Available online at www.sciencedirect.com

SciVerse ScienceDirect

journal homepage: www.elsevier.com/locate/crvasa

Case Report

Coincidence of bicuspid aortic valve presence and hypertrophic cardiomyopathy, and significance of magnetic resonance in its diagnostics

Branislav Obžut, Peter Blaško, Martin Porzer*

Cardiovascular Departement, University Hospital Ostrava, Czech Republic

ARTICLE INFO

Article history:
Received 18 July 2012
Received in revised form
12 September 2012
Accepted 13 September 2012
Available online 28 September 2012

Keywords:
Hypertrophic cardiomyopathy
Nuclear magnetic resonance
Percutaneous transluminal septal myocardial ablation
Bicuspid aortic valve

ABSTRACT

Hypertrophic cardiomyopathy (HCM) is a complex heart disease with a typical pathophysiological characteristic, and with a wide scale of morphologic, functional, and clinical symptoms. The presented case study describes a case of coincidence of a patient with HCM and bicuspid aortic valve, on which we wanted to point out the benefit of the NMR examination and efficiency of alcohol septal ablation with a positive result in a long-term monitoring.

1. Introduction

Hypertrophic cardiomyopathy (HCM) was first described in detail by London forensic pathologist Donald Teare in 1958, who published pathologic findings of young patients with sudden death. The oncoming decades brought a large number of publications describing HCM from many perspectives and labelling this disease with tens of terms—idiopathic hypertrophic subaortic stenosis, muscular subaortic stenosis, or hypertrophic obstructive cardiomyopathy. In the 1970s came globally into use the term "hypertrophic cardiomyopathy".

Historically, the HCM was defined as a heart disease, characteristic with myocardium hypertrophy with consequent absence of any other disease which would initiate the hypertrophy (aortic stenosis, hypertension, amyloidosis, impacts of excessive sports training, etc.). At present, we know that an important proportion of patients suffer concurrently from intraventricular obstruction. Concerning the occurrence of intraventricular obstruction of bloodstream can be in a simplified way said that 1/3 of patients is without the obstruction, 1/3 has latent obstruction which will develop only during stress, and 1/3 of patients has obstruction even in rest conditions. From the genetic point of view, the HCM is in 60% (which is a limitation of this definition) an autosomal dominant disease with incomplete penetration, caused by mutation of genes coding the cardiac sarcomeric proteins. Morphologic image at macroscopic level is characteristic with myocardium hypertrophy, particularly of the left ventricle, which is asymmetric in majority

*Correspondence to: University Hospital Ostrava, Cardiovascular Departement, 17th Listopadu 1790, 708 52 Ostrava, Czech Republic. Tel.: +42 05973 73216 (Office), +42 07761 80979 (Private).
E-mail address: brano.obzut@post.sk (B. Obžut).

0010-8650/$ - see front matter
http://dx.doi.org/10.1016/j.crvasa.2012.09.004

of patients. Histopathologic characteristics of HCM include myocyte disarray, disrupted myofibrillar architecture, and defect of intercellular connection with fibrosis. Prevalence of this disease is approximately 0.2% and is thus the most frequent hereditary cardiovascular disease—in the Czech Republic there are approx. 20,000 patients with hypertrophic cardiomyopathy, out of which circa 2000–4000 are carriers of the highest risk of sudden cardiac death [1]. Annual mortality in the non-selected population was reported to be 1% [2]. The most important aspect of the novel interpretation of HCM classification should be in the future particularly the prognostic seriousness of a given HCM form.

The clinical progress of the disease is very diverse. Some patients will remain asymptomatic in the course of their life; with some will develop symptoms of cardiac failure or angina pectoris, while with other patients the sudden cardiac death can be the first display of the disease.

2. Case study

In the presented study is described a case of a 47-year-old male, referred to our workplace with diagnosis of a significant aortic stenosis for selective coronary angiography and other tests, and preparation before cardio-surgical solution of the valve defect.

Apart from significant overweight, chronic venous insufficiency, and moderate mixed dyslipidemia, the patient had no important internal anamnesis. The patient has no children and in his family neither this diagnosis was made nor sudden death appeared. The patient used perindopril 10 mg p.o. and metoprolol 50 mg p.o. for about one year. Clinically, the patient showed dominant expressions of cardiac insufficiency in class NYHA III, negated the syncope, stenocardia appeared after a greater stress. ECG examination indicated hypertrophy of left ventricle together with inverted T waves and fixed ST depressions in leads V3-6 and II, III, aVF.

Due to his significant obesity with BMI 45, it was very difficult to examine the patient by transthoracic echocardiogram. In spite of this, there was evident significant hypertrophy of left ventricle with septum thickness 31 mm, the present gradient (gradient at rest)—110 mmHg—was localized in the left ventricle outflow tract (LVOT), not on the aortic valve. As there was suspected presence of bicuspid aortic valve, a trans-oesophageal echocardiography examination was carried out, which confirmed the morphologic finding. Further on was executed NMR examination of heart, confirming the diagnosis of hypertrophic cardiomyopathy with evident SAM (systolic anterior movement) (Fig. 1). From the trans-oesophageal echocardiography, as well as NMR (Fig. 2), was determined the area of the bicuspid aortic valve area (AVA) exceeding 4 cm^2 (4.5 according to NMR) without serious degenerative changes. The ascending aorta was without signs of dilatation.

After eliminating a significant aortic stenosis and confirming the HCM diagnosis we decided to execute percutaneous transluminal septal myocardial ablation (PTSMA). First the patient underwent an coronary angiography—only unimportant wall

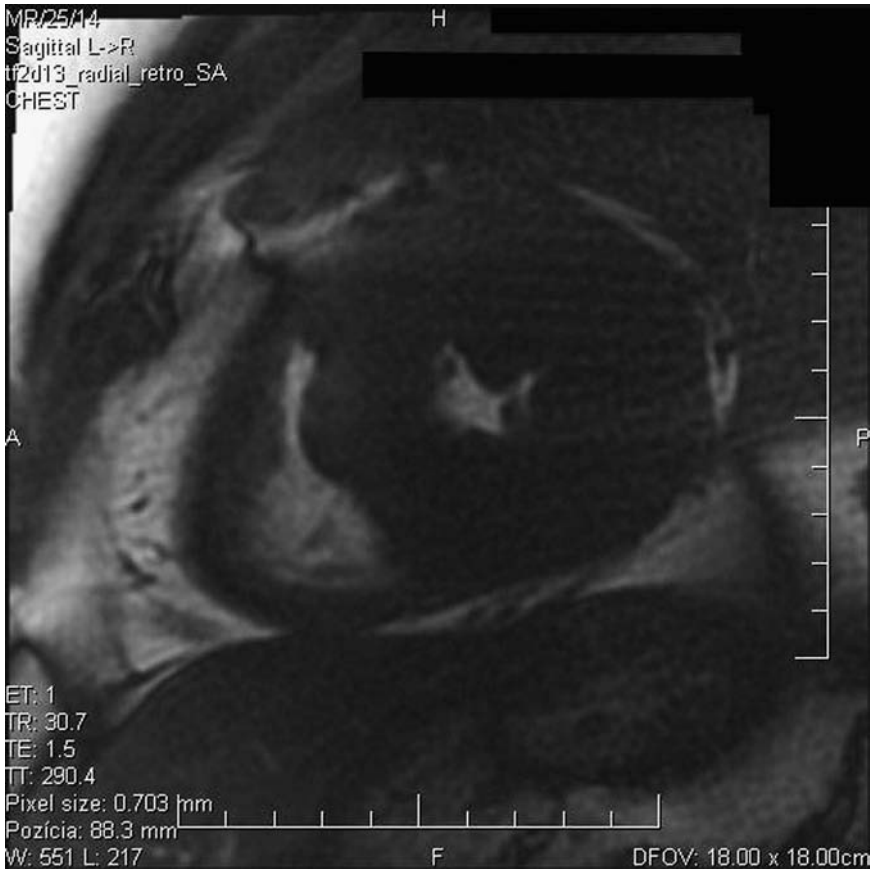

Fig. 1 – NMRI: left ventricle hypertrophy before the percutaneous transluminal septal myocardial ablation (PTSMA) (parasternal long axis equivalent projection).