CASE REPORT

An unusual cause of sustained ventricular tachycardia in a 27-year-old man

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SUMMARY

We present an interesting, unusual and complex case of a young man who initially presented with symptoms suggestive of tuberculosis and later developed malignant ventricular arrhythmias. A diagnosis of cardiac sarcoidosis was made only after histological evidence was paired with his presentation of monomorphic ventricular tachycardia. In this case we highlight the current challenges faced in the choice of investigations and diagnostic criteria. Additionally, we have identified the difficulties in treatment and long-term management of such a multisystem disorder. Ultimately by doing so, we hope to encourage clinicians to be aware of making a diagnosis of cardiac sarcoidosis.

BACKGROUND

Ventricular tachycardia (VT) can be defined as heart rate that exceeds 100 bpm originating from the lower chambers of the heart (ventricles), associated with a wide QRS complex ≥ 120 ms. Monomorphic VT is distinguished from polymorphic VT by a stable QRS morphology. This can be in the absence or presence of structural heart disease.¹ When this arrhythmia occurs in the context of structural heart disease, the commonest cause is from scar tissue as a result of a previous myocardial infarction. Other important causes come from the group of cardiomyopathies, for example, arrhythmogenic right ventricular cardiomyopathy (ARVC), dilated cardiomyopathy and hypertrophic cardiomyopathy.²

Sarcoid is a multisystem granulomatous disorder. It is important to be aware that although sarcoidosis is rare, it is an important aetiological factor for ventricular arrhythmias. Cardiac involvement has been shown to produce symptoms in 5% of patients with sarcoidosis and is often missed. In such cases, there is a high risk of recurrence of the VT and sudden cardiac death. Acute myocardial infiltration can take many forms, one of which can be the presence of granulomatous nodules that can be the foci for malignant arrhythmias.³

CASE PRESENTATION

A 27-year-old Asian man initially presented to the chest clinic with a 6-month history of dry cough, weight loss, shortness of breath and night sweats. This was in the absence of any recent foreign travel or known exposure to infectious contacts. His chest X-ray revealed subtle left upper lobe opacities and predominant left hilar lymphadenopathy (figure 1).

The pleural fluid aspirate analysis demonstrated an exudate (protein 35.2 g/dL), and there were no acid-

fast bacilli (AFB). Gram stain was negative and no organism was grown on culture. Blood tests for a viral screen (HIV, hepatitis B virus, hepatitis C virus), tumour markers (lactate dehydrogenase, human chorionic gonadotropin, α -fetoprotein), full blood count, rheumatoid factor, serum protein electrophoresis, serum ACE, immunoglobulins and autoantibody screen returned within normal ranges. His renal and liver function was mildly deranged: urea 5.8 mol/L, creatinine 141 μ mol/L, bilirubin 19 μ mol/L, alkaline phosphatase 143 IU/L, alanine transaminase 44 IU/L, and corrected calcium 2.73 mmol/L. CT of the chest/abdomen with contrast reported conglomerate mediastinal lymphadenopathy, with pleural thickening associated with effusions and old scarring in the left upper lobe. He was also found to have bilateral ill-defined nodules with mild diffuse bronchial wall thickening, an abnormal liver enhancement with subscapular soft tissue and evidence of left ureteric obstruction. An ultrasound of his renal tract went on to confirm a moderate left hydronephrosis thought to be secondary to extrinsic compression from surrounding lymphadenopathy. He was treated empirically for a presumptive diagnosis of tuberculosis (TB) with isoniazid, rifampicin, ethambutol and pyrazinamide.

He proceeded to a bronchoscopy which showed general inflammatory changes. Three samples of

Figure 1 Chest radiograph demonstrated subtle left upper lobe opacity and predominant left hilar lymphadenopathy.

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To cite: Ali S, Rathod KS, Deaner A, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-201517

washings for AFB were smear negative. Biopsy of the left lower lobe of the lung showed multiple non-caseating granulomas with microcalcifications within multinucleated giant cells.

Several months later, he presented to the emergency department with palpitations and presyncope. He was found to be in sustained monomorphic VT at a rate of 186 bpm and was haemodynamically compromised with hypotension. The VT was successfully treated with synchronised direct current cardioversion, but his baseline ECG in sinus rhythm was notably abnormal with ST depression in septal leads (figure 2).

His echocardiogram exhibited a dilated right heart (right ventricle, right atrium) with reduced systolic function, evidence of pulmonary hypertension (estimated pulmonary artery systolic pressure of 62 mm Hg+right atrial pressure) and a moderate global pericardial effusion 1.5 cm max across the apical region. His left ventricular ejection fraction was preserved at >55% and all valves were normal except for moderate tricuspid regurgitation.

Sustained VT recurred during his in-patient stay and he was transferred to the tertiary cardiac centre for further investigation and management. He proceeded to a cardiac MRI which demonstrated a dilated pulmonary artery, mildly impaired left and right ventricular systolic function, and global dyskinesia of the left ventricle with flattening of the septum. A lesion 20×11 mm appeared firmly attached to the myocardium at the level of the mid-lateral wall of the left ventricle with focal fibrosis not thought to be part of any myopathic process. A small pericardial effusion was also identified.

The complex imaging paired with the histological finding of non-caseating granulomas supports a unifying diagnosis of systemic sarcoidosis rather than TB. The substrate for VT is suspected to be granulomatous infiltration of the myocardium or perhaps the mass lesion attached to the left ventricle. The pulmonary artery hypertension was attributable to extrinsic compression of the pulmonary artery by enlarged lymph nodes and scarring of the upper lobe of the left lung. He was ultimately managed with steroids and implantation of an implantable cardioverter defibrillator (ICD) for secondary prevention.

DIFFERENTIAL DIAGNOSIS

Sarcoidosis is the most likely diagnosis given the bronchoscopy findings of multiple non-caseating granulomas with microcalcifications within multinucleated giant cells.

However, TB is an essential differential diagnosis to consider in a young Asian man presenting with a chronic history of weight loss, shortness of breath and night sweats.

Lymphoma and malignancy are less likely to be the cause in this case given the histological evidence but should be considered as important diagnoses to exclude. Thus far it is not possible to exclude the concealed phase of ARVC as a cause of VT, but this condition would not be associated with pulmonary infiltrate/hilar lymphadenopathy.

Ethambutol is also rarely reported to cause VT, but does not cause structural heart disease.

TREATMENT

The treatment in this case was implantation of a cardioverter defibrillator to reduce the risk of sudden cardiac death due to ventricular arrhythmia. Antiarrhythmic medication may suppress VT but is not effective in reducing mortality. On confirmation of the diagnosis with histological evidence, the patient was promptly treated with steroids. This is to be reviewed and gradually tapered down over time. He is also on bone protection and has regular review of his fasting glucose readings.

He remains on small doses of β -blocker bisoprolol and ACE inhibitor ramipril.

OUTCOME AND FOLLOW-UP

The cardiology and respiratory teams are jointly following up this patient. He has regular outpatient follow-up with the cardiology team including ICD checks to monitor for arrhythmias. Review of the structural and functional capacity of this young man's heart is important. He is at risk from further infiltration of his myocardium that can result in congestive cardiac failure, pericardial effusions (rarely tamponade), valvular pathology and cor pulmonale (from lung involvement).

The respiratory team will be assessing the need for steroid treatment in terms of dosage and length of treatment as well as

Figure 2 ECG demonstrating sustained monomorphic ventricular tachycardia with a ventricular rate of 186 bpm.

the long-term effects of the condition and additional medication.

DISCUSSION

Most commonly, sarcoidosis is thought to affect the lungs. However, myocardial involvement was first established in 1952 by Longcope and Freiman. The left ventricular free wall is the most frequently affected section of the myocardium.⁴ VT is the commonest first presentation of arrhythmic disturbance in patients with cardiac sarcoidosis. This can arise from sarcoid granulomas that form the foci of abnormal conduction exhibiting automaticity.⁵ In the case of our patient, the lesion of the left ventricle found on cardiac MRI could be the underlying cause.

This patient was treated for TB given his suggestive symptoms. However, the diagnosis of sarcoidosis did not become apparent until the patient presented with an arrhythmia. This shows the difficulty of identifying such patients at the onset of symptoms. The guidelines developed in 1993 by the Japanese Ministry of Health and Welfare identify patients with cardiac sarcoidosis through two groups. The first group has histological confirmation of cardiac sarcoidosis. The second group has extra-cardiac histological confirmation of sarcoidosis with common clinical manifestations, for example, cardiac arrhythmias.⁶ These guidelines are all that are available to date and unless there is a high clinical suspicion of cardiac involvement or sarcoidosis is included in the list of differential diagnoses when relevant patient groups present with arrhythmias, the diagnosis is often missed or delayed.

In this case, the diagnosis of cardiac sarcoidosis was eventually reached when histological evidence from a transbronchial biopsy was paired with the presentation of monomorphic VT. To add further to the diagnostic challenge of cardiac sarcoidosis, endomyocardial biopsy usually has a low yield for tissue confirmation as myocardial infiltration is patchy.⁷

There are several cardiac imaging modalities that can be used to achieve the diagnosis of cardiac sarcoidosis, each of which is useful for different areas of investigation and management, the most sensitive being contrast-enhanced MRI which correlates well with disease activity.⁸

In the case of this 27-year-old Asian man, treatment was initiated, once histological confirmation from a transbronchial biopsy was obtained, with steroids. He did not go on to have an endomyocardial biopsy and all TB medications were stopped. He is currently doing well and his symptoms are stable. The main limitation in his further management is that he will not be able to have repeat MRI to determine response to treatment because of his implanted defibrillator. Further studies and subsequent guidelines have yet to be established for

the long-term management of such patients. Multidisciplinary teams will remain involved to assess symptomatic control and clinical response.

Learning points

- ▶ Monomorphic ventricular tachycardia secondary to structural heart disease increases the risk of sudden cardiac death.
- ▶ Sarcoidosis is a rare but important cause of life-threatening ventricular arrhythmia.
- ▶ ECHO, cardiac magnetic resonance imaging and transbronchial biopsy are important in making the diagnosis of sarcoidosis, where as endomyocardial biopsy may not yield diagnostic material.
- ▶ There is a need for updated guidelines for the investigation and management of cardiac sarcoidosis to deal with the current diagnostic challenges.
- ▶ A multidisciplinary team approach should always be adopted when managing such complex cases of multisystem disease.

Contributors SA and KSR prepared the first draft of the manuscript. FF and AD proof read and amended the manuscript and prepared the final draft for publication.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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