Electronic Journal of General Medicine
2022, 19(5), em397
e-ISSN: 2516-3507
https://www.ejgm.co.uk/

MODESTUM

Case Report **OPEN ACCESS**

A case report: Rapid progression of arrhythmogenic cardiomyopathy with biventricular failure

Mae-Sing Lim-Cooke [1], Federico Fedel [1], Mengfei Lu [1]*, Debashis Roy [1,2]

[1] Princess Alexandra Hospital, Harlow, UK
[2] Department of Cardiology, Princess Alexandra Hospital, Harlow, UK
***Corresponding Author:** m.lu@nhs.net

Citation: Lim-Cooke M-S, Fedel F, Lu M, Roy D. A case report: Rapid progression of arrhythmogenic cardiomyopathy with biventricular failure. Electron J Gen Med. 2022;19(5):em397. https://doi.org/10.29333/ejgm/12263

ARTICLE INFO

Received: 2 Jun. 2022

Accepted: 7 Jul. 2022

ABSTRACT

Background: Arrhythmogenic cardiomyopathy (ACM) is an inherited condition. It is sub-characterized into arrhythmogenic right, left, and bi- ventricular cardiomyopathies. The Padua task force criteria is used in the diagnosis of ACM.

Case report: A 41-year-old presented with palpitations was found to be in ventricular tachycardia. He was reverted back into sinus rhythm following cardioversion. Electrocardiogram showed widespread T-wave inversions. Initial troponin was raised with unobstructed coronaries on coronary angiogram. Transthoracic echocardiogram (TTE) seven months prior showed reduced left ventricular systolic function (LVSD) with unremarkable right ventricular (RV) function. TTE on this admission showed new right sided failure with worsening LVSD. Cardiac magnetic resonance imaging (MRI) appearance was consistent with biventricular ACM based on morpho-functional criteria.

Discussion: This case highlights how RV function can decline rapidly in ACM. Widespread T-wave inversion is a minor criterion in the task force criteria. If TTE is indicative of underlying cardiomyopathy, cardiac MRI is the gold-standard investigation to confirm the pathology.

Keywords: arrhythmogenic cardiomyopathy, bi-ventricular failure, cardiac arrhythmias

INTRODUCTION

Arrhythmogenic cardiomyopathy (ACM) is an inherited condition that predominantly affects the younger demographic. It is characterized by the replacement of myocytes with fibrofatty deposits. ACM commonly presents with ventricular arrhythmias. However, patients can also present with sudden death. ACM is more commonly known to affect solely the right ventricle. However, literature and case reports have shown this to be no longer accurate, and as such, ACM can be divided into: arrhythmogenic right ventricular cardiomyopathy (ARVC), biventricular, and arrhythmogenic left ventricular cardiomyopathy (ALVC) [1]. The diagnosis of ARVC is aided by the use of the 'task force' criteria (**Figure 1**).

This includes major and minor criteria surrounding cardiac morphology and dysfunction, as well as electrocardiogram (ECG) changes and family history. Diagnosis of biventricular ACM is based solely on morpho-functional criteria of the left and right ventricle without need for ACM gene mutation, whilst this becomes necessary in ALVC dominant-left diagnosis [2]. Because of the risk of sudden death, there is an indication for implantable cardioverter-defibrillators (ICD) in these patients.

ACM is previously known to solely affect the right ventricle. However, literatures and case reports are demonstrating further categorizations of ACM with new diagnostic criterion. This case report provides an unusual insight into how rapidly the RV function can decline in the context of ACM and progress into bi-ventricular failure within months

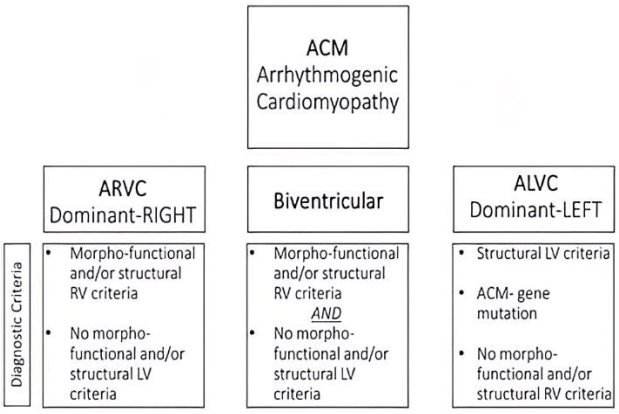

Figure 1. Diagnosis of ACM: The Padua criteria [2]

CASE REPORT

A 41-year-old male presented to emergency department with palpitations and associated dizziness. He was found to have ventricular tachycardia (VT) (**Figure 2**), and initial dose of 300 mg amiodarone intravenously was administered. Despite

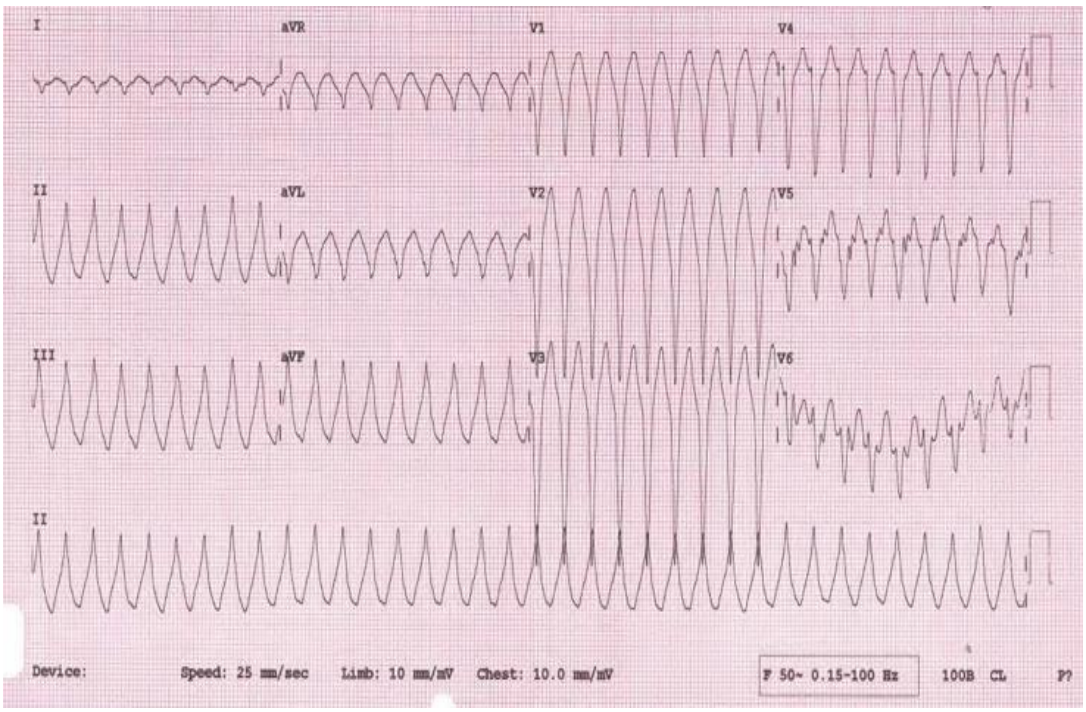

Figure 2. ECG on admission showing VT

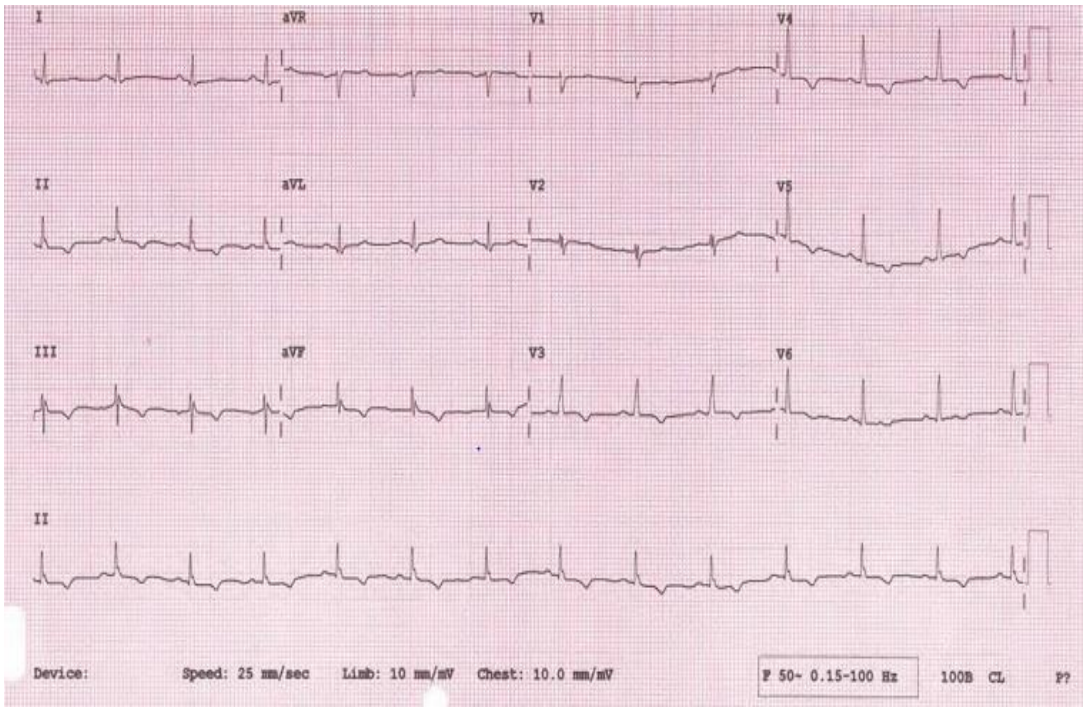

Figure 3. ECG following direct current cardioversion showing sinus rhythm with T-wave inversion in the anterolateral and inferior leads and incomplete RBBB

the amiodarone, he did not revert back to sinus rhythm. Therefore, one shock of 150 Joules was delivered and he was successfully cardioverted back into sinus rhythm. However, his ECG showed T-wave inversion in the anterolateral and inferior leads with incomplete right bundle branch block (RBBB) and ventricular ectopics (**Figure 3**). As per protocol, he was then started on an amiodarone infusion of 900 mg over 23 hours.

On admission, his bloods were unremarkable apart from a raised troponin, which was measured at 388 ng/mL and 469 ng/mL. Because of this, he was treated for a possible diagnosis of acute coronary syndrome (ACS). As such, he was started on ACS protocol, and received loading doses of aspirin and clopidogrel. However, his coronary angiogram showed unobstructed coronaries, therefore the dual antiplatelet therapy was stopped.

This gentleman previously had a transthoracic echocardiogram seven months prior to this admission, where he was found to have a left ventricular ejection fraction of 40-45%, borderline mild septal hypertrophy and mild general hypokinesis and anterior wall hypokinesis. Apart from this, his echocardiogram was unremarkable. The clinical indication for the echocardiogram was an incidental finding of widespread T-

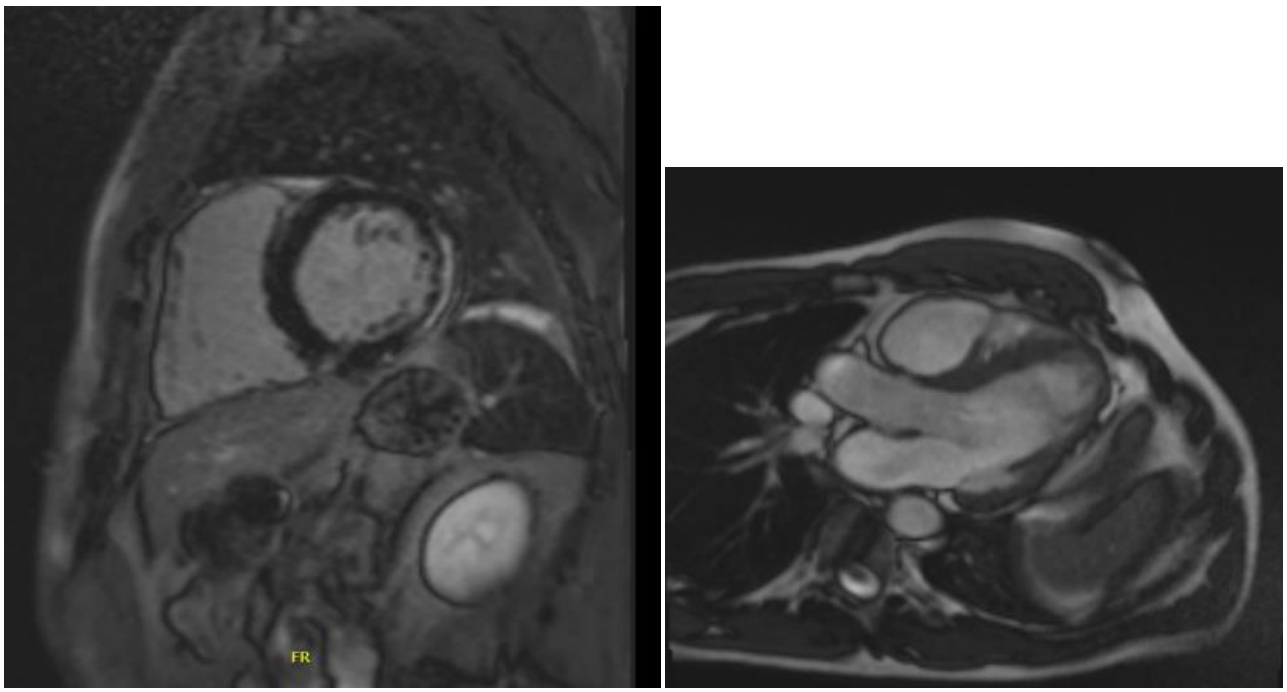

Figure 4. Late gadolinium enhancement and correspondent fibrosis replacement with known ischaemic pattern (subepicardial) clearly evident in LV chamber in 2-chamber view (left) & 4-chamber view (right)

wave inversion in the anterolateral leads. No treatment was initiated at this stage as the patient was completely asymptomatic. A strict follow up with cardiology team was put in place. However, the T-wave inversion was also present on this admission, which in hindsight is a minor criterion in the task force criteria.

In term of his background this gentleman was fit and well with no prior past medical history, nor any family history of cardiac disease. On examination, there were no obvious signs of heart failure, nor did the patient reports any symptoms in keeping with this. There were no stigmata of palmoplantar keratodermas or other signs of Carvjal or Naxos syndrome (desmoplakin mutations). He denied any mild or moderate infection during the intercurrent time from the first echocardiogram and this admission.

An echocardiogram was repeated during this admission (seven months after the initial one) because of the new ventricular arrhythmia to check for any signs of cardiomyopathy. It showed reduced left ventricular systolic function (LVSD) with an ejection fraction of 35-40%, and globally severe hypokinesia. It also showed new right sided failure with a moderately dilated right ventricle and reduced systolic function, as well as a mildly dilated right atrium. There was also new moderate tricuspid regurgitation and a mildly dilated pulmonary artery.

To further investigate these findings, a cardiac magnetic resonance imaging (MRI) was carried out. This showed biventricular dilatation, regions of akinesia and microaneurysm in the right ventricle. It also showed extensive subepicardial scarring at the left ventricle lateral wall and mid to apical inferior left ventricle, as well as patchy RV enhancement in the apical inferior right ventricle and anterior right ventricle at mid ventricular level (**Figure 4**).

There was also evidence of limited fatty infiltration in the subepicardial mid lateral wall and superior right ventricle at the mid ventricular level (**Figure 5**). These findings were

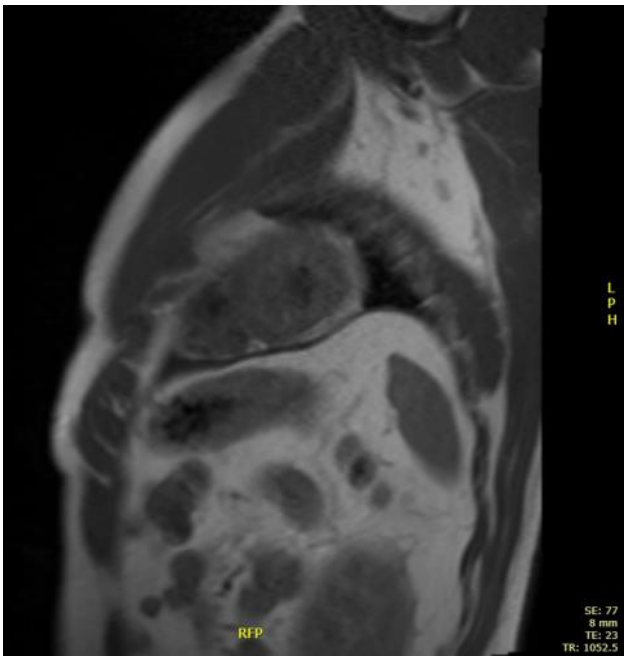

Figure 5. Fatty infiltration inferior apical LV chamber

consistent with a biventricular ACM based on morpho-functional criteria.

In view of the above, this patient was fitted with an ICD and referred to a specialist center for ongoing management, genetic family screening and follow up.

DISCUSSION

As shown in this case, young patients presenting with suspicious ECG changes such as widespread T -wave inversion, with no prior cardiac issues should be further investigated with an echocardiogram to look for cardiomyopathies as a possible

cause. If the echocardiogram results are indicative of underlying cardiomyopathy, then a cardiac MRI should be considered for the patient as the gold-standard to confirm such pathology.

In this case, the cardiac MRI confirmed the diagnosis and showed a relevant amount of fibrosis replacement, which is normally a late presentation characterized by a monomorphic arrhythmia and ventricular dysfunction. It is well established that at the beginning of this pathology, arrhythmias are normally more unstable and life-threatening because they behave and present more as a channelopathy.

An ICD is indicated in these patients due to risk of ventricular arrhythmias despite medical therapy and risk of sudden death. Medical therapy is used to prevent the occurrence of arrhythmias and hence prevent the needed for ICD activation. Such therapies include class II and III anti-arrhythmic drugs, for example sotalol and amiodarone respectively. For patients with recurrent presentations with VT, scar ablation can be considered. However, this has been shown to have varying levels of success.

Ultimately, risk factors do play a fundamental role in the progression of the disease, most of these are still not fully recognized and understood. So far, there are only few risk factors known: systemic infections, sport activity, myocarditis, etc. Physical activities despite ICD implantation needs to be restricted, and familiar screening also need to be implemented for early recognition, surveillance and treatment.

CONCLUSION

This case report of biventricular ACM provides an unusual insight into how rapidly the RV function can decline in the context of ACM. This patient entered biventricular failure within seven months of his first echocardiogram. This supports the notion that ACM is not only a myopathy affecting the right ventricle but can affect both ventricles. However, it questions whether biventricular failure occurs in later stages of disease, and the timeline between the stages of ACM. The main limitation of this report is that we are unable to comment on the long-term outcome. Large scale retrospective studies will enable further characterization of the stages of ACM and evaluate the prognosis in relation to patients' age, demographics and genetic information.

Author contributions: All authors contributed to the drafting the case report and revision of manuscript. All authors were directly involved in patient care or have undertaken or interpreted patient investigations.

Funding: No funding source is reported for this study.

Declaration of interest: No conflict of interest is declared by authors.

Data sharing statement: Data supporting the findings and conclusions are available upon request from the corresponding author.

REFERENCES

1. Marcus FI, McKenna WJ, Sherrill D, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D). Circulation. 2010; 121(13):1533-41. https://doi.org/10.1161/CIRCULATIONAHA.108.840827
2. Corrado D, Marra MP, Zorzi A, et al. Diagnosis of arrhythmogenic cardiomyopathy: The Padua Criteria. Int J Cardiol. 2020;319:106-14. https://doi.org/10.1016/j.ijcard.2020.06.005