A 28 year old woman with ventricular tachycardia and an abnormal chest radiograph

M Heath, H Deubner, E J Stern, J E Cooke, E A Gill

A 28 year old white woman with no medical history presented to the emergency room with symptomatic non-sustained ventricular tachycardia. She was asymptomatic up to a few days before presentation. Her physical examination was essentially normal and hence did not help with the differential diagnosis of the problem. Bronchoscopic transbronchial biopsy led to the final diagnosis of cardiac sarcoidosis.

The following case report is presented and discussed by the authors.

CASE PRESENTATION
Michele Heath
A 28 year old woman with no medical history presented to the emergency room with complaints of five days of recurrent palpitations and occasional midepigastric chest discomfort. She had no associated symptoms of dyspnea, nausea, vomiting, diaphoresis, lightheadedness, or syncope.

Her review of systems was negative for fever, rash, cough, haemoptysis, myalgias, or arthralgias or for a history of cardiac, pulmonary, or renal disease. She reported normal exercise tolerance and played tennis regularly. She did report a 9 kg (20 lb) intentional weight loss over the previous three months.

The patient was originally from Moscow, Russia and had immigrated 21 years previously. She worked as a food handler, smoked half a pack of cigarettes a day, and did not use alcohol or injection drugs. She had no other HIV risk factors. She was taking no medications.

On physical examination, she was a young, fit appearing woman without distress. She was afebrile, with a blood pressure of 94/51 mm Hg, pulse of 80 beats/min, and oxygen saturation of 99% on room air. Sclera were anicteric and oropharynx was without lesions. Skin was without rash. No cervical, axillary, or inguinal lymphadenopathy was noted. Lungs were clear. On precordial examination, the rhythm was regular and no murmurs, gallops, or rubs were present. The cardiac point of maximal intensity was brisk and non-displaced. Abdominal examination was benign, without organomegaly. There was no clubbing or peripheral oedema. Neurological examination was intact. Admission laboratory results of creatinine, blood urea nitrogen, thyroid stimulating hormone, white blood cell count, haematocrit, platelets, and urinalysis testing were normal. Urine toxicology screen and β human chorionic gonadotropin were negative. A chest radiograph (fig 1) showed a diffuse interstitial infiltrative pattern. ECG (fig 2) showed left bundle branch block. During her evaluation in the emergency department palpitations recurred and ECG...
The patient was admitted to the telemetry unit and metoprolol administration was started. An echocardiogram (fig 4) was obtained and showed mild mitral regurgitation, mild left ventricular dilatation, and an estimated left ventricular ejection fraction of 40%. The middle and basal portions of the anterior and inferior intraventricular septum were thin and dyskinetic. The right ventricle was normal and the pulmonary artery pressures were normal. Normal coronary arteries were seen on cardiac catheterisation.

During her hospital course she continued to have recurrent episodes of non-sustained ventricular tachycardia. She remained hemodynamically stable. On hospital day 4 she developed sustained ventricular tachycardia and was mildly symptomatic; the tachycardia resolved spontaneously and lasted slightly longer than 30 seconds.

Further diagnostic investigation found an erythrocyte sedimentation rate of 2 mm/h, negative rheumatoid factor, and serum angiotensin converting enzyme (ACE) concentration of 38 U/l (normal range 14–87 U/l).

Chest computed tomography (CT), including high resolution images (fig 5), showed numerous very small nodular opacities throughout the middle and upper lungs in a perilymphatic distribution typical of an interstitial granulomatous process. There was no mediastinal or hilar lymphadenopathy.

A diagnostic procedure was performed.

DISCUSSION
Edward A Gill

The patient is a young white woman who presented with symptomatic non-sustained ventricular tachycardia. She was asymptomatic up to a few days before presentation. Her physical examination was essentially normal and hence did not help with the differential diagnosis of the problem. Although not mentioned, the patient should have had, based on the conduction defect, a paradoxically split second heart sound. The ECG is helpful in that it shows a conduction defect of the left bundle branch type. The combination of the conduction defect, the septal wall motion abnormality on the echocardiogram, and the interstitial pattern on her chest radiograph suggests a systemic disease.

Ventricular tachycardia is always a worrisome rhythm. Sustained ventricular tachycardia can be life-threatening, regardless of its cause. Except for unusual causes of ventricular tachycardia, such as would be seen in a re-entrant tachycardia involving the right ventricular outflow tract, ventricular tachycardia is associated with and caused by structural heart disease. Structural heart disease is most commonly caused by myocardial infarction and subsequent scar formation.

The presence of a septal wall motion abnormality consisting of paradoxical motion and thinning on the echocardiogram suggests an old myocardial infarct. However, a myocardial infarct secondary to atherosclerotic coronary artery disease would be distinctly unusual in a 28 year old woman, unless risk factors such as familial hypercholesterolaemia were present. She had no family history of hypercholesterolaemia or myocardial infarction occurring at a young age. Other causes of myocardial infarction in a young woman would include cocaine use, Kawasaki disease with subsequent coronary aneurysm formation, and spontaneous dissection of a coronary artery. The patient denied cocaine use and a...
the disease affects mainly whites. The coronary angiogram did not show the characteristic aneurysm formation that would be seen in Kawasaki disease. Dissection of the coronary arteries has its highest incidence in young women who are pregnant. The patient had no history of pregnancy and a ß human chorionic gonadotropin test was negative.

Non-ischaemic causes of a wall motion abnormality on echocardiography would include myocarditis, infiltrating cardiomyopathy, metabolic related cardiomyopathy such as that secondary to thyroid disease or pheochromocytoma, and an early idiopathic cardiomyopathy. The patient did not describe a recent viral illness to suggest a viral mediated myocarditis, although the lack of such history certainly does not exclude the diagnosis. Viral serologies were not performed, but these rarely establish a diagnosis. The patient’s thyroid stimulating hormone concentration was normal. Idiopathic cardiomyopathy is a diagnosis of exclusion. With regard to infiltrating cardiomyopathy, the two most likely causes of this uncommon condition are amyloidosis and sarcoidosis. I will dismiss amyloidosis at this point, as there was no plasma cell dyscrasia and no evidence of a chronic inflammatory process that would cause amyloidosis. In addition, the patient’s young age makes amyloidosis unlikely.

The findings on echocardiography are compelling for the diagnosis of sarcoidosis. Cardiac sarcoidosis should always be considered in a young patient with ventricular tachycardia and a conduction disorder. Sarcoidosis is a granulomatous disease of unknown cause that is relatively rare. The incidence is 10–40/100 000 in the USA and Europe. There is of course a remarkable diversity in the prevalence of sarcoidosis between racial groups, with African Americans being particularly affected. In the USA the ratio of affected African Americans to affected whites ranges from 10:1 to 17:1. In Europe, however, the disease affects mainly whites.1 Although infiltration of the lungs, reticuloendothelial system, and skin dominate its clinical presentation, virtually any tissue can be involved, and the heart is no exception. Despite this, only 5% of patients have clinical manifestations of cardiac disease.2,3 Conduction defects and ventricular arrhythmias are frequent clinical manifestations of cardiac sarcoidosis. However, conduction defects are considerably more common than ventricular tachycardia. Indeed, sarcoidosis is a relatively rare disease and ventricular tachycardia is a rare presentation of sarcoidosis.2,4 Conduction system disease varies from first degree atrioventricular block to bundle branch block to complete heart block.5 At necropsy, cardiac involvement has been shown in 20–30% of cases, most of which were cases of generalised sarcoidosis.6 Interestingly, myocardial sarcoidosis often affects young or middle aged patients without sex predilection.7 Particularly relevant to the present case is that the granulomatous infiltration of sarcoid can be patchy but it preferentially involves the upper septum,8 as seen in this case.

Ventricular arrhythmias caused by cardiac sarcoidosis can be serious and even fatal. Roberts and colleagues7 found that sudden death is the terminal event in 67% of sarcoïd heart disease deaths. The risk of sudden death has been related to the extent of cardiac involvement.4 In a series by Winters and colleagues,7 seven patients with cardiac sarcoïd, ejection fraction < 45%, and inducible ventricular tachycardia on programmed stimulation were studied. There was no improvement in arrhythmia with corticosteroids. In this era, antiarrhythmic treatment was more frequently used for malignant ventricular arrhythmias than today when implantable defibrillators have become standard of care. In this series, two patients had sudden cardiac death and four had recurrence of ventricular tachycardia on medical treatment. Four patients had an automatic cardioverter-defibrillator placed and received appropriate shocks.

Chest radiography and CT findings are consistent with pulmonary sarcoidosis. As far as can be discerned, the patient had no respiratory symptoms. Of note is that the serum ACE concentration was normal. However, serum ACE concentrations are not highly sensitive for sarcoïdosis10 and may be even less sensitive for cardiac sarcoïdosis.11,12

Several diagnostic procedures could have been considered, as well as further imaging studies. Perfusion defects have been noted in cardiac sarcoïdosis using thallium 201.13 Positron emission tomography has recently been advocated to diagnose cardiac sarcoïdosis.14,15 An endomyocardial biopsy might have been considered, especially since the interventricular septum appeared to be involved, the area where endomyocardial biopsy specimens are taken. Having said this, the rate of a false positive biopsy still would have been substantial, as the area of involvement was the proximal septum and endomyocardial biopsy specimens are more frequently taken from the mid to distal septum. In addition, the diagnostic rate achieved with endomyocardial biopsy is said to be low even in the setting of a high suspicion of cardiac sarcoïdosis.16 A transbronchial biopsy would probably yield diagnostic results, given the radiological evidence of pulmonary involvement.

In summary, I believe this patient suffered from cardiac sarcoïdosis and the diagnostic procedure was a transbronchial biopsy. If sarcoïdosis were confirmed by transbronchial biopsy, the involvement of the heart with sarcoïdosis would be essentially certain given the findings on echocardiography and the malignant ventricular arrhythmia, both characteristic of cardiac sarcoïdosis.

**RADIOLOGICAL FINDINGS**

**Eric J Stern**

In the case presented, the studding of the pleural surfaces is compelling evidence of granulomatous involvement of the subpleural lymphatics and is typical of sarcoïdosis, despite the lack of lymphadenopathy. In a patient with no respiratory symptoms, the high resolution CT features are very strongly suggestive of sarcoïdosis.

In the lung, sarcoïd granulomas are distributed primarily along the lymphatics and therefore in the peribronchovascular
The patient had an implantable cardioverter defibrillator and prednisone 60 mg/day for one month followed by tapering. Somewhat anecdotal and the patient was treated with prednisone did not seem to affect the arrhythmias. Sotalol was added to help suppress the arrhythmias. At last follow up one month after hospitalisation, the patient continued to have non-sustained episodes of ventricular tachycardia that are short and have not required use of the implantable cardioverter defibrillator.

**FINAL DIAGNOSIS**

The final diagnosis was cardiac sarcoidosis.

**REFERENCES**

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