A rare but life threatening cause of syncope

Shaun Khanna, Kartheek Garikapati, Henry Chen, Aditya Bhat, Gary Gan
Department of Cardiology Blacktown Hospital, Sydney NSW ; Western Sydney University

Clinical History

A 63 year old gentleman presented to our facility with a syncopal episode. He denied any history of preceding chest pain or palpitations. He was at work (as a carpenter) and was found on the ground by the owner of the house he was constructing. His past medical history consisted of only depression managed with duloxetine.

His neurological and cardiac examination was unremarkable. His ECG showed sinus rhythm with no evidence of acute ischemia. His troponin was negative at 4 ng/L and the rest of his laboratory tests were also unremarkable. He had a CT Brain which showed no evidence of any acute intracranial haemorrhage and an MRI DWI which showed no evidence of diffusion restriction or stroke. Furthermore his EEG was negative for seizure activity. He was admitted under cardiology for further investigation of this syncope. During his inpatient stay he was monitored on telemetry for 24 hours showing no arrhythmias. He had a CT Coronary Angiogram (CTCA) which showed normal course of coronary arteries with mild disease. He also had a transthoracic echocardiogram (TTE) which showed a normal left cavity size with mildly impaired systolic function. There was however marked left ventricular hypertrophy with anteroseptal hypokinesis. **See Figure 1A & 1B.**

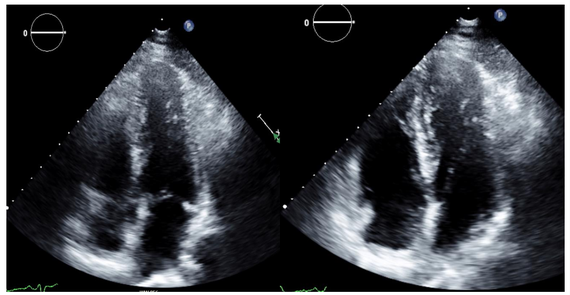
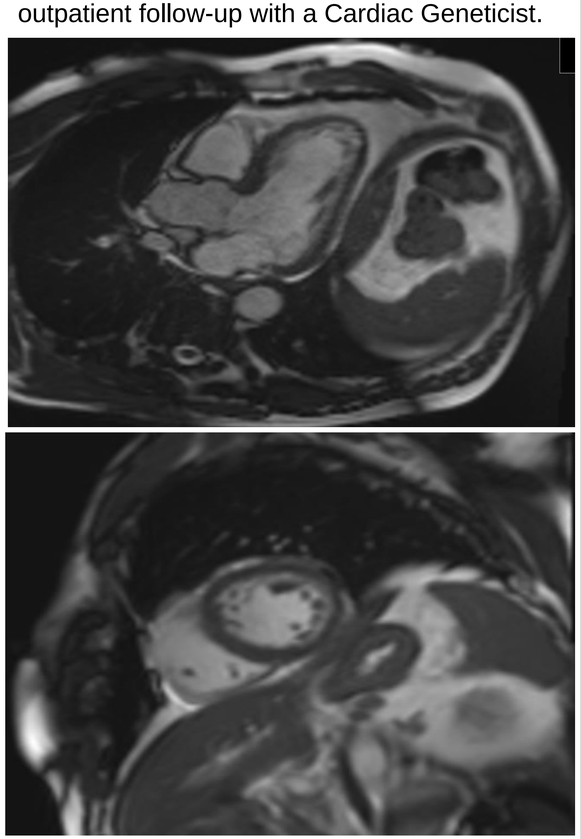


Figure 1A+ 1B: Echocardiogram showing marked LVH

Given no conclusive aetiology for the syncope, along with the suspicious transthoracic echocardiogram findings, the patient underwent an inpatient Cardiac MRI. The results of the Cardiac MRI indicated regional left ventricular non-compaction in the mid-distal lateral wall and apex with preserved LV size and function. There was no evidence of myocardial scar, infiltration, oedema or dysplasia. **See Figures 2 & 3.** Given this significant finding, the patient underwent a Cardiac Electrophysiology Study (EPS) which was positive and hence the patient subsequently had an automated implantable cardioverter-defibrillator (AICD) inserted for prevention of life threatening arrhythmias. The patient was subsequently discharged with outpatient follow-up with a Cardiac Geneticist.



Figures 2 &3. Axial slice CMRI showing non-compaction and hyper-trabeculation in the anterior, septal and lateral walls. Parasternal short view showing noncompacted myocardium.

Discussion

This is an interesting case of a Left Ventricular noncompaction (LVNC) which a rare entity in the clinical setting with prevalence described as affecting approximately 8 – 12 patients per million per year. [1] This congenital cardiomyopathy results in a spongy myocardium and deep trabeculations in the myocardial wall. These changes can predispose patients to life threatening ventricular arrhythmias, chronic heart failure and systemic embolic events. [2] Patients can present symptomatic with shortness of breath, fatigue, syncope, palpitations and peripheral oedema. [3] The initial diagnostic approach uses echocardiography and the most common criterias used are by Chin, Jenni and Stöllberger, which assess trabeculations, LV wall thickness and NC:C ratio of >2: 1. [4] Peterson et al has additionally described diagnosis of LVNC using CMR. The ratio of noncompacted myocardium to compacted myocardium during diastole should be greater than 2.3 (sensitivity of 86% and specificity of 99%). [5] As per the Cardiac MRI report, our patient's greatest ratio of noncompacted to compacted myocardium was 4.3, indicating the high specificity of LVNC. Management of LVNC involves treatment of arrhythmias, thrombus formation in trabeculations, treatment of heart failure and genetic counselling. [3-5]

Conclusion

Left Ventricular noncompaction is a rare congenital cardiomyopathy and should be considered in cases of syncope. Diagnosis is largely through imaging modalities such as echocardiography and cardiac MRI. Patients at high risk for ventricular arrhythmias require AICD insertion to prevent sudden cardiac death.

On reflection, this patient was appropriately treated with AICD insertion and referral to cardiac geneticist for further monitoring and management. Further studies are required to better explore the pathogenesis of LVNC and optimise the treatment paradigm.

References

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4. Bennett CE and Freudenberger R. The Current Approach to Diagnosis and Management of Left Ventricular Noncompaction Cardiomyopathy: Review of the Literature. Cardiol Res Pract. 2016; 5172308

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Contact information

Shaun Khanna
MBBS, M.Med (Adv)*
In Progress
Blacktown Hospital, NSW Australia
T: +61 401852696
E: shaun.khanna@health.nsw.gov.au