

A Rare Case of Relapsed Refractory T-Cell Lymphoma with Cardiac Metastases

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Introduction

Peripheral T-cell lymphoma is an aggressive and rare form of Non-Hodgkin's lymphoma (NHL), carrying a poor prognosis with a tendency to relapse. Most patients present with painless lymphadenopathy and constitutional symptoms including night sweats, weight loss and pyrexia. A small proportion develop 'extranodal' symptoms which may affect the skin, bones, liver and gastrointestinal tract. However, cardiac manifestations of T-cell Lymphoma are extremely rare and sparsely reported in literature. Here we present a case of relapsed refractory peripheral T-cell lymphoma in a 66-year old male with cardiac metastases.

Case Presentation

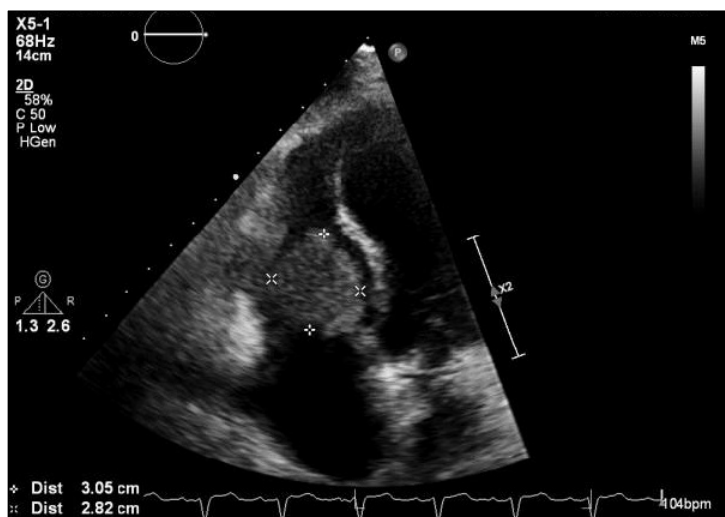
The patient was initially diagnosed with T-cell NHL in 2002 and was in remission following CHOP chemotherapy. He re-presented in 2016, and was found to have relapsed with large volume lung lesions and extensive lymphadenopathy. He was treated with 2 cycles of ESHAP chemotherapy. Following a 4 further cycles of chemotherapy, the patient opted to stop treatment due to frequent hospital admissions for neutropenic sepsis. His clinical course was further complicated by pulmonary emboli, treated with Rivaroxaban, and hypercalcaemia.

Prior to starting palliative Alemtuzumab, the patient presented in late 2018 with 2 weeks of exertional dyspnoea and orthopnoea. He did not report any chest pain or palpitations. Examination revealed normal first and second heart sounds but a new diastolic murmur heard loudest at the left sternal edge. He also had poor air entry at his lung bases, an elevated visualised jugular venous pressure (JVP) and bilateral pitting ankle oedema.

A trans-thoracic echocardiogram (Figure 1) revealed a new large infiltrating homogenous mass on his tricuspid valve (TV) causing inflow obstruction (pseudo-tricuspid stenosis). Potential differential diagnoses included cardiac thrombus or tumour. Subsequent Cardiac Magnetic Resonance Images (Figures 2 and 3) confirmed these lesions were consistent with lymphomatous infiltration.

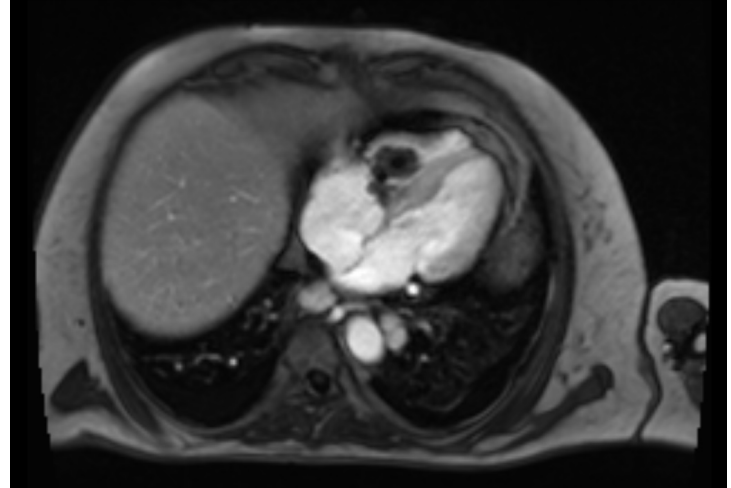
Following discussion with the patient, his family and the multidisciplinary team, the patient opted to cease all active treatment and to focus on symptom control. He died a few weeks later.

Figure 1



This is an apical 4 chamber view of this patient's echocardiogram. A large mass is seen in the right ventricle attached to tricuspid valve leaflets. There was no prolapse of the mass, but it appeared to cause significant obstruction to TV inflow. There was no significant tricuspid regurgitation. The right ventricle was mildly dilated at the base with good systolic function. The left ventricle was non-dilated with no hypertrophy and good systolic function (ejection fraction 55%).

Figure 2



This axial image taken from this patient's Cardiac Magnetic Resonance scan demonstrates a small pericardial effusion and bilateral pleural effusions. The mass in the right ventricle is seen attached to the tricuspid valve measuring 31 x 35mm. Another smaller mass (10 x 18mm) is also attached to the tricuspid valve. Both masses have some enhancement on early gadolinium imaging, which demonstrates perfusion. This is in keeping with lymphomatous infiltration of the tricuspid valve, and excludes the other differential diagnosis of a cardiac thrombus.

Discussion

In theory the heart can be metastasised by any malignant neoplasm, with a reported incidence of 2.3% to 18.3%. The pericardium is the most commonly affected site, followed by the epicardium, myocardium and endocardium.

The potential for cardiac metastasis is dependent on the characteristics of the primary tumour but also on the functional and histological features of the heart. Tumours can spread to the heart via direct extension, haematogenous spread, lymphatic spread or via intracavitary diffusion. The right side of the heart, as seen in this patient, is more frequently involved.

The majority of patients do not develop symptoms from their cardiac disease, and so diagnosis during life is rare. Potential clinical manifestations can be due to valvular incompetence, cardiac failure, rhythm disturbance, syncope, embolism or pericardial effusion.

Many imaging techniques can be used to diagnose cardiac metastases, with echocardiography being a good screening tool although lacking sensitivity. Computed tomography can determine the morphology, extent and location of a lesion, but magnetic resonance images with contrast will identify anatomy, blood flow, cardiac function, and importantly characterise a mass. Gadolinium enhanced MRI confirmed the diagnosis in this case.

Treatment options are limited, and are restricted to aggressive chemotherapy and radiotherapy. This is unsurprising given identification of cardiac metastases is often delayed until the point at which cardiac symptoms develop. Without treatment, median survival following diagnosis of cardiac metastases is less than 6 months.

This case illustrates a cardiac manifestation of peripheral T-cell lymphoma which has infiltrated tricuspid valve leaflets. Identification of cardiac involvement in lymphoma can help to guide prognosis.

References

- Bussani R, De-Giorgio F, Abbate A, Silvestri F. Cardiac Metastases. *J Clin Pathol.* Jan 2007; 60(1): 27-34
- Abdullah H, Nowalid W. Infiltrative cardiac lymphoma with tricuspid valve involvement in a young man. *World J Cardiol.* Feb 2014; 6(2): 77-80
- Agha A et al. B-cell lymphoma in the tricuspid valve. *Cureus.* Dec 2016; 8(12): e930