CT and MR Findings of Primary Cardiac Lymphoma: Report upon 2 Cases and Review

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Although primary cardiac lymphoma was a near fatal disease in the past, recent advances in imaging diagnosis and chemotherapy have dramatically improved survival. We describe the CT and MR findings of primary cardiac lymphoma in two immunocompetent subjects who recovered after chemotherapy. The primary cardiac lymphomas manifested as a poorly enhancing mass on CT, relatively hyperintense on a T2-weighted image and a heterogeneously enhanced mass on contrast-enhanced MR. The tumor also showed a predilection for right heart chambers and contiguous infiltration along adjacent structures.

Key Words: Heart, neoplasms-lymphoma, Heart, computed tomography, Heart, magnetic resonance imaging

INTRODUCTION

In patients with malignant lymphoma, metastatic deposits are frequently found at postmortem; however, lymphoma can also have a primary effect on the heart. Primary cardiac lymphoma (PCL) is classically defined as an extranodal non-Hodgkin's lymphoma, which is exclusively located in the heart and/or pericardium. Recently, this definition has been extended to include other localizations less important than cardiac sites, but heart remains the most important during the illness course. PCL is extremely rare in immunocompetent patients, accounting for 1.3% of all cardiac tumors and 0.5% of all extranodal lymphomas, but it is being encountered with increasing frequency in patients with AIDS or other immunocompromised conditions.

The rarity and nonspecific clinical presentation of PCL has made antemortem diagnoses difficult in the past. The prognosis of PCL used to be grave due to delayed diagnosis and the difficulty in locating the disease. However, recent advances in modern imaging techniques and management have dramatically improved the survival rate. To our knowledge, there have been few reports about the CT and MR features of PCL. We report upon two recently experienced cases of pathologically proven primary cardiac lymphoma and review their CT and MR appearances.

CASE REPORT

CASE 1

A 64-year old man complained of dyspnea of 1 month's duration and had experienced a short syncopal attack some three days previously. He had smoked 40 pack-years of cigarettes and was a social drinker. Coarse breath sounds were auscultated over the lung bases. Lymph nodes were not palpable anywhere. The EKG was normal except for sinus bradycardia (55 beats/min). There was no abnormality on laboratory examination. A chest radiograph showed a bulging contour at the right cardiac border and a 1.5 cm-sized, ill-defined pulmonary nodule, in the right upper lobe. Both transthoracic and transesophageal echocardiography revealed an isoechoic mass at the right atrial wall. The mass was...
contiguously infiltrating the inferior vena cava. Minimal pericardial effusion was also in evidence.

A contrast-enhanced CT scan (Fig. 1) showed a focal bulging mass at the postero-inferior wall of the right atrium and diffuse infiltration of almost the entire right atrial free wall and the interatrial septum. The mass contiguously infiltrated the wall of the superior and inferior vena cavi, and was more homogeneous and more poorly enhanced than the normal myocardium. A 1.5 cm-sized, ill-defined nodule was seen at the right upper lobe. No hilar or mediastinal lymphadenopathy or pericardial effusion was evident. On conventional spin-echo MRI under ECG gating (Fig. 2A-C), the mass was isointense on the T1-weighted image and mildly hyperintense on the T2-weighted image, compared to the signal intensity of the normal myocardium. On dynamic study after Gadolinium-DTPA administration, the mass became progressively enhanced with a non-enhancing central portion. Right pleural effusion and stenosis of the right inferior pulmonary vein due to mass invasion, and stagnation of the contrast material in the right lower lobe were demonstrated. No evidence of abdominopelvic involvement was apparent in ultrasonographic or CT scans.

Biopsy was performed during cardiac catheterization, using a 7.5 Fr biopsy forcep, guided fluoroscopy and transesophageal echocardiography. The biopsy material was obtained from the interatrial septum of the right atrium. Immunohistochemistry revealed positive results for L26 and

![Fig. 1. Case 1. 64-year-old man. Contrast-enhanced CT scan showing diffuse wall thickening of the entire free wall of the right atrium (arrows) and interatrial septum (arrowhead). CT attenuation of the mass is homogeneous and less enhanced compared to the enhanced normal myocardium.](image)

![Fig. 2. Case 1. A, B; ECG-gated, spin-echo MRI. C; ECG-gated, gradient echo MRI. A: On the axial T1-weighted image (TR/TE/NEX, 1276/10/2), the right atrial mass is homogenous and isointense compared to the normal myocardium. B: An axial T2-weighted image (TR/TE/NEX, 3829/80/2) reveals a hyperintense right atrial mass with several foci of lower signal intensity (arrow). Right pleural effusion is associated. C: Dynamic study after administration of Gd-DTPA. Late phase axial T1-weighted image demonstrates a progressive enhancing pattern at the peripheral area of the right atrial mass, leaving the non-enhancing region (arrow).](image)
CD79α, which are indicative of B-cell origin. The final diagnosis was diffuse large B-cell type malignant lymphoma. The histology of the nodule on the right upper lobe was not proven. The patient was treated with a CHOP regimen (cytoxan, doxorubicin, vincristine, and prednisone). On follow-up chest CT scan after three months (Fig. 3), showed that the cardiac mass had almost completely disappeared, and showed minimal nodular wall thickening of the right atrial wall. The ill-defined nodule in the right upper lobe also markedly decreased in size.

CASE 2

A 31-year-old woman had suffered from chest discomfort for three months, and had developed dyspnea one week prior to presentation. Physical examination demonstrated borderline hypertension (130/95 mmHg). Her breath sound was clear and her lymph nodes were not palpable anywhere. Elevation of the liver enzymes (AST=511U, ALT=1451U) and a weak positive troponin-T (0.452 U) were found during laboratory examination. Sinus tachycardia (90 beats/min) and a complete right bundle branch block were observed on the ECG. The chest radiograph revealed nonspecific cardiomegaly and interstitial pulmonary edema. On transthoracic echocardiography, a large infiltrating intracardiac mass was found mainly at the interatrial and the interventricular septa, and the aortic root and the tricuspid valve. The echotexture of the mass was relatively homogeneous, and a moderate amount of pericardial effusion was present.

Conventional, ECG-gated, spin-echo MRI (Fig. 4A-D) depicted a slightly lobulated main mass at the crux cordis. Infiltrative lesions were observed at the interatrial septum, the interventricular septum, the tricuspid valve ring, the aortic root and the inferior free wall of the left ventricle. The mass was homogeneous and isointense on the T1-weighted image and slightly hyperintense on the T2-weighted image compared to the normal myocardium. The periphery of the mass showed slight enhancement, which was similar to that normally associated with enhanced myocardium after Gadolinium-DTPA administration, though the central portion was unenhanced. A small amount of pericardial effusion was detected, but no definite pericardial thickening was observed. The contrast-enhanced CT scan (Fig. 5) showed that the mass was less enhanced than the normal myocardium. Bilateral pleural effusion and interstitial pulmonary edema were combined secondary to progressive congestive heart failure. Hilar or mediastinal lymphadenopathy was not seen, and no evidence of abdominopelvic involvement was detected by ultrasonography, CT or MR, but dilatation of the inferior vena cava and mild hepaticomegaly caused by congestive heart failure were in evidence.

Open thoracotomy and biopsy through the right atrial wall were performed. A 4 × 5cm slightly lobulated and myxoid mass covering the septal and posterior leaflets of the tricuspid valve was observed. Immunohistochemical study proved positive for L-26 and CD79α, the B-cell marker. The final diagnosis was a large B-cell primary cardiac lymphoma. Chemotherapy with CHOP regimen was administered. Follow-up chest CT scan (Fig. 6), taken seven months later, revealed no evidence of any residual tumor mass.

DISCUSSION

PCL is mainly a B-cell lymphoma of the diffuse large cell type in around 80% of reviewed cases.\textsuperscript{4,7} PCL is difficult to diagnose clinically, because of its non-specific clinical manifestations. Frequent
Fig. 4. Case 2. 31-year-old man. A, B, C and D; Spin-echo MRI under ECG-gating. A and B. T1-weighted axial images (TR/TE/NEX, 1325/12/2). A slightly lobulated, relatively isointense mass was found located mainly at the crux cordis, and extended to the tricuspid valve (arrow in A), interventricular septum (arrowhead in A) and periaortic root (arrow in B). C. T2-weighted axial image (TR/TE/NEX, 3756/80/2). A moderate amount of pericardial effusion can be seen (arrow). D. Gd-DTPA-enhanced, fat-suppressed T1-weighted axial image. The mass showed a peripherally enhancing feature, which was similar to that of normally enhanced myocardium, leaving the nonenhancing central portion (arrow).

Fig. 5. Case 2. Contrast-enhanced CT scan 17 days after the MRI. The main bulk of the mass was found at the crux cordis, and showed homogeneously nonenhanced characteristics compared to the normally enhanced myocardium. Extension to the interventricular septum (arrowhead) and the attachment site of tricuspid valve (arrow) are also shown. Bilateral pleural effusion and consolidation at the left lower lobe were newly developed.

Fig. 6. Case 2. Follow-up contrast-enhanced CT scan, 7 months later chemotherapy revealed no evidence of residual tumor mass.
clinical signs are unresponsive right heart failure, arrhythmia, precordial pain and cardiac tamponade,\(^5\) which in some instances may need urgent treatment. PCL diagnosed antemortem was extremely rare in the past due to the limited use of non-invasive diagnostic techniques and the difficulties of applying invasive biopsy methods. However, the advent of cross-sectional imaging has allowed early and accurate detection of primary cardiac malignancies. Transthoracic echocardiography is the most useful initial screening method for suspected cardiac masses, but it has limitations due to its narrow acoustic window. Thus, the tumor lesion in pulmonary vessels, the superior vena cava and the upper part of the right atrium may not be depicted.\(^8\) Transesophageal echocardiography can overcome such limitations and routinely offer excellent images of the cardiac base.\(^9\)

CT and MR are advantageous for the simultaneous visualization of the great vessels, heart, pericardium, mediastinum, and the lung parenchyma. Both modalities have better contrast resolution than echocardiography and can give a specific diagnosis for a number of tumors depending on their CT number and/or signal intensity (SI) characteristics, these include, cardiac lipoma, osteochondrosarcoma and pericardial cysts.\(^10\)\(^,\)\(^11\)

The reports of the specific characteristics of CT radiologic imaging for PCL are rare. Reports by Ceresoli et al.\(^7\) and Dorsay et al.\(^12\) show that PCL presented as an evenly and poorly enhanced mass on the CT scan, as was found in our cases. Lymphomas involving the central nervous system reveal various CT patterns according to their histologic characteristics, but solid portions are relatively well enhanced. The exact reason why PCL has poor enhancement, however, is not clear.\(^13\) PCL is known to have a predilection for sites as do other types of cardiac tumors, and most frequently arises from the right cardiac chambers, with similar prevalence in the right ventricle and right atrium.\(^4\)\(^7\)\(^9\) In contrast, unclassified sarcoma, leiomyosarcoma and fibrosarcoma are more commonly located in the left atrium. Rhabdomyosarcoma is the most common cardiac tumor in infants and children and tends to occur multicentrically.\(^11\)\(^,\)\(^14\)\(^,\)\(^17\) It also demonstrates infiltrative feature and may involve more than one cardiac chamber.\(^18\) The two cases reported here also revealed this tendency, as the tumors were mainly located in the right atrium and infiltrated the adjacent chambers and great vessels. Previous reports also suggested that PCL rarely involve the valves compared to other sarcomas,\(^19\) though some involved the tricuspid valve leafets as in our Case 2. PCL has been reported to involve more than one cardiac chamber in 75% of cases.\(^14\)

We believe these CT features, i.e., the intracardiac location in right cardiac chambers, a tendency to extend to and infiltrate adjacent chambers and structures and poor enhancement after contrast administration, are characteristic of PCL. Furthermore, CT is superior to echocardiography and MR in determining the extent of tumor in the lung parenchyma, particularly in case of lymphoma.

Of various advantages of MR, its multiplanar capability is capable of precisely defining the extent of tumor involvement in the heart, and in this respect it has a capability comparable to trans-esophageal echocardiography. Transthoracic echocardiography and CT could not definitively detect tumor extension along the superior vena cava and the right inferior pulmonary vein in the case 1, while MR imaging did. The most distinct advantage of MR imaging as compared to the other modalities is its ability to characterize tissue or tumor with or without use of contrast materials, because of its inherent superb contrast resolution. Different primary cardiac malignancies may have different imaging features. In general, lymphoma is known to be less likely than other sarcomas to have necrosis or hemorrhage, therefore, the signal intensity (SI) of lymphoma is relatively homogenous.\(^11\) Reports on the characteristics of SI in PCL are relatively scarce, but iso- or hypo-intensity on T1-weighted image, and hyp, iso- or mild hyper-intensity on T2-weighted image compared with myocardial SI, and homogenous or heterogenous enhancement after Gadolinium-DTPA administration were observed.\(^12\)\(^,\)\(^14\)\(^,\)\(^19\) In our cases, the mass had homogenous isointensity to myocardium on the T1-weighted image, and higher SI than myocardium on the T2-weighted image with a peripheral enhancement pattern. A high SI on the T2-weighted image or peripheral enhancement were related with histologic evidence of necrosis of the tumor in primary lym-
phomas of the central nervous system,21 but the radiologic-histologic correlation has not been reported in PCL. Heterogeneity of SI on the T2-weighted image and after contrast enhancement were also reported to be related with a malignancy grade of lymphoma.22 PCL, unlike other primary cardiac malignancies, responds to chemotherapy and early suspicion by imaging diagnosis is helpful in terms of avoiding open thoracotomy and biopsy of the lesion under an imaging guide.5,7,23 Early institution of chemotherapy may be effective,4,6,7 as it was in our cases and MR imaging proved useful in assessing the response.

In conclusion, CT and MR imaging workup allow PCL to be suspected at an early stage. In patients with PCL, CT and MR imaging may demonstrate a cardiac mass with a predilection for right heart chambers and contiguous infiltration along adjacent structures. The PCL is also seen as a poorly enhanced mass on CT, has relatively high SI on T2-weighted images and heterogeneous enhancement on postcontrast MR imaging. CT seems to be a better diagnostic modality for staging the tumor in the lung parenchyma, while MR imaging is excellent for the precise assessment of intracardiac extent and the characterization of tumors.

REFERENCES