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"Burned-out" hypertrophic obstructive cardiomyopathy presenting as acute coronary syndrome

Authors:
A Siama1, E Hamodraka1, P Fountoulakis1, A Kalogeris1, A Tsoukas1, AJ Manolis1, 1Asclepeion Voulas Hospital - Athens - Greece,

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Introduction: Hypertrophic cardiomyopathy is defined as left ventricular hypertrophy not solely explained by abnormal loading conditions. Besides the macroscopic picture of tissue hypertrophy, the disease is microscopically characterized by disarray and fibrosis and associated with small vessel disease.

Case presentation: A 49 year old male was admitted to the Emergency Department of our hospital with symptomatic sustained ventricular tachycardia which converted to sinus rhythm in response to intravenous amiodarone administration. Cardiac enzymes were elevated and troponin showed a rise and fall pattern. Ultrasound examination revealed a non-dilated left ventricle with moderately reduced ejection fraction. Basal and mid wall hypertrophy was observed with pronounced apical thinning and akinesis resulting in hourglass left ventricular remodelling. Intracavitary mid-ventricular gradient of 30 mmHg at rest and upon Valsalva manoeuver was recorded. The patient underwent coronary angiography which demonastraed atheromatous coronary network with minor non-obstructive lesions. Cardiac magnetic resonance confirmed the regional wall motion abnormalities described by the ultrasound; in particular an hourglass shaped left ventricle with hyperdynamic mid segments and thin akinetic to dyskinetic apical segments. Moreover subendocardial late gadolinium enhancement was demonstrated indicative of extensive fibrosis, compatible with "burned-out" hypertrophic cardiomyopathy that has entered a dilative phase. The patient was programmed for genetic family counselling and implantable cardioverter-defibrillator placement.

Conclusion/Discussion: Hypertrophic cardiomyopathy is a genetically and phenotypically heterogeneous disease with varied penetrance, ranging from asymptomatic "grey-zone" hypertrophy to severe left ventricular obstruction and/or end-stage dilated left ventricle with reduced ejection fraction. Prognosis and sudden cardiac death risk assessment takes into account several structural and functional parameters, including wall thickness, left atrial size, maximum left ventricular outflow tract gradient, episodes of non-sustained ventricular tachycardia, unexplained syncope and family history of sudden death. However physicians should be alert to recognize more atypical presentations of the disease.
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