Reminder of important clinical lesson

A man awaiting a double transplant

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Summary

A 40-year-old gentleman previously fit and well presented to hospital on Christmas day in 2003 with dyspnoea, indigestion and 'pins and needles' down his left arm. Investigations in the emergency department were normal, so the patient was discharged from hospital with a diagnosis of panic attacks. A month later he re-presented to hospital, again with an indigestion-like pain in the chest, worsening dyspnoea, poor exercise tolerance and paroxysmal nocturnal dyspnoea. Myocardial infarction was diagnosed, as well as dilated cardiomyopathy. The patient was subsequently put on the heart transplant register. First he had a pacemaker put in, and 4 months later a defibrillator was inserted which dramatically improved the patient's signs and symptoms. He felt well in himself for 7 years; however began to deteriorate in August 2010. No heart transplants were available at the time, so the patient was offered a ventricular assist device in September 2010. The patient went into kidney failure in December 2010 and haemodialysis was commenced in January 2011. He is currently at home awaiting a double heart and kidney transplant.

BACKGROUND

I think this case is important because a missed diagnosis of dilated cardiomyopathy in the emergency room is worth reminding readers about. This patient's cardiomyopathy was diagnosed as a 'panic attack' the first time he presented to hospital, and he was discharged home without follow-up. It is important to be aware of the vague signs and symptoms that cardiomyopathies can present with, therefore this is a reminder of an important clinical lesson.

CASE PRESENTATION

Presenting complaint

Shortness of breath, 'pins and needles' down the left arm, and indigestion.

History of presenting complaint

On 25 December 2003, a fit and well 40-year-old gentleman presented to the emergency department with severe indigestion, dyspnoea and paraesthesia affecting the left arm. Following a full history, examination and unremarkable investigations, including an ECG, chest x-ray and a blood test which revealed a negative troponin, the patient was reassured and sent home from the emergency department with a diagnosis of panic attack.

The patient re-presented to hospital a month later again with indigestion, dyspnoea and paraesthesia in the left arm, as well as worsening exercise tolerance and paroxysmal nocturnal dyspnoea. The blood test on this occasion revealed a raised troponin level and so a diagnosis of myocardial infarction was made. He was admitted to hospital for treatment.

Further cardiac investigations revealed dilated cardiomyopathy and persistent atrial fibrillation. The patient had a biventricular pacemaker inserted, followed by an implantable cardioverter-defibrillator (ICD) device 4 months later. His symptoms were under control for 7 years; however he

began to deteriorate in August 2010 and was in end-stage cardiac failure. At the time there were no heart transplants available, so the patient was offered an electrically powered heart ware left ventricular assist device (LVAD). This was implanted on 20 September 2010 with ASD closure, with a permanent electrical line that exists through his abdomen (figures 1–3).

Prior to LVAD implantation, the patient had had two episodes of acute kidney injury requiring haemofiltration. However following the LVAD implantation he had rapidly deteriorating renal function with acute on chronic renal impairment. Postsurgery he was found to have worsening peripheral oedema, increasing shortness of breath, six pillow orthopnoea and occasional paroxysmal nocturnal dyspnoea due to his severe cardiac failure. He required inotropic support and was initially treated with intravenous diuretic

Figure 1 This image shows the midline sternotomy scar from the sternotomy incision that was made to insert the ventricular assist device during open heart surgery. This image also shows a long-term tunneled catheter access for haemodialysis.

Figure 2 A cable connects the external control unit and internal ventricular assist device through a small incision in the abdomen.

Figure 3 Control unit.

therapy, followed by a 2 day course of oral Metolazone, a thiazide-like diuretic. This had resulted in the removal of too much fluid causing volume depletion and hypotension, causing symptoms of dizziness and fainting. He was found to be in acute renal failure and emergency haemofiltration was started.

His postoperative complications consisted of persistent renal impairment requiring on-going renal replacement therapy, which was complicated by the fact that he was requiring epinephrine in order to maintain his mean arterial pressure (MAP) within a range that could be filtered. He also suffered a bleeding duodenal ulcer in October

2010, which was injected with epinephrine under endoscopic guidance and started on empirical *Helicobacter pylori* eradication therapy.

In December 2010, the patient was receiving haemofiltration four times per week on the ward. Since 13 January 2011 this patient has been at home, receiving haemodialysis through a Tessio line four times per week, lasting 6 h each time, as a day case at the hospital's haemodialysis unit.

The patient remains grossly fluid overloaded, with tense ascites, hepatic congestion and pedal oedema. At present he can manage to walk a few hundred yards and is in NYHA class III, but he does become breathless on heavy exertion.

Family history

The patient was adopted and therefore is not aware of his family's medical history.

Social history

The patient lives at home with his long-term partner. He admits to regular cocaine use in the past, although he has not used any illicit drugs for more than 8 years. He is an ex-smoker.

On examination

His jugular venous pressure is elevated by 3 cm. VAD sounds are normal. His chest has sparse left basal crackles but is otherwise clear on auscultation. Minimal pedal oedema is evident and although his abdomen is distended, it is tympanic and there are only mild bilateral flank dullness.

INVESTIGATIONS

Cardiac investigations

ECG, ambulatory ECG, echocardiography and exercise testing.

Renal investigations

Blood tests, urinalysis, osmolality studies, renal ultrasound. The patient did not have a renal biopsy.

Blood test

On 19 December 2011: Na=132, K=5.4, creatinine=652, urea=16.5, Hb=9.6, white cell count=12.5.

On 21 November 2011: Na=131, K=5.8, creatinine=651, urea=16.3, Hb=9.

On 15 March 2011: Hb=10.2, plasma Hb=0.3, white cell count=9.6, platelets=357, urea=11.1, creatinine=388, Na=3.137, K=5.9, bilirubin=28, albumin=32, C reactive protein=45.

On 12 October 2010: creatinine=203, urea=6.1, K=5.1, Hb=7.2, mean corpuscular volume=85.4, platelets=335

Echocardiogram

Mitral and aortic valves are thin with no obvious masses and pacing wires in the right place. Overall left ventricular (LV) systolic function is severely impaired with best contraction seen from the basal posterior segment. Markedly dilated and severely impaired right ventricle. Minimal central AR. Possible mild collapse of the anterior leaflet tip. Mild to moderate eccentric MR persists. Pacing wire in

right heart. No obvious pericardial effusion. Dilated and poorly responsive inferior vena cava.

Renal ultrasound

Kidneys are of normal echogenicity and of normal morphology measuring approximately 11.5 cm each. There is no evidence of obstructive uropathy.

DIFFERENTIAL DIAGNOSIS

- ▶ Previous MI x2 (December 2003 and January 2004)
- ▶ Dilated cardiomyopathy – cardiac resynchronisation therapy defibrillator implanted
- ▶ Heart ware left ventricular assist device implanted on 20 September 2010
- ▶ Renal failure – on haemodialysis
- ▶ Warfarinised
- ▶ Nasal septal perforation and re-current epistaxis
- ▶ End stage renal failure requiring dialysis four times a week at the Hammersmith Hospital
- ▶ Permanent atrial fibrillation
- ▶ Dyslipidaemia
- ▶ Peptic ulcer disease
- ▶ It is important to exclude a common condition affecting both the heart and kidneys in this patient, such as Fabry's disease.

TREATMENT

- ▶ Left ventricular assist device
- ▶ ICD
- ▶ Biventricular pacemaker
- ▶ Haemodialysis

Drug history (prior to renal failure); enalapril, spironolactone, furosemide, bisoprolol, atorvastatin, warfarin and (currently) bisoprolol 1.25 mg once daily, pravastatin 40 mg once daily, omeprazole 40 mg once daily, piriton 4 mg three times a day, amiodarone 200 mg once daily, warfarin as per international normalised ratio (INR) for the LVAD (target INR 1.7–2.3), erythropoietin (EPO) once per week.

OUTCOME AND FOLLOW-UP

His most recent tissue typing suggested that he is positive for HLA antibodies. HLA antigens are present in approximately 19% of the UK donor population. He is listed for a combined heart and kidney transplant.

DISCUSSION

Dilated Cardiomyopathy

Dilated cardiomyopathy is one of the commonest heart muscle diseases in developed countries. It is characterised by dilatation and impaired systolic function of either the right, left, or both ventricles, in the absence of coronary artery disease, hypertension, valveular disease and congenital heart disease. This leads to progressive cardiac enlargement and hypertrophy, a process known as remodelling.¹ The decreased cardiac function can affect other organs in the body such as the lungs.

Dilated cardiomyopathy is likely to be the result of damage to the myocardium caused by various toxic, metabolic, or infectious agents. It may be due to fibrous changes of the myocardium following a myocardial infarction. Other potential causes include congenital (X linked), postviral

infection, autoimmune, thyrotoxicosis, connective tissue disease, alcohol or cocaine abuse, haemochromatosis, peri or postpartum. However, in most patients it is described as idiopathic as no cause is apparent.^{2 3}

Dilated cardiomyopathy occurs more frequently in men than in women. It can occur in children; however it is most common between the ages of 20 and 60 years. Presentation is generally with congestive cardiac failure. About one in three cases of congestive cardiac failure is due to dilated cardiomyopathy.¹

Other associated symptoms include vague chest pain, syncope due to arrhythmias, and systemic embolism. There is a risk of sudden death with dilated cardiomyopathy.

Management of dilated cardiomyopathy

The aim of treatment is to relieve symptoms, slow down disease progression and prevent complications. Treatment involves bed rest, treatment of heart failure, anticoagulation, bi-ventricular pacing, implantable cardioverter-defibrillator and cardiac transplantation.

Pharmacological management of heart failure involves ACE inhibitors, β -blockers, spironolactone, loop diuretics and digoxin.⁴ ACE inhibitors antagonise the renin-angiotensin system, therefore lowering systemic vascular resistance and venous pressure. β -blockers inhibit the sympathetic nervous system, decreasing the heart rate and force, therefore improving symptoms, exercise tolerance, left ventricular function and mortality. β -blockers may also help prevent arrhythmias. Diuretics are used to relieve symptoms of fluid overload by promoting renal excretion of salt and water.^{2 3} Digoxin increases heart muscle contraction. When combined with ACE inhibitors and diuretics, they reduce death and hospitalisation resulting from progressive heart failure.⁵ Anticoagulants may also be used as patients with heart failure are at increased risk of thromboembolic events as their poor ventricular function promotes stasis.⁴

Non-pharmacological treatments include cardiac re-synchronisation therapy. In approximately 25% of patients with heart failure, intraventricular conduction delay is evident,⁶ resulting in asynchronous ventricular contraction. Asynchrony may further decrease systolic function and cardiac output in the already challenged heart.⁷ In light of this, cardiac re-synchronisation therapy has become a well-established therapy for patients with advanced heart failure. Cardiac stimulation re-synchronises ventricular contraction thereby improving left ventricular systolic function.

Pacemakers are the re-synchronisation therapy of choice used in patients with intraventricular conduction delay, and implantable cardioverter-defibrillators in those at risk of arrhythmias. However, one third of patients with cardiac re-synchronisation therapy do not show any clinical improvement.⁸ The gold standard treatment for heart failure is heart transplantation in patients with advanced disease refractory to medical therapy and cardiac re-synchronisation therapy.

Mortality studies have shown there has been an improvement in the survival of patients with dilated cardiomyopathy over the years. Statistics show that the majority of patients died within 3 years of the onset of symptoms. However, the prognosis has improved dramatically in

recent years with earlier diagnosis and better treatment which can slow down the progression of the disease. Death is commonly due to either congestive cardiac failure or ventricular arrhythmias.⁹

Learning points

- The vague symptoms that dilated cardiomyopathy can present with management of dilated cardiomyopathy.

Competing interests None.

Patient consent Obtained.

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