CASE REPORT

Atrial myxoma presenting as infective endocarditis

Gerald Paul Fitzgerald, John Joseph Coughlan, Zahir Satti, Samer Arnous

Department of Cardiology,
University Hospital Limerick,
Limerick, Ireland

Correspondence to
Dr Gerald Paul Fitzgerald,
gearoidwork@gmail.com

Accepted 20 February 2018

SUMMARY

A 23-year-old Asian student presented to our service with a 1-month history of fever, weight loss of 10 kg, night sweats, fatigue and general malaise. He was previously well with no significant medical or family history. He had a low-grade pyrexia and cardiac auscultation revealed a diastolic murmur consistent with 'tumour plop'. He had no sequelae of endocarditis. He had low-grade pyrexia of 37.7°C, and ECG showed sinus tachycardia at 130 bpm. He had raised inflammatory markers and was started on broad spectrum antibiotics. Blood cultures grew *Streptococcus viridans* twice. Transthoracic and transo-oesophageal echocardiography revealed a large mobile mass attached to the interatrial septum, suspicious for atrial myxoma, flopping into the left ventricle but not causing left ventricular outflow tract obstruction. All valves looked normal in appearance. He was treated with antibiotics for 2 weeks until inflammatory markers normalised. The patient was referred for cardiothoracic surgery where a large atrial myxoma (5 cm×3 cm) was excised just superior to the mitral valve. It had areas of necrosis and was colonised with *S. viridans*. He had an unremarkable postoperative course and made a complete recovery.

BACKGROUND

The presenting complaint of general malaise, fever, weight loss and night sweats is concerning for a number of serious pathologies including malignancy, tuberculosis and infective endocarditis. Classically, cardiac neoplasms present with one or more of the triad of constitutional, obstructive and embolic symptoms.¹ For a cardiac myxoma to present as infective endocarditis is a rare occurrence, with limited cases reported in the literature.

Initially treated empirically as endocarditis, we discovered a large atrial myxoma attached to the interatrial septum and prolapsing thorough the mitral valve. Unusually, on excision the myxoma was found to have focal areas of necrosis and superimposed infection resulting in the initial presentation mimicking infective endocarditis.

Definitive treatment with surgical resection results in favourable outcomes in the majority of cases, with success rates of over 90% and disease-free survival greater than 80%.¹

Prompt diagnosis and treatment is important as cardiac myxomas are recognised sources of emboli, and this risk persists until resection.²

CASE PRESENTATION

A 23-year-old Asian man was referred by his general practitioner to our hospital in November 2017 with

a 1-month history of anorexia, 10 kg weight loss, non-productive cough, night sweats, fatigue and general malaise.

He had no significant medical or surgical history. Dentition was excellent. Family history was unremarkable. He is non-smoker, does not use illicit drugs and drinks alcohol only occasionally. He is studying in Ireland and lives with other two classmates.

He had a low-grade fever of 37.7°C and a diastolic murmur with characteristic 'tumour plop' of atrial myxoma. ECG showed sinus tachycardia with a rate of 130 bpm. The rest of the examination was unremarkable.

Empiric broad spectrum antibiotics were initiated (amoxicillin+gentamycin), and after blood cultures were reported positive for *Streptococcus viridans* sensitive to benzylpenicillin, treatment was rationalised. The patient was treated for presumed infective endocarditis and improved clinically. His inflammatory markers also improved with serial reductions in white cell count (WCC), C reactive protein (CRP) and erythrocyte sedimentation rate (ESR). Echocardiography was arranged promptly. This showed a large atrial myxoma measuring 5 cm×3 cm attached to the interatrial septum just superior to the mitral valve. It was prolapsing into the left ventricle but there was no evidence of left ventricular outflow tract obstruction.

The patient was transferred to a cardiothoracic centre for surgical intervention. He had a successful operation and recovery. He completed a 6-week course of benzylpenicillin and inflammatory markers returned to normal.

INVESTIGATIONS

Initial laboratory investigations revealed a high WCC of $17.28 \times 10^9/L$, neutrophils $15.18 \times 10^9/L$, haemoglobin of 11.1 g/dL (normocytic and normochromic) and platelet count of $421 \times 10^9/L$. ESR was elevated at 91 mm/hour, CRP was also elevated at 113 mg/L. Albumin was low at 29 g/L. Renal and liver function tests were normal. *S. viridans* was isolated from two separate blood cultures from both aerobic and anaerobic bottles, highly sensitive to penicillin with minimum inhibitory concentration of $<0.1 \mu g/mL$.

Nasopharyngeal swab was negative for influenza A and B, parainfluenza, adenovirus, rhinovirus, coronavirus and respiratory syncytial virus. HIV 1 and 2 virus and hepatitis screens were also negative. Legionella and pneumococcal urinary antigens were negative. Mantoux test was negative. Chest radiograph was unremarkable.

To cite: Fitzgerald GP, Coughlan JJ, Satti Z, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-223656

Figure 1 Transthoracic echo, parasternal long axis view, showing a large mass almost filling the left atrium.

Transthoracic echocardiography showed a large mass in the left atrium attached to interatrial septum and prolapsing through mitral valve (figures 1 and 2), (videos 1 and 2). Urgent transoesophageal echocardiography confirmed a large left atrial myxoma. There was no evidence of mitral regurgitation. Other valves were grossly normal and left ventricular ejection fraction was $>55\%$.

Pathological analysis of the excised sample confirmed cardiac myxoma with no mitotic activity or atypia. There were focal active inflammatory and necrotic areas with gram-positive cocci cultured (figure 3)

DIFFERENTIAL DIAGNOSIS

- ▶ Benign cardiac neoplasm such as myxoma, fibroma;
- ▶ Malignant cardiac neoplasm such as sarcoma;
- ▶ Bacterial endocarditis;
- ▶ Tuberculosis;
- ▶ Malignancy.

Figure 2 Transthoracic echo, apical four chamber view, showing a large mobile mass in the left atrium prolapsing through the mitral valve.

Figure 3 Excised specimen showing a large atrial myxoma with superimposed necrosis.

TREATMENT

- ▶ Supportive measures;
- ▶ Psychological and social support;
- ▶ Intravenous benzylpenicillin;
- ▶ Surgical intervention.

OUTCOME AND FOLLOW-UP

- ▶ Surgery with good outcome;
- ▶ Postoperative care and rehabilitation, physiotherapy, dietician;
- ▶ Out patient follow-up with yearly echocardiography.

Video 1 Transthoracic echocardiogram parasternal long axis loop.

Video 2 Transthoracic echocardiograph apical four chamber loop.

DISCUSSION

Cardiac myxoma is the most common primary cardiac tumour; however, it is still a relatively rare pathology which is not very well described in the literature. Postmortem, primary cardiac tumours have a frequency of approximately 0.02%,³ and myxoma accounts for 50%–80% of primary cardiac neoplasms.^{4,5} The most common location is the left atrium, with right atrial involvement being rare.⁶ Complications can involve mechanical complications such as valvular obstruction, and embolisation has been reported in 30%–50% of patients.⁷ By far the most common embolic complication is stroke, which accounts for over 80% of embolic phenomenon.⁸

In contrast, metastatic involvement of the myocardium through haematogenous spread, direct invasion or tumour growth through the vena cava into the right atrium is relative common with an incidence of 8% in one large postmortem study of patients who died from cancer, with a particularly high frequency of myocardial involvement observed in squamous cell carcinoma of the lung (62%), urinary tract tumours (60%) and melanoma (45%).⁹

Cardiac myxoma presenting as infective endocarditis without valvular involvement is a rare phenomenon, and not a lot is known about the effect this has on the course of the illness. A case series published in 1998 examined 40 cases and suggested that embolic phenomenon as well as fever and raised inflammatory markers may be more common in infected cases.¹⁰ An updated review of 39 cases between 1998 and 2014 had similar

findings, with infected myxomas having relative more occasions of high-grade fever, multiple embolic events and the presence of refractory microorganisms.¹¹

Prognosis postexcision is excellent, with survival rates consistently quoted at greater than 90% with reoccurrence rates of approximately 5%.^{12,13} There are no definitive guidelines but yearly surveillance echocardiography is a reasonable strategy as early diagnosis and intervention reduces potential complications.

Correction notice This article has been corrected since it was published online. Videos 1 and 2 have been edited in order to remove personal information about the patient.

Acknowledgements We would like to acknowledge the patient for his kind cooperation and also our colleagues not mentioned as authors here as well as the nursing staff and the Cardiac Physiology Department for their excellent quality images. We would also like to acknowledge the surgical team of Mr J Hinchion in Cork University Hospital.

Contributors All of the authors named above have contributed to the diagnosis and management of the patient and were active in the authorship, research and proofing of the final manuscript. The case was planned with SA and he provided consultant oversight. GPF was involved in the day-to-day care and performed the TTE and TOE. ZS wrote a first draft and GPF and SA edited it. JJC got consent and explained the process to the patient as well as edited the images and did a literature review.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Learning points

- Cardiac neoplasms (of which atrial myxoma is the most common) can present with vague constitutional symptoms.
- Prompt echocardiography is required to make a definitive diagnosis and facilitate referral to cardiothoracic surgery for definitive treatment.
- Atrial myxomas can rarely become necrosed and infected and mimic infective endocarditis.

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