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Images in cardiology

Microvascular ischaemia after cardiac arrest in a patient with hypertrophic cardiomyopathy

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BACKGROUND

An 11-year old boy was admitted to our intensive care unit following a successfully resuscitated ventricular fibrillation (VF) cardiac arrest during mild physical activity. Six months earlier he was diagnosed with non-obstructive hypertrophic cardiomyopathy (HCM) after an ECG at a pre-sport participating screening had shown left ventricular hypertrophy, marked repolarization abnormalities, and ST depression (Figure 1). There was no family history of HCM or sudden cardiac death (SCD). The child had been completely asymptomatic before the event; specifically, he had never experienced syncopal episodes. Maximal left ventricular wall thickness on echocardiography was 18 mm (z-score 4,5) and an Echo-bike and a 24h Holter monitor had been completely unremarkable. He was on no medications.

During hospitalization, there was a marked increase in Troponin I (from 1.57 ng/ml on admission to 80.5 ng/ml 14 hours later, n.v. $<$ 0.07 ng/ml). He was extubated after 6 days and had a complete neurological recovery. A CMR was requested prior to ICD implantation and performed 12 days after the cardiac arrest. At the time of CMR, Troponin I had almost returned to normal (0.27 ng/ml).

SSFP Cine images showed asymmetric septal hypertrophy with maximal wall thickness of 18 – 19 mm (Movie 1 and 2). T2-weighted FSE images showed marked increased signal in the septum, highly suggestive of oedema (Figure 2, left panel), which co-localized with areas of extensive fibrosis at late-gadolinium enhancement sequences (Figure 2, right panel) and hypoperfusion at rest (Movie 3). A 3D-whole heart SSFP view showed normal origin of the coronary arteries (Figure 3). Five months after the cardiac arrest, the patient remains asymptomatic and free of arrhythmic events.

The multimodality approach of CMR was able to describe all the aspects of a severe post-hypoxic state superimposed on preexisting microvascular ischaemia, including oedema, hypoperfusion and fibrosis in a young HCM patient without conventional risk factors for SCD,[1,2] who suffered a VF cardiac arrest. In addition CMR excluded an abnormal origin of the coronary arteries, avoiding the need for invasive or CT angiography. To our knowledge, acute post-cardiac arrest abnormalities have not been previously described in this disease. It is interesting that the oedema was localized to the hypertrophic areas, likely reflecting predisposition to microvascular hypoperfusion and ischemia. Despite prompt resuscitation and young age, the acute damage, paralleled by elevated Troponin I levels, was extensive. These findings support a central role of microvascular dysfunction in the genesis of ischemia in HCM hearts,[3] and illustrates their enhanced susceptibility to acute necrosis.

The case highlights the complexity of risk stratification for SCD in children with HCM. A large multicenter study would be desirable to establish the clinical role of CMR in this context and to better define risk predictors for SCD in young HCM patients.

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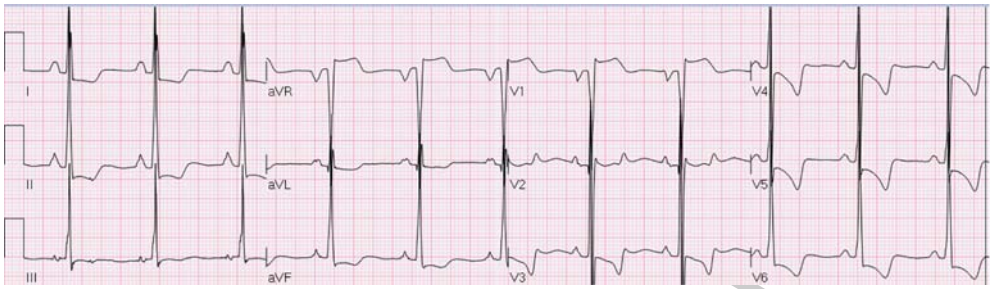
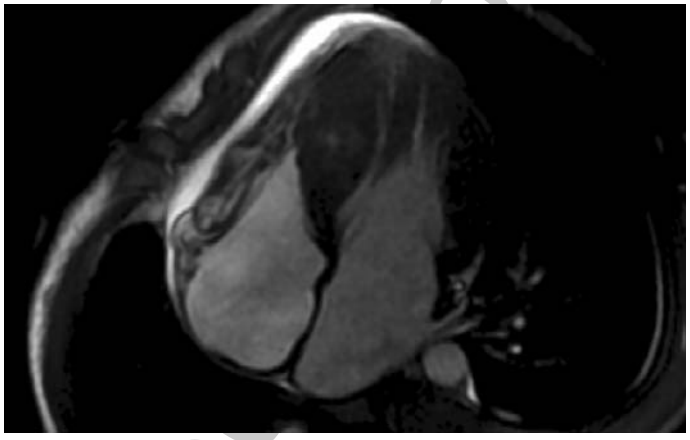
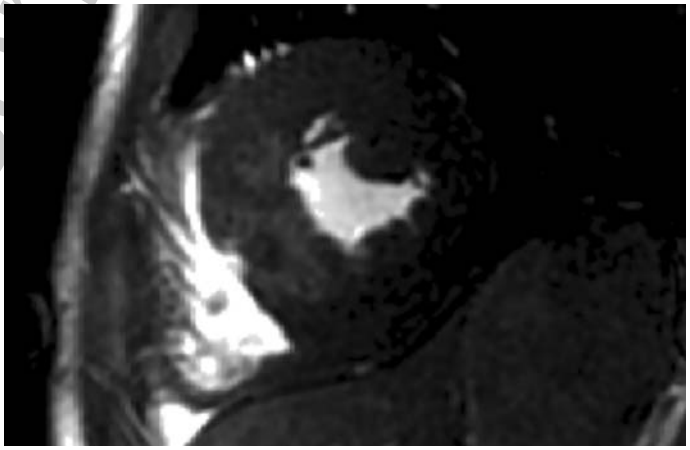


Figure 1. Electrocardiogram showing left ventricular hypertrophy, marked repolarization abnormalities and ST depression.



Movie 1. SSFP 4 chamber (Movie 1) cine views showing asymmetric left ventricular (LV) hypertrophy and good LV systolic function. Please note the dys-homogeneous signal within hypertrophied septum corresponding to eodema.



Movie 2. Short axis (Movie 2) cine views showing asymmetric left ventricular (LV) hypertrophy and good LV systolic function. Please note the dys-homogeneous signal within hypertrophied septum corresponding to eodema.

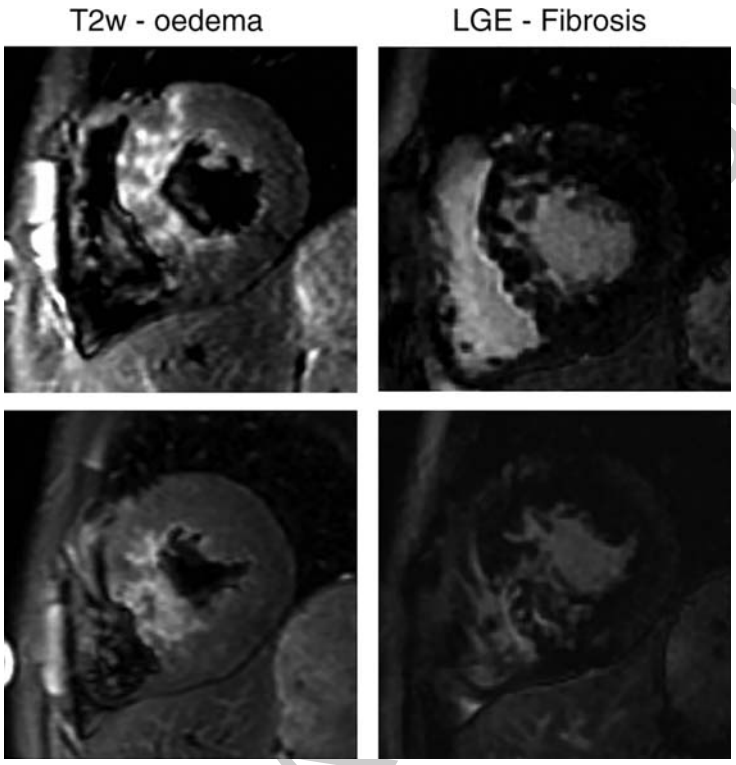
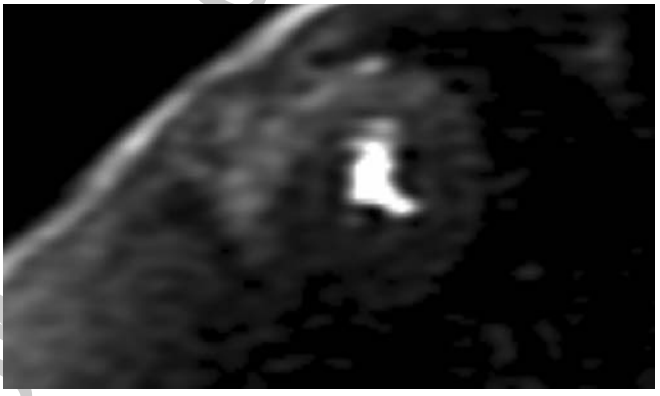


Figure 2. Left panel: T2-weighted fast spin echo images showing significant patchy hyperintense areas indicative of oedema in the hypertrophic interventricular septum. Right panel. Late Gadolinium images demonstrating areas of extensive fibrosis, co-localized to the oedematous regions. T2-W: T2-weighted. LGE: late gadolinium enhancement.



Movie 3. Rest perfusion defects are shown within the hypertrophic areas.

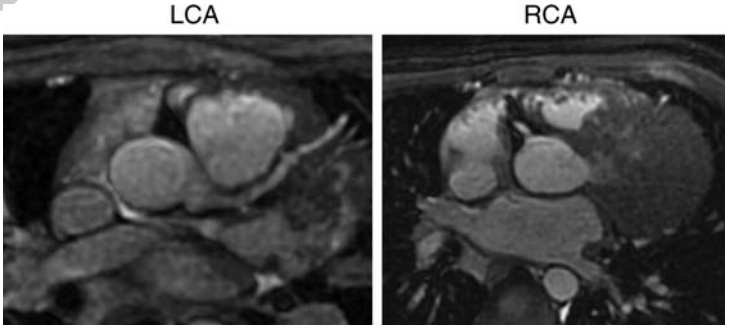


Figure 3. 3D SSFP images showing normal coronary origins. LCA: left coronary artery. RCA: right coronary artery.

Disclosures

There are no conflicts to disclosure.

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