Hypertrophic cardiomyopathy is a primary genetic heart disorder with variable phenotype that involves myocardial thickening and obstruction of the ventricular outflow tract. In contrast to left ventricular pathology, the involvement of the right ventricle is uncommon, occurring in up to 15% of patients. Histological findings appear to be similar to those in the left ventricle, suggesting similar pathogenesis, but the rarer right-side flow obstruction may result in more severe symptoms. Occasionally, predominant right ventricular disease can be seen.

A 52-year-old male with a history of alcohol and psychotropic drug abuse was referred to our department with New York Heart Association class III symptoms of heart failure after a resuscitated cardiac arrest. An ECG showed left atrial abnormality, left axis deviation, and left ventricular strain pattern (Figure I). Transesophageal echocardiogram revealed marked symmetric left ventricular hypertrophy (diastolic thickness 21 to 25 mm) without signs of outflow tract obstruction. Normal contractile function of both ventricles was found (left ventricular ejection fraction 61%). A significant thickening of the right ventricular free wall was present (6 to 7 mm in diastole) with an unusual thickened moderator band and turbulent flow in the right ventricular outflow tract. Doppler echocardiography suggested a significant mid-right ventricular pressure gradient (Figure 2, top left and top right; Movie I). Measurements of gradient were imprecise because of the suboptimal angle between flow direction and ultrasound beam. More detailed imaging of the right ventricular outflow tract was performed during transesophageal echocardiography. Irregular thickening of the right ventricular free wall was most pronounced in the subvalvular region, with the peak systolic mid-ventricular gradient estimated at 80 mm Hg and the mean at 40 mm Hg (Figure 2, bottom right and bottom left; Movie II). Because of directional Doppler limitations, however, cardiac magnetic resonance imaging was performed to specify right ventricular abnormalities. Cardiac magnetic resonance imaging confirmed marked hypertrophy of both the left and right ventricles (maximum 23 to 24 mm in anterior and septal segments of the left ventricular wall) without a significant left outflow tract obstruction (left ventricular outflow tract diameter 15.5 mm) (Figure 3, top left and top right; Movies III through and V). Delayed enhancement imaging after gadopentetate dimeglumine administration revealed no abnormal findings. A subvalvular narrowing of the right ventricular outflow tract had a minimal area of 60 to 65 mm² (Figure 3, bottom right). Mean peak and maximal flow velocities in the subvalvular narrowing of the right ventricular outflow tract were estimated at 210 to 220 cm/s and 530 cm/s, respectively, with a corresponding maximal gradient of 112 mm Hg (Figure 3, bottom left).

The patient received beta-adrenolytic medical therapy and remains in New York Heart Association class II/III. He has been scheduled for surgical consultation regarding the possibility of right ventricular myectomy; cardiac transplantation remains an option if his heart failure becomes aggravated.

Disclosures

None.

References


From the Department of Rapid Cardiac Diagnostics, Medical University of Lodz, Biegianski Hospital, Lodz, Poland (R.K., P.L., L.C., J.D., M.K.-P., J.D.K.); Department of Nuclear Medicine, I Chair and Department of Cardiology, Medical University of Warsaw, Warsaw, Poland (D.P.-K.); and Department of Nuclear Medicine, Medical University of Warsaw, Warsaw, Poland (L.K.).

The online-only Data Supplement, which contains Movies I through V, can be found at http://circ.ahajournals.org/cgi/content/full/116/23/e551/DC1.

Correspondence to Radoslaw Krecki, MD, II Chair of Cardiology, Medical University of Lodz, Bialasiewicza 1/5, 91–347 Lodz, Poland. E-mail rkrecki@gazeta.pl

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Figure 1. Twelve-lead ECG on admission showing pathological left-axis deviation, ST depression, P mitrale, and T-wave inversion in leads with a predominant R wave, suggesting left ventricular hypertrophy.

Figure 2. Top left, Transthoracic echocardiography. Modified apical 4-chamber view showing pronounced hypertrophy of left and right ventricular muscle. Top right, Transthoracic echocardiography. Modified parasternal short-axis views with color Doppler flow mapping indicate the presence of turbulent flow in the right ventricular outflow tract (arrows). Inset, Continuous-wave Doppler confirms a significant systolic pressure gradient in the right ventricular outflow tract, potentially underestimated because of the high incident angle of the ultrasound beam. Bottom left, Transesophageal echocardiogram showing a subvalvular narrowing of the right ventricular outflow tract (arrow). Bottom right, Transesophageal echocardiogram (in a view corresponding with bottom left panel) with color Doppler flow mapping indicates the presence of turbulent flow in the right ventricular outflow tract (arrow). Angle-corrected measurement of the right ventricular flow obtained with pulsed-wave Doppler reveals a peak systolic gradient of 80 mm Hg and mean gradient of 40 mm Hg. LV indicates left ventricle; RV, right ventricle; RA, right atrium; LA, left atrium; Ao, aorta; AoV, aortic valve; and PA, pulmonary artery.
Figure 3. Magnetic resonance imaging of the heart. Top left, A sagittal section through the right ventricular outflow tract in late systole. A subvalvular narrowing of the right outflow tract (arrow) results from the hypertrophy of the interventricular septum and right ventricular free wall. Bottom left, Mean flow velocity (210 to 220 cm/s) within the subvalvular narrowing of the right ventricular outflow tract over the cardiac cycle measured using phase contrast-flow magnetic resonance imaging technique. Top right, A transversal section through the right ventricular outflow tract in late systole. Demonstration of the obstruction of the right ventricular outflow tract with a dark, high-velocity jet of blood passing through the stenosis into the pulmonary artery (arrow). Bottom right, Magnetic resonance estimation of cross-sectional area of the right ventricular outflow tract over the cardiac cycle (minimal area 60 to 65 mm²).
Predominant, Severe Right Ventricular Outflow Tract Obstruction in Hypertrophic Cardiomyopathy

R. Krecki, P. Lipiec, D. Piotrowska-Kownacka, L. Chrzanowski, L. Krolicki, J. Drozdz, M. Krzeminska-Pakula and J.D. Kasprzak

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