Burned out myocardium in biventricular hypertrophic cardiomyopathy presenting with congestive heart failure: Importance of ECG changes

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A 60 year old man was found to have a heart murmur and ECG features of ventricular hypertrophy on a medical check up for military recruitment at age of 20, despite having swimming as the only exercise. His mother had 3 survived children out of 9 pregnancies.

The patient was referred for cardiac investigations at the Umeå university hospital, where a phonocardiogram showed split 2nd heart sound without respiratory variation and a loud holosystolic murmur which peaked in early systole with

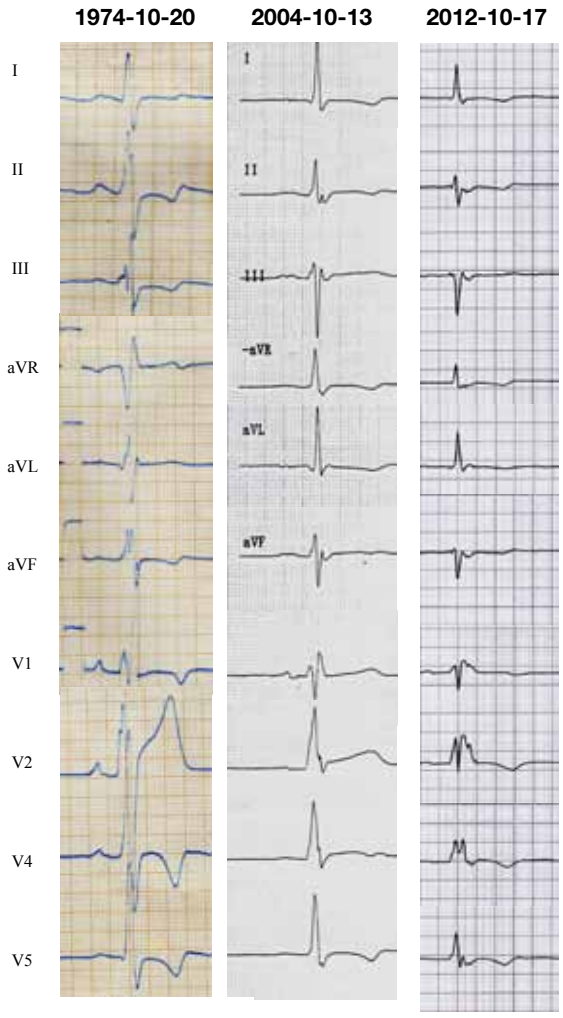

Figure 2: 12 Lead ECG showing progressive QRS broadening and fall of voltage

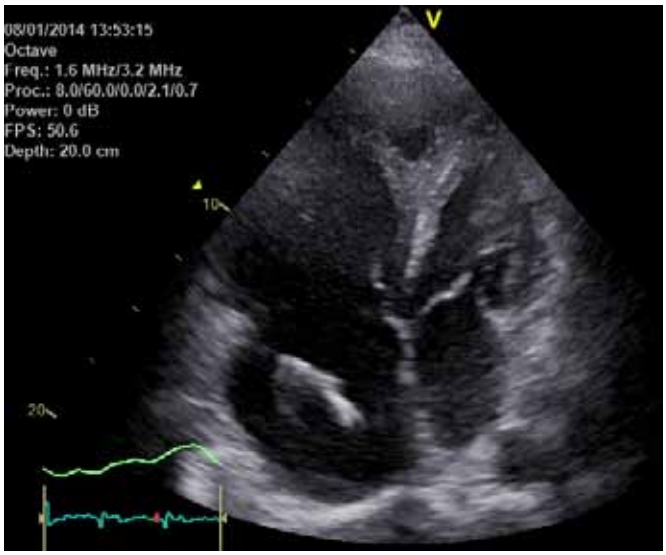

Figure 1: Apical 4 chamber view showing disproportionately enlarged right heart and biventricular burned out myocardium

isoprenaline, but the carotid pulse remained normal. Echocardiography (M-mode) showed a thickened intraventricular septum (16 mm) but no SAM. A heart catheterisation showed raised biventricular end-diastolic pressures: RV 32/13 and LV 104/18 mmHg at rest which slightly increased to 37/17 and 140/18 mmHg, respectively with 80 Watt supine exercise. There was no intraventricular gradient at rest but developed with isoprenaline: LV 143 and aorta pressure dropped to 92 mmHg. The patient was discharged on propranolol 40 mg tds. Four years later, he complained of arrhythmia but 24 hour Holter monitor showed only premature SVE. A repeat biventriculogram revealed a normal size but triangular shaped LV due to extensive apical hypertrophy and concentric hypertrophy of 16 mm. There was also RV hypertrophy with slight obstruction of the outflow tract. Despite the pronounced RV trabeculations and slightly enlarged cavity, EF was maintained as was that of the LV.

Ten years later the patient developed atrial fibrillation, which responded initially to DC cardioversion but eventually required a Maize procedure which kept him in sinus rhythm for 4 years. Pre-op echocardiography showed slightly enlarged left atrium. At age of 50 years he developed non-sustained ventricular tachycardia and received ICD implantation in addition to

amiodarone and flecainide. An Echocardiogram showed insignificant LVOT obstruction and a coronary angiogram confirmed normal arteries. Eight years later the patient developed moderate mitral regurgitation associated with regressed LV hypertrophy to 12 mm, but he denied symptoms. Two years later he was admitted with fluid retention and clear signs of right ventricular failure. At that stage ECG voltage fell from 33 to 6 mV and its duration prolonged from 120 to 160 ms (V4)

Discussion

Dilated-hypokinetic evolution of hypertrophic cardiomyopathy is a rare development and is known to be related to early phenotypic presentation of the disease and in those with family history of sudden death*. Our patient fulfils these criteria having been diagnosed with HCM at the age of 20 then soon developed manifestations of stiff LV and recurrent atrial fibrillation, in addition to the family history of terminated pregnancies or early infant deaths in 6 family members. The natural history of this condition 'burned out myocardium' is less promising compared to conventional HCM, with shorter cardiac events related survival. Indeed, our patient developed ventricular tachycardia at age 50, consistent with arrhythmogenic myocardium, which required ICD. In addition to the arrhythmia complications the patient recently presented with congestive heart failure, with moderate mitral and tricuspid regurgitation and particularly enlarged right heart with clear signs of regressed hypertrophy. The 12 lead ECG, showed significant reconfiguration of the depolarisation pattern; regressed voltage criteria and delayed depolarisation. Thus, this case highlights the need for regular follow up of HCM patients, not only for detecting signs of LV outflow tract obstruction but also, early signs of deterioration of ventricular structure and function and left atrial enlargement which eventually would result in intractable arrhythmia. Also, progressive ECG changes in such case, could be potentially used as a monitoring tool. Finally, in the absence of evidence based recommendation for managing such condition a CRT treatment could optimise ventricular pumping capacity and reduce the substrates of systemic congestion.

** Biagini E, Coccolo F, Ferlito M, Perugini E, Rocchi G, Bacchi-Reggiani L, Lofiego C, Boriani G, Prandstraller D, Picchio FM, Branzi A, Rapezzi C. Dilated-hypokinetic evolution of hypertrophic cardiomyopathy: prevalence, incidence, risk factors, and prognostic implications in pediatric and adult patients. J Am Coll Cardiol. 2005 Oct 18;46(8):1543-50. Epub 2005 Sep 28.*

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