

Case report and images in cardiology

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Primary cardiac tumours are extremely rare,⁽¹⁾ occurring with a lifetime incidence of 0.0017% to 0.02% according to various autopsy series.⁽²⁾ Benign primary cardiac tumours constitute approximately 75% of those originating from the heart.⁽³⁾ Cardiac lipomas account for a mere 8.4% of benign primary cardiac tumours.⁽⁴⁾ According to the available literature, 25% of cardiac lipomas are intramyocardial, 25% are extracavitary with epicardial origin and 50% are intracavitary of subendocardial origin.⁽⁵⁾ They can cause cardiac dysfunction by compression of the left ventricle, valvular dysfunction, peripheral embolisation and cardiac arrhythmia, either by virtue of intramyocardial position or involvement of the conduction system.⁽⁶⁾ Some arrhythmias have paroxysmal occurrence and can cause sudden death. Arrhythmogenic right ventricular cardiomyopathy (ARVC) represents another rare condition amongst cardiac diseases and remains an under-recognised entity.⁽⁷⁾ ARVC is characterised macroscopically by a fatty appearance of the right ventricular free wall with resultant thinning of the wall (<3mm) in the fibrofatty variant of the disease.⁽⁸⁾ The partial or total replacement of the myocardium by adipose or fibro-adipose tissue leads to progressive right ventricular failure and ventricular arrhythmias.

Whilst the incidence of ARVC remains unknown, its prevalence in the general population is estimated to be approximately 1:1000.⁽⁷⁾ It is an important cause of sudden cardiac death in young adults accounting for approximately 8 percent of cases overall and 22 percent in athletes.⁽⁹⁾ Diagnosis remains challenging because most patients are asymptomatic, or have symptoms limited to paroxysmal palpitations occasionally leading to syncope.⁽⁸⁾ Approximately 50 percent of patients with ARVC present with symptomatic ventricular arrhythmias.⁽¹⁰⁾ The most common arrhythmia is sustained or nonsustained monomorphic VT that originates in the right ventricle and therefore has a left bundle branch block (LBBB) pattern, similar to that seen in idiopathic RV outflow tract tachycardia.

We describe the clinical case of a young man presenting with ventricular tachycardia of left bundle branch block morphology, originating from the right ventricle, and an apical intra-myocardial lipomatous right ventricular mass in the setting of Arrhythmogenic Right Ventricular Cardiomyopathy. A combination of two extremely rare cardiac conditions for which, to our knowledge, there exists a single report in the literature.

CASE REPORT

A 23-year-old male was admitted to our cardiac unit after an episode of palpitations with associated dyspnoea and central chest discomfort without syncope. He gave a history of paroxysms of palpitations experienced over a four year period for which he had sought medical advice at another institution. Unfortunately, after having been informed that he may require corrective cardiac surgery in the future he had not returned for follow up. No family history of premature cardiac disease was obtained. There were no clearly identifiable precipitants for his episodic palpitations and he could recall only one associated syncopal episode.

On admission, he was asymptomatic. Examination revealed a regular pulse rate of 75 beats per minute with good volume, blood pressure 128/80 mmHg and no pyrexia. There were no clinically significant abnormal findings on physical examination. Twelve-lead ECG demonstrated sinus rhythm, PR interval of 160

milliseconds, no evidence of pre-excitation and a QRS width of 100 milliseconds. The axis is difficult to determine accurately in his ECG, lead I is noted to be negative and we have calculated it to be +105 degrees. The QT interval is not prolonged and there is T-wave inversion from V1 through V5 with epsilon waves in V1, V2 and the inferior leads (Figure 1). Twelve-lead ECG done at the referring day clinic/hospital demonstrated a regular, broad complex tachycardia (QRS width 160 ms) of left bundle branch block morphology with a QRS axis of between +30 to 45 degrees. (Figure 2).

Chest radiograph revealed a normal cardiothoracic ratio and clear lung fields. Basic blood tests including electrolytes and thyroid

function tests were all within normal limits. On continuous cardiac telemetry, frequent runs of non-sustained ventricular tachycardia of left bundle branch block morphology were documented despite oral B-blockade. Transthoracic echocardiography revealed a thin and extremely trabeculated right ventricle with a prominent moderator band and a hyperechoic mass measuring 1.7cm situated in the right ventricular apex with evidence of an apical aneurysm (Figure 3). A clearly thin and dyskinetic segment with diastolic bulging was demonstrated over the right ventricular free wall proximal to the apex representing an aneurysm (Figure 4). Cardiac computerised tomography confirmed the apical mass, measuring a transverse diameter of 24.5 millimetres and between +26 to +50 Hounsfield units compatible

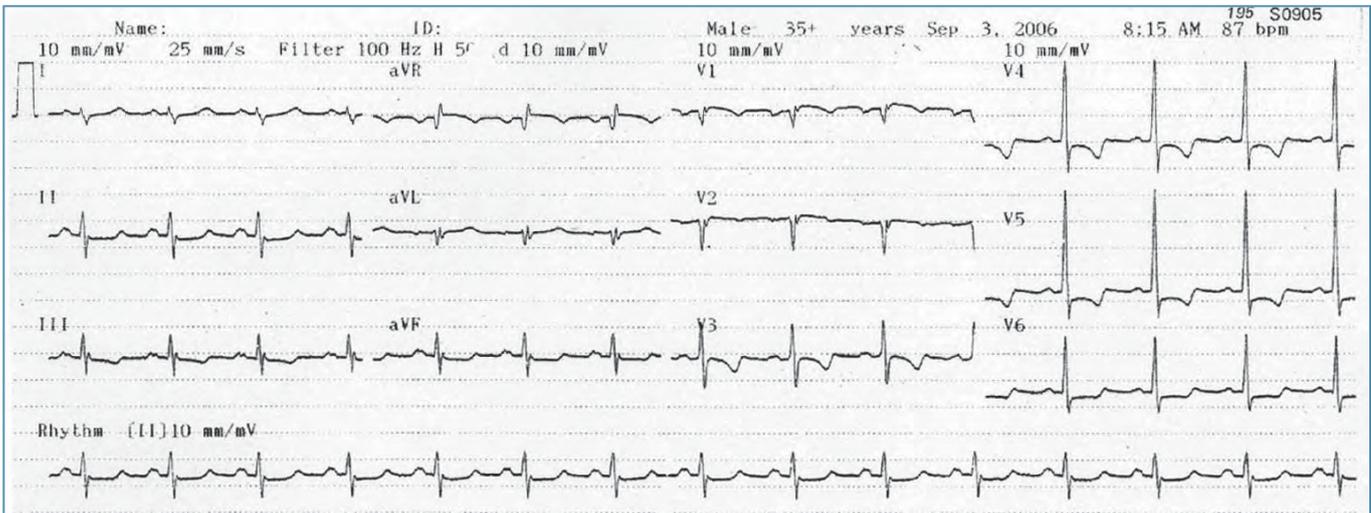


FIGURE 1: 12 lead ECG in sinus rhythm displaying epsilon waves in leads V1, V2 and the inferior leads

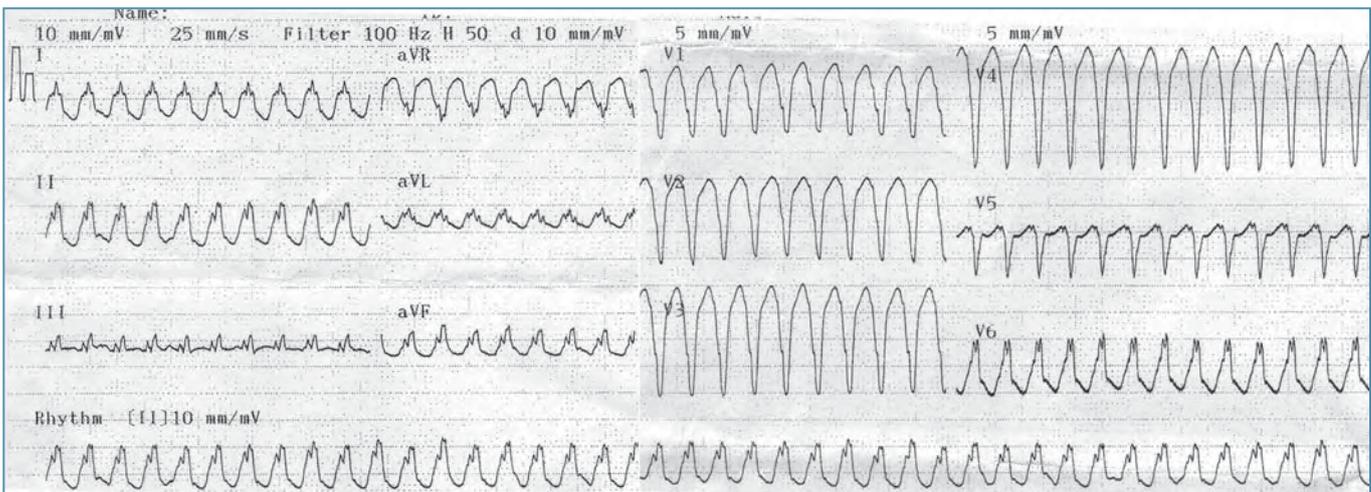


FIGURE 2: 12 lead ECG during ventricular tachycardia of right ventricular origin

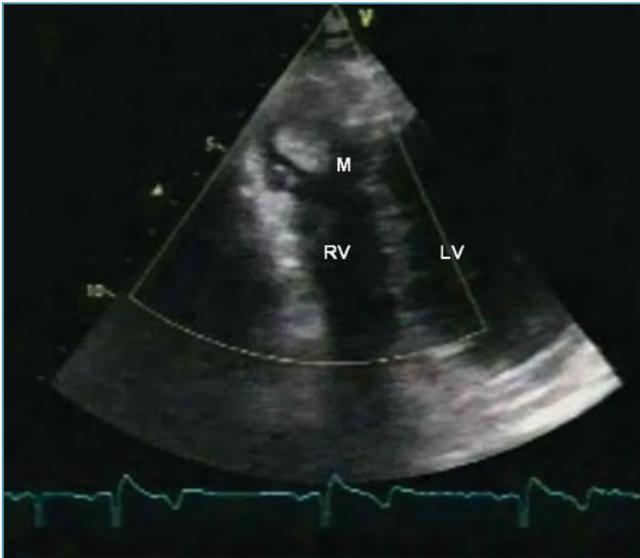


FIGURE 3: Echocardiogram demonstrating RV aneurysm and intramural apical mass

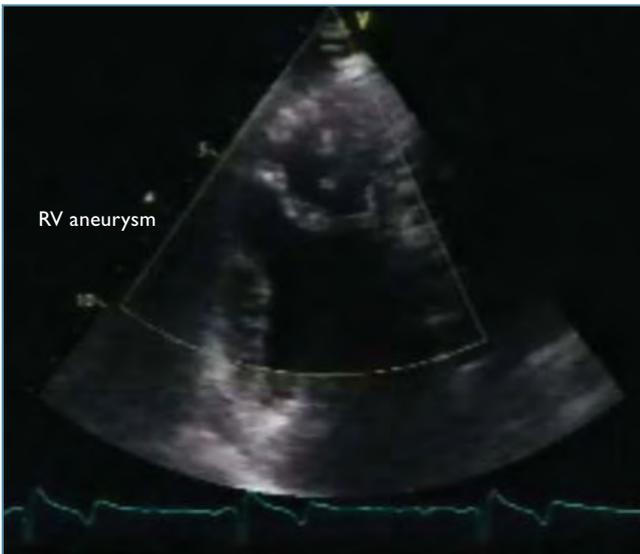


FIGURE 4: Echocardiogram of the right ventricle demonstrating the proximal RV aneurysm

with soft tissue. The right ventricular free wall measured between 1,5 and 2 millimetres in thickness and diffuse fatty infiltration was evident (Figure 5).

In view of the thin walled right ventricle free wall cardiac surgery was preferred over endomyocardial biopsy in order to determine the nature and histology of the right ventricular apical mass. The possibility of the mass being an organised thrombus was considered in the differential diagnosis on the echocardiographic

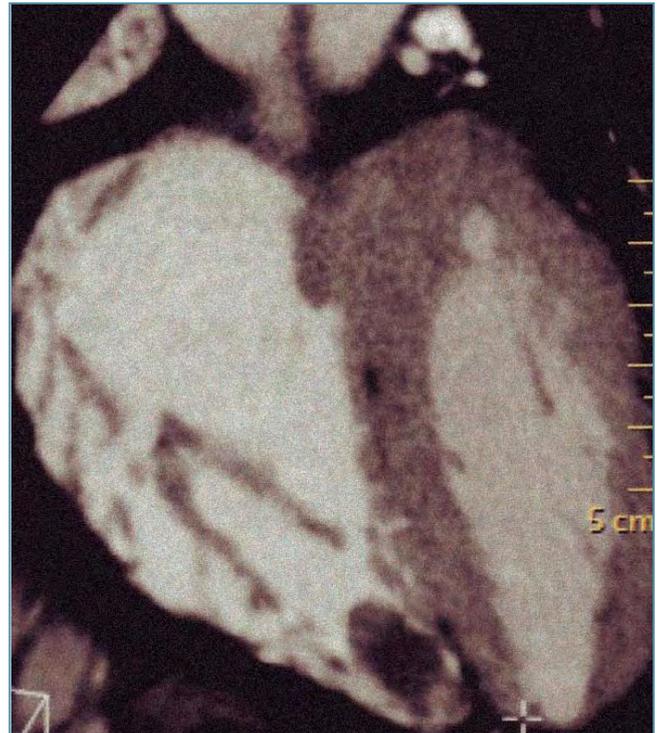


FIGURE 5: CT scan of the heart demonstrating fat infiltration of the RV free wall and an inhomogenous RV apical mass arising from within the myocardium

views (Figure 3). However, once a well defined inhomogenous mass was identified within the right ventricular myocardium at its apex on cardiac CT, we considered this unlikely. He underwent cardiac surgery via median sternotomy for diagnostic and therapeutic purposes. Via a right atriotomy approach, a yellow, well-circumscribed intramural mass was identified in the apex of the right ventricle and completely excised. Macroscopically, the right ventricular free wall was found to be unusually thin with an appearance of diffuse fatty infiltration (parchment like). The patient was not anticoagulated pre-operatively and surgery took place shortly after the CT heart as it was our opinion that had the mass been found to be malignant this would have significantly altered further management of our patient. No thrombus or other mass was identified within the right ventricular cavity at surgery.

His surgery was uncomplicated and he made a good postoperative recovery. Histology of the excised mass demonstrated mature adipose tissue confirming a diagnosis of cardiac lipoma. The discrepancy between the measured Hounsfield units of the mass on CT heart and the histological findings require comment. The scattered myocardial fibres present within the mass at

histology may have contributed to the spread in measured Hounsfield units and inhomogeneity of the mass on cardiac CT. No fibrous tissue or dysplasia was found. Myocardial biopsy was not performed during the procedure in view of the marked thinning of the ventricular wall.

Upon re-evaluation of our patient we found that he met the following two major and two minor criteria for the diagnosis of arrhythmogenic right ventricular cardiomyopathy as published by McKenna et al. in 1994:⁽¹¹⁾

- Major:**
1. Epsilon waves of the right precordial leads (V1 to V3); and
 2. Localised right ventricular aneurysm (dyskinetic area with diastolic bulging)
- Minor:**
1. Sustained left bundle branch block type ventricular tachycardia; and
 2. Inverted T waves in right precordial leads (V2 and V3) in a patient older than 12 years in the absence of right bundle branch block.

Access to cardiac magnetic resonance imaging pre-operatively was unavailable. This investigation was planned for follow up with a view to referring the patient to a centre for electrophysiological studies and consideration of ablative therapy versus ICD placement. Unfortunately the patient did not return to clinic.

DISCUSSION

An extensive literature search has yielded only one other such description of a cardiac lipoma associated with ARVC.⁽¹²⁾ We are also not aware that fatty changes in ARVC can be so extensive as to include the development of distinct lipomas.

In light of the fact that the mass may have been responsible for his paroxysmal ventricular arrhythmias, previously associated with syncope, and that we needed to exclude a mass of malignant origin, surgical resection was the preferred approach. Whilst the single 12 lead ECG recorded during an episode of ventricular tachycardia (Figure 2) has an axis which is not necessarily in keeping with an origin from the right ventricular apical mass, we were of the opinion that the mass may have acted as a substrate for VT during his prior paroxysms for which there are no documented ECG's. The fact that the mass was found to be intramural and not intracavitary at surgery increases the likelihood that it was indeed a potential substrate for VT.

Ideally, one would like to have access to cardiac MRI (CMRI) to more accurately define the tissue characterisation of a cardiac mass and in this case the morphology of the right ventricle. Unfortunately this is not always possible in a resource limited environment where radiological expertise in interpreting CMRI is limited. Multidetector cardiac CT has been shown to hold promise as an alternative imaging modality to CMRI.⁽¹³⁾

Cardiac CT has become available as an imaging modality at our hospital more recently and the availability of cardiac MRI is planned for the future. The diagnosis of arrhythmogenic right ventricular cardiomyopathy was made retrospectively in this patient as most of our attention was initially focused on the clearly defined mass in the right ventricular apex. South Africa has a paucity of electrophysiologists and not every tertiary centre has on site access to expertise in this field. As highlighted in a prior issue this is a sub-discipline of Cardiology that should be cultivated to allow for more widespread access to it in our academic tertiary referral centres.

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