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Multimodality imaging of right ventricular outflow tract obstruction in hypertrophic cardiomyopathy

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SOUHRN

Obstrukce výtokového traktu pravé komory je u hypertrofické kardiomyopatie vzácný nález; obvykle je důsledkem hypertrofie pravé komory, u níž se předpokládá, že je způsobena stejnými mutacemi genu pro kódování srdečních sarkomer vedoucími k fenotypové expresi hypertrofie levé komory. V námi popsaném případě v této kasuistice se nejspíše jednalo o přítomnost subpulmonální membrány současně s hypertrofií pravé komory. Vzájemně se doplňující vyšetření transthorakální echokardiografií, magnetickou rezonancí a výpočetní tomografií umožnilo přesně popsat anatomické, funkční a strukturální charakteristiky levé i pravé komory.

ABSTRACT

Right ventricular outflow obstruction is a rare finding in hypertrophic cardiomyopathy; it is usually due to hypertrophy of the right ventricle, which is considered caused by the same cardiac sarcomere mutations leading to the phenotypical expression of left ventricular hypertrophy. In the case described in the present report, the presence of a sub-pulmonary membrane likely concurred to right ventricular hypertrophy. The complementary use of transthoracic echocardiography, magnetic resonance imaging and computed tomography allowed to precisely evaluate the anatomical, functional and structural characteristics of both the left and right ventricle.

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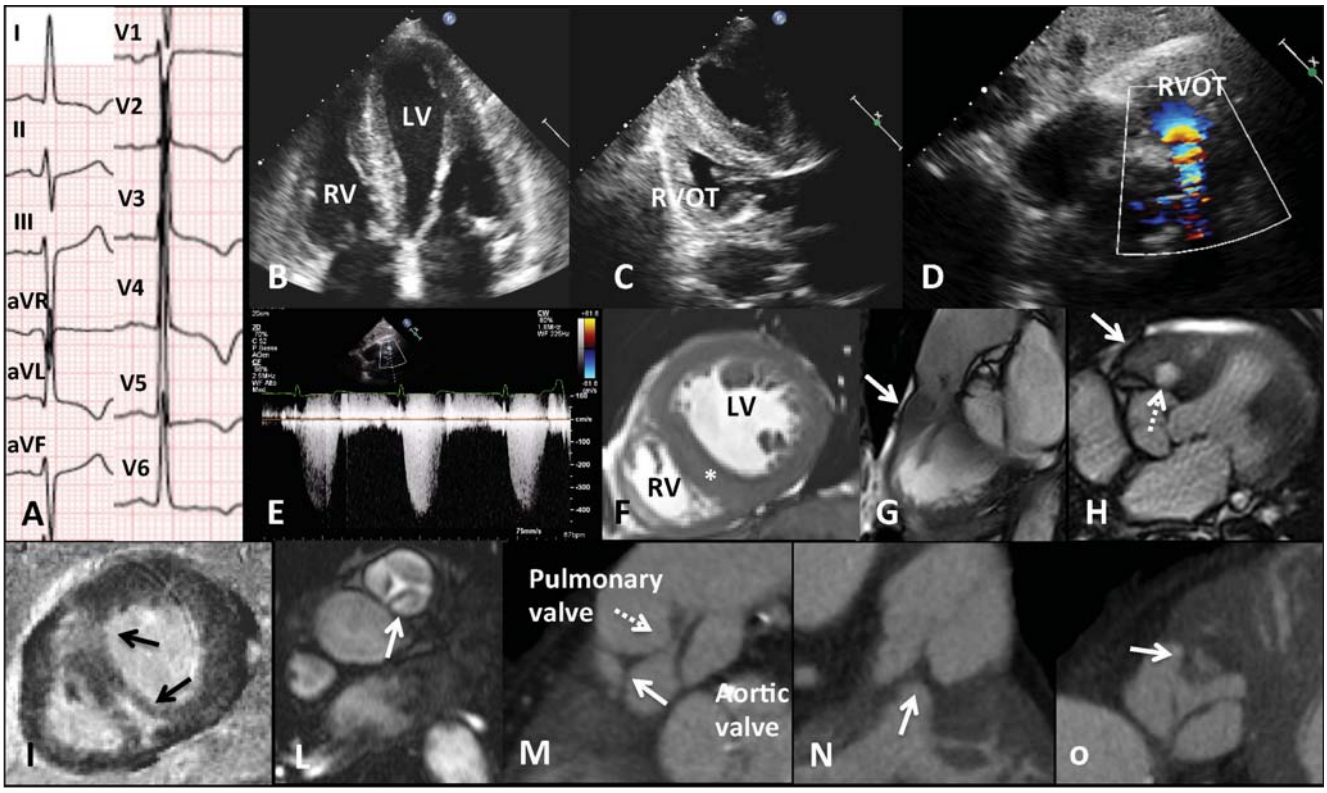

Fig. 1 – Panel A. 12-lead electrocardiogram; panel B–C. Two-dimensional transthoracic echocardiography; panel D. Color-Doppler echocardiography; panel E. Continuous-wave Doppler echocardiography; Panel F–L. Cardiac magnetic resonance images. Asterisk in Panel F indicates hypertrophy of interventricular septum. Arrows in Panel G and H indicate hypertrophy of RVOT. Dashed arrow in Panel H indicates narrowed RVOT area. Arrows in Panel I indicate patchy foci of late gadolinium enhancement (i.e. myocardial fibrosis) at the insertion points of RV. Arrow in Panel L indicates the pulmonary valve. Panel M–O. Computed tomography images. Arrows in Panel M–O indicate the presence of a membrane connecting the RVOT wall to the anterior cusp of the pulmonary valve. LV – left ventricle; RV – right ventricle; RVOT – right ventricular outflow tract.

A 58-year-old man was referred to our Institution for clinical evaluation before non-cardiac surgery. Familiar and personal medical history was unremarkable except for a systolic heart murmur known since childhood. The ECG showed left axis deviation and left ventricular (LV) hypertrophy pattern (Panel A) in the absence of an obvious cause. Transthoracic echocardiography revealed biventricular hypertrophy (Panels B–C) with predominant involvement of the right ventricle (RV) and interventricular septum (IVS). The thickened walls of the RV outflow tract (RVOT) (Panel C) caused a functional obstruction with a peak gradient of 60 mmHg, as demonstrated by color-Doppler and continuous-wave Doppler echocardiography (Panels D–E). Cine magnetic resonance imaging (MRI) confirmed the hypertrophy of IVS (Panel F, asterisk) and RVOT (Panel G–H, arrow) and demonstrated a significantly narrowed RVOT area (Panel H, dashed arrow). Late gadolinium enhancement imaging revealed patchy foci of myocardial fibrosis at the insertion points of RV (Panel I, arrows). Cine-MRI aroused the suspicion of a membrane-like structure in the RVOT in connection with the pulmonary valve (Movie), which appeared tricuspid (Panel L; arrow). For a better anatomic definition, a computed tomography scan was performed, which confirmed the presence of a membrane connecting the RVOT wall to the anterior cusp of the pulmonary valve (Panel M–N–O; arrow) (Fig. 1).

RVOT obstruction is a rare finding in hypertrophic cardiomyopathy; it is usually due to hypertrophy of the RV, which is considered caused by the same cardiac sarcomere mutations leading to the phenotypical expression of LV hypertrophy. In the present case, the presence of a subpulmonary membrane likely concurred to RV hypertrophy. The complementary use of transthoracic echocardiography, MRI and computed tomography allowed to precisely evaluate the anatomical, functional and structural characteristics of both LV and RV.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.crvasa.2015.05.003.

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