

Unusual Focal Hypertrophy of the Left Ventricle in a Patient with ECG Abnormalities but a 'Normal' Echocardiogram

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Clinical Presentation

A 20-year old South Asian man was admitted with a 6 week history of chest pain radiating to the left arm. Clinical examination was unremarkable. His 12 lead ECG showed T- wave inversion in the anterolateral leads with suggestion of dynamic changes in the inferior leads. His blood tests, including a troponin, were normal.

A transthoracic echocardiogram was reported to be normal. A coronary angiogram demonstrated normal coronary arterial flow with no obvious cause for his symptoms. He was referred to our center for a cardiovascular magnetic resonance (CMR) scan.

Investigations

CMR short axis SSFP cines showed marked focal, mass like, left ventricular hypertrophy (LVH). This selectively involved the areas adjacent to the right ventricular insertion points (fig 1) with a maximal LV wall thickness of 18mm. This was not clear from the initial long axis cine views (fig 2). Reversible stress perfusion defects were present during first pass gadolinium images in these focal areas (fig 3). There was also corresponding patchy late gadolinium enhancement (LGE) involving these same regions of greatest LV wall thickness (fig 4). These findings were most consistent with hypertrophic cardiomyopathy (HCM).

Retrospective review of the echocardiogram identified evidence of LVH that was seen by CMR however the images were suboptimal and the finding was likely missed at the time the study was initially reported.

Further prognostic risk assessment for HCM has been carried out and the importance of family screening highlighted.

Discussion

Focal areas of mass like hypertrophy can occasionally be seen in HCM. Tumour masses may have similar appearances but should not be contractile and the signal intensities is usually different to that of the myocardium. First pass perfusion images and LGE images are also usually different from normal myocardium.

In the context of cardiac chest pain with ECG abnormalities, CMR should be part of the diagnostic pathway, particularly where an echocardiogram is non-diagnostic or equivocal.

Suboptimal scans should have statements about quality clearly stated in the report. Simply labeling them as normal may have significant consequences as important diagnoses may be missed. Systems for highlighting potential false negative reports should be part of the clinical governance framework to prevent serious clinical incidents.



ECG - Abnormal with T wave inversions

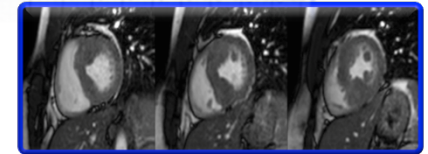


Figure 1. SAX cine still including basal, mid and apical slices with Focal LV hypertrophy involving RV insertion points

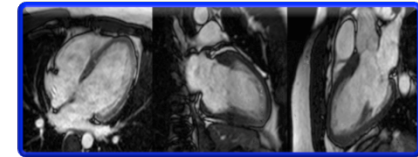


Figure 2: Long axis cine still at end diastole with no obvious LVH

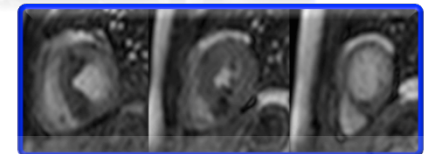


Figure 3. Perfusion defects seen in the area of focal hypertrophy during first pass perfusion images following adenosine infusion. This was not present at rest.

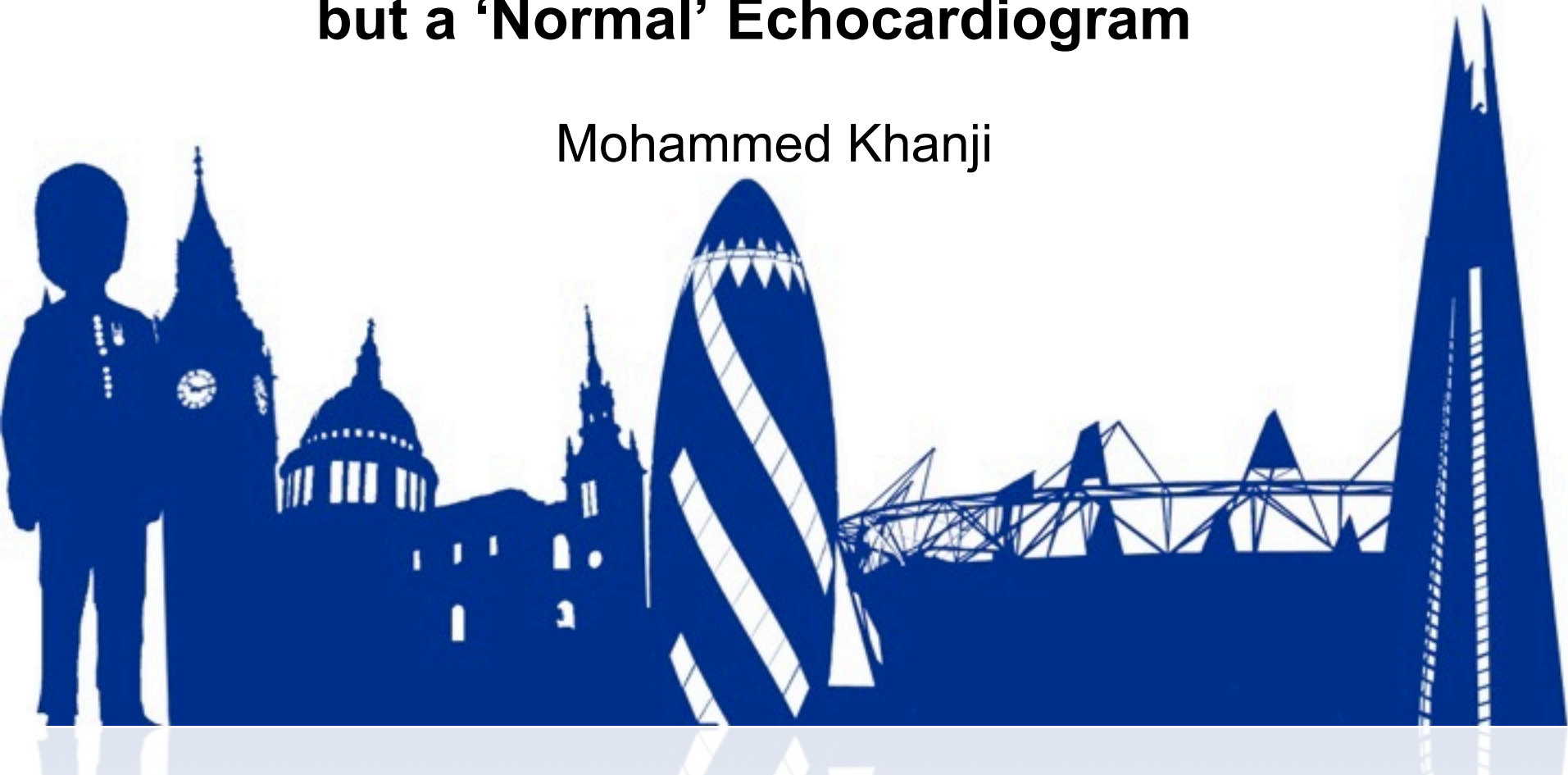


Figure 4. Late gadolinium enhancement in the areas of focal hypertrophy.

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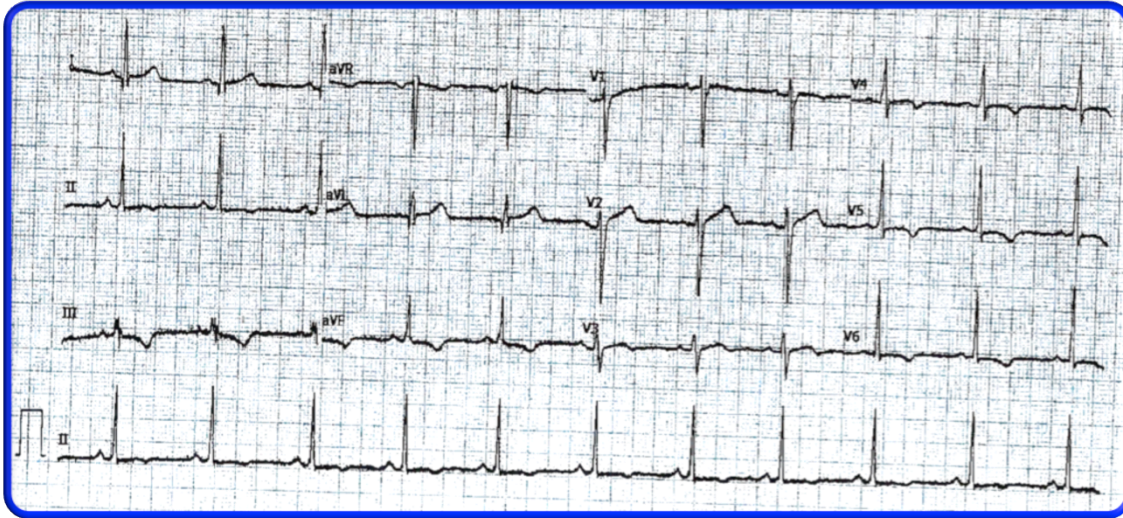


Case

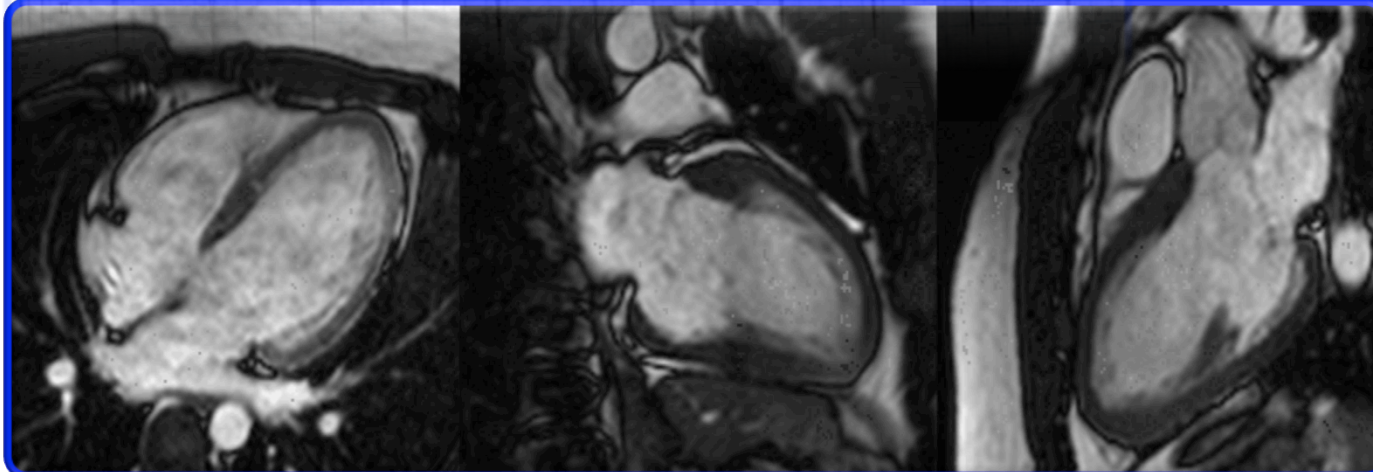
- 20 year old Asian man
- Admitted with cardiac sounding chest pain for 6 weeks
- Abnormal ECG
- Troponin negative
- Normal echo and angiogram

CMR requested for further assessment

Long axis views

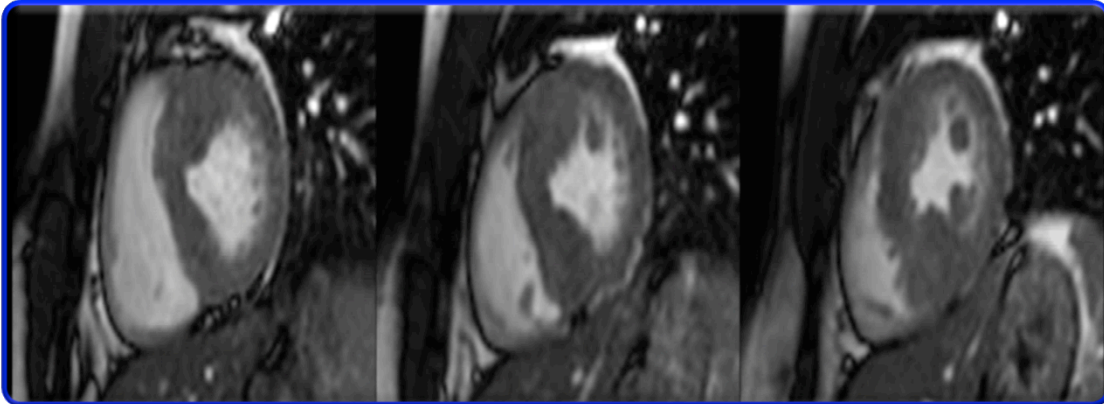


T- wave inversion in the anterolateral ECG leads with suggestion of dynamic changes in the inferior leads

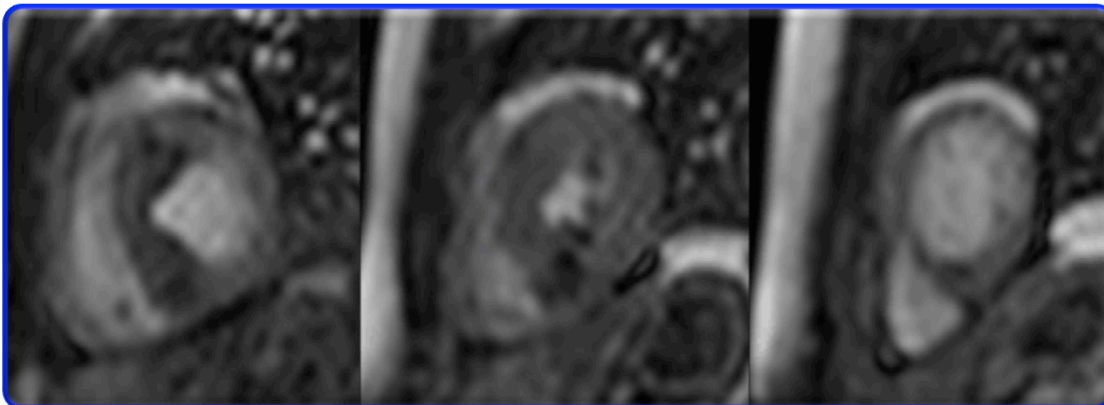


No obvious LVH at end diastole in long axis views

Short axis stack – focal hypertrophy

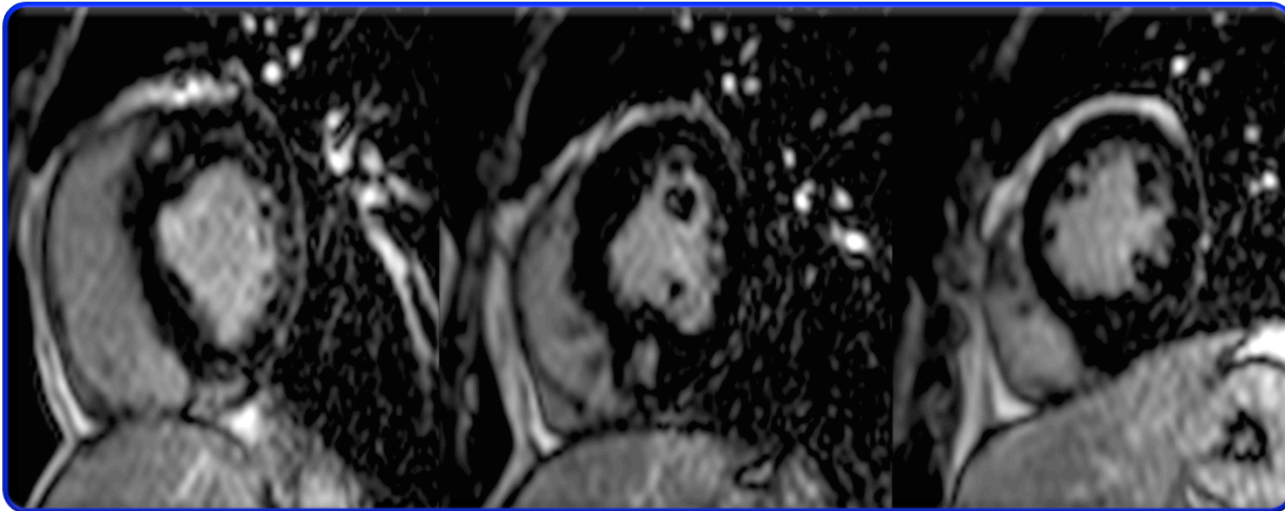


Focal LV hypertrophy involving RV insertion points



Perfusion defects seen in the area of focal hypertrophy during first pass perfusion images following adenosine infusion. This was not present at rest.

Late Enhancement

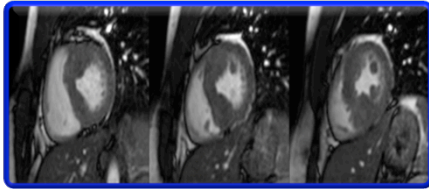


Late gadolinium enhancement in the areas of focal hypertrophy.

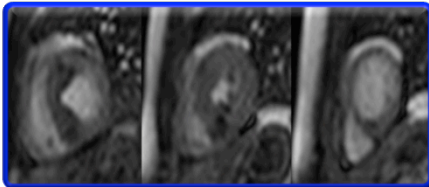
Diagnosis

Hypertrophic cardiomyopathy

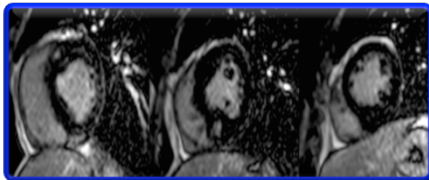
- Asymmetric LVH



- Perfusion defect in hypertrophied area



- RV insertion point LGE



- Further risk stratification and family screening

CMR Risk Stratification

- CMR parameters alone **currently** not major criteria for risk stratification in hypertrophic cardiomyopathy eg LGE, perfusion defects
- “... Nonetheless, the present data would support a potential role of LGE as an arbitrator in decision making for primary prevention ICDs in patients in whom risk status remains uncertain after assessment of conventional risk markers.” (AHA 2011)

Learning points

- Focal hypertrophy in HCM - contractile
- Check the images not just the report
- CMR in chest pain with abnormal ECG, especially when echo non diagnostic
- CMR useful in diagnosis and risk stratification HCM
- Suboptimal scans should not be reported as normal