Postcardiotomy syndrome: recurrent cardiac tamponade and an exquisite steroid response

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SUMMARY

A 26-year-old woman presented moribund with fever and pleuritic chest pain 3 times in 4 months following elective aortic root surgery. She was admitted 41 days after surgery with cardiac tamponade requiring surgical drainage twice within 1 week. Despite this, she was re-admitted for a second time 4 days after discharge with persistent pericardial effusion. High fevers and an incidental regurgitant murmur were extensively investigated for and treated as possible endocarditis or graft infection without conclusive results. The patient spent a total of 61 days in hospital during this period, receiving seven different antibiotic courses. Her third admission, with most severe clinical features, nearly led to further surgery and removal of her aortic graft but instead culminated in a multidisciplinary team decision to initiate steroid therapy for postcardiotomy syndrome. A short course of oral prednisolone saw her pericardial effusion and symptoms resolve completely.

BACKGROUND

The case is particularly instructive from diagnostic and treatment perspectives. Postcardiotomy syndrome likely accounts for significant morbidity in patients who undergo instrumentation of their heart or pericardium. However, it is not typically associated with life-threatening pathology.¹ This case describes severe cardiac tamponade secondary to postcardiotomy syndrome. Further, the syndrome is generally regarded as an acute process presenting within the first month of instrumentation.² This case illustrates that the disease can be chronic with severe features after 3 months of initial presentation. Perhaps, the most relevant and practical lesson from this case centres on treatment. There is very limited trial data on treatment of postcardiotomy syndrome,³ especially in the context of life-threatening disease. Prolonged courses of steroids with their associated adverse effects, or surgical instillation of anti-inflammatory agents,⁴ are both unattractive treatment options that are among the guidelines for severe and chronic postcardiotomy syndrome. This case demonstrates how a 7-day course of steroids can treat severe and chronic disease with almost immediate effect and without recurrence up to at least 5 months of follow-up.

CASE PRESENTATION

A 26-year-old woman was diagnosed with Turner's syndrome when she was 2 years old and athetoid cerebral palsy at 3 years. Despite significant physical disability requiring assistance with most activities of daily living, and significant dysarthria, the patient was a bright and vivacious young lady with

high-functioning cognitive abilities. Owing to her athetoid movement disorder, it is important to note that the patient was electively intubated for all CT, nuclear medicine and trans-oesophageal echocardiogram (TOE) imaging.

She underwent aortic root replacement on 25 January 2012 with sparing of a well-functioning bicuspid valve. She was discharged on day 9 post-surgery. She received no therapeutic anticoagulation therapy.

She was seen in the grown-up congenital heart disease clinic on the 6 March, some 41 days post-surgery. She reported palpitations, malaise and bed-sheet soaking night sweats since discharge. Clinically, she was afebrile but appeared unwell. No pericardial or pleural rub, or new murmur, were documented. Her blood pressure (BP) was normal and heart rate (HR) was 120 beats per min (bpm), and her ECG demonstrated sinus tachycardia. Trans-thoracic echocardiography (TTE) showed severe cardiac tamponade. The replaced aortic root appeared well seated; there was a mild degree of new aortic valve regurgitation. Her C reactive protein (CRP) was 14 with a normal white cell count (WCC). She was admitted to the hospital and underwent surgical pericardial drainage via the lower end of her sternotomy wound. The appearance of the fluid was serous; microbiology and cytology studies were unremarkable. The pericardial drain was removed at 48 h after minimal output. The patient continued to feel unwell with vomiting, left-sided pleuritic chest pain and palpitations. She remained afebrile and haemodynamically stable. Within 4 days, TTE demonstrated re-accumulation of the pericardial effusion and tamponade. She underwent re-do surgical pericardial drainage. She completed 5 days of high-dose aspirin therapy. The pericardial drains were removed for the second time on 19 March. TTE demonstrated a small localised residual pericardial effusion but no right ventricular collapse. She felt subjectively better. She was discharged on the 28 March feeling well, afebrile and with a CRP of 11. She was due to complete a further 14 days of ibuprofen from the day of discharge, and advised to monitor her temperature at home.

On 1 April, she was re-admitted for the second time since her surgery. Drenching night sweats had returned with left-sided pleuritic chest pain, vomiting and malaise. She appeared unwell clinically and was febrile to 38.9°C. She was tachypnoeic, her BP was 99/58 and her ECG showed sinus tachycardia at a rate of 128 bpm. There were no clinical stigmata of endocarditis. Urinalysis was negative for blood and nitrites. Neither pericardial nor pleural rub was documented. The diastolic murmur of

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aortic regurgitation was identified clinically. Her CRP climbed to 286 by day 3 from 86 on admission. Her WCC peaked at 12 on admission, but normalised thereafter. Chest radiography was unremarkable. On TTE, her pericardial effusion appeared unchanged compared to the previous images 4 days prior. White cell scan demonstrated diffuse non-specific pulmonary uptake. Positron emission tomography (PET) CT imaging showed mild fludeoxyglucose (FDG) uptake at the aortic root which was described as inflammatory. No pericardial or pleural effusion was identified, and no FDG avid disease above or below the diaphragm was found that might explain the fevers. She was initiated on broadspectrum antibiotics. Review by the infectious disease microbiology consultants suggested that culture-negative endocarditis or graft infection were the most likely diagnoses. Mild splenomegaly on ultrasound scanning (USS) of the patient's abdomen supported the diagnosis of possible endocarditis. However, TOE revealed no clear vegetation or abscess despite moderate aortic regurgitation. Incidentally a left atrial appendage thrombus was identified. The pericardial effusion remained static. Teicoplanin was added to her antibiotic regimen and she initiated warfarin therapy for the newly diagnosed thrombus without complications. She improved clinically with progressive reduction in her inflammatory markers. However, 9 days later, she spiked a temperature of 38.9°C and her CRP rose once more to 131. Her TTE appearances did not change. She reported feeling well. On microbiology advice, these fevers were attributed to her teicoplanin therapy. The course was stopped on day 15. To this point, no blood cultures had yielded significant growth. She was discharged on 25 April, feeling well, afebrile and her CRP was 3.

Within the week, she was re-admitted for the third time from home after self-measuring a temperature of 38°C. On admission, she was very unwell. She was moribund, clammy and in tears. She described heavy sweats, severe weakness and left-sided pleuritic chest pain. Her HR was 110 bpm and BP 100/60 mm Hg. The decision was made to monitor clinically and wait the results of further microbiology screening. However, she continued to spike temperatures over 39°C and clinically deteriorated further, 'aching all over' with severe lassitude. Her HR peaked at 130 bpm but her BP did not alter significantly. Her CRP peaked at 152. Repeat TTE and TOE were unchanged with a persistent small pericardial effusion. She was started on vancomycin, and then moved to linezolid and gentamicin, given the severity of her clinical features. Her fevers initially improved on antibiotic therapy, but she continued to spike fevers and feel unwell. At this point, the team remained concerned that aortic root graft infection was the most likely explanation. The surgical team planned to re-open her chest wall for the third time for a re-do aortic root replacement. However, in view of this high-risk operation and with the extensive but inconclusive data available, a multidisciplinary discussion between cardiology, cardiothoracic surgery and microbiology departments concluded that she should commence a trial of steroid treatment for postcardiotomy syndrome (PCS) before proceeding to surgery. She stopped antibiotics and initiated oral prednisolone at a dose of 1 mg/kg for 7 days. Her symptoms improved with astonishing immediacy and her CRP fell synchronously within 24 h. On day 6 of steroid therapy, her CRP had fallen to 9 and her TTE showed complete resolution of the pericardial effusion. She was kept in hospital for a further 48 h after stopping the short course of steroids during which time she remained asymptomatic. She was discharged on 14 May with PR 92 bpm and CRP 2.

She was seen as an outpatient on 21 May. She reported no symptoms in the intervening period. TTE on the day again showed no pericardial effusion. She remains well at the time of article submission.

INVESTIGATIONS

Multiple TTEs: Two TOEs requiring elective intubation. Radiology: white cell scan, PET CT, CT chest, abdomen, pelvis, Ultrasound scan abdomen, multiple chest x-rays. Multiple blood cultures.

DIFFERENTIAL DIAGNOSIS

- ▶ Endocarditis
- ▶ Aortic root graft infection

TREATMENT

7 days 40 mg oral prednisolone.

OUTCOME AND FOLLOW-UP

The patient abruptly responded to high-dose oral steroid treatment within 24–48 h. The course was truncated to 7 days given her startling recovery. Since completing her treatment, she has remained completely well without TTE evidence of even sub-clinical disease up to 3 months of follow-up.

DISCUSSION

There are a handful of case reports that describe cardiac tamponade as a complication of PCS in the absence of anticoagulation therapy. However, the range of cardiac instrumentations represented in these reports is broad: pacemaker implantation,⁵ open heart surgery,⁷ and even penetrating cardiac trauma.⁸ In all of these cases, initial presentation is the most common between the second and fourth week postprocedure.

Published cases of patients continuing to present with PCS symptoms despite apparent definitive management of cardiac tamponade are vanishingly rare. One such case report in Mexico describes cardiac tamponade in the context of PCS in a patient presenting 35 days after atrial septal defect repair.⁹ Despite pericardiocentesis and low-dose prednisolone therapy on that first admission, the patient was re-admitted 1 month later with persistent fever, chest pain and pericardial effusion. Similar to the case we describe, there was rapid resolution of the patient's symptoms and effusion on initiation of higher dose (40 mg/day) prednisolone within 10 days. Other case series have recorded the 'exceptional' need for repeated pericardiocentesis to manage tamponade in the setting of PCS,¹⁰ but this is within a single admission.

According to the European Society of Cardiology, Systemic corticosteroid therapy should be restricted to those patients 'with poor general condition' or who experience 'frequent crises'. The recommended regimen is: prednisone 1–1.5 mg/kg, for at least 1 month. Long-term (3–6 months) oral steroids or preferably pericardiocentesis and intrapericardial instillation of triamcinolone (300 mg/m²) are therapeutic options in refractory forms.⁴ Our case report suggests that even severe and persistent disease may respond very effectively to a much shorter course of treatment. There are case reports for other therapies. Single doses of intravenous methylprednisolone¹¹ and intravenous immunoglobulin have been used effectively for relapsing PCS.¹² In the longer term, methotrexate has also been used successfully in one steroid-dependent paediatric patient with PCS.¹³

Learning points

- ▶ Postcardiotomy syndrome (PCS) is a life-threatening condition.
- ▶ Severe disease may present late and develop chronic features. Trans-thoracic echocardiography should be a first-line modality of any effective monitoring.
- ▶ It is a diagnosis of exclusion that means extensive and invasive investigations, for other life-threatening diagnoses cannot be overlooked. Diagnosis should be a multidisciplinary team decision.
- ▶ Severe and chronic PCS that is refractory to simple anti-inflammatory treatments may remain exquisitely sensitive to only a short course of oral steroids.
- ▶ Despite severe and chronic PCS, recovery may be rapid and complete.

Competing interests None.

Patient consent Obtained.

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