Diffuse large B cell lymphoma presenting as a cardiac mass and odynophagia

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Abstract

Cardiac involvement as an initial presentation of malignant lymphoma is a rare occurrence. We describe the case of a 77-year-old man who had initially been diagnosed with a left atrial mass on an echocardiogram, presenting with progressive dyspnea, dysphagia, odynophagia and fevers. The cardiac mass had been managed as an outpatient with full anticoagulation for the suspicion of clot. On admission, cardiac magnetic resonance imaging revealed a large mediastinal mass invading the left atrium that originated from the oesophagus. A barium oesophagram revealed an apple core lesion involving the distal third of the oesophagus. A subsequent computed tomography scan demonstrated a large mediastinal soft tissue mass and paratracheal lymphadenopathy. A flexible upper endoscopy revealed an oesophageal mass that was approximately 10 cm in length, irregular at the margins, and with a very necrotic appearance. This was biopsied, revealing findings consistent with high grade diffuse large B cell lymphoma. This case illustrates lymphoma presenting with dyspnea, odynophagia and a left atrial mass. To our knowledge, there are no reported cases of diffuse large B cell lymphoma presenting as odynophagia and a cardiac mass. (Cardiol J 2008; 15: 471-474)

Key words: atrial mass, odynophagia, echocardiogram, lymphoma

Introduction

Gross tumour formation in any of the cardiac chambers is rare, particularly at the time of presentation and diagnosis of lymphoma [1]. Symptoms are usually very subtle and non-specific, particularly in the setting of co-existing comorbidities [2]. We report an unusual case of a 77-year-old Caucasian man presenting with a gross intracardiac mass and symptoms of odynophagia, who was ultimately diagnosed with diffuse large B cell lymphoma.

Case report

A 77-year-old Caucasian man with emphysema and hypertensive kidney disease presented with

a three-week history of generalized fatigue, progressive dyspnea on exertion, decreased appetite, subjective fevers and a ten-pound weight loss. Three weeks prior to presentation, the patient had obtained a two dimensional echocardiogram for his dyspnea that revealed a large left atrial mass. Consequently, the patient was fully anticoagulated and discharged with the presumed diagnosis of a left atrial clot versus a clot overlying a mass. After discharge, the patient continued to decline, developing dysphagia to solids, odynophagia to cold liquids that led to food aversion and a hoarse voice.

On admission, he was afebrile with a blood pressure of 92/32 mm Hg and a pulse rate of 100/min, and appeared somewhat cachectic. The laboratory findings are shown in Table 1. Chest radiograph

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Peripheral blood	
White blood cell	7.3 thou/μL
Neutrophil	77.1%
Lymphocyte	7.4%
Monocyte	12.2%
Eosinophil	2.3%
Red blood cell	3.4 mil/μL
Hematocrit	28%
MCV	84 fL
Hemoglobin	9.2 g/dL
Platelet	267 thou/µL
Coagulation	
PT	22.7 s
INR	2.0
PTT	52.1 s
Blood chemistry	
Na	137 mmo/L
К	4.5 mmol/L
CI	109 mmol/L
Urea nitrogen	46 mg/dL
Creatinine	2.5 mg/dL
Total bilirubin	0.6 mg/dL
Total protein	5.5 g/dL
Albumin	3.0 g/dL
AST	6.0 U/L
ALT	16 U/L
ALK	61 U/L

Table 1. Laboratory data on admission.

revealed a moderate increase in cardiac size and mild pulmonary oedema. A 12-lead EKG showed normal sinus rhythm with occasional premature ventricular complexes. A transesophageal echocardiogram that was performed prior to admission revealed an extensive left atrial tissue density mass with some obstruction to both left and right filling. The left atrium appeared to be enlarged and was almost entirely filled with a multilayered tissue density mass. Only the left atrial area directly above the mitral valve and the left atrial appendage appeared to be free of the mass. The mass appeared to be contained within the enlarged left atrium with no evidence of invasion into the adjacent tissue (Fig. 1). Cardiac magnetic resonance demonstrated a large mediastinal mass invading the left atrium posteriorly. The mass appeared to originate from and actually encircle the oesophagus, extending back towards the aorta and the thoracic spine, measuring 9.2 cm \times 6.2 cm in its largest dimensions (Fig. 2). At this point, anticoagulation was halted.



Figure 1. Transesophageal echocardiogram revealing a mass that appears to be contained within the enlarged left atrium (LA) with no evidence of invasion into the adjacent tissue; LV — left ventricle; Ao — aorta.



Figure 2. Cardiac magnetic resonance with 4 chamber and 2 chamber views revealing a left atrial mass that appears to encircle the oesophagus, measuring 9.2 cm \times \times 6.2 cm in its largest dimensions.



Figure 3. Oesophagram revealing an oesophageal mass appearing as a distal apple core lesion.

A barium oesophagram revealed a large, near circumferential filling defect involving the distal third of the oesophagus, with greater than 75% narrowing of the oesophageal lumen. The filling defect had a typical apple core appearance that spared the posterior wall of the oesophagus. There was a mucosal irregularity involving the left lateral oesophageal wall proximal to the lesion, as well as prominent shouldering proximal and distant to the narrowed region (Fig. 3).

A flexible upper endoscopy revealed a small area of irregularity in the gastric cardia that was biopsied and sent for histological diagnosis. At the gastroesophageal junction, a large 10 cm long mass with irregular margins and a necrotic appearance was visualized and biopsied. Histopathology revealed diffuse infiltration with a large number of intermediate to large pleomorphic neoplstic lymphoid cells that were positive for CD20, PAX-5, CD45, subset BCL-6, with Ki-67 proliferation rate approximately 50-60%. These findings were consistent with a high-grade diffuse large B-cell lymphoma (Fig. 4). Computed tomography revealed a large midline soft tissue mass in the inferior portion of the posterior mediastinum, as well as paratracheal lymphadenopathy (Fig. 5).

A bone marrow biopsy revealed normocellular bone marrow without lymphoma infiltrate. Immediately after the diagnosis, the patient underwent a cycle of R-CHOP chemotherapy — Rituximab/ /Cyclophosphamide/Adriamycin/Vincristine/Prednisolone. Unfortunately, on day fifteen of chemo-



Figure 4. Histological exam of the biopsied specimen showing diffuse infiltration with a large number of intermediate to large pleomorphic neoplastic lymphoid cells, consistent with a high grade lymphoma (Giemsa stain \times 60, Ki-67 stain \times 60).



Figure 5. Chest computed tomography revealing a mediastinal mass with paratracheal lymphadenopathy.

therapy, the patient developed a seizure that progressed into status epilepticus and hypoxic respiratory failure. Given his poor overall prognosis, his goals of care were switched to comfort only. The patient passed away 12 hours later.

Discussion

Cardiac involvement as an initial presentation of malignant lymphoma is a rare occurrence [1]. At autopsy, cardiac involvement is evident in about 1% of patients with advanced cancer, and the primary tumour encountered is most frequently bronchogenic or oesophageal carcinoma [3]. Secondary involvement of the heart is seen in 8.7–27.2% of documented clinical cases of lymphoma [1, 4, 5]. Despite its life-threatening nature [6], the cardiac manifestations of lymphomatous involvement of the heart are often subclinical [7–9]. Signs and symptoms of cardiac dysfunction such as chest pain, dyspnea, and arrhythmia may be clinically undetectable, and cardiac involvement is often undetermined prior to death. The initial presenting complaint in this patient was progressive dyspnea, which ultimately led to the detection of the intracardiac mass by echocardiogram.

In many cases, an intracardiac mass is difficult to distinguish between a clot and a tumour. Left atrial thrombi is most often associated with atrial fibrillation and/or rheumatic mitral stenosis, accounting for over 45% of cardiogenic thromboemboli. One study of 2,894 consecutive patients who underwent transesophageal echocardiogram for various indications, found a left atrial thrombus in 94 (3%); 83 of these patients were in atrial fibrillation [10]. Gross tumour formation in any of the cardiac chambers is rare, particularly at the time of presentation and diagnosis [1]. This study and others also note that the majority of intracavital tumours occur on the right side of the heart, the reason for which is yet to be found [1, 11].

The case presented here illustrates a rare presentation and complication of diffuse large B cell lymphoma, and is unique in several respects. The patient presented initially with dyspnea then odynophagia along with a recent finding of an intracardiac mass. The case revealed a rare cause of an intracardiac mass that was initially presumed to be a clot. The mass occurred in the left atrium, although the majority of intracavital tumours occur on the right side of the heart [1, 11]. Finally, the use of cardiac magnetic resonance imaging was instrumental in guiding the ultimate diagnosis of lymphoma.

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