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Devices for Resident Physicians

Successful dual chamber ICD implantation via a persistent left superior vena cava after ratchet syndrome

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1. Introduction

Device implantations may be challenging in patients with venous abnormalities. The most common congenital variation, frequently associated with other congenital abnormalities, is a persistent left superior vena cava (PLSVC), and successful atrial and ventricular lead implantation via a PLSVC have both been reported [1].

“Ratchet syndrome” has also been reported as a rare complication after a newly implanted pacemaker or implantable cardioverter-defibrillator (ICD) and involves lead dislodgement and retraction due to a ratchet-like movement through the suture sleeve that causes a continuous short pull [2,3]. Here, we present a rare case in which successful dual-chamber ICD implantation via a PLSVC was achieved after the occurrence of ratchet syndrome.

2. Case

A 63-year-old woman with arrhythmogenic right ventricular cardiomyopathy (ARVC) experienced a sustained monomorphic ventricular tachyarrhythmia (VT) unresponsive to drug therapy. She underwent implantation of a dual chamber ICD (TELIGEN 100 F111, Boston Scientific Inc., St. Paul, MN, USA) using active fixation leads for both the atrial (DEXTRUS 4136, Boston Scientific Inc.) and ICD leads (ENDOTAK RELIANCE G, Boston Scientific Inc.) for the secondary prevention of VT. Venography confirmed a PLSVC and

revealed no small branch that would allow the PLSVC to communicate with the right-sided venous drainage (Fig. 2A). Thus, ICD implantation was performed from the right subclavian vein because of the PLSVC. The suture sleeves were secured with two non-absorbable monofilament sutures for each. The active fixation lead was placed into the right atrial appendage (stimulation threshold measured as 1.4 V at 0.4 ms, and atrial wave amplitude 2.1 mV), and the ICD lead was placed into the right ventricular apex (stimulation threshold measured as 1.0 V at 0.4 ms, and R-wave amplitude 7.1 mV). The lead position after the operation was identical. After 2 months, an atrial lead dislodgement was documented (Fig. 1A). Eventually the lead retracted fully and spontaneously into the ICD pocket (Fig. 1B). The electro-cardiogram revealed right ventricular pacing without atrial pacing (Fig. 2B). After the loss of atrial pacing, chest radiography revealed cardiomegaly. We considered that progressive heart failure had occurred due to the high frequency of ventricular pacing (Fig. 1B).

Six months after the first implantation, a reimplantation procedure was performed. The atrial lead was easily removed from the pocket, and a new lead placement was attempted. Using a venogram as a guide, the axillary vein was found to be almost totally occluded (Fig. 2C) with collateral development, and the guidewire could not pass through the axillary vein after the puncture. The patient had right-arm edema 2 months before admission, so this venous trouble was strongly suspected to have coincided with the emergence of edema. The previous ICD lead was smoothly extracted without forcibly tugging, with a back-up cardiac surgical team. After extraction of the previous lead, we attempted to implant both the atrial and ICD leads from the left pectoral region via the PLSVC (Fig. 2D). The atrial lead was

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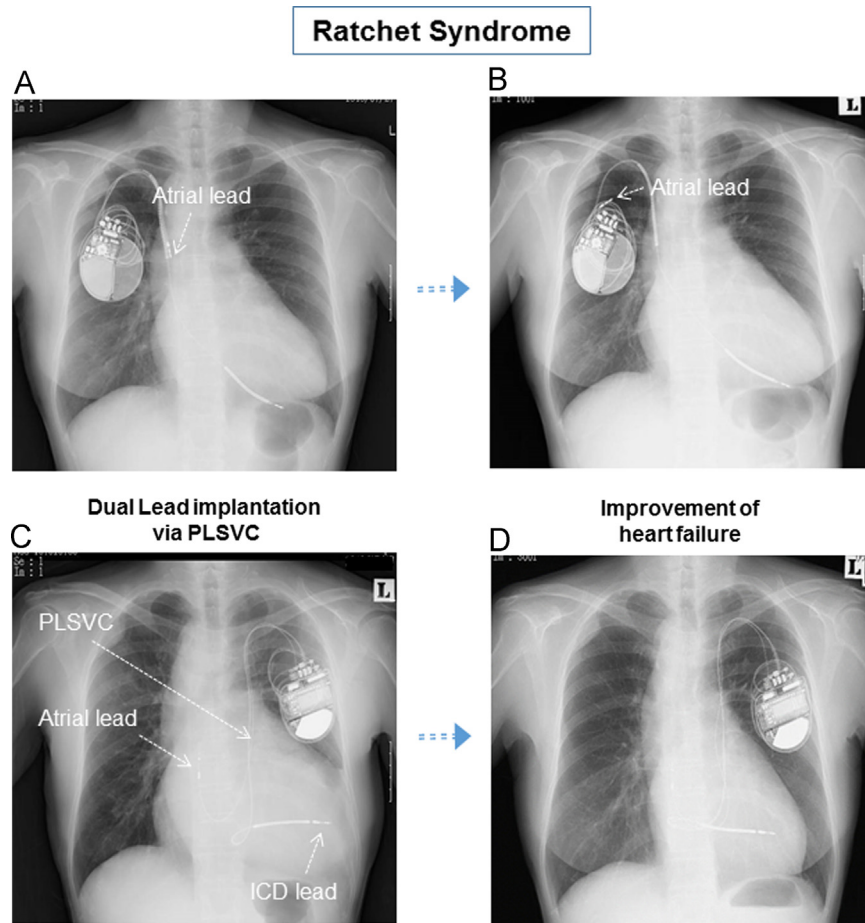


Fig. 1. (A) A chest radiograph taken 2 months after the operation. The tip of the atrial lead is located at the mid portion of the superior vena cava (indicated by the arrow). (B) Full retraction of the atrial lead into the pocket (arrow) at the time of subacute exacerbation of her heart failure. (C) The alpha configuration of the implantable cardioverter defibrillator lead and atrial lead implanted via the persistent left superior vena cava (PLSVC). (D) Dramatic improvement in heart failure after the successful dual lead implantation, with complete atrial pacing and without any ventricular pacing.

smoothly placed in the right atrial appendage. To implant the ICD lead, access to the right ventricle was achieved using a hand-shaped stylet, first advancing the lead towards the lateral wall of the right atrium and subsequently with the tip of the lead pointing towards the atrium into the right ventricle. Eventually, the ICD lead was placed with a loop on the lead across the tricuspid valve (Fig. 1C). After successful implantation, the patient's heart failure dramatically improved with complete atrial pacing and without any ventricular pacing (Fig. 1D).

3. Discussion

Lead dislodgement with loss of the pacing and sensing function is a serious complication after the implantation of cardiac implantable devices [4–6]. “Ratchet syndrome” is recognized as a rare complication involving lead dislodgement and retraction that occurs by a ratchet-like mechanism. A previous paper has suggested that the underlying ratchet-like mechanism is a consequence of repetitive upward and downward movements of the shoulder and/or arm along with weak tightening of the fixation-sleeve suture. Although the right hand was dominant in this patient, we had implanted the device system in a right pectoral site due to the existence of the PLSVC. Furthermore, the oval-shaped device used in this case had square and flat edges, and this may have allowed surplus space for the suture and lead to move within the device pocket. Therefore, it is quite possible that a

continuous “pushing and pulling” of the lead may have constituted the proposed ratchet-like mechanism in this case [6]. In addition, only the atrial lead was dislodged, and it was suspected that loosening of the suture on only the atrial lead fixation sleeve had occurred during the first implant procedure.

This patient had sick-sinus syndrome and also took sotalol for the prevention of VT related to ARVC. Therefore, constant atrial pacing was indispensable for maintaining a physiological atrial rhythm. Frequent right ventricular pacing is known to cause left ventricular dysfunction, myocardial perfusion defects, abnormal septal contractility, and heart failure in some patients [7,8]. Therefore, it appears possible that the dominant ventricular pacing that occurred after the dislodgement of the atrial lead caused exacerbation of her chronic heart failure.

ICD lead implantation through a PLSVC is often challenging and sometimes unsuccessful [9]. A loop on the lead across the tricuspid valve can result in higher mechanical stress, making it prone to lead failure [10,11]. Furthermore, this patient may need an upgrade from an ICD to a cardiac resynchronization therapy device in the near future because of the presence of cardiomyopathy, but a left ventricular (LV) lead implantation via a PLSVC seems difficult. A recent paper has demonstrated both successful ICD [12] and LV [13,14] lead implantation through a small branch communicating between a PLSVC and the right-sided venous drainage. However, as in our case, the PLSVC and right superior vena cava are not connected by a communicating branch in 40% of cases (Fig. 2A) [15]. Thus, before the implantation procedure, the PLSVC

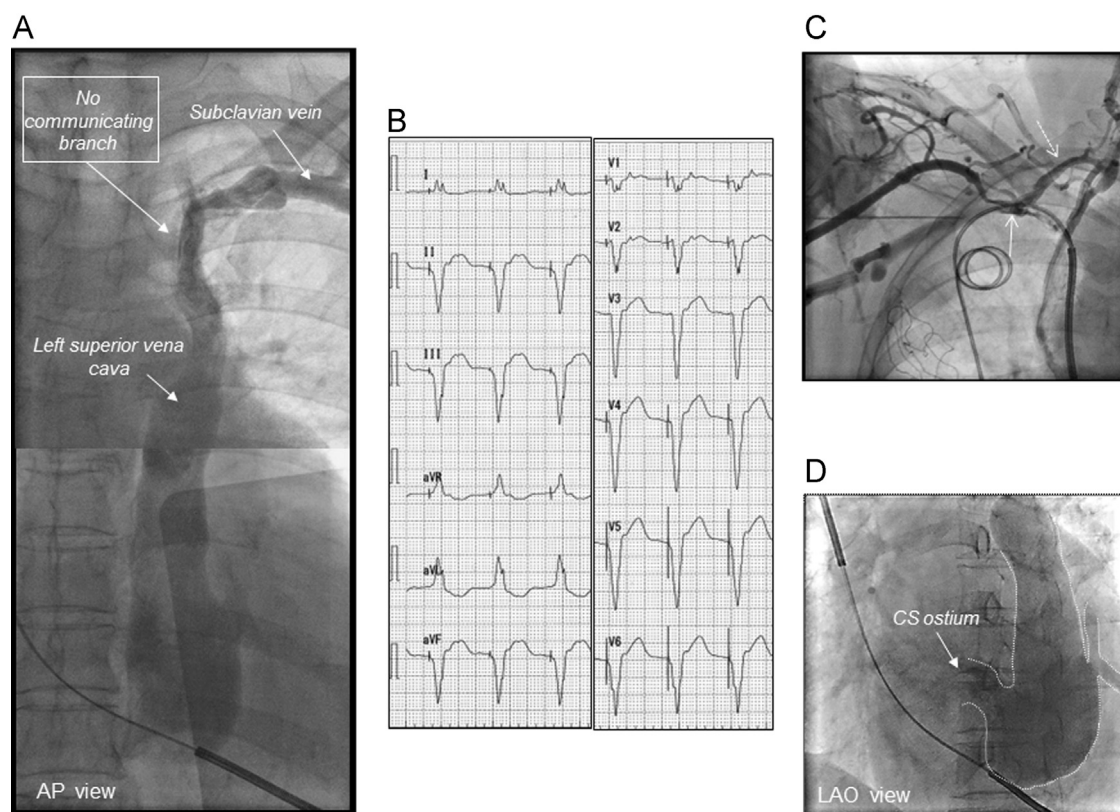


Fig. 2. (A) Venogram from the left subclavian vein showing the persistent left superior vena cava without a communicating branch. AP, anteroposterior. (B) Twelve-lead electrocardiogram during complete ventricular pacing, which appeared after the dislodgment of the atrial lead. (C) Venogram showing partial occlusion of the axillary vein (white arrow) with the development of collaterals (white dotted arrow). (D) Venogram showing the persistent left superior vena cava and the coronary sinus (CS). LAO, left anterior oblique.

system should be carefully inspected for any communicating branches or appropriate lateral coronary veins using detailed venography.

The current generation of ICDs is also capable of achieving acceptable defibrillation thresholds when placed in the right pectoral region [16]. However, the easier method of performing a right-sided ICD implantation in patients with both a PLSVC and dextral dominance may cause an unusual form of lead dislodgment. To prevent an iatrogenic complication after an unavoidable right-sided implantation, such as that reported here, tightening of the suture on the lead fixation sleeve should be recommended.

Conflict of interest

All authors declare no conflicts of interest related to this study.

Disclosures and funding sources

None.

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