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A casuistic coexistence

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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 provicable 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 the fibro-muscular membrane. It constitutes only 0.1% to 0.4% of congenital heart defects [1]. CTS is frequently associated by tetralogy of Fallot, septal defects or anomalous pulmonary vein return. Nonetheless, a coexistence of HCM and CTS is a casuistic finding. Pellaton et al. [2] most probably described the first HCM-case with
CTS. Bahlmann et al. [3] reported on CTS and apical-HCM. Uemura et al. [4] reported on the prenatal diagnosis of Costello syndrome expressed by CTS and HCM. Finally, Tatco [5] presented cardiac magnetic resonance (CMR) images of a female adult with these 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). Systolic anterior motion of the mitral leaflets with moderate mitral regurgitation were visualized. Maximal LVOT-gradient was 80–90 mm Hg. Moreover, right ventricular systolic pressure was elevated to 57 mm Hg. 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 (BMI 36.1 kg/m²) conservative treatment strategy was chosen.

TTE is a first choice tool for diagnosis of CTS (providing satisfactory imaging quality) and diagnosis of CTS in the adulthood is rarer. A systematic review revealed that the median age of diagnosis was 43 years [6], as opposite 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 ones [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, 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).

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

REFERENCES


**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. White arrow shows the linear echogenic structure across the left atrium.