CASE REPORT

Resolution of an intracardiac mass with chemotherapy

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SUMMARY

Right atrial intracardiac tumours are uncommonly seen during echocardiography. Our patient presented with primary mediastinal large B-cell lymphoma with intracardiac involvement. The tumour was seen by echocardiography and the extent of the tumour was defined by CT scan of the chest. Following chemotherapy directed to her specific tumour cell type, there was complete resolution of the intracardiac mass.

BACKGROUND

First recognised in the 1980s primary mediastinal large B-cell lymphoma (PMBL) normally presents with a bulky tumour in the anterior mediastinum that is rapidly progressive and may cause local compression of the mediastinal structures giving rise to local compressive effects including dyspnoea, cough, dysphagia and superior vena cava (SVC) obstruction.¹ By the time of diagnosis, many patients with PMBL show invasion of the lungs, pleura or pericardium, while distant spread to the bone marrow or other extra thoracic sites is uncommon.²

A case of PMBL compressing the SVC and extending through the wall of the right atrium into the right atrium on echocardiogram is presented in this case report. This case presents shows a rare cardiac involvement by PMBL and is one of few reported cases where an intracardiac mass was treated successfully with chemotherapy alone.

CASE PRESENTATION

A young Caucasian woman presented to the urgent care clinic with the chief complaint of decreasing energy level, palpitations, dyspnoea, and swelling of the face and neck. Facial swelling was initially thought to be owing to angio-oedema. Pertinent findings on physical examination revealed a markedly elevated jugular venous pressure that did not vary with posture and a musical grade 3/6 systolic murmur heard best in the pulmonary area suggestive of compression of the right ventricular outflow region. In addition, there was a continuous murmur (grade 2/6) heard in the right subclavicular area near the sternum. In the clinic (ECG) showed sinus tachycardia and the chest x-ray showed mediastinal widening (figure 1). Owing to these x-ray findings, the patient was scheduled for a chest CT and an echocardiogram. On echocardiography, there was a large (37×30 mm) irregular solid and fixed mass in the right atrium (figure 2). Other images showed the mass surrounding the aorta in the ascending and arch portions. Doppler evaluation of the pulmonary artery showed a high-velocity jet in the right pulmonary artery (approximately 3 m/s). No mass in the inferior vena cava was noted. The CT scan of the chest revealed an ill-defined mass within the mediastinum obscuring the SVC and invading the right atrium (figure 3A,B). The CT also showed the presence of confluent bilateral hilar lymphadenopathy. Owing to

Figure 1 Chest x-ray showing mediastinal widening (arrows).

Figure 2 Two-dimensional echocardiogram in the apical four chamber view showing the right atrial mass measuring 37×30 mm (arrow). LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

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Figure 3 (A) Coronal section from the CT scan of the chest showing a large anterior mediastinal mass completely obscuring the superior vena cava and extending into the right atrium (broad arrow). The mediastinal mass is outlined by white arrows. Ao, aorta; LV, left ventricle; RV, right ventricle. (B): Horizontal section from the chest CT showing infiltration of the right atrium by the lymphoma. (C): Coronal section more anteriorly than (A) showing large contrast filled collaterals arising from the right subclavian vein toward the lower veins in the chest.

complete occlusion of the SVC, collaterals were seen extending from the right subclavian vein to the azygous and the hemiazygous to the inferior vena cava (figure 3C).

INVESTIGATIONS

The patient was admitted to the haematology/oncology service for SVC syndrome and a CT-guided biopsy of the mediastinal mass performed. Histological examination of the mass showed a diffuse growth pattern, comprising large cells with polymorphic nuclei and an abundant rim of clear cytoplasm. Fibrosis was noted and compartmentalised the neoplastic cells into small packets (figure 4). Immunophenotyping demonstrated the presence of B-cell antigens (CD19, CD20, CD22 and CD79a). These findings were consistent with the diagnosis of primary large B-cell lymphoma. A bone marrow biopsy was without evidence of lymphoma.

Figure 4 Micrograph showing a diffuse growth pattern, comprising large cells with polymorphic nuclei that have an abundant rim of clear cytoplasm and fibrosis (arrow) compartmentalising the cells which is characteristic feature of primary mediastinal large B-cell lymphoma.

TREATMENT

The patient was started on chemotherapy (Rituximab – Etoposide, Prednisone, Vincristine, Cyclophosphamide, Doxorubicin).

OUTCOME AND FOLLOW-UP

Serial echocardiograms performed following chemotherapy showed that the right atrial mass progressively decreased in size and was completely absent a month after the first cycle of chemotherapy (figure 5). The patient was discharged from the hospital after the first cycle and had noticed significant improvement in her symptomatology and suffered no more episodes of palpitations.

DISCUSSION

Most intracardiac masses are either primary cardiac tumours such as myxoma, papillary fibroelastoma, lipoma, fibroma, or tumours spreading through the inferior vena cava such as renal cell carcinoma, carcinoid tumour or thrombi.³ At the same time, lymphomatous involvement of the heart, which is seen in 9–27% of patients with advanced lymphoma,⁴ may invade the pericardium and present as pericardial effusions but rarely extend into the myocardium of the heart.⁵

Figure 5 (A) Two-dimensional echocardiogram in the apical four-chamber view showing complete resolution of the mass 1 month following chemotherapy.

Also despite its life-threatening nature,⁴ the cardiac manifestations of lymphomatous involvement of the heart are often sub-clinical.^{6–8} Signs and symptoms of cardiac dysfunction such as chest pain, dyspnoea and arrhythmia may be clinically undetectable, and cardiac involvement is often undetermined prior to death. Tachycardia, dyspnoea along with symptoms of SVC obstruction were the initial presenting complaints of our patient which eventually led to detection of intracardiac mass by echocardiogram.

This case presented illustrates a very rare type of cardiac involvement from PMBL. PMBL is known to locally invade the pericardium and present as a pericardial effusion.² The recognition of this unusual presentation is critically important since these masses may be misdiagnosed as a right atrial thrombus. In these cases, patients may have anticoagulation initiated which could lead to significant bleeding events. In our case, combined with a strong suspicion on echocardiography and confirmed by other imaging modalities that the mass was continuous with a mediastinal mass leading us to recognise this was likely an intracardiac lymphoma. This realisation was the impetus to treat with chemotherapy rather than open resection of the tumour.

In conclusion, through recognition that the cardiac mass was contiguous with the mediastinal mass, which was later determined to be PMBL, helped guide appropriate management of our patient to a regimen that used chemotherapy alone resulting in complete resolution of the intracardiac and extracardiac masses.

Learning points

- An unusual case of primary mediastinal large B-cell lymphoma (PMBL) invading the cardiac chambers (right atrium) and presenting as an intracardiac and extracardiac masses on echocardiography.
- PMBL should be in a differential diagnosis in patients presenting with superior vena cava syndrome.
- Chemotherapy alone can treat both the extracardiac and intracardiac involvement of PMBL.

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