Incidental Detection of Coronary Sinus Agenesis in Two Patients with Surgically Repaired Atrial Septal Defect

Cerrahi Olarak Onarılan Atriyal Septal Defektli İki Hastada Rastlantsal Koroner Sinüs Agenezisi Tespiti

Case 1
A 42-year-old female patient, who underwent surgical atrial septal defect (ASD) repair in 1984, was admitted with exertional angina persisting for six months. Her family history was notable for premature coronary artery disease (CAD). She had been taking metoprolol 25 mg/day due to palpitations for three years. Physical examination revealed normal findings. An electrocardiogram (ECG) showed sinus rhythm and T wave inversion in the anterolateral derivations. Echocardiography was unremarkable. Coronary computerized tomography angiography (CCTA) ruled out CAD. However, an incidental finding of coronary sinus (CS) agenesis was detected, with the great cardiac vein and marginal vein joining in the posterior atrioventricular (AV) groove and draining into the lateral left atrium (LA) (Figure 1A–D). She was diagnosed with vasospastic angina, and metoprolol was replaced with diltiazem treatment.

Case 2
A 51-year-old female patient, who underwent surgical ASD repair in 2001, was admitted to the emergency room with acute dyspnea. Her past medical history included diabetes for five years, dual-chamber pacemaker implantation for symptomatic tri-fascicular block in 2021, and mitral valve replacement for severe rheumatic mitral regurgitation in 2022. Physical examination, ECG, echocardiography, and pacemaker interrogation revealed no abnormalities. Despite a positive high-sensitive troponin value, triple-rule-out computed tomography angiography (CTA) ruled out CAD. However, an incidental finding of CS agenesis was detected, with all coronary veins draining into the LA separately (Figure 1E). Furthermore, a remnant of a persistent left superior vena cava (PLSVC) connected to the lateral LA was observed. She was diagnosed with atypical pneumonia and discharged with oral antibiotic treatment.

Congenital CS anomalies are rare, and therefore they have attracted less attention. CS agenesis is frequently associated with a PLSVC, an ASD, and other additional anomalies. It usually presents with a right-to-left shunt at the LA level as part of the complex functional abnormality. Moreover, it is important to consider in CS-related procedures, including cardiac resynchronization therapy, and diagnostic/therapeutic electrophysiological procedures. Operators should consider the possibility of CS agenesis in cases with a history of ASD closure and difficulty in CS cannulation. Herein, we presented two cases of CS agenesis in patients who had undergone surgical ASD repair.

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Figure 1. The coronary sinus (CS) was not observed at the atrioventricular (AV) groove in axial and sagittal planes and 3-dimensional volume rendering views of the cardiac computerized tomography angiography. The great cardiac vein and marginal vein were joining in the posterior AV groove and draining into the lateral left atrium (LA) (white arrows) (A–D). In case 2, a 3-dimensional volume rendering view showed that all coronary veins were draining into the LA separately because of the CS agenesis (white arrows) (Figure 1E).