Transcatheter Closure of an Atrial Septal Defect with Single Coronary Artery and Retro-Aortic Right Coronary Artery

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Isolated single coronary artery in the presence of an atrial septal defect (ASD) is a rare congenital anomaly. There are no clear guidelines regarding the management of ASD in these patients. Transcatheter closure of the ASD in the presence of a single coronary artery that has a retro-aortic course is a matter of concern because of the possible risk of coronary impingement by the left atrial disc. Here we report the first case of an ASD device closure in a patient with a retro-aortic right coronary artery. The immediate results were good; however, long-term follow up is warranted to look for late complications.

Case presentation

A 56-year-old man presented with a history of class 2 exertional dyspnoea and a recent history of bacterial pneumonia. The patient had a past history of left lung lobectomy. On clinical examination, a 2/6 systolic murmur was present in the second left intercostal area. The ECG showed right bundle branch block. Transthoracic echocardiography showed an ostium secundum ASD with a pulmonary to systemic flow ratio (Qp:Qs) of 2.5. Right ventricular systolic pressure was 25 mmHg. Prior to planned ASD closure, transoesophageal echocardiography (TOE) was performed. The anatomy of the interatrial septum was considered suitable for ASD device closure. A coronary angiogram performed before ASD device closure showed a single coronary artery arising from the left sinus with the RCA having a retro-aortic course (type L 2 P; Figure 1A,B). The patient was not a good surgical candidate because of his history of pulmonary lobectomy. In view of the non-dominant RCA, we decided to close the ASD, and a 24 mm Cocoon ASD occluder (Vascular Concepts Limited, Deans Hall Business Park, Halstead, Essex) was deployed under TOE and fluoroscopy guidance with the utmost care to avoid impingement on the RCA. The patient tolerated the procedure well, with no evidence of electrical or haemodynamic instability. The post-procedure CT coronary angiogram showed a minimum distance of 3.4 mm between the left atrial disc and the retro-aortic RCA (Figures 2A, 2B, 2C).

Discussion

Coronary artery anomalies occur in 0.3-0.9% of the population without structur-
al heart defects and in 3-6% of those with congenital heart defects. Isolated single coronary artery (SCA) is a rare congenital anomaly occurring in approximately 0.024% of the population, and ASD with SCA is even rarer. ASD device closure in patients who have an SCA with a retro-aortic course is a matter of concern, given the risk of post-interventional fatalities due to the anatomical proximity of the left atrial disc and the anomalous coronary vessel. To avoid complications, these cases are normally referred for surgical closure. However with careful imaging during the procedure, an ASD can be closed with a device, provided that imaging modalities show enough space between the anomalous artery and the left atrial disc. A pre-procedure CT angiogram can also help by establishing the three-dimensional anatomical relationship between the ASD and the anomalous artery.

Figure 1. A. Coronary angiogram showing a single coronary artery from the left sinus. B. CT coronary angiogram showing the retro-aortic course of the right coronary artery. LAD – left anterior descending; LCx – left circumflex; RCA – right coronary artery.

Figure 2. A. Post-procedure CT image showing closely related atrial septal defect (ASD) device and the right coronary artery (RCA). B. CT curved planar image revealing the minimum distance of 3.4 mm between the left atrial disc and the RCA. C. CT reconstruction image showing the three-dimensional anatomical relationship between the ASD device and the anomalous RCA.
We have very little information about ASD device closure among patients with anomalous coronaries. Theoretically, in the case of an anomalous retroaortic coronary artery supplying a large myocardial area, a large ASD device may prove fatal as a result of possible coronary compression. A few case reports have shown concern about the increased cardiac risk in patients who have an ASD in combination with an anomalous coronary artery. However, these patients can be managed with small devices if careful imaging by TOE or selective coronary angiography is used during the procedure. The myocardial area at risk, supplied by the anomalous artery, is an equally important factor that must be considered before taking a decision. We have reported a case of SCA with ASD device closure that showed good immediate results. However this patient needs to be followed up to determine the long-term outcome.

References