Hypertrophic obstructive cardiomyopathy and cor triatriatum sinistrum. A casuistic coexistence

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Hypertrophic cardiomyopathy (HCM) is characterized by increased thickness of the left ventricular myocardium, not explained by abnormal loading conditions. Up to 70% of HCM patients have either resting or easily provoked left ventricle outflow tract (LVOT) obstruction-hypertrophic obstructive cardiomyopathy (HOCM).

Cor triatriatum sinistrum (CTS) is a rare congenital anomaly in which the left atrium (LA) is divided into proximal and distal chambers by a fibromuscular membrane. It constitutes only 0.1% to 0.4% of congenital heart defects. CTS is frequently associated with the tetralogy of Fallot, septal defects, or anomalous pulmonary vein return. Nonetheless, a coexistence of HCM and CTS is a casuistic finding. Pellaton et al. [1] most probably described the first HCM case with CTS. Bahlmann et al. [2] reported on CTS and apical-HCM. Uemura et al. [3] reported on the prenatal diagnosis of the Costello syndrome expressed by CTS and HCM. Finally, Tatco [4] presented cardiac magnetic resonance (CMR) images of a female adult with both anomalies.

A 59-year-old male HOCM-patient with metabolic syndrome, atrial fibrillation, and chronic heart failure, after implantation of implantable cardioverter-defibrillator for secondary prevention of sudden cardiac death, was admitted for further treatment. Both transthoracic echocardiography (TTE) and CMR showed asymmetrical hypertrophy of the interventricular septum (25 mm). The systolic anterior motion of the mitral leaflets with moderate mitral regurgitation was visualized. Maximal LVOT-gradient was 80–90 mm Hg. Moreover, right ventricular systolic pressure was elevated to 57 mm Hg. The systolic function of both ventricles was preserved. Assessment of the enlarged LA by TTE and CMR was somehow ambiguous (Figure 1), thus chest computed tomography was done and showed non-restrictive CTS: LA was divided by a fibrous and fibromuscular membrane with duplication of the right part of the membrane. Wide communication between proximal and distal LA chambers was present in the lower LA part. All four pulmonary veins drained into the proximal LA chamber. Alcohol septal ablation was considered for the reduction of LVOT obstruction. However, inappropriate anatomy of the septal branch precluded alcohol septal ablation. Surgical treatment was considered, however, due to increased perioperative risk (body mass index [BMI], 36.1 kg/m^2) conservative treatment strategy was chosen.

TTE is a first choice tool for diagnosis of CTS (providing satisfactory imaging quality) and diagnosis of CTS in adulthood is rarer. A systematic review revealed that the median age of diagnosis was 43 years [5], as opposed to 59 years of our patient. Multimodality imaging is advisable in suspicious LA appearance and recommended in HCM patients. Next, LVOT obstruction (which was visible in our patient) was most probably reported only once [5] among the HCM-CTS population.

Finally, CTS may lead to the obstruction of LA flow and create pulmonary hypertension (PH). The pathophysiology is similar to that of mitral stenosis namely, PH may result from backward transmission of the increased LA pressure. In the context of our patient no significant gradient across the LA membrane was observed (Supplementary material, *Figure S1*). Thus, PH is likely to be post-capillary due to significant LVOT obstruction, mitral regurgitation, diastolic dysfunction, or their combination. Nonetheless, the method of choice to definitely differentiate between the pulmonary artery hypertension and post-capillary PH is the right heart catheterization (not performed in our patient).

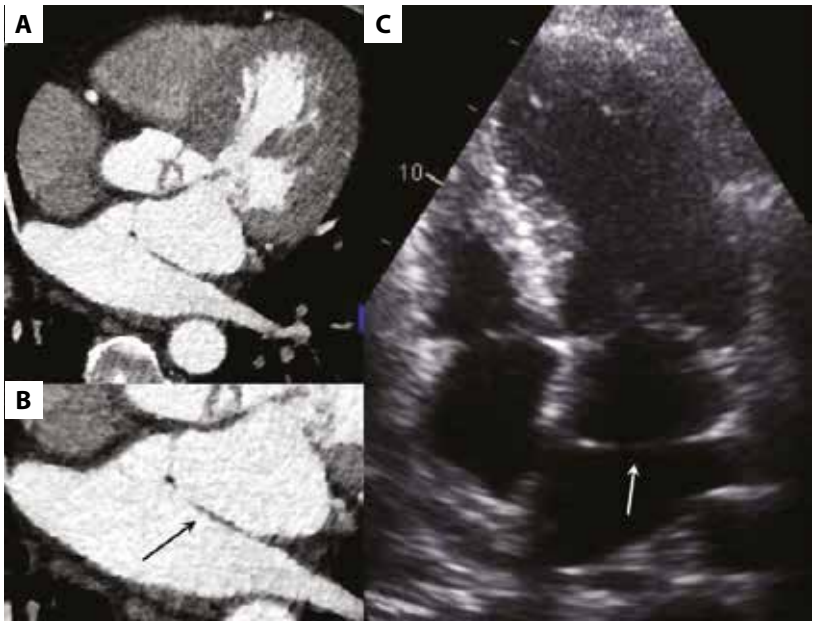

Figure 1. A. Computed tomography angiography. **B.** Magnification of the panel "A" with focus on the left atrium — white arrow indicates the fibromuscular membrane. **C.** Transthoracic echocardiography — apical view. The white arrow shows the linear echogenic structure across the left atrium

In conclusion, this report adds to the very limited literature on HOCM coexisting with CTS.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska.

Article information

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