Sudden Shortness of Breath and Bradycardia Leading to Dual-Chamber Pacemaker Placement in a 78-Year-Old Man

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On physical examination, the patient’s blood pressure was 140/60 mm Hg. Cardiac examination revealed marked bradycardia with a heart rate of 30 bpm and a regular rhythm. He had bibasilar rales on lung auscultation and pitting edema in both legs. Resting electrocardiogram showed complete heart block. Troponin I was within normal limits. He was immediately admitted to the cardiovascular unit where his blood pressure remained stable overnight. The following morning, the patient underwent dual-chamber pacemaker placement due to persistent third-degree atrioventricular block. Chest radiograph obtained after the procedure showed the pacing wires diverging into 2 well-delineated directions (Figure).

WHAT IS YOUR DIAGNOSIS?

(A) Extravascular malposition of the pacer wire with mediastinal rupture
(B) Insertion of the lead in a left pericardiophrenic vein
(C) Insertion of the pacer wire in a persistent left superior vena cava
(D) Insertion of the pacer wire in the descending aorta

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ANSWER

The correct answer is (C), insertion of the pacer wire in a persistent left superior vena cava (PLSVC).

DISCUSSION

On closer inspection of the posteroanterior view of the chest radiograph (Figure), the ventricular lead is seen on the right side of the mediastinum following the usual course through the superior vena cava to the right ventricle. On the other hand, the atrial lead is seen on the left side of the mediastinum traversing a path en route to the right atrium. These radiographic findings are consistent with the presence of a PLSVC. Extravascular malposition with mediastinal rupture can be excluded by the uneventful postoperative course and the absence of mediastinal widening on the chest radiograph. Careful attention to the course of the lead can help rule out insertion in the descending aorta. In the case of insertion in the descending aorta, the lead would follow a course from the left border of the heart and then along the spine, whereas a lead in a PLSVC will curve medially near the left atrium. Alternatively, if a central line was inserted, an arterial wave form would be present if the pacer wire had been inserted into the aorta. Occasionally, diagnostic modalities such as echocardiography or computed tomography may be necessary to rule out aortic malposition. When the catheter is inserted in the left pericardiophrenic vein, the wire is expected to travel along the left border of the mediastinum and then curve laterally along the left border of the heart. In contrast, a wire that passes through a PLSVC curves medially near the left atrium.

PERSISTENT LEFT SUPERIOR VENA CAVA

Prevalence

Although relatively rare, PLSVC is the most common thoracic venous anomaly, with a prevalence of approximately 0.05% to 0.5% in the general population. When PLSVC is associated with congenital heart disease (most commonly with tetralogy of Fallot), prevalence is higher, approaching 2.1% to 4.3%. One study documented that 4 of 1139 patients undergoing pacemaker implantation had PLSVC. Patients with this condition typically do not have symptoms or experience hemodynamic insufficiency. Rather, PLSVC is usually detected as an incidental finding on chest radiographs following placement of pacemakers or pulmonary artery catheters.

Embyology

During embryologic development, venous blood from the head and upper limbs drains into the right atrium via the left and right anterior cardinal veins. At 8 weeks of gestation, the left brachiocephalic vein develops as a bridge between the left and right cardinal veins. The portion of the left anterior cardinal vein caudal to the left brachiocephalic vein usually degenerates, whereas the right cardinal vein stays patent and becomes the superior vena cava. PLSVC results from nonregression of the caudal portion of the left anterior cardinal vein. At least 67% of cases of PLSVC are associated with a patent right anterior cardinal vein, resulting in bilateral superior vena cava. PLSVC empties into the right atrium via the coronary sinus in most cases or may occasionally drain into the left atrium through an unroofed coronary sinus.

Diagnosis and Clinical Implications

Diagnosis of PLSVC can be made definitively by echocardiography, and pulsed and color Doppler can further illustrate venous flow pattern. Other noninvasive diagnostic modalities include multislice spiral computed tomography or magnetic resonance imaging. Invasive procedures such as cardiac catheterization are typically unnecessary.

PLSVC does not require treatment; however, recognition is important for several reasons, especially in view of the rising number of cardiac electrophysiologic interventions being performed. First, it is technically more difficult to insert pacemakers or pulmonary artery catheters through the left internal jugular or subclavian vein in the presence of a PLSVC. The orifice of the coronary sinus lies inferior and posterior to the tricuspid valve. Thus, it becomes more difficult to pass a pulmonary artery catheter from the superior vena cava through the coronary sinus into the right atrium and then into the right ventricle. In these cases, alternative routes of central venous cannulation (eg, femoral, right internal jugular, or right subclavian veins) may be considered.

The risk of cardiovascular complications during catheterization is higher in patients with PLSVC. There is an increased incidence of arrhythmias, particularly supraventricular tachycardia, likely caused by catheterization of the coronary sinus. Cardiac arrest has also been reported during cardiac catheterization. Awareness of PLSVC also may help to accurately position the atrial pacing lead near the coronary sinus to prevent atrial arrhythmias. Postprocedure chest radiographs may falsely suggest misplacement of the atrial lead if the physician is not aware of this abnormality.

CLINICAL COURSE OF CASE PATIENT

Our patient had an unremarkable postprocedure course and was discharged with stable vital signs. He

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did not require repositioning of the pacemaker leads and has had no pacemaker-related complications on follow-up.

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