant the pacing lead through the right subclavian vein revealed an unusual course of the pacemaker lead on the left side of the spine, indicating the presence of persistent left superior vena cava (PLSVC). Indeed, phlebography performed through the right subclavian vein revealed the presence of an isolated PLSVC and the absence of the right superior vena cava (SVC) (Fig. 1A & 1B). A VDD pacemaker lead was then inserted through the left subclavian vein (Fig. 1C), but despite our intense efforts to advance it to the right heart cavities, that was not feasible, and the patient was referred for an epicardial pacemaker lead implantation. An echocardiogram was performed, which disclosed a dilated coronary sinus in the absence of elevated right ventricular filling pressures. The injection of normal saline into the left arm vein revealed the enhancement of the dilated coronary sinus before the right atrium (Fig. 2).

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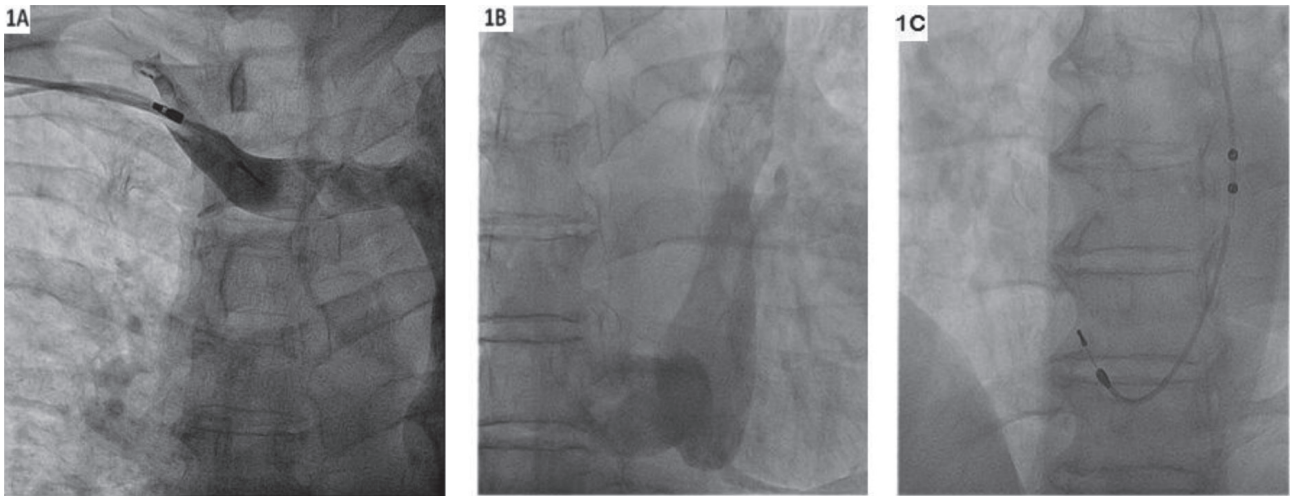

FIGURE 1. Phlebography through the right subclavian vein showed the absence of the right superior vena cava (1A), and the presence of a left persistent superior vena cava with a dilated coronary sinus (1B). Panel 1C shows the pacemaker lead having an unusual course on the left side of the spine.

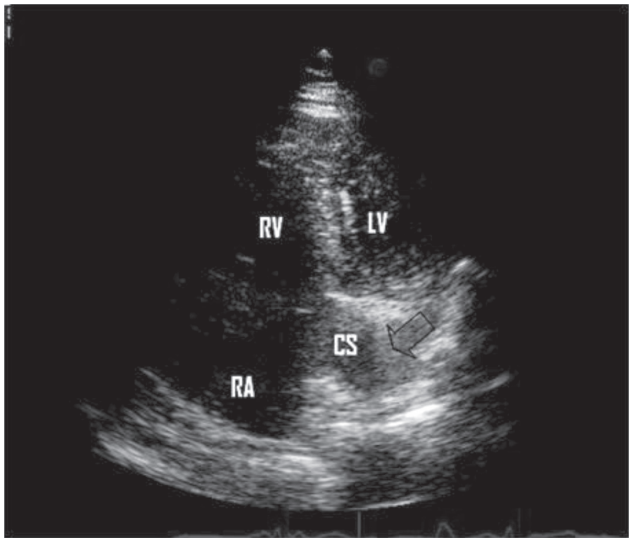

FIGURE 2. Echocardiographic appearance of early enhancement of the large coronary sinus (arrow) following normal saline injection from the left brachial vein. LV = left ventricle; RV = right ventricle; CS = coronary sinus; RA = right atrium.

No other cardiac abnormalities were detected.

DISCUSSION

A persistent left superior vena cava (PLSVC) constitutes the most common anatomic variation of the thoracic venous system. Since the associated clinical finding and symptoms are rare, a PLSVC is incidentally identified usually during device implantation. A PLSVC constitutes an embryological remnant, resulting from the persistence of the embryonic left anterior cardinal vein [1] with a prevalence of 0.3% in healthy individuals and 4.4% in patients with congenital heart disease. [2,3] In the most common form of PLSVC, both the left and right SVCs are present. A bridging innominate vein may or may not be present. Less frequently, the caudal right superior cardinal vein regresses resulting in absent right SVC with PLSVC (isolated PLSVC), with a reported incidence of 0.1%.[4]

A routinely performed chest X-ray after a central venous access has been established, and a dilated coronary sinus (>1 cm) on echocardiography, raise the suspicion of the anomaly. The echocardiographic evaluation should be followed by agitated saline application from the left or right arm. Normally, the infusion of saline injection from the left or right antecubital vein results in opacification of the right atrium. In cases where the right SVC accompanies the PLSVC, contrast given from the left arm first appears in the coronary sinus, whereas contrast given from the right arm first appears in the right atrium. In 10% of patients, a PLSVC may drain into the left atrium either directly or via an unroofed coronary sinus; in such a situation, contrast first appears in the left atrium.[5] The transesophageal echocardiographic findings are focused on the appearance of the PLSVC near the left atrial appendage and left upper pulmonary vein, in mid-esophageal views, while in the bicaval view, the absence of a right SVC can be demonstrated. Other imaging modalities, like venous computed tomography or magnetic resonance imaging, directly visualize the venous anatomy and confirm the diagnosis.[6]

The presence of isolated PLSVC is associated with various clinical problems,[5] such as rhythm abnormalities (atrioventricu-

lar block because of stretching of the atrioventricular node and His bundle secondary to dilatation of the coronary sinus), a high incidence of accompanying congenital heart defects[7] (ventricular and atrial septal defects), and the inability to insert central venous catheters and transvenous leads through the right internal jugular/subclavian vein. Furthermore, in cases of right-sided open-heart surgical procedures, a PLSVC has to be drained by inserting a separate cannula into it.[5] Additionally, in open heart surgery the delivery of retrograde cardioplegia is not feasible, because of inability to obtain tight seal by the balloon of cardioplegia catheter.[5]

Implantation of pacemaker leads through a persistent left superior vena cava constitutes a challenging procedure since the lead enters the right ventricle through the coronary sinus, making the maneuver into the right ventricle extremely difficult.[8] The existence of an acute angle between the coronary sinus ostium and the tricuspid valve makes it necessary that the lead should be looped in the right atrium in order to enter the right ventricle. Successful pacemaker implantation in cases of isolated PLSVC have been reported in the literature, whereby a long (85 cm) active fixation lead could be manipulated to enter the right ventricle by the use of the wide-loop technique.[9,10] In case of dual pacemaker implantation a second lead is screwed into the right atrial wall.[9] Finally, pacing through the tributaries of the coronary sinus or a middle cardiac vein in patients with PLSVC have also been reported.[11,12]

A dilated coronary sinus on echocardiography should raise the suspicion of PLSVC. The diagnosis should be confirmed by saline contrast echocardiography. Physicians should consider the presence of PLSVC whenever a catheter or a guide wire inserted via the left subclavian vein takes an unusual left-sided downward course. A PLSVC, especially in the absence of a right SVC, presents technical difficulties and risks during the insertion of leads into the right heart chambers for implantation of devices (pacemakers or implantable cardioverter defibrillators). Long active fixation leads have overcome many of the implantation technical obstacles.

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