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STUDIUM PRZYPADKU / *CLINICAL VIGNETTE*

Left ventricular non-compaction associated with hypertrophic cardiomyopathy in the same patient

Niescalenie lewej komory i kardiomiopatia przerostowa u tego samego pacjenta

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A 19-year-old man was referred for lipothymia, episodes of palpitations, and dyspnoea New York Heart Association class II. He had no previous personal medical history; however, his brother experienced sudden cardiac death at the age of 16 years. The physical examination showed a heart rate of 65 bpm and a blood pressure of 130/70 mm Hg; no signs of heart failure were found. Electrocardiogram (ECG) revealed sinus rhythm, incomplete left bundle branch block, and no ST-T segment abnormalities. Two-dimensional echocardiogram showed a dilated left ventricle (LV) with mild systolic dysfunction (ejection fraction [EF] 42%) and hypertrabeculated LV apex, dilated left atrium (area 27 cm^2), and bicuspid aortic valve with mild aortic insufficiency (effective regurgitant orifice 15 mm^2). Cardiac magnetic resonance imaging (MRI) confirmed the impairment of LVEF (37%) and dilatation of the left atrium (Fig. 1B). It also revealed localised mid lateral and basal inferior segmental LV hypertrophy (thickness 16 mm) (Fig. 1A) with a bilayered aspect of the lateral wall, and diastolic ratio of non-compacted-to-compacted myocardial wall of 2.3 (Fig. 2). According to the echo and MRI findings, the diagnosis of LV non-compaction (LVNC) associated with hypertrophic cardiomyopathy (HCM) was considered. Holter ECG monitoring showed the presence of two episodes of non-sustained ventricular tachycardia. Beta-blocker (bisoprolol) and amiodarone were prescribed. Although no arrhythmia was induced at stress test, the HCM sudden cardiac death score was 16.2%. Therefore, an implantable cardioverter defibrillator was indicated. Screening of the relatives was performed; HCM was diagnosed in a first-degree cousin. LVNC is a relatively new disease, first described in 1984 and not yet fully understood. The LV is characterised by multiple trabeculations with deep intertrabecular recesses in communication with the ventricular cavity. LVNC may either be isolated or associated with other cardiac abnormalities such as Ebstein's anomaly, congenitally corrected transposition, bicuspid aortic valve (as in our patient), coronary arteries abnormalities, and septal defects. Both HCM and LVNC are genetically determined and familial diseases with variable but overlapping genetic defects. Indeed, previous literature has mostly reported their separate occurrence in different members of the same family. Recently, mutations in genes encoding sarcomeric proteins, which have previously been implicated in the pathogenesis of HCM, have been identified in patients with LVNC and no LV hypertrophy. This case and a few others reported in literature (especially of apical location) confirm that phenotypic expression of both diseases can occur in the same patient.

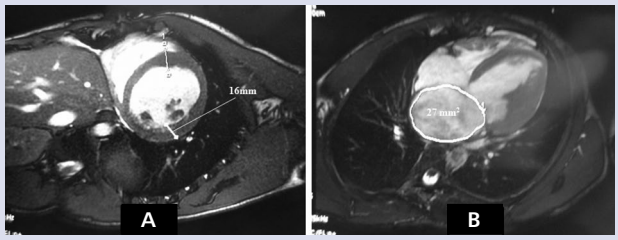

Figure 1. Cardiac magnetic resonance imaging showing a hypertrophic basal inferior segment (**A**) and a dilated left atrium (**B**)

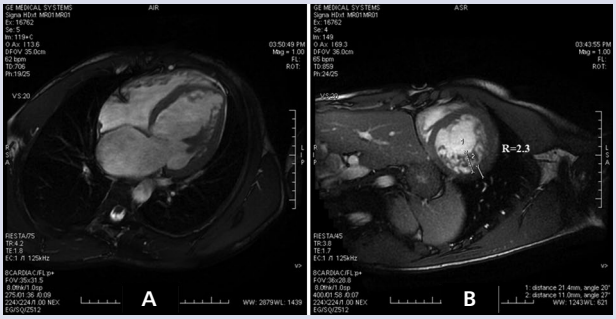

Figure 2. Cardiac magnetic resonance imaging revealing a bilayered aspect of the lateral wall (**A**), with diastolic ratio of non-compacted-to-compacted myocardial wall of 2.3 (**B**)

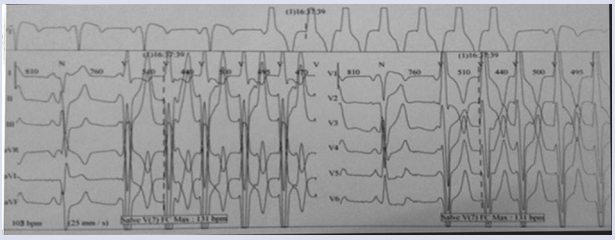

Figure 3. Episode of non-sustained ventricular tachycardia on 24-h Holter electrocardiography recording

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Conflict of interest: *none declared*