Cardiac involvement by lymphoma: diagnostic difficulties

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A patient presenting with recurrent tamponade was subsequently shown to have non-Hodgkin's lymphoma. Catheterization demonstrated obstruction of the tricuspid valve by tumour and shunting through a patent foramen ovale.

The difficulties in making an ante-mortem diagnosis of cardiac lymphoma are discussed.

Introduction

Cardiac presentation of lymphoma is rare, although post mortem studies have shown involvement in up to 25% of cases[1]. Cardiac manifestations include congestive failure, tamponade, conduction disturbance and arrhythmias. Ante-mortem identification of cardiac lymphoma and assessment of progression by echocardiography has been advocated previously[2]. Cytological examination of pericardial effusions in diagnosing lymphoma is recommended although its sensitivity is not established[3].

We report a case of lymphoma presenting with recurrent tamponade which was complicated by the development of right ventricular inflow tract obstruction and intra-atrial shunting confirmed at cardiac catheterization. Cytology of the effusion was repeatedly negative and echocardiography failed to demonstrate extensive intra-cardiac invasion.

Case report

A 67-year-old man was referred for investigation of recurrent tamponade. Six months previously a permanent pacemaker had been inserted for intermittent complete heart block following an inferolateral infarction. Angiography performed 4 months later, for exertional dyspnoea, showed proximal occlusion of the right coronary artery, insignificant disease in the left coronary system and normal left ventricular function. An area of tumour circulation was demonstrated in the right atrium (Fig. 1). Subsequent echocardiography revealed a moderate pericardial effusion, but no intracavity tumour.

During outpatient follow-up he deteriorated and was admitted to a local hospital with hepatomegaly, jaundice and cardiac tamponade. Echocardiography confirmed the diagnosis and 500 ml of serous fluid was aspirated, giving relief of symptoms. Cytological and microbiological examination of the pericardial fluid was negative. Haematological, biochemical and immunological screens were normal. Abdominal ultrasound and liver biopsy showed congestion only. Three weeks post discharge he re-presented with cyanosis and tamponade; at this stage he was transferred to our care.

On admission he was deeply cyanosed (pO₂ 3-6 kPa, increasing to 5-6 kPa in 100% O₂), hypotensive 80/30 mmHg with 20 mmHg of paradox and his apex was palpable. Pericardiocentesis of 350 ml serous fluid failed to improve his haemodynamic condition and inotropes were commenced. Cytology was again negative and post-aspiration echocardiography did not demonstrate any intra-cardiac tumour (Fig. 2) or shunting. At right heart catheterization the right ventricular and pulmonary systolic pressures were low. Simultaneous right atrial and ventricular pressure recordings showed a consistent 8 mmHg end-diastolic gradient (Fig. 3). Saturations demonstrated right to left shunting at atrial level and withdrawal from left to right atrium showed a step up in pressure (Fig. 4). Contrast injection into the right atrium showed a large mobile filling defect within the right atrial and ventricular cavities (Fig. 5).

As the tumour aetiology was unknown the patient was referred for urgent surgical relief of the
obstruction. An explorative median sternotomy revealed a superior mediastinal tumour invading and replacing the right atrium and ventricle, infiltrating the floor of the left ventricle and surrounding both the superior and inferior venae cavae. The situation was deemed inoperable and the chest was closed after taking biopsies. The patient died 1 h later.

Histological findings were of a non-Hodgkin's lymphoma.

Discussion

Post-mortem studies report an incidence of cardiac involvement in lymphomas of between 9 and 25%[1,4] and suggest that the severity of the involvement is unrelated to the clinical picture[4]. There has been no prospective study to evaluate the frequency and severity of cardiac involvement by non-invasive methods or to correlate these with the subsequent clinical course. Early involvement appears to be either missed clinically or not reported. Presentation as cardiac involvement is rare, with less than 40 cases in the literature. Almost invariably a dramatic presentation, such as tamponade or rupture[9], is described when the lymphoma is well-advanced and the prognosis poor. In recent cases tamponade was the commonest presenting feature; other forms of presentation included right heart failure[6], complete heart block[7], arrhythmias[8] and non-specific ECG changes.

The paucity of information regarding the true incidence of cardiac lymphoma probably reflects the difficulty in achieving an ante-mortem diagnosis. The ECG changes that may occur lack specificity[9] and although echocardiography has been advocated by some[10] its sensitivity has not been evaluated. Others like ourselves have reported significant cardiac invasion without echocardiographic demonstration of myocardial involvement[7]. The examination of serous fluids in general is positive in 88% of cases[11], but no study has specifically studied pericardial effusions. More recently the use of isotopes, to demonstrate either selective uptake by lymphoma[9] or the presence of a space-occupying lesion[12] has been advocated, but these have still to be evaluated.

This case is interesting as it demonstrates some of the rare features of cardiac lymphoma, especially the presence of shunting through a patent foramen ovale which although once described retrospectively[7] has never been demonstrated ante-mortem.
Diagnosis of cardiac lymphoma

by cardiac catheterization. The failure of echo (in three different establishments) and cytology on two occasions to provide the diagnosis underlines the uncertainty in establishing clinically the presence of cardiac lymphoma. How earlier diagnosis of cardiac involvement might affect the patients prog-

nosis or optimal treatment is not known, but if this is to be assessed then the sensitivity of screening methods must be established.

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References


