Sudden death: hypertrophic cardiomyopathy with myocardial scarring and extremely rare coronary origin

Flavio Zuccarino1*, Sergio Moral2, and Elisabet Pujol2

1Department of Radiology, IDI Institut de Diagnostic per l’Imatge Hospital Universitari Josep Trueta, Avenida de France SN, Girona 17007, Spain and 2Department of Cardiology, Hospital Universitari Dr Josep Trueta, Avda. De França, sn, 17001 Girona, Spain
* Tel: +34 626661911, Fax: +34 972486960, Email: flaviozuccarino@yahoo.it

A 23-year-old man, with no relevant medical or family history of sudden death (SD) was admitted to the Coronary Care Unit after resuscitation because of a ventricular fibrillation. Haemodynamic parameters and physical examination were correct, the electrocardiogram only showed Q waves in the inferior leads and cardiac markers curve were positive. Echocardiography revealed asymmetric non-obstructive hypertrophic cardiomyopathy (HCM) (septal wall: 21 mm/posterior wall 13 mm) and a preserved ejection fraction. Coronariography (Panels D and E) showed permeable coronary arteries but with an extremely rare anomalous origin of the right coronary artery (arrows), which arose from the distal segment of left circumflex artery. Cardiac magnetic resonance (CMR) (Panels A–C) confirmed the presence of severe HCM (asterisks) with an infrequent localization affecting the anterior and lateral free wall (31 mm thickness) of the middle portion of the left ventricle. Delayed-enhancement gadolinium sequences (Panels F and G) showed no areas of subendocardial fibrosis and a focal contrast enhancement (asterisks) inside the hypertrophic myocardium with evident subendocardial and subepicardial sparing (arrows), consistent with myocardial scarring (MS).

This case presents an unreported combination of two extremely rare findings, both potential causes of SD. Anomalous origin of right coronary artery is described like one of these, although the aetiological mechanism is unknown in our case. MS is associated with ventricular arrhythmogenicity and represents another important risk factor in HCM, probably not only for its presence but also for its extension. For these reasons, recent studies conclude that CMR should acquire a fundamental role in the management and risk-stratification of these patients.