



## CASE REPORT

**Cardiac Pseudotumor: Tissue Characterization by Cardiovascular Magnetic Resonance****James C. C. Moon,<sup>1</sup> Mary N. Sheppard,<sup>1</sup> G. Lloyd,<sup>2</sup> Nikhil R. Patel,<sup>2</sup>  
Dudley J. Pennell,<sup>1</sup> and Raad H. Mohiaddin<sup>1,\*</sup>**<sup>1</sup>Centre for Advanced Magnetic Resonance in Cardiology (CAMRIC), Royal Brompton Hospital, London, UK<sup>2</sup>Department of Cardiology, Eastbourne District General Hospital, Eastbourne, UK**ABSTRACT**

We present serial cardiovascular magnetic resonance (CMR) in a case of cardiac pseudotumor (inflammatory myofibroblastic tumor). The diagnosis proved difficult and was helped by myocardial tissue characterization using CMR. Intrinsic contrast and extrinsic contrast (using gadolinium-DTPA perfusion, early and late imaging) differentiated between normal and pathological tissues and demonstrated the changes in properties of cardiac pseudotumor over time. The case illustrates the versatility of CMR in tissue characterization, which is unavailable by other cardiac imaging modalities.

*Key Words:* Cardiovascular magnetic resonance (CMR); Pseudotumor; Tissue characterization; Gadolinium.

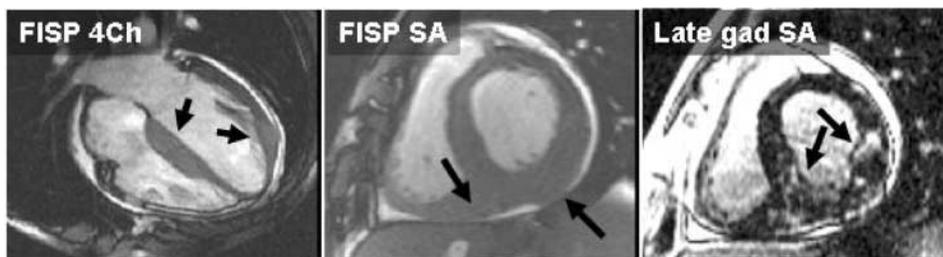
**CASE HISTORY**

A 29-year-old man presented to hospital with sustained monomorphic ventricular tachycardia (VT). After electrical cardioversion, the history revealed a background of 18 months of night sweats and a 3-week history of exertional left arm and chest pain. He was a nonsmoker with no history of recent foreign travel, tuberculosis, or a family history of premature coronary artery disease. Over the next week, he developed repeated episodes of VT and a pericardial friction rub.

Inflammatory markers (CRP) became raised as did troponin T. The echocardiogram (ECG) demonstrated inferior ST and T wave changes consistent with inferior ischaemia. Coronary angiography was normal. Echocardiography demonstrated asymmetrical hypertrophy of the infero-basal septum, suggestive of hypertrophic cardiomyopathy and a small pericardial effusion.

Because of the diagnostic uncertainty, cardiovascular magnetic resonance (CMR) was performed on a Siemens Sonata scanner (Erlangen, Germany) at 1.5T with high gradient performance. Cine imaging demonstrated unusual

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**Figure 1.** First CMR scan. Regional hypertrophy with abnormal gadolinium hyperenhancement. The hypertrophy is very localized (arrows) around the inferior interventricular groove, involving the RV, septum, and inferior wall of the LV. The gadolinium pattern is also abnormal and indicates a myopathic process.

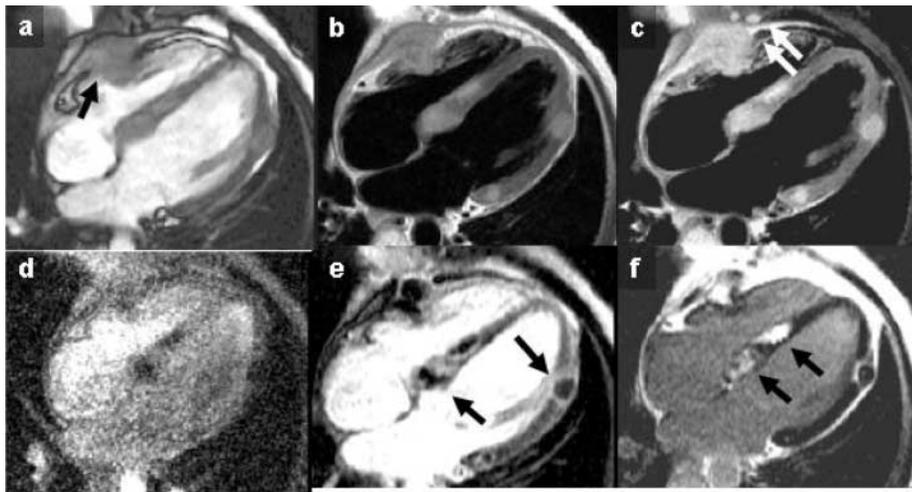
regional cardiac hypertrophy associated with regional hypokinesia, Fig. 1. At the base, the hypertrophy of the septum and inferior wall merged into hypertrophy of the inferior wall of the right ventricle (RV), which was at max 18 mm thick. The edges of the hypertrophy were seen to have a shoulder. Late gadolinium inversion recovery imaging demonstrated patchy hyperenhancement throughout the hypertrophied region. This pattern was not thought consistent with hypertrophic cardiomyopathy and the diagnosis of an infiltrative malignant process was considered.

On the basis of the above findings, transvenous right ventricular biopsy was performed. Four adequate samples were taken, but histology demonstrated normal myocardium only. Over the next month the patient was treated

conservatively with anti-inflammatory drugs for symptomatic pericarditis and amiodarone for arrhythmic control, and the clinical state of the patient improved. The inflammatory markers fell, the rub resolved, and the echo normalized. Six weeks after the first scan, repeat CMR was performed. This scan was performed using the full range of tissue characterization sequences available, Table 1. In the 5 weeks since the previous scan, there had been a profound change in the scan appearances (Fig. 2). Some areas of regional thickening on the previous scan appear thinned but remain hypokinetic. There was new thickening of the RV wall, and further areas in the basal septum and apical antero-lateral wall of the left ventricle (LV). The thickening towards the base of the RV breached

**Table 1.** Sequences used in tissue characterization.

Technique	Name	Parameters	Reason
Cine imaging	True FISP	TE/TR 1.6/3.2 ms 2.3 × 1.4 × 7 mm $\alpha = 60^\circ$ , 20 phases	Function. Fluid appears bright, blood: myocardial contrast good
Intrinsic contrast	T1w TSE:	TE 7 ms TR = RR interval 1.4 × 1.4 × 5 mm turbo 15	High resolution tissue differentiation
Intrinsic contrast	T2w TSE:	As above but TR = 2 × RR interval	High resolution tissue differentiation edema and fluid bright
First pass perfusion	Saturation recovery FLASH, Oppelt et al. (1986)	TE/TR 1.45/456 ms 1.4 × 1.4 × 10 mm $\alpha = 12^\circ$ , TI 110 ms	First-pass rest defects reflect capillary density or severe epicardial coronary stenosis
Post gadolinium: early avascularity imaging	Early fixed long TI inversion recovery, Moon et al. (2003)	Fixed TI at 440 ms 2.3 × 1.4 × 8 mm Alternate heartbeats 23 segments $\alpha = 20^\circ$ FLASH Readout	Dark areas are regions of avascular tissue: necrosis, thrombus, and microvascular obstruction
Post gadolinium: late infarct imaging	Late variable TI inversion recovery, Simonetti et al. (2001)	As above but: variable TI to null remote myocardium (typically 340–400 ms)	Bright areas are regions of focal interstitial expansion. Infarction, edema, other noncellular infiltration



**Figure 2.** Second CMR scan 5 weeks later. Tissue characterization (4-chamber view) with multiple techniques. (a) FISP demonstrating focal lesions with RV pericardial breach (arrow); (b) and (c) T1 and T2 weighted turbo spin echo show that these regions possess the same intrinsic contrast; note satellite lesions (arrow) and pericardial spread; (d) rest perfusion and; (e) early fixed long TI inversion recovery imaging demonstrates first-pass regions confirmed to be avascular, denoting necrosis and microvascular obstruction (arrows); (f) late gadolinium imaging demonstrates hyperenhancement from focal interstitial expansion likely to be fibrosis or edema.

the pericardial fat layer and was contiguous with a mass outside the pericardium and spreading along it. Two further small adjacent discrete masses were observed.

Myocardial tissue characterization using a variety of techniques demonstrated at least five areas of involvement with identical tissue characteristics. On fast imaging with steady-state precession (FISP) and T1 weighted images these areas were similar to normal myocardium. High signal on T2 weighted imaging showed a higher water content, and the rest perfusion deficit indicated low regional blood flow. Persistent hypoenhancement over the next 5 minutes indicated avascularity, which, given that these are not fluid filled (or FISP would have been high signal), implies necrosis and microvascular obstruction. Finally, late gadolinium imaging demonstrated extensive hyperenhancement indicating focal interstitial expansion, likely to be edema or fibrosis.

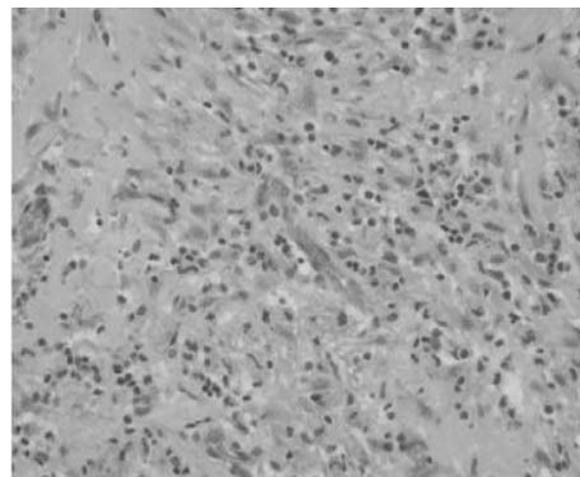
Thus the scan demonstrated a rapidly progressing, pericardial-based multifocal, infiltrative process that crosses tissue planes, resulting in satellite lesions and necrotic foci in the larger areas.

A diagnosis of a malignant pericardial process was considered likely despite the clinical improvement and negative biopsy. Surgical referral was made for open biopsy.

At operation, a red, friable 3 cm lump from the anterior wall of the RV was removed. The histology (Fig. 3) consisted entirely of vascular granulation tissue rich in myofibroblasts and histiocytes. Many blood

vessels were thrombosed with infarction. There were no malignant cells, granuloma, or organisms, and polymerase chain reaction (PCR) for tuberculosis was negative.

The diagnosis of primary pericardial pseudotumor, also known as inflammatory myofibroblastic tumor, was made and the patient was started empirically on immunosuppressive therapy with high dose oral steroids



**Figure 3.** Hematoxylin- and eosin-stained section showing admixed histiocytes, blood vessels, lymphocytes, and plasma cells. This tissue was infiltrating and destroying the myocardium and overlying pericardium. Magnification  $\times 400$ .



(prednisolone 40 mg). Eight weeks later, the patient was well, although functionally in New York Heart Association (NYHA) II. A third CMR scan was performed.

This scan demonstrated the changes associated with the regional biopsy and overall regression of the regions of thickening. The T2 weighted imaging showed less pronounced regional heterogeneity, and the microvascular obstruction was no longer apparent. However, there was new abnormal tissue outside the pericardium posterior to the LV suggesting regional progression. Currently, the patient remains clinically stable. His steroids are being reduced, baseline investigation for possible cardiac transplantation are being performed, although it is unclear whether this will be required.

### DISCUSSION

Inflammatory pseudotumor or inflammatory myofibroblastic tumor are rare proliferative lesions occurring most commonly in the lung, but have been reported in almost every major organ of the body (Coffin et al., 1998). Cardiac involvement is rare with fewer than 13 previous cases reported, most typically in children (Rose et al., 1996). Whilst many features of the condition suggest an inflammatory process, the potential for local recurrence, local infiltration, vascular invasion, and the development of new, noncontiguous sites are characteristic of neoplasia, and some tumors have been found to be clonal (Biselli et al., 1999). In this case, the constitutional features have resolved with time and it is to be hoped that the condition is self-limiting, although the most recent scan suggests slower but ongoing activity.

Cardiovascular magnetic resonance in this case dramatically illustrated the key features of the condition and its progress. Using a combination of FISP cine, T1 and T2 weighted spinecho, rest perfusion, early fixed long inversion recovery technique for avascularity, and late infarct imaging allowed both accurate tissue characterization and highlighted disease progression culminating in the decision for open biopsy, even with an initial negative biopsy, symptoms resolution, and echo/blood test normalization.

This use of CMR intrinsic and extrinsic contrast techniques for tissue characterization, combined with high spatial resolution, allowed differentiation from hypertrophic cardiomyopathy in the early stages and from disease regression on the second scan, and in the third scan demonstrated a mix of regression and progression at

different sites. It would not have been possible to obtain this information without CMR, emphasizing its role in myocardial and cardiac tissue characterization.

### ADDENDUM

Since the submission of this article, the patient developed a rash which at biopsy proved to be a cutaneous T cell lymphoma. The cardiac histology was reviewed with additional stains and sections. Abnormal intra-vascular, non-migratory T cell were detected occluding vessels, suggesting that the cardiac pathology was granulation tissue in response to malignant microvascular obstruction, a process detected by the CMR imaging.

### ACKNOWLEDGMENTS

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