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Case Report

CASE REPORT

An unusual case of disseminated intravascular coagulation

Dmitri Pchejetski[1,2,*], Mojiba Kenbaz[1], Heba Alshaker[2,3], Dharmesh Rajput[1] and Kiruparajan Jesudason[1]

[1]James Paget University Hospital, Great Yarmouth, UK, [2]School of Medicine, University of East Anglia, Norwich, UK, and [3]Department of Pharmacology and Biomedical Sciences, Faculty of Pharmacy and Medical Sciences, University of Petra, Amman, Jordan

*Correspondence address. School of Medicine, University of East Anglia, Bob Champion Research & Educational Building, James Watson Road, Colney, Norwich, NR4 7UQ, UK. Tel: +44 (0)1603-59-1623; E-mail: d.pshezhetskiy@uea.ac.uk

Abstract

The use of cardiac pacemakers is increasing worldwide. Infective endocarditis from a pacemaker lead is a rare, but one of the most severe complications of pacemaker insertion. The diagnosis of pacemaker-related infective endocarditis is usually delayed due to unspecific clinical signs and symptoms at presentation compared to native valve infective endocarditis. Several factors can increase the risk of cardiac pacemaker-related infective endocarditis including cachexia, malignancy, diabetes mellitus, immunosuppression and corticosteroid treatment. This case report is about a 70-year-old diabetic male who presented to the emergency department with disseminated intravascular coagulation (DIC), cardiac and liver failure. He was diagnosed with pacemaker infective endocarditis, which was ultimately fatal.

CASE REPORT

History

A 70-year-old male presented to the Accident and Emergency Department with a 3-day history of painless jaundice, pruritus, non-productive cough and increasing shortness of breath with an exercise tolerance limited to 50 m. He also had diarrhoea, unintentional weight loss and felt generally unwell and lethargic. He denied any nausea, vomiting, urinary symptoms, chest or abdominal pain. There was no history of recent travel.

His past medical history included a cerebrovascular accident (5 years ago), Type 1 diabetes mellitus, ocular hypertension, depression, previous alcohol excess (three bottles of whisky a day for 8 years stopped 15 years ago (but no history of liver cirrhosis)) and a greater than 50 pack years smoking history. He had a significant cardiac history, suffering with angina, two previous myocardial infarctions, a coronary artery bypass graft and a biventricular pacemaker which was inserted in 2015. Additionally, he also suffered from aortic stenosis, mitral regurgitation and moderate to severe left ventricular (LV) impairment. His current medications included aspirin, spironolactone, furosemide, carvedilol, candesartan, sildenafil, clopidogrel, simvastatin, citalopram and insulin.

On examination

He was alert and oriented, looked unkempt, had gynaecomastia, proximal muscle wasting, was tanned but had no buccal pigmentation. He had a tattoo on his right arm. At rest he was apyrexial, respiratory rate of 18, O_2 saturations of 95% on air. His heart rate was 64 beats per minute (BPM), a normal jugular venous pressure and a capillary refill time of 3 s. On chest

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examination, he had bibasal crepitations and a pan-systolic murmur. His abdomen was distended with shifting dullness. There were no signs of peritonitis, no masses, no organomegaly and normal bowel sounds. There was no focal neurology. His lower limbs had evidence of venous eczema with mild pitting oedema and hyperpigmentation.

The following day the patient's temperature fell to 35.5°C, blood pressure dropped to 86/50, urine output dropped to 10 ml/h and he became more confused.

Investigations

At presentation the patient's electrocardiogram showed a ventricular paced rhythm with broad QRS complexes at 65 BPM. Urine dipstick was negative. Two sets of blood cultures were negative (patient did not have antibiotics prior to admission or prior to blood cultures being taken). His initial blood tests are shown in Table 1.

Computed tomography (CT) revealed: biventricular cardiac failure with reflux into inferior vena cava and hepatic veins, bilateral pleural effusions, moderate abdominal ascites, fatty liver, gallbladder sludge, but no stones or biliary tree dilatation, normal kidneys with tiny simple cysts and enhancement of both adrenal glands. No lymphadenopathy or masses.

Following the CT, a transthoracic echocardiogram was performed. It showed a 5 × 5 mm mobile echogenic lesion in the right atrium associated with the LV pacing lead, 15 mm from the coronary sinus ostium (images in Fig. 1 and video 1). Other findings included, severe impairment of LV systolic function consistent with a previous myocardial infarction and an endothelialised apical thrombus. While transoesophageal echocardiogram would have been an ideal investigation, the patient was deemed unfit for the procedure.

Differential diagnosis

Based on the history, blood analyses and the CT scan, a diagnosis of congestive heart failure with decompensated liver disease and disseminated intravascular coagulation (DIC) was made. The cause for the DIC was unclear. Malignancy (with liver involvement) was suspected, however, no masses were identified on the CT scan. Adrenal insufficiency (Addison's disease) was one of the top differentials on admission, so the patient was initially treated with supportive therapy, dexamethasone and vitamin K and kept for further investigations (shown in Table 2). A differential diagnosis of hepatorenal syndrome was considered; however, initial urea of 20.8 and Creatinine 135 together with low LFTs (normal ALP, ALT 185 and Alb 32) was not indicative.

Table 1: Blood analyses on Days 1 and 5 post admission

Test	Day1	Day 5	Reference range
C-reactive protein	24	20	<1 mg/l
White cell count	8.6	16.4	3.6–11.0 × 10^9/l
Haemoglobin	178	166	130–180 g/L
Platelets count	24	46	140–400 × 10^9/L
Mean cell volume	86		80–100 fl
Ferritin	486		25–350 ng/ml
INR	2.38	1.64	
Activated partial thromboplastin time	53		24–37 S
D-dimer	2147		<250 ng/ml
Fibrinogen	0.43		1.50–4.50 S
Sodium	122	129	135–145 mmol/l
Potassium	4.8	4.8	3.5–5 mmol/l
Urea	20.8	29.2	2.5–6.5 mmol/l
Creatinine	135	227	55–120 μmol/l
Glucose	8.0		3.2–6.0 mmol/l
Bilirubin	214	224	0–20 μmol/l
Alkaline phosphatase	106	85	20–140 U/l
Alanine transaminase	185	67	10–49 U/l
Albumin	32	26	32–48 g/l
Adjusted calcium	2.36		2.20–2.60 mmol/l
Prostate specific antigen		1.3	0.1 to 4.0 ng/ml

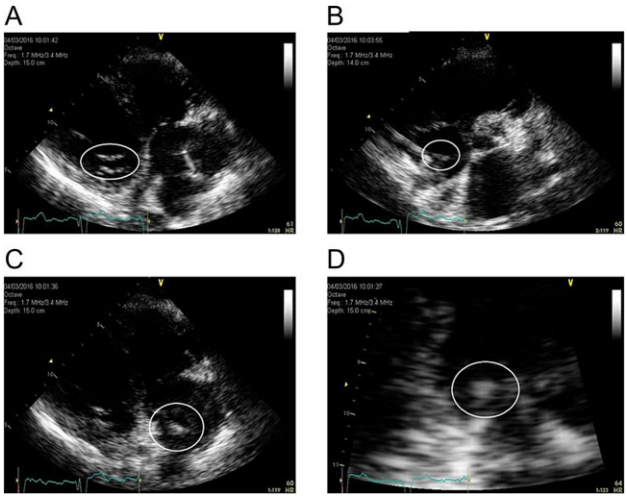

Figure 1: Transthoracic echocardiography images. (**A**) A two chamber view, long axis parasternal with posterolateral wall close to the LV lead. An echogenic mobile mass is circled. (**B**) A short axis view showing dilated right atrium and right ventricle with an echogenic mass circled. (**C**) An apical four chamber view, pacing lead can be seen at the right atrium encircled with an echogenic mass crossing the tricuspid valve. (**D**) A zoomed image C.

Table 2: Further investigations

Test	Value	Reference range
Serum virology hepatitis A	Negative	
Serum virology hepatitis A	Negative	
Serum virology hepatitis A	Negative	
Serum virology hepatitis A	Negative	
Serum virology hepatitis A	Negative	
Serum virology hepatitis EBV	Negative	
Alpha-feto protein (TM)	3.5	0.0–10.0 kU/
Alpha 1 Antitrypsin	1.56	1.10–2.10 g/l
Caeruloplasmin	0.38	0.20–0.60 g/l
Anti-mitochondrial antibodies	Negative	
Anti-smooth muscle antibodies	Negative	
Anti-liver, kidney microsomal antibodies	Negative	
Anti-gastric parietal cell antibodies	Negative	
Anti-reticulin R1	Negative	
Coombs test	Negative	
Blood cultures	Negative	
Sputum cultures	Negative	
Urine culture	Negative	
Faeces *cryptosporidium*	Negative	
Faeces *Giardia Lamblia*	Negative	
Faeces *Escherichia coli* 0157	Negative	
Faeces *Salmonella* species	Negative	
Faeces *Shigella* species	Negative	
Faeces *Campylobacter* species	Negative	

As the patient continued to deteriorate (hypothermia, low systolic pressure and low urine output), a diagnosis of sepsis causing DIC was made, which was narrowed to infective endocarditis based on echocardiogram results. Synacthen test and ascitic tap were, therefore, not performed.

Treatment and Outcome

Once the diagnosis of sepsis was made, the patient was treated with piperacillin–tazobactam, gentamicin and clarithromycin, supported by pabrinex, sliding scale insulin, IV cryoprecipitate and folate. Following echocardiogram findings, antibiotic therapy was changed to flucloxacillin and rifampicin and later switched to teicoplanin. Ideally the patient would have been transferred to a specialized cardiac unit for pacemaker extraction; however, given that the patient was haemodynamically unstable this was not possible. The patient continued to deteriorate with refractory hypotension and low urine output, despite active fluid resuscitation. He continued to have persistent hypothermia, desaturations and his blood tests showed worsening kidney function. Given the patient's poor prognosis a discussion between the medical team, patient and the family took place. A joint decision was reached that the patient was not for any further escalation of treatment and that he should not be resuscitated. Eight days later the patient died from multi-organ failure caused by aggressive pacemaker sepsis with associated DIC. Autopsy was not performed according to family wishes.

DISCUSSION

Endocarditis associated with a pacemaker lead infection is a rare but serious complication of pacemaker insertion [1]. The reported incidence varies from 0.13 to 7% [2]. Medical treatment alone is rarely successful, and current consensus is that the infected leads should be removed urgently [3]. Several factors can increase the risk of cardiac pacemaker-related infective endocarditis including cachexia, malignancy, diabetes mellitus, immunosuppression and corticosteroid treatment [4]. Staphylococci are involved in the majority of these infections. The natural history of such infections is devastating, with a mortality rate of 33%. Recent evidence suggests that the rate of infection seems to be rising [5], and together with the increased use of cardiovascular implantable electronic devices [6] this represents a new significant clinical challenge.

The presentation of such infections can be varied, including non-specific signs and symptoms of systemic infection such as fevers, chills, night sweats, malaise and anorexia. In up to 50% of cases, blood culture results are negative for growth [7]. In 2013, Ward *et al.* [8] described a case of pacemaker-related bacterial endocarditis in a patient who presented to the local emergency department with diarrhoea and productive sputum and then rapidly progressed to DIC and death. Fewer than 10% of patients present with septic shock. Patients may also present with secondary foci, such as spinal or pulmonary infection [9]. Clinical diagnosis can, therefore, be challenging and is often delayed.

It is crucial to suspect a diagnosis of endocarditis related to pacemaker lead infection in the presence of fever, complications or pulmonary lesions in patients with permanent pacemakers. Transoesophageal echocardiography should be performed to look for vegetations [9]. One clinical study proposed to use modified Dukes criteria for diagnosis of infective endocarditis on pacemaker leads [7], and this has been accepted by the recent British Society for Antimicrobial Chemotherapy, British Heart Rhythm Society, British Cardiovascular Society, British Heart Valve Society and British Society for Echocardiography joint guidelines [9]. The proposed management includes early and total explantation of pacing material and appropriate antibiotic therapy pursued for 6 weeks [10].

LEARNING POINTS

- Importance of including bacterial endocarditis in the early differential diagnosis in patients with implanted pacemaker devices presenting with signs of cardio/respiratory decline even in the absence of sepsis or evidence of infection.
- Transoesophageal echocardiography should be performed for diagnostic purposes.
- Management should be done according to the Duke's criteria and recent guidelines [9].

SUPPLEMENTARY MATERIAL

Supplementary material is available at *Oxford Medical Case Reports* online.

CONFLICT OF INTEREST STATEMENT

None declared.

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ETHICAL APPROVAL

Not required.

CONSENT

From patient's son.

GUARANTOR

Dmitri Pchejetski.

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