Persistent Left Superior Vena Cava during Aortic Valve Replacement

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A 57-year-old African American man with a 6-month history of chest pain presented for aortic valve replacement. He had documented severe aortic stenosis, moderate aortic insufficiency, left ventricular hypertrophy and an ejection fraction (EF) of 50% on a preoperative transthoracic echocardiography (TTE). After an uneventful induction, a prebypass transesophageal echocardiography (TEE) revealed a calcified tricuspid aortic valve with severe stenosis (aortic valve area 0.7 cm$^2$; AV mean gradient 40 mmHg), severe aortic insufficiency, and an EF of 35%. His coronary sinus was significantly dilated. The X-plane view at the midesophageal (ME) 2-chamber revealed a diameter of 2.09 cm (Figure 1). The diagnosis of persistent left superior vena cava (PLSVC) was made by a bubble study (Figure 2). This finding was further confirmed by the inability to perform retrograde cardioplegia. The enlarged coronary sinus (CS) is well demonstrated in 3-Dimension (3-D) TEE (Figure 3). Once the surgeon was informed of the findings and other potential associated congenital anomalies ruled out, the cardioplegia solution was injected directly into the coronary arteries and then proceeded with the 21-mm On-X aortic valve replacement. The rest of the procedure was uneventful and the patient was transported to the Surgical intensive care unit (SICU) in stable condition. Later, the patient had computer tomography (CT) scan of his chest for evaluation of his lung nodules. It confirmed the intraoperative TEE diagnosis (Figure 4).

PLSVC is the remnant of the embryologic left sinus horn found in 0.1% to 0.2% of the general population and 2% to 9% of patients with congenital heart disease [1]. However, Bartram et al. found 56 out of 121 patients (46%) with PLSVC were accompanied with congenital diseases [2-4]. PLSVC in association with congenital cardiac malformation increases the risk of mortality in cardiac surgery on cardiopulmonary bypass (CPB) [5]. If it drains into the left atrium or through an unroofed persistent left superior vena cava, the risk of death is significantly increased [5].

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CS or partially unroofed CS, or CS ostial atresia, a significant right-to-left shunting, cyanosis, and even brain abscess secondarly to paradoxical embolization can occur. It should be surgically corrected by redirect the PLSVC to the right atrial appendage [5,6]. In patients with CS ostial atresia, the exclusion of PLSVC may cause severe coronary ischemia during cardiac surgery [6].

PLSVC could be missed by echocardiography [5]. Diagnosis of PLSVC is usually made as an incidental finding during cardiovascular imaging or surgery as in our case. In most cases, PLSVC drains into the right atrium through CS. It has no hemodynamically significance and no clinical symptoms. Thus, no surgical management is needed [7]. Because of that reason, we did not attempt to make surgical correction or direct visual inspection of CS because it requires right atrium incision and two cannulations for the veinous drainage instead of one. However, it can cause technical difficulties during procedures such as right cardiac catheterization performed by using left subclavian vein, such as pace-maker application, implantable cardioverter defibrillator administration, biventricular cardiac pace or electrophysiologic studies as such as pace-maker application, implantable cardioverter defibrillator in cardiac congenital surgery [6]. The presence of PLSVC is a relative contraindication to the administration of retrograde cardioplegia during cardiac surgery [3,5-7]. Cardiogpegia may be inadequate, since it comes back through the PLSVC. If a failed retrograde cardioplegia goes unrecognized, this could result in failed myocardial protection, which in turn would cause heart failure. Arrhythmia and cardiogenic shock. Therefore, we use antergrade cardioplegia by injecting the cardioplegia solution directly into the coronary arteries ostia.

Full TEE exam and index of suspicion are the keys to making an earlier diagnosis of PLSVC. 80% of PLSVC is associated with dilation of the coronary sinus [5] as seen in this case. The diagnosis can be made by injection of enhanced saline contrast in the left antecubital vein. The contrast will be seen in the dilated CS before the right atrium (RA). There is normal transient contrast in the RA after injection through the right SVC. Combining 3D and 2D echocardiography is important because of the strong association between PLSVC and other congenital abnormalities, such as coronary AV fistula, partial anomalous pulmonary venous return, or “unroofed” CS affording the shunt between CS and left atrium. If there is any doubt, a venogram, contrast-enhanced CT or magnetic resonance imaging should be performed to obtain the precise diagnosis of PLSVC [8]. In our case, we confirmed the diagnosis of PLSVC by CT scan.

References