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Giant Cell Myocarditis

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Abstract

Presentation

A pre-morbidly well 57-year-old lady presented to the Emergency Department in acute respiratory distress.

Diagnosis

The patient had an angiogram which was normal. Further investigation with an echocardiogram demonstrated cardiomyopathy. Within days of admission, she rapidly deteriorated into fulminant heart failure. Unfortunately, our patient succumbed to torsades de pointes arrhythmia. A post mortem revealed her underlying cardiac pathology: Giant Cell Myocarditis (GCM).

Treatment

She was provisionally treated for acute decompensated heart failure (ADHF) secondary to acute coronary syndrome (ACS), requiring diuretics, inotropes, and non-invasive ventilation in ICU.

Discussion

GCM mimics myocardial infarction and can cause heart failure and arrhythmias. It is rare but should be considered in young, fit patients. Cardiac MRI is a useful screening tool and biopsy confirms the diagnosis. Steroids and other immunosuppressing therapies improve survival and can avoid the necessity for heart transplant which is the most definitive treatment.

Introduction

GCM is a rare but fatal disease that afflicts a young and healthy population. Giant cells infiltrate the myocardium causing inflammation and fibrosis in this T-cell mediated autoimmune disease. It can present as acute heart failure, heart block or arrhythmia - often refractory to standard therapy. GCM is clinically indistinguishable from other more common presenting illnesses and diagnosis can only be confirmed by endomyocardial biopsy (EMB). Given its rarity, fulminant course and diagnosis necessitating histology, it is commonly found at autopsy. Our case illustrates the danger of GCM.

Case Report

Our patient presented with one day history of acute onset paroxysmal nocturnal dyspnoea. She also complained of chest tightness in the Emergency Department. The patient was a forty pack-year smoker and had no other known risk factors for cardiovascular disease. On examination, she was tachypnoeic and tachycardic but haemodynamically stable. On auscultation there was reduced air entry in the right lung base, heart sounds were normal and gross oedema was absent. Early differential diagnosis included ACS, decompensated cardiomyopathy, arrhythmia, and pulmonary embolism.

ECG demonstrated sinus tachycardia with premature ventricular contractions. Troponin was >1200 ng/L, peaking at >5000 ng/L on serial measurement. CRP measured 60 mg/L. An echocardiogram revealed a left ventricular hypokinetic basal region and moderate concentric hypertrophy, restrictive diastole, and reduced ejection fraction <30%. Angiogram performed was unremarkable.

Unfortunately, our patient deteriorated into respiratory failure from pulmonary oedema. She rapidly became critically unwell and had a cardiac arrest on day 9 of admission from which resuscitation was unsuccessful. Torsades de Pointes was seen on telemetry. Post mortem reported the heart was grossly abnormal, mottled and weighing 460g. Histologically severe diffuse infiltrates of lymphocytes and multi-nucleated giant cells with fibrous replacement of myocardial fibres (figures 1 and 2) were seen. The findings were consistent with GCM as the cause of death.

Figure 1: *High power histology section showing multi-nucleated giant cells (arrow), lymphocytes (arrowhead) and myocyte damage.*

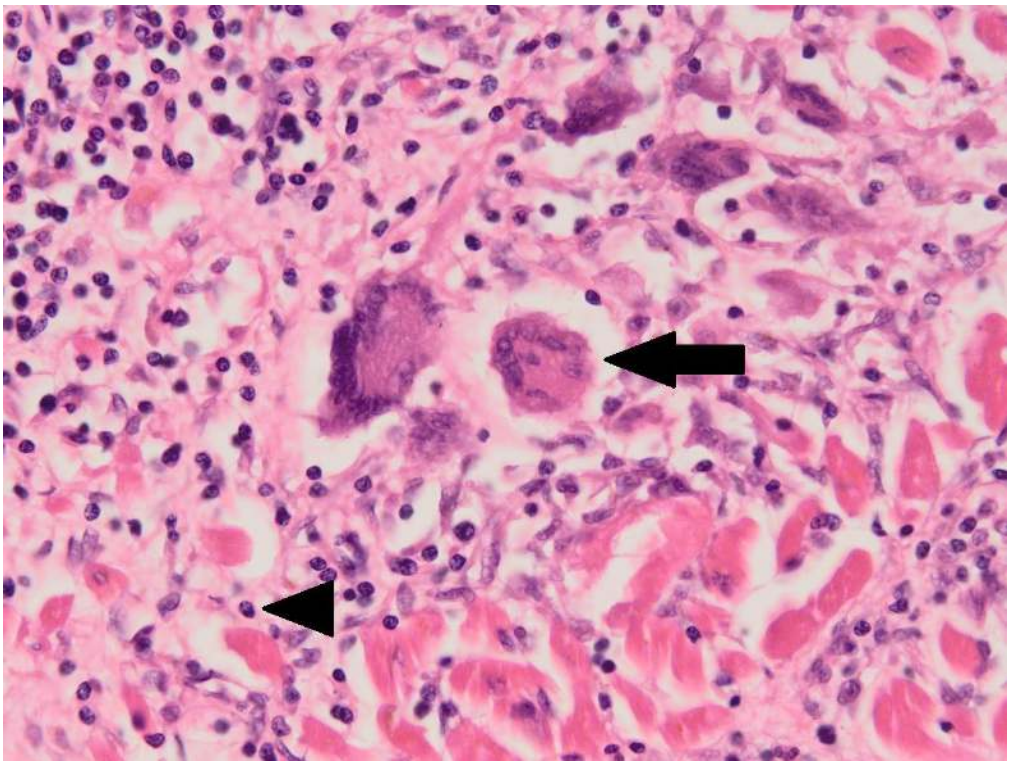
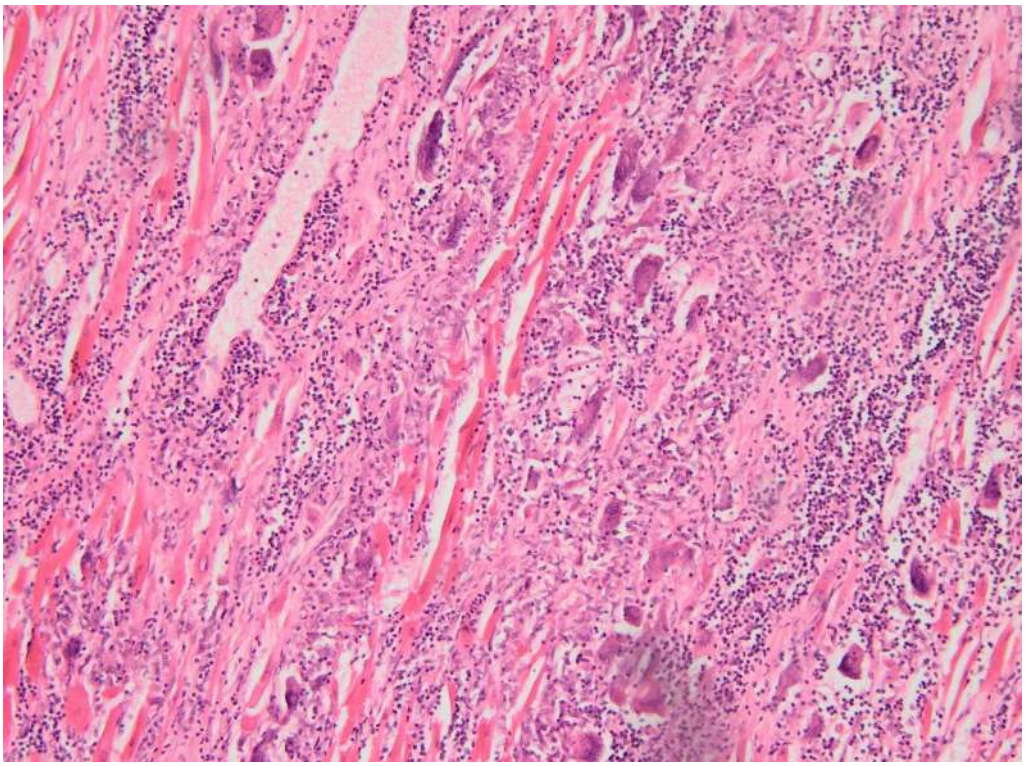


Figure 2: *Low power histology section of myocardium showing diffuse infiltrates of lymphocytes with scattered multi-nucleated giant cells. There is extensive damage to myocardial muscle fibres.*



Discussion

Our patient presented with a picture of ADHF that mimicked acute myocardial infarction. In addition, she was subject to arrhythmia, another common feature of GCM. Our patient had a normal angiogram which is a classic finding in GCM. There is a wider differential diagnosis than GCM in patients with similarly favourable demographics. Alternative myocardial pathology merits consideration in such patients who present with constitutional cardiac signs and symptoms. These broadly include viral, eosinophilic, lymphocytic, granulomatous, rheumatoid, and other autoimmune aetiologies.

One retrospective review of 32 patients recommended cardiac MRI or PET as a screening tool[1]. Cardiac MRI detects local myocardial disease and thus improves the yield for EMB. In the same study, patients with a high index of suspicion for GCM were subjected to repeat biopsies. This increased the sensitivity from 68% to 93%. Samples were typically taken from the right ventricle which is perceived as safer. An early multicentre database identifying 63 patients revealed a transplant-free survival of 5.5 months[2]. A more recent analysis of 46 patients would estimate a 42% survival rate at 5 years in the medically treated, transplant-free group[3].

Immunosuppression with high dose steroids should be initiated early in the management of GCM. This is typically combined with agents such as cyclosporine and azathioprine while the steroid dose is tapered. Mechanical circulatory support (MCS) has a role in fulminant presentations, but survival is still related to disease severity at presentation. Extra-corporeal membrane oxygenation (ECMO) or ventricular assist devices can serve to stabilise patients and bridge towards cardiac transplant, which is the most definitive treatment. In the largest multicentre cohort study to date examining the treatment outcomes of MCS, 9 of 13 patients were alive at 1 year[4]. Notably, all surviving patients had been transplanted before 1 year. ICDs have a place as arrhythmias due to myocardial scarring pose a risk which is not mitigated by immunotherapy. A systematic review and meta-analysis found disease recurred in 8% of transplanted patients, in line with its autoimmune pathogenesis. Continuing a regime of steroids and two immunosuppressive agents is advisable[5].

Declaration of Conflicts of Interest:
The authors have no conflicts of interest to declare.

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