

잔존 좌측 상대정맥을 통한 심실제동기의 성공적인 삽입

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Successful Implantation of an Implantable Cardioverter-Defibrillator Through a Persistent Left Superior Vena Cava

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The prevalence of persistent left superior vena cava (PLSVC) in the general population has been estimated to be approximately 0.3%, as determined by autopsy. PLSVC is hemodynamically insignificant if it is not associated with other congenital cardiac anomalies, and usually goes unrecognized until a left superior approach to the heart is required. Here, we report a 60-year-old male diagnosed with dilated cardiomyopathy, who had survived an episode of sudden cardiac arrest with documented ventricular fibrillation. PLSVC was recognized incidentally while implanting an implantable cardioverter-defibrillator (ICD). The ICD was successfully implanted through the left superior vena cava via the coronary sinus using a straight stylet and an active fixation device. (Korean J Med 2014;87:328-333)

Keywords: Persistent left superior vena cava; Implantable cardioverter-defibrillator

INTRODUCTION

Persistent left superior vena cava (PLSVC) was first described by Edwards and DuShane in 1950 [1]. The prevalence of PLSVC has been estimated to be 0.3% of the general population through an examination of a large sample of unselected autopsies, and is much higher in patients with congenital cardiac anomalies, ranging from 3% to 9% [2,3]. The PLSVC generally

drains into the right atrium (RA) through the coronary sinus (CS) [4]. An isolated PLSVC is usually not recognized until a left cephalic or subclavian approach is used in diagnostic or therapeutic transcatheter procedures. The transvenous introduction of a lead from the RA to the right ventricle (RV) may be a technically demanding procedure in subjects with a PLSVC. Here, we report a patient with a PLSVC, in whom an RV implantable cardioverter- defibrillator (ICD) lead was implanted

Received: 2008. 4. 23

Revised: 2008. 9. 10

Accepted: 2008. 10. 7

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successfully. To our knowledge, this is the first case report of ICD implantation through the left side in a patient with a PLSVC in Korea.

CASE REPORT

In July 2007, a 60-year-old male with no relevant prior medical history felt heavy in his chest and collapsed at Incheon International Airport. There was no history of any cardiopulmonary symptoms or family history of heart disease or sudden death. He was not taking any medication. The basic life-support team found the patient lying on the ground and initiated cardiopulmonary resuscitation immediately. Upon arrival at the emergency department, the patient was intubated and ventricular fibrillation (VF) was documented (Fig. 1A). Multiple electrical countershocks resulted in a stable pulse and the patient was transferred to the cardiac intensive care unit for observation. On the subsequent day, the patient again went into hemodynamically unstable VF and underwent successful cardioversion. Although amiodarone infusion began to stabilize the patient, he experienced several episodes of VF thereafter, each of which was treated successfully with cardioversion. Twelve episodes of VF were recorded within 48 hours of admission.

After conversion, cardiac evaluation revealed a normal sinus rhythm on 12-lead electrocardiogram (Fig. 1B) and an

enlarged cardiac silhouette without pulmonary congestion on chest X ray. Echocardiography showed that the left ventricle was dilated and the left ventricular function was severely depressed with an ejection fraction of 18%. Cardiac catheterization revealed no stenosis in the coronary arteries. Routine laboratory tests, including metabolic panel and electrolytes, were within the normal limits. A diagnosis of dilated cardiomyopathy was made. As the patient had survived an episode of sudden cardiac death with documented VF, and he had advanced heart disease, it was decided to implant an ICD considering the results of a previous meta-analysis of AVID, CIDS, and CASH results [5].

Under local anesthesia, the left subclavian vein was punctured using Seldinger's technique and a guide wire was passed. Advancement of the wire under fluoroscopy raised suspicion of PLSVC. The wire was removed and contrast medium was injected, which confirmed the presence of a PLSVC (Fig. 2A). A defibrillating lead for ICD (model SPRINT FIDELIS, 6931-651 Medtronic Inc., Minneapolis, MN) was inserted and further advanced through the PLSVC into the CS, and then into the RA. Advancement of the lead to the RV through the tricuspid annulus was technically challenging due to the acute angle between the CS ostia and tricuspid annulus, which is different from advancement from the right superior vena cava to the RV. Due to the angle of the pre-shaped stylet itself, the lead could not be advanced through the

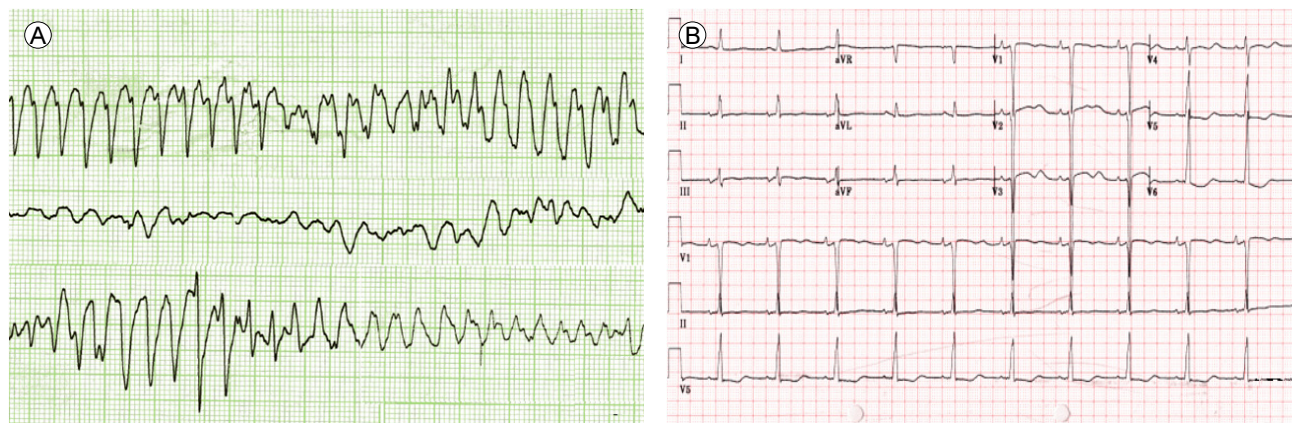


Figure 1. (A) Electrocardiogram recorded in the emergency department showing ventricular fibrillation. (B) Twelve-lead electrocardiogram after conversion showing a normal sinus rhythm in the same patient.

tricuspid annulus. Although careful manual shaping of a curved stylet in various ways was attempted, the lead failed to be advanced into the RV; instead it went in the opposite direction, reaching the high RA. After several unsuccessful attempts, we changed to a straight stylet. It was advanced smoothly through the PLSVC into the CS, reached the RA wall, and then formed a loop on itself using the lateral RA wall for support within the atrium and was advanced through the tricuspid annulus. The lead was placed successfully in the RV and anchored to the apex by extending the screw to ensure lead stability.

The pacing threshold was 0.5 V at 0.5 ms with a pacing impedance of 504 Ω . The sensed R wave activity was 15.6 mV. After placing the defibrillating lead, it was connected to an ICD generator (model Maximo VR; Medtronic Inc.) in a subpectoral pocket. VF was induced using a T-wave shock after sedation with sodium thiopental and the device successfully

reverted the VF into a sinus rhythm with an energy of 35 J. The patient was discharged on the third postoperative day with amiodarone. Follow-up chest X rays 1 week later revealed stable lead positioning (Fig. 2B).

DISCUSSION

PLSVC is due to abnormal development of the sinus venosus in the early stages of fetal life. In a 4-mm embryo, this structure is comprised of three distinct parts, the right horn, the transverse part, and the left horn, which collects three pairs of veins (the omphalomesenteric veins, the umbilical veins, and the common cardinal veins). Due to the rightward direction of blood flow, the right horn undergoes preferential growth and ultimately constitutes the intercaval (nonmuscular) part of the RA. The transverse part and the proximal left horn of the sinus venosus invaginate from the

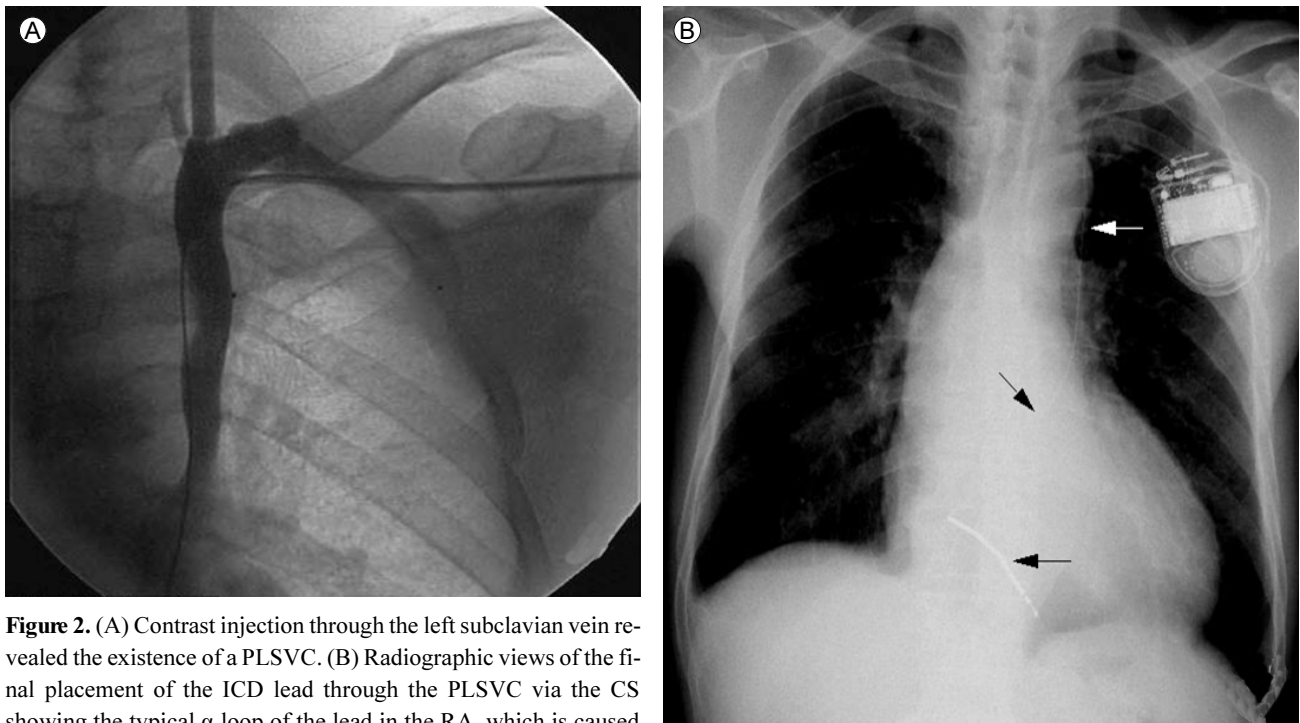


Figure 2. (A) Contrast injection through the left subclavian vein revealed the existence of a PLSVC. (B) Radiographic views of the final placement of the ICD lead through the PLSVC via the CS showing the typical α loop of the lead in the RA, which is caused by the close proximity of the orifice of the CS and the tricuspid valve (arrows). The ICD generator is placed in the left pectoral region. PLSVC, persistent left superior vena cava; ICD, implantable cardioverter-defibrillator; RA, right atrium; CS, coronary sinus.

left atrium (LA) and forms the CS. The distal left horn and the left cardinal vein are obliterated, and form the so-called *ligament of Marshall* in adult subjects [6]. If the left cardinal vein persists, it drains into the RA through the CS in 92% of cases, and forms a PLSVC. In the remainder of cases, drainage occurs in the LA because of failure to form the CS [4].

The ontogenetic development of the sinus node, the atrioventricular (AV) node, and the His bundle may be heavily influenced by the lack of regression of the left cardinal vein because these structures are located at the junctions of the right and left cardinal veins with the sinus venosus [7]. James et al. reported that a PLSVC altered the histological organization of both the sinus node and the AV junction, causing a small and poorly formed sinus node, fetal dispersion of the AV node, and a His bundle within the central fibrous body, small diameter of the His bundle, and poor arterial supply to either the AV node or sinus node [8,9]. On the other hand, Anderson et al. reported that the PLSVC deviated the AV node from a horizontal to a vertical position without changing the cellular morphology of the node [10]. Overall, these histological and anatomical findings indicate the basis of various types of cardiac electrical instability and predispose the patients to arrhythmias and sudden death. Therefore, they should be considered in evaluation of any patient with PLSVC, particularly if there is clinical evidence suggesting arrhythmia or a conduction disturbance, as in our patient.

Generally, the condition is asymptomatic and hemodynamically insignificant if it is not associated with other congenital cardiac anomalies. Moreover, unless specifically searched for, the presence of a PLSVC may be overlooked, including on two-dimensional echocardiography [11]. Therefore, when isolated, PLSVC is usually not recognized until transvenous procedures, such as right heart catheterization, pacemaker or ICD implantation, biventricular pacing, or when electrophysiological assessment via the left cephalic or subclavian approach is required. The unexpected presence of a PLSVC draining into an enlarged CS may cause technical

difficulties and lead to misplacement of the catheter and injury to the vessel wall. Serious complications, including angina, arrhythmia, cardiogenic shock, and even cardiac arrest, have been reported when a guide wire or catheter is manipulated through a PLSVC. Despite these risks, several investigators have already reported the successful implantation of an ICD in patients with PLSVC [12-17]. The most common problems encountered were reaching a convenient pacing site and ensuring stable lead placement. Various techniques have been used to obtain reliable function of the implanted system in these patients. In particular, exchanging or hand-shaping of the stylet in a predefined manner and using active fixation leads have been reported to be useful to overcome the technical difficulties encountered in procedures of ordinary durations [12-14]. In the case of an inability to maneuver from the left side, right-sided access is used after confirming the presence of a right superior vena cava by right peripheral venography or echocardiography. However, a change in procedure to the right side necessitates a longer procedure time with increased radiation exposure, and echocardiography or contrast venography to ensure the presence of a right superior vena cava, which may cause greater discomfort to the patient [12-15]. In addition, it was reported that the defibrillation thresholds were increased by right-sided vascular access at the time of the initial ICD implantation [18].

Between 1982 and April 2007, five patients with isolated PLSVC combined with arrhythmia or conduction disturbance were reported in Korea. Among them, two patients were diagnosed with sick sinus syndrome. One underwent pacemaker implantation via the right subclavian approach, and the other through a PLSVC [19,20]. In two patients with AV nodal reentry tachycardia, successful radiofrequency catheter ablation was performed with energy delivery at the enlarged CS through the PLSVC [21]. The fifth patient with chronic atrial fibrillation did not undergo the transvenous approach, but was diagnosed with a PLSVC by contrast echocardiography using agitated saline [22].

Although the QT intervals corrected for heart rate using Bazette's formula varied over a wide range from 400 to 600

ms during sinus rhythm in our patient, there was no T wave alternans, notched T wave in three leads, or low heart rate for age, which are known as unique electrocardiogram characteristics in long QT syndrome. Moreover, he denied any history of syncope or congenital deafness, and there was no history of long QT syndrome or sudden cardiac death in family members <30 years of age without defined cause. In this case, the PLSVC was recognized incidentally while implanting an ICD. A defibrillating lead was inserted and further advanced smoothly through the PLSVC into the CS and then into the RA. However, despite several attempts to hand-shape the pre-shaped stylet in various ways, which has been reported to be helpful, the lead could not be advanced into the RV. The lead was placed successfully in the RV with a straight stylet by having the lead form a loop on itself using the lateral RA wall for support, and active fixation of the lead was accomplished. The induced VF could be terminated with an energy of 35 J. Statistically significant predictors of high defibrillation threshold have been reported [23]. The most common predictors are large cardiac size, large body size, wide QRS, high New York Heart Association functional class, VF as the manifesting arrhythmia, and low ejection fraction. VF was documented in our patient and echocardiography showed that the left ventricle was dilated and left ventricular function was severely depressed with an ejection fraction of 18%. Moreover, it should be noted that the patient had been taking amiodarone for 14 days when we implanted the ICD, because short-term administration of amiodarone has been reported to raise defibrillation threshold [24].

There were several important aspects of this case. First, PLSVC is an important anatomical finding when a left superior approach to the heart is considered. Therefore, the internist or cardiologist should be aware of the possibility of a PLSVC when a guide wire or catheter takes an unusual left-sided downward course. In addition, a literature review as well as this case report emphasized the fact that although manipulating the catheter is technically difficult, a PLSVC does not prevent successful device implantation. Finally, the present case showed that the usage of a straight stylet may

help overcome the technical difficulties encountered when introducing a lead from the RA to the RV, which has not been reported previously. We suggest use of a straight stylet, which allows easy access to the RV in our experience.

In summary, we reported a patient with PLSVC in whom successful ICD implantation was possible. To our knowledge, this is the first case report of ICD implantation by the left side in a patient with PLSVC in Korea.

중심 단어: 잔존 좌측 상대정맥; 심실제세동기

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