Successful treatment of an aneurysmatic aorta-right atrial tunnel by vascular plug: a very rare case

Anevrizmatik aorta-sağ atriyal tünelin vasküler plakla başarılı tedavisi: Çok nadir görülen bir olgu

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A 57-year-old female patient was admitted to the clinic with dyspnea. She was found to have a grade 3/6 continuous murmur over the right parasternal area. Transthoracic echocardiography revealed a tunnel stemming from the right sinus of Valsalva (RSV) and draining into the right atrium (Figures 1A-D). Pulmonary artery pressure and left-to-right shunt ratio were 65 mmHg and 2.2 respectively. Coronary and computed tomography angiography showed a giant aneurysmatic tunnel stemming from the aorta (Figures E, F and Video 1*), and also revealed the right coronary artery (RCA) originating from the proximal segment of the tunnel and the normal nature of the other coronary arteries (Figure G-J and Video 1*). The anomaly was successfully treated by closing the middle of the tunnel using an Amplatzer vascular plug II (12x9 mm) (Figures J-L and Video 2*). Aorta-right atrial tunnel (ARAT) is an abnormal tubular connection between the ascending aorta and right atrium, and is a very rare congenital cardiac anomaly leading to the left-to-right shunt. ARAT generally originates from the left sinus of Valsalva, and only very rarely from the right. It should be treated due to the risk of cardiac failure, coronary steal, aneurysm formation or rupture, and endocarditis. Surgical closure of the tunnel is usually suggested. However, if there are no other cardiac anomalies and the RCA does not arise from the tunnel, trans-catheter techniques offer an alternative procedure. A few cases with ARAT arising from the right sinus of Valsalva have been reported in the literature. They were treated surgically, except for a 10-year-old patient treated with coil occlusion and a 4-year-old treated using an Amplatzer Duct Occluder II. However, in these two cases the RCA stemmed from the aorta, not from the tunnel. In the case we report, the ARAT arose from the right sinus of Valsalva, and the RCA originated from the proximal segment of the tunnel. The patient was successfully treated by placing a vascular plug in the middle of the ARAT. The case suggests that in the diagnosis of continuous murmur and left-to-right shunts, ARAT should be considered, and therefore it should be carefully screened. In addition, the catheter-based treatment of ARAT should be given priority over surgery in appropriate cases.

Figures– Transthoracic echocardiography **(A, B)** showing the proximal portion of the tunnel arising from the right sinus of Valsalva on the parasternal long-axis view (with and without color Doppler), **(C, D)** showing the distal portion of the tunnel draining into the RA on the apical four-chamber view (with and without color Doppler). Ao: Aorta; LA: Left atrium; LV: Left ventricle; RA: Right atrium; RV: Right ventricle. Multi-detector computed tomography **(E)** demonstrating the giant tunnel arising from the RSV **(F)** three-dimensional reconstruction (volume rendering technique) demonstrating the extra-cardiac course of the giant and tortuous tunnel and distal RCA. Ao: Aorta; LCx: Left circumflex artery; LSV: Left sinus of Valsalva; NCSV: Non-coronary sinus of Valsalva; RCA: Right coronary artery; RSV: Right sinus of Valsalva. Coronary angiography displaying **(G)** normal left coronary arteries, **(H-J)** the ARAT, non-occluded RCA originating from it and drainage of the tunnel into RA, **(K)** the vascular plug and **(L)** complete tunnel closure after release of the vascular plug. ARAT: Aorta-right atrial tunnel; LAD: Left anterior descending artery; LCx: Left circumflex artery; RA: Right atrium; RCA: Right coronary artery. *Supplementary video files associated with this presentation can be found in the online version of the journal.