Case Report: Cardiovertor-Defibrillator Implantation in an Isolated PLSVC with Hypertrophic Cardiomyopathy

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Abstract: Persistent Left Superior Vena Cava (PLSVC) has an estimated incidence of 0.2-0.8% in the general population and 3-10% in patients with known congenital heart disease [1]. A rare anatomic variation of LSVC occurs in up to 0.13% of otherwise normal individuals where both brachiocephalic venous systems drain into a persistent LSVC [1]. We describe a patient with hypertrophic cardiomyopathy (HCM) and malignant ventricular arrhythmias in whom this rare malformation was discovered at the time of implantable cardioverter–defibrillator (ICD) implant.

Keywords: Persistent left superior vena cava, isolated superior vena cava, absent right superior vena cava, combined superior vena cava, Hypertrophic cardiomyopathy, Cardioverter-defibrillator, Anomalies of superior vena cava.

INTRODUCTION

Despite being the most commonly reported thoracic vein anomaly, Persistent Left Superior Vena Cava (PLSVC) has an estimated incidence of only 0.2-0.8% in the general population and 3-10% in patients with known congenital heart disease [1]. PLSVC, caused by failure of regression of the left cardinal vein into the innominate vein, is characterized by the entire left brachiocephalic venous system draining into a markedly enlarged coronary sinus. The SVC is normally developed and drains the right brachiocephalic system in the RA. However, rarely an anatomic variation of this situation occurs in which the RSVC is absent and both the right and left superior systems are drained solely by the Left Superior Vena Cava (Isolated PLSVC). The incidence of Isolated PLSVC is estimated to be 0.07-0.13% [2], and 50% of these cases are accompanied by other cardiac malformations including atrial septal defect, endocardial cushion defect, and tetralogy of Fallot [3]. Here, we describe a novel association of Isolated PLSVC with hypertrophic cardiomyopathy (HCM).

CASE

A 50-year-old male with asymmetric septal hypertrophy (Figure **1**) without significant outflow gradient presented with recurrent syncope. Non-sustained ventricular tachycardia (VT) was documented on Holter and sustained polymorphous VT was easily induced in an electrophysiology study. He was subsequently referred for implantable cardioverter–defibrillator (ICD). The left subclavian vein was accessed successfully, but as the guide wire was advanced, its abnormal course suggested the presence

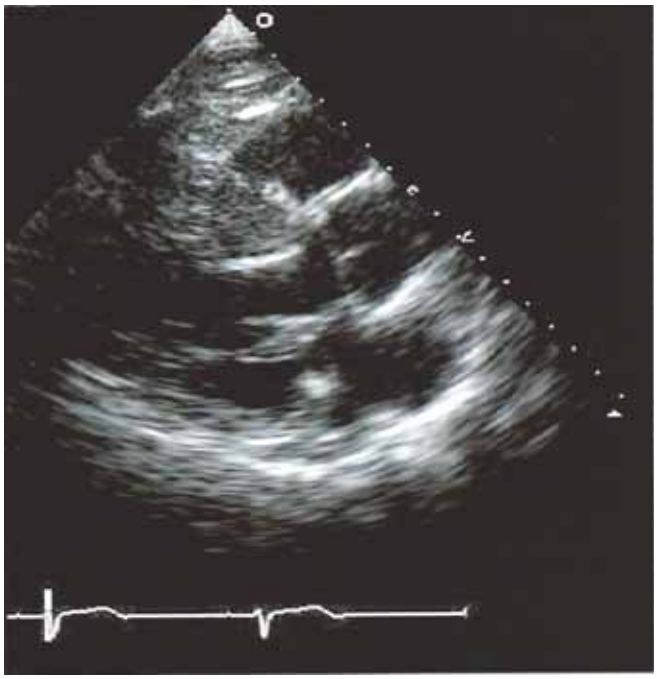

Figure 1: Echocardiographic Parasternal Long-axis view showing asymmetrically hypertrophied ventricular septum compared to the inferior-lateral wall.

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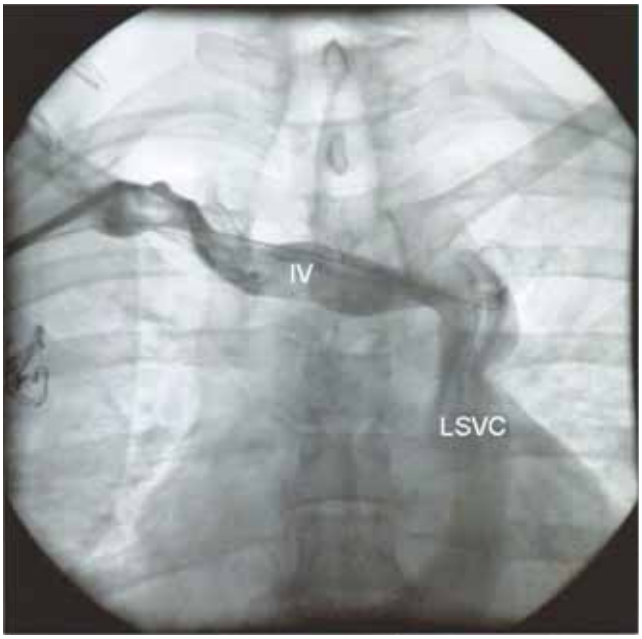

Figure 2: Contrast venography in the anterior-posterior view. Contrast material injected through the Innominate Vein (IV) showed absent Right Superior Vena Cava with the contrast material entering the Right Atrium through the Left Superior Vena Cava (LSVC) and the Coronary Sinus (CS).

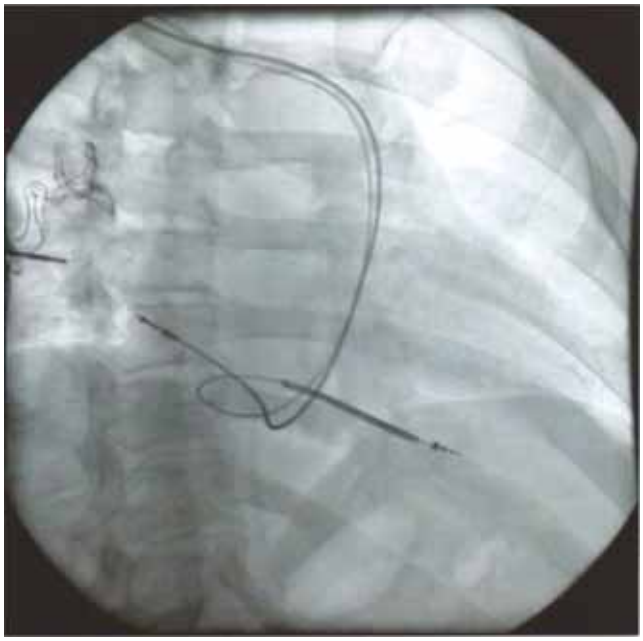

Figure 3: Right Anterior Oblique (RAO) view showing the course of the intra-cardiac leads. The Atrial and the Ventricular leads course through the Innominate Vein to the Left Superior Vena Cava to the Coronary Sinus, and then into the Right Atrium and Right Ventricle, respectively.

of a persistent left superior vena cava. In order to decrease the chance of lead dislodgment, implantation was instead pursued on the right side. The right subclavian vein also showed an anomalous course, this time consistent with an absent vena cava. Contrast injection confirmed this anomaly (Figure **2**). Leads were inserted using standard peel-away sheaths. Exiting from the coronary sinus the lead faces the lateral wall of the right atrium rendering the placement of the atria lead unproblematic. A curved stylet inserted in the ventricular lead permitted us to torque it clockwise to the level of the mid right ventricular septum. The lead was secured in this location by extending the fixation screw. This maneuver stabilized the lead allowing the curved stylet to be exchanged with a straight one and after retracting the fixation mechanism deploy the lead in the right ventricular apex (Figure **3**).

DISCUSSION

Persistence of the left superior vena cava is the most frequently reported anomaly of the systemic venous return. Most commonly, a right-sided superior vena cava opening into the right atrium is present. In our patient a more complex anomaly existed in which the innominate veins carried blood from left and right upper body venous systems to the coronary sinus. Despite this anatomic anomaly, using standard tools and a modified manipulation technique both defibrillator leads were secured in a stable position. Developmental anomalies of the superior vena cava are often observed in conjunction with other congenital anomalies, but the association of HCM and isolated PLSVC was never reported before. This is more likely to have occurred as chance event but the possibility of a single genetic defect explaining both abnormalities cannot be discounted.

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