Ethical approval

The National Institute for Cardiovascular Research (NICOR) which includes the Myocardial Ischaemia National Audit Project (MINAP) (Ref: NIGB: ECC 1-06 (d)/2011) has support under section 251 of the National Health Service (NHS) Act 2006. The current study obtained the ethical approval from the Faculty of Medicine & Health Sciences Research Ethics Committee, University of East Anglia.

Supplementary material

Supplementary material is available at European Heart Journal online.

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References

The list of references is available in the online version of this paper.

CARDIOVASCULAR FLASHLIGHT

Myocardial infarction in a teenager

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An 18-year-old male presented to the Emergency Department with a 3-h history of chest pain. On physical examination, he had a blood pressure of 150/95 mmHg, mild obesity, and otherwise normal examination. An electrocardiogram showed sinus rhythm with short PR interval, ventricular preexcitation, and non-specific repolarization abnormalities (Panel A). There was a mild peak troponin elevation (3.2 μg/L). Cardiac catheterization showed normal coronary arteries. An echocardiogram showed massive hypertrophy of the left ventricle (LV) (Panel B). One week later a cardiac magnetic resonance with delayed contrast enhancement imaging showed localized gadolinium uptake in the interventricular septum, consistent with a myocardium infarction (arrows, Panel C). An LV septal thickness was 44 mm. A fasciculo-ventricular pathway was diagnosed at electrophysiological study. An endomyocardial biopsy was performed and the anatomical pathology revealed myocyte vacuolization consistent with a storage disease, preserved myocyte architecture, and absence of inflammation and fibrosis (Panels D and E). Genetic analysis identified a PRKAG2 gene mutation (R302Q). PRKAG2 cardiac syndrome causes a cardiomyopathy due to glycogen storage which may mimic classic hypertrophic cardiomyopathy. It is important to correctly differentiate between these entities because prognosis and treatment are different. We speculate the patient’s acute septal infarct resulted from a demand–supply mismatch of a severely hypertrophied septum, which has not been previously reported in storage diseases of the myocardium.

The patient underwent clinical treatment, had an uneventful recovery, and is doing well 2 years later.