Abstract
Persistent left superior vena cava (LSVC) with absent right SVC (RSVC) is a rare congenital anomaly. If undetected, the condition may pose difficulties in central venous catheter insertion, pacemaker electrode insertion, and cannulation during cardiopulmonary bypass. We describe a case of persistent LSVC with absent RSVC, who was diagnosed to have bicuspid aortic valve with aortic stenosis.

Keywords: Absent right superior vena cava, aortic stenosis, bicuspid aortic valve, persistent left superior vena cava

Introduction
A bicuspid aortic valve (BAV) is the most common congenital cardiovascular anomaly, occurring in 1%–2% of population.[1] It may be associated with other abnormalities in 20% of cases. Association of BAV with persistent left superior vena cava (PLSVC) and agenesis of right SVC (RSVC) is extremely rare. Majority of these remain undetected until the patient presents with the symptoms of associated heart disease. However, it may be detected during central venous catheterization (CVC) or at the time of surgery for other conditions.

Case Report
A 65-year-old woman was admitted with diagnosis of severe aortic stenosis (AS). She had a history of breathlessness and palpitations on exertion (NYHA III). Comorbidities included hypertension and diabetes mellitus on treatment. Transthoracic echocardiography (TEE) revealed severe calcific AS, BAV, an aortic valve area of 0.6 cm² and mean gradient of 42 mmHg, Grade I left ventricular diastolic dysfunction with increased end-diastolic pressure, mild tricuspid regurgitation, and minimal pericardial effusion. Coronary angiogram showed normal coronaries. Other biochemical and hematological investigations were normal.

The patient was scheduled for AV replacement. Patient’s induction and intubation were uneventful. There was difficulty encountered during CVC through the right internal jugular vein (IJV) as the guidewire could not be advanced beyond an acceptable length. Finally, the external jugular vein was cannulated with ease of passage of guidewire and catheter.

On TEE examination, the coronary sinus was found to be dilated, more than 1 cm in diameter. The injected drugs through the CVC catheter appeared in the right atrium (right atrium) through the coronary sinus.

During surgery, the innominate vein was found to be absent. The CVC catheter was seen at the termination of the vein proceeding toward the left side. This was checked with slight withdrawal of 1.5 cm, which brought the catheter tip into view. RSVC was found to be absent and a large LSVC was found draining into the coronary sinus. No other congenital cardiac abnormality was detected.

The inferior vena cava and RA were cannulated. After antegrade cardioplegia, aorta was opened. The AV was bicuspid and heavily calcified. It was excised and replaced with 21 mm Edwards Perimount Magna Ease valve.

Postoperatively, she remained hemodynamically stable. Postbypass, there was a problem to measure the central venous pressure. A separate cannula was easily placed in the left IJV.

She made an uneventful recovery and was discharged on the 5th postoperative day.

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Chest X-ray [Figure 1] (retrospectively) showed a vertical structure in the left mediastinum, most likely the persistent LSVC. Venography through both the catheters in situ confirmed the presence of persistent LSVC and absent RSVC [Figures 2 and 3].

Discussion

Persistence of LSVC occurs in approximately 0.3% to 0.5% of the general population.[1] Its incidence is 10-fold higher in patients with congenital heart malformation. PLSVC with absent RSVC occurs in only 0.09%–0.13% of patients with congenital heart defects.[2-4] Previously, only two cases have been reported of BAV with PLSVC and absent RSVC.[5,6]

Diagnosis is to be suspected if the X-ray of the chest does not show a clear-cut right upper cardiac border. In addition, it may be suspected on TTE if the LSVC is large and drains into a dilated coronary sinus. There is generally no difficulty in performing AV surgery. However, any procedure requiring opening of RA/right ventricle may require cannulation of LSVC. Hemodynamically, there is generally no abnormality. It may pose a problem with placement of a CVC. Inability to deliver retrograde cardioplegia can be a problem because of large size LSVC draining into the coronary sinus.

On suspicion of this anomaly, the available diagnostic modalities are TEE (injecting agitated saline or contrast in the left antecubital vein), contrast upper venous digital subtraction cavography, or computed tomography/magnetic resonance imaging. An absent RSVC can lead to inability to insert central venous cannula, pulmonary artery catheter, and transvenous pacing leads through the right internal jugular or subclavian vein.

Conclusion

Absent RSVC with a PLSVC in the patient of AS with BAV is an extremely rare anomaly and has several clinical and operative implications. Anticipation and proper imaging is important to avoid any complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.
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