CLINICAL VIGNETTE

Hypertrophic obstructive cardiomyopathy with anomalous left circumflex coronary artery

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Hypertrophic obstructive cardiomyopathy (HOCM) with resting or provocable left ventricular outflow tract (LVOT) gradient is observed in approximately 70% of all patients with hypertrophic cardiomyopathy (HCM). Anomalous aortic origin (AOO) of coronary arteries is frequently an accidental finding on coronary angiography or computed tomography angiography (CTA). Both HCM and coronary artery with AOO are common cardiac anomalies (0.1%-0.2% and 0.1%-0.7%, respectively). Nevertheless, only casuistic reports (around 10) have presented the coexistence of HCM and anomalous left coronary artery (LCA) [1] or right coronary artery (RCA) [2] originating from opposite Valsalva's sinuses or the left main stem originating from the pulmonary trunk [3]. What is clinically important, both HCM with significant LVOT obstruction and anomalous course of the coronary artery with exercise-inducible ischaemia (especially LCA coursing between the aorta and pulmonary trunk) may separately increase the risk of sudden cardiac death. A 61-year-old female HOCM patient in functional class II/III according to the New York Heart Association was admitted for further evaluation and treatment. Transthoracic echocardiography (TTE) revealed left ventricular hypertrophy up to 20 mm in the sub-aortic segment of the interventricular septum and systolic anterior motion (SAM) of the anterior mitral leaflet. Mitral regurgitation was assessed as moderate in TTE and as significant in cardiac magnetic resonance. Maximal resting LVOT gradient was 100 to 116 mmHg. Preoperative coronary angiography showed a large left circumflex coronary artery (LČx) originating from RCA ostium and single left anterior descending coronary artery with non-significant coronary irregularities (Fig. 1A, B). Invasive measurement of LVOT gradient was 150 to 200 mmHg. Successful mitral valve replacement with mechanical prosthesis together with LVOT myectomy was performed. CTA done six years after surgery for exclusion of pulmonary embolism confirmed the presence of AAO-LCx with retroaortic course (Fig. 2A, B). To the best of our knowledge, only one report showed coexistence of HCM with AAO-LCx [4]. While both cases shared the same type of anomalies, some parameters distinguished our female patient from the previously reported male one. Sex differences were raised previously, with 60% to 90% male predominance across HCM-population studies [5]. Also, thicker interventricular septum (20 mm vs. 14 mm) and the presence of SAM resulted in higher LVOT gradient (100-116 mmHg vs. 70 mmHg) in our patient. Of



Figure 1. A. Angiography of the right coronary artery with anomalous take-off of left the circumflex coronary artery (arrow); B. Single left anterior descending coronary artery with non-significant irregularities



Figure 2. A. Computed tomography angiographic image showing anomalous left circumflex coronary artery (arrow; posterior-superior view); **B.** Three-dimensional volume-rendered reconstruction — superior view

note, midventricular obstruction dominated in the previous report, whereas in our patient mainly LVOT obstruction was present. Thus, corrective surgery was the treatment of choice in our patient (and not in the previously reported patient). Surgical correction of AAO-LCx is usually not needed or controversial (as opposed to when AAO-LCA is significantly compressed between the aorta and pulmonary trunk), unless clear compromise of the artery is present. The choice of interventional treatment type is discussed elsewhere. Briefly, the presence of full, prolonged SAM is important for a decision of mitral valve repair or replacement. In conclusion, this report adds to the very limited literature on HCM coexisting with coronary anomalies.

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