CASE REPORT

Broadening the scope of thoracic oncological intervention: a novel minimally invasive method for the diagnosis of primary cardiac lymphoma

Karen Sayal,¹ Tariq Ali,² Angela Tasker,² Nicholas Carroll³

¹Department of Thoracic Oncology, Papworth Hospital NHS Foundation Trust, Cambridge, UK

²Department of Radiology, Papworth Hospital NHS Foundation Trust, Cambridge, UK

³Department of Radiology, Cambridge University NHS Foundation Trust, Cambridge, UK

Correspondence to

Dr Karen Sayal,
karen.sayal@cruk.cam.ac.uk

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SUMMARY

Primary cardiac lymphoma is a rare and aggressive form of non-Hodgkin's lymphoma. It presents with non-specific symptoms which depend on the degree of cardiac infiltration. Appropriate management and early initiation of therapy depends on an understanding of the imaging characteristics and early histological diagnosis. Obtaining histology can be challenging. This is more widely performed using endomyocardial biopsy. However, this technique is associated with recognised morbidity and mortality. We present a novel diagnostic method for the investigation of myocardial and pericardial lesions. This is the first documented case of the diagnosis of primary cardiac lymphoma using endobronchial ultrasound. Endobronchial ultrasound has a well-documented and excellent safety profile.

BACKGROUND

This is the first documented case of the diagnostic utility of endoscopic ultrasound with bronchoscope-guided fine-needle aspiration (EUS-B-FNA) for myocardial and pericardial lesions.

CASE PRESENTATION

A 65-year-old Caucasian man was transferred to our hospital following an 11-month history of progressive dyspnoea secondary to biventricular failure. He had initially presented 1 year previously to his local hospital with acute onset of chest pain and dyspnoea and was referred to our care for a coronary angiogram. Angiography revealed a mild-to-moderate stenosis of his left anterior descending artery. He was discharged back to his local hospital and advised to continue with medical treatment.

The patient re-presented to his general practitioner (GP) 11 months later with persistent orthopnoea, paroxysmal nocturnal dyspnoea and newly progressive bilateral ankle oedema. On ECG, he had new T wave depression in his inferior leads.

The patient was admitted to his local hospital with clinical signs of biventricular failure. An inpatient echo revealed significant right ventricular infiltration with moderate to severe obstruction of the right ventricular outflow tract. He was transferred to our care for further diagnostic workup.

The patient's medical history is significant for rheumatoid arthritis which was well controlled on sulfasalazine, methotrexate and etanercept. He was also on warfarin for a deep vein thrombosis identified 3 months previously.

On examination, the patient's Glasgow Coma Scale score was 15/15 and he was able to mobilise independently. He was afebrile with a blood pressure of 123/54, in sinus rhythm at 88 beats per minute and with oxygen saturations of 96% on air. Heart sounds I and II were present with no additional heart sounds. The patient had fine bibasal crepitations, jugular venous pressure 3 cm above the clavicle and pitting oedema to the sacrum. There was no axillary or inguinal lymphadenopathy.

INVESTIGATIONS

A chest radiograph on admission showed mild cardiomegaly with clear lungs and a small right pleural effusion with no other significant findings.

The transthoracic echocardiogram was repeated on admission showing pericardial infiltration throughout the region of the atrioventricular groove and inferiorly to the right ventricle. There was moderate to severe obstruction of right ventricular inflow owing to compression by the pericardial infiltration.

On the basis of these findings, the patient was referred for a cardiac MRI.

DIFFERENTIAL DIAGNOSIS

Cardiac MRI showed diffuse soft tissue thickening encasing the inferior and lateral aspects of the heart with diffuse enhancement and signal characteristics similar to the heart muscle (figure 1A–E). Inferiorly, there was also a pericardial lentiform mass. Biventricular function was normal and the inferior right ventricular wall and both atria were hypokinetic. CT showed a similar appearance (figure 2). Primary cardiac lymphoma was suggested as the most likely cause of the imaging findings.

OUTCOME AND FOLLOW-UP

There was extensive discussion regarding the most appropriate route to obtain tissue for histological diagnosis.

Transjugular endomyocardial biopsy is the most commonly described method for cardiac biopsy.¹ Our case presented additional challenges due to the left-sided predominance of the infiltrative mass and the need for histological confirmation of the separate pericardial mass.

Endomyocardial biopsy is performed routinely in the UK for right-sided lesions only. In contrast, left-sided cardiac lesions require a large arterial sheath (8F) which confers a high risk of bleeding.

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Figure 1 (A) Four-chamber T2-weighted image (B) four-chamber balanced steady-state free precession (SSFP) and (C) three-chamber balanced SSFP images: encasement of the left ventricle, left atrium, septum and the atrioventricular grooves (including the right coronary and circumflex) by material which is isointense to the myocardium. (D) T1-weighted short axis precontrast demonstrates a lentiform mass with a low signal margin in keeping with calcification. (E) T1-weighted short axis postcontrast (delayed) image demonstrates enhancement of the affected myocardium and diffuse peripheral enhancement of the lentiform mass.

Crossing the aortic valve with the biptome is also accompanied by a small risk of leaflet damage and stroke (S Pettit, personal communication, October 2014).

Figure 2 Enhanced CT diffuse soft tissue encasing the inferior and lateral aspects of the heart with further peripherally calcified mass posteroinferior to the left atrium.

Endomyocardial biopsy is appropriate only for sampling the internal surface of the heart. It is not appropriate for pericardial lesions. Invasive methods such as a pericardial biopsy via thoracotomy would have exposed the patient to the morbidity and mortality associated with an invasive procedure.

Therefore, endobronchial ultrasound was selected to provide a safe, efficient and minimally invasive method to obtain tissue from the myocardial and pericardial lesions via a posteroinferior approach.

Endobronchial ultrasound (EBUS)-guided transbronchial needle aspiration integrates flexible bronchoscopy with a linear ultrasound probe to visualise and sample structures beyond the tracheobronchial mucosa. It is indicated as a minimally invasive method for obtaining a tissue diagnosis in suspected lung cancer or for mediastinal staging.² Few serious complications have been reported in reviews of efficacy and safety for diagnostic EBUS with only a single documented case of mortality.³

In this case, an EBUS bronchoscope via the oesophagus was used for tissue sampling of the myocardium and pericardium. This is the first documented case of the diagnostic utility of endoscopic ultrasound with EUS-B-FNA for myocardial and pericardial lesions.

The oesophagus was intubated with an EBUS-equipped bronchoscope. The left atrium was observed to be concentrically infiltrated by abnormal tissue replacing the normal myocardium.

Figure 3 Fine-needle biopsy of soft tissue mass encasing the heart at endoscopic ultrasound using the endobronchial ultrasound bronchoscope.

A further area of mixed echogenicity was seen inferior to the pericardium. Fine-needle aspiration samples were acquired, using 21-gauge and 22-gauge needles, from the myocardium and the abnormal pericardial tissue inferior to the heart (figure 3). There was no haemodynamic instability during the procedure.

Histological examination of the EUS-B-FNA samples revealed fragments of the myocardium, some diffusely infiltrated by small hyperchromatic cells, which on immunostaining were positive for CD45, CD20 and CD79a and negative for thyroid transcription factor-1, synaptophysin and MNF116 (figure 4A, B).

Ki67 confirmed a high-proliferation rate. Some admixed reactive CD3 positive T cells were also present. The appearances were consistent with those of a diffuse B-cell lymphoma.

Biopsy from the separate pericardial mass showed a fibrin clot with occasional fibrous tissue.

DISCUSSION

Primary cardiac lymphoma is a rare condition which primarily involves the heart and/or lymph nodes, accounting for 1.3% of primary cardiac tumours and 0.5% of extranodal lymphomas.⁴

It typically presents in elderly males and predominantly involves the right-sided cardiac chambers, in particular the right atrium.⁴ Symptoms depend on the degree of infiltration of the heart and surrounding structures. Presenting symptoms are therefore non-specific and include cardiac failure, arrhythmias, conduction disorders, superior vena cava obstruction and tamponade.⁴

The diagnostic workup usually includes a transthoracic echo, which provides a basic tool for assessment but is limited by its sensitivity and narrow acoustic window. Transoesophageal echocardiography is more sensitive in providing better visualisation of the heart with improved evaluation. CT and MRI are primarily used for tumour detection and extent, providing better contrast resolution than an electrocardiogram.

Obtaining tissue for diagnosis in such cases as above is difficult. Invasive methods such as open thoracotomy and mediastinoscopy provide a definitive diagnosis.⁵ However, they are associated with significant morbidity and mortality.

We have demonstrated a novel application of EUS-B-FNA in a previously undocumented setting. The key benefits of this increasingly available technique are multifold. It has an excellent safety profile. It is performed under sedation, permitting streamlined patient discharge several hours after uncomplicated cases. EUS-B can be performed quickly, which facilitates ease in clinical implementation.

Figure 4 (A) Fragment of the myocardium infiltrated by lymphoma H&E x200 magnification. (B) Fragment of the myocardium infiltrated by B-cell lymphoma CD20 expression.

Therefore, greater awareness is required among clinicians regarding its diagnostic potential. We postulate that this technique could be used for monitoring allograft rejection post cardiac transplant in cases where endomyocardial biopsy is not feasible.

In this report, there were no serious immediate complications. There is a theoretical risk of infection on accessing the mediastinum via the oesophagus. This could be mitigated by appropriate antibiotic cover for the procedure. However, prospective studies may be required to evaluate its safety profile in this setting.

In conclusion, primary cardiac lymphoma is difficult to diagnose. Given the rapid tumour progression, early diagnosis with

Learning points

- ▶ Primary cardiac lymphoma is a rare form of primary cardiac malignancy.
- ▶ It is associated with characteristic imaging and histological features.
- ▶ Early histological diagnosis to allow prompt initiation of treatment is essential given the rapid tumour progression.
- ▶ Current diagnostic modalities are associated with morbidity and mortality.
- ▶ We present the first documented diagnosis of primary cardiac lymphoma by bronchoscope-guided endoscopic ultrasound.

biopsy allows prompt initiation of therapy. We present the first case report using bronchoscope-guided endoscopic ultrasound in the diagnosis of myocardial and pericardial disease in a patient with lymphoma.

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