New breathlessness in a young patient with rheumatoid arthritis

Introduction

This article presents the case of a 35-year-old man with a longstanding history of rheumatoid arthritis maintained on disease-modifying therapy. He presented with a short history of increasing breathlessness and was found to have a large pericardial effusion. Anti-inflammatory agents and antibiotics were initiated and then, following clinical deterioration, pericardiocentesis was performed successfully. After 3 weeks he represented with pleuritic chest pain, an abnormal electrocardiogram and was found to have developed new systolic left ventricular impairment with an apical thrombus.

A cardiac magnetic resonance study showed features consistent with a diagnosis of eosinophilic myocarditis and sub-epicardial fibrosis. He was anticoagulated with warfarin and responded well to systemic corticosteroid treatment. Follow-up cardiac magnetic resonance and echocardiogram showed resolution and he is currently doing well.

Discussion

Eosinophilic myocarditis, also known as Loeffler's endomyocardial fibrosis, is a rare form of immune-mediated myocardial inflammation (Kuchynka et al, 2016). Eosinophilic myocarditis may occur in the context of parasitic infections, drug reactions, hypereosinophilic syndrome and vasculitis (Li et al, 2015). In 36% of cases the eosinophilic myocarditis is idiopathic (Brambatti et al, 2017).

As approximately 10% of patients with rheumatoid arthritis have hypereosinophilia, there is a higher rate of eosinophilic myocarditis in this population (Diny et al, 2017).

Dr Holly Morgan, Cardiology Registrar, Department of Cardiology, Royal Glamorgan Hospital, Llantrisant CF72 8XR

Dr Abbas Zaidi, Cardiology Consultant, Department of Cardiology, University Hospital of Wales, Cardiff

Dr Richard Anderson, Cardiology Consultant, Department of Cardiology, University Hospital of Wales, Cardiff

Dr Rhian Goodfellow, Rheumatology Consultant, Department of Rheumatology, Royal Glamorgan Hospital, Llantrisant

Dr Gethin Ellis, Cardiology Consultant, Department of Cardiology, Royal Glamorgan Hospital, Llantrisant

Correspondence to: Dr H Morgan (morganhp7@gmail.com)

CASE REPORT

A 35-year-old man with a history of well-controlled rheumatoid arthritis presented to his GP with a 10-day history of breathlessness and pleuritic chest pain. He was prescribed oral antibiotics for presumed lower respiratory tract infection but did not respond and self-presented to the emergency department with worsening symptoms. Drug history included sulfasalazine, methotrexate and folic acid. He was a council worker and current smoker.

On examination he was morbidly obese (body mass index 57 kg/m^2), clammy, respiratory rate 35 breaths/minute and blood pressure 170/110 mmHg. Blood results showed a C-reactive protein level 120 mg/litre, high-sensitivity cardiac troponin T 656 ng/ml (normal <14 ng/ml), white cell count 15.4x10^9/litre, neutrophil count 10.2x10^9/litre and eosinophil count 0.7x10^9/litre (normal 0.0–0.4x10^9/litre). Electrocardiogram showed sinus tachycardia. Chest X-ray showed cardiomegaly and right basal atelectasis. A computed tomography scan of the thorax identified a large pericardial effusion (32 mm) with minor thickening and enhancement of the pericardium, mediastinal lymphadenopathy and areas of ground glass opacification bilaterally. Bedside echocardiogram confirmed a large pericardial effusion.

Broad-spectrum antibiotics, colchicine and ibuprofen were commenced, and his regular medications withheld. On day two he developed atrial fibrillation with a fast ventricular response. His condition deteriorated, blood pressure dropped to 105/70 mmHg and he underwent urgent pericardiocentesis – 900 ml of blood-stained fluid drained over the following 24 hours. Analysis revealed mixed inflammatory cells, predominantly neutrophils. There was no bacterial growth and no malignant cells detected.

He was given 3 mg intrapericardial dexamethasone and after 24 hours his pain and haemodynamic status improved and the pericardial drain was removed. Repeat echocardiogram showed a significant decrease in the effusion and well-preserved biventricular function. On day 8 he was discharged home with oral antibiotics and re-started his regular medication. High-sensitivity cardiac troponin T at discharge was 13 ng/ml.

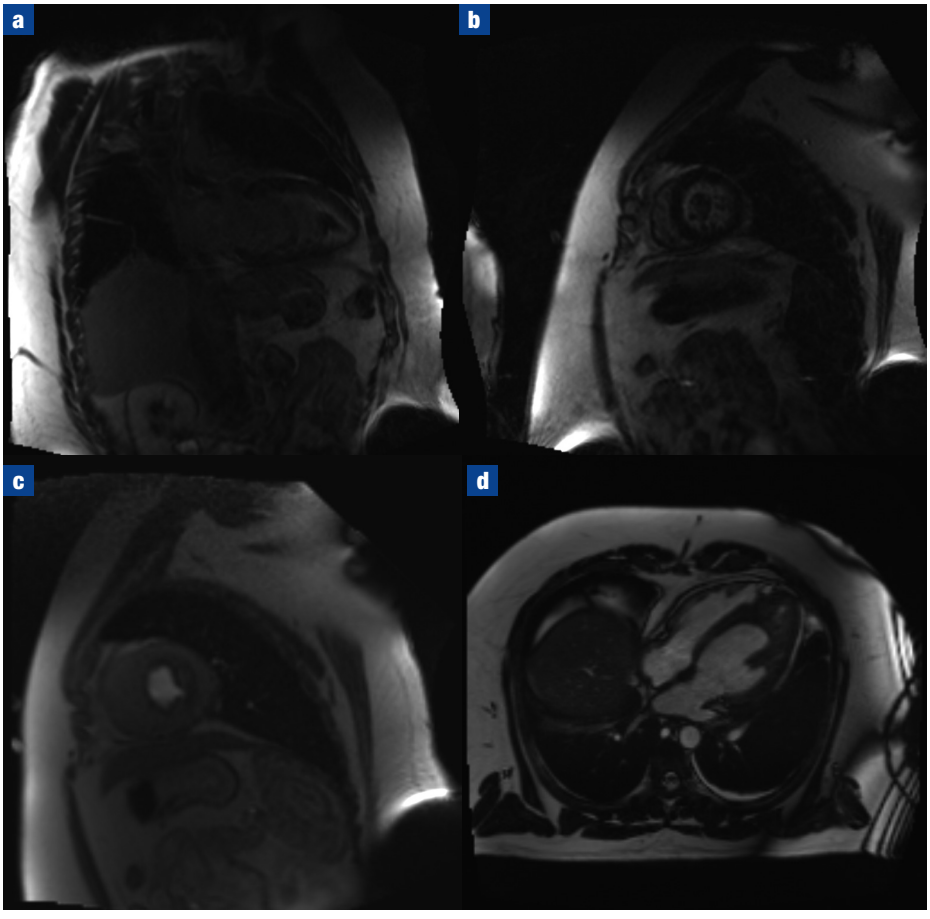
He re-presented 20 days later, reporting a 1-week history of increasing breathlessness with a recurrence of chest pain. Electrocardiogram showed sinus tachycardia with new inferolateral ST depression and ST elevation in aVR. Repeat echocardiogram showed minimal pericardial effusion but a large dense area in the left ventricle apex suggestive of thrombus with apical hypokinesia and mildly reduced LV systolic dysfunction. High-sensitivity cardiac troponin T was 1600 ng/ml and C-reactive protein 59 mg/litre.

The patient was transferred to the local tertiary hospital where he underwent invasive coronary angiography, showing normal coronary arteries. He underwent cardiac magnetic resonance which confirmed left ventricle apical thrombus, widespread sub-endocardial late gadolinium enhancement most prominent towards the apex (*Figure 1*) and a circumferential resting perfusion defect. These features were strongly suggestive of Loeffler's endocarditis with sub-epicardial fibrosis.

Heparin was initiated and, on the advice of rheumatology colleagues, so was methylprednisolone 1 g daily. After 3 days this was switched to oral prednisolone and warfarin. Fourteen days later he was clinically well and was discharged. He was followed up by cardiology and rheumatology and underwent repeat cardiac magnetic resonance 4 months after discharge which showed normal left ventricle function, minimal pericardial effusion, resolution of the sub-endocardial delayed enhancement and apical thrombus, and mild left ventricular hypertrophy.

© 2019 MA Healthcare Ltd

Figure 1. Cardiac magnetic resonance imaging scan. **a.** Apical two-chamber view and **(b)** short axis view – both late gadolinium images showing widespread subendocardial late gadolinium enhancement and the apical thrombus. **c.** Resting perfusion image showing circumferential subendocardial first-pass perfusion defect. **d.** Apical four-chamber cine view showing filling defect at the apex.



LEARNING POINTS

- Eosinophilic myocarditis can prove fatal if not diagnosed and treated early.
- The condition can mimic acute myocardial infarction.
- Diagnostic criteria are a useful aid for the clinician.
- Use of cardiac magnetic resonance imaging in diagnosis of eosinophilic myocarditis is now considered vital.
- Multidisciplinary team involvement is important in patients with organ-specific complications of chronic disease.

Eosinophilic myocarditis may present with shortness of breath (68%) or chest pain (41%) (Brambatti et al, 2017). A pericardial effusion is found in 38% of cases, with tamponade developing in approximately 5%. In 18% an endoventricular thrombosis is also found (Brambatti et al, 2017). Eosinophilic myocarditis can mimic acute myocardial infarction and therefore approximately 50% of patients undergo coronary angiography (Brambatti et al, 2017). Peripheral eosinophilia occurs in about 75% of cases (Morimoto et al, 2003). The diagnostic criteria for eosinophilic myocarditis include an eosinophil count above 500/mm^3 (or >0.5x10^9/litre) (JCS Joint Working Group, 2011).

Endomyocardial biopsy was previously the gold standard diagnostic investigation (JCS Joint Working Group, 2011), but developments in imaging (cardiac magnetic resonance and cardiac positron emission tomography–computed tomography) have allowed clinicians to opt for non-invasive alternatives (Brambatti et al, 2017), as demonstrated in this case.

Management involves use of analgesia, anticoagulation and treatment of the underlying cause (Kuchynka et al, 2016). In this case systemic corticosteroids were effective. Of interest, this patient developed eosinophilic myocarditis on dual immunosuppressive therapy when his disease was quiescent, raising questions about the immune-mediated aetiology of eosinophilic myocarditis.

Recurrent disease is common and difficulties in reducing immunosuppression are recognized, with newer therapies being considered including tyrosine kinase inhibitors and interleukin-1 (Kuchynka et al, 2016; Song et al, 2017). The role of long-term anticoagulation is poorly defined.

Conclusions

The challenging management of patients with rheumatoid arthritis presenting with breathlessness and chest pain is exemplified in this case. The significant differential causes of a pericardial effusion and the usefulness of non-invasive cardiac imaging modalities to support management are also highlighted. **BJHM**

Brambatti M, Matassini MV, Adler ED, Klingel K, Camici PG, Ammirati E. Eosinophilic myocarditis. J Am Coll Cardiol. 2017 Nov;70(19):2363–2375. https://doi.org/10.1016/j.jacc.2017.09.023

Diny NL, Rose NR, Čiháková D. Eosinophils in autoimmune diseases. Front Immunol. 2017 Apr 27;8:484. https://doi.org/10.3389/fimmu.2017.00484

JCS Joint Working Group. Guidelines for Diagnosis and Treatment of Myocarditis (JCS 2009). Circ J. 2011;75(3):734–743. https://doi.org/10.1253/circj.CJ-88-0008

Kuchynka P, Palecek T, Masek M, Cerny V, Lambert L, Vitkova I, Linhart A. Current diagnostic and therapeutic aspects of eosinophilic myocarditis. BioMed Res Int. 2016;2016:1–6. https://doi.org/10.1155/2016/2829583

Li H, Dai Z, Wang B, Huang W. A case report of eosinophilic myocarditis and a review of the relevant literature. BMC Cardiovasc Disord. 2015 Dec;15(1):15. https://doi.org/10.1186/s12872-015-0003-7

Morimoto S, Kubo N, Hiramitsu S et al. Changes in the peripheral eosinophil count in patients with acute eosinophilic myocarditis. Heart Vessels. 2003 Sep 1;18(4):193–196. https://doi.org/10.1007/s00380-003-0721-0

Song T, Jones DM, Homsi Y. Therapeutic effect of anti-IL-5 on eosinophilic myocarditis with large pericardial effusion. BMJ Case Rep. 2017 May 24;2017:bcr-2016-218992. https://doi.org/10.1136/bcr-2016-218992

© 2019 MA Healthcare Ltd