Rare coronary anomaly coexisting with atrial septal defect: contraindication to Amplatzer occluder implantation

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A 39-year-old man presented with a history of gradually progressing exertional dyspnoea. He had 2/6 systolic murmur in the second left intercostal space. ECG revealed right bundle branch block. Transthoracic echocardiography showed ostium secundum atrial septal defect (ASD) with pulmonary to systemic flow ratio of 2.5. Right ventricular systolic pressure was 47 mmHg. Prior to planned ASD closure with an Amplatzer occluder transoesophageal echocardiography (TEE) was performed (Panels A and B).

TEE confirmed the presence of ASD II. The size of the defect assessed with 3D reconstruction was 2 × 3 cm. The anatomy of interatrial septum was considered suitable for Amplatzer implantation. However, atypical linear echo-free space was noted by aortic root next to the ASD. Detailed evaluation revealed that it was most probably anomalous right coronary artery (RCA) originating from left circumflex artery (LCx) and surrounding the aortic root between the non-coronary sinus of Valsalva, the atria, and interatrial septum (Panel A).

Anomalous origin of the RCA from proximal segment of LCx was confirmed with 64-slice multislice computed tomography (MSCT) (Panel C). RCA was coursing very close to the ASD (Panel D).

Coronary angiography is not routinely performed prior to ASD closure in young persons without history of coronary artery disease. If the coronary anomaly was missed in the TEE study, the anomalous RCA could be compressed by the Amplatzer occluder, causing myocardial ischaemia or even infarction. Such complications were previously described in the literature. The patient was referred for surgical ASD closure. The surgeon was informed on the course of RCA and safely placed the sutures leaving the artery intact.

In conclusion, physicians performing TEE evaluation in ASD patients should be aware of this very rare but potentially dangerous coronary anomaly. In our opinion, in such a case, surgical strategy of ASD closure should be recommended.

Panel A. TEE showing the abnormal linear echo-free structure (RCA) located very close to the ASD.
Panel B. Three-dimensional TEE reconstruction. View from the left atrium (LA). Interatrial septum with large ASD is seen.
Panel C. MSCT. Reconstruction of the coronary arteries confirms abnormal RCA origin from the left coronary artery (LCA).
Panel D. MSCT slice showing RCA located close to the ASD.