Primary cardiac lymphoma (PCL) is an extremely rare entity that accounts for only 0.5% of extranodal lymphomas and 1.0% of cardiac tumors.[1] Because the tumor is clinically aggressive and fatal, it is of great significant to diagnose this disease and treat it promptly. Nevertheless, The diagnosis of PCL is very difficult due to its nonspecific clinical manifestations.[1] Herein, we report the imaging findings in two cases of PCL.

A 73-year-old man presented to our hospital on November 25, 2014, with breathlessness and chest distress for 2 months. The initial laboratory results were as follows: prothrombin time, 23.4 s; C-reactive protein (CRP), 6.83 mg/L; albumin, 30.9 g/L; and lactate dehydrogenase (LDH), 399 U/L. Electrocardiogram (ECG) demonstrated third-degree atrioventricular block [Figure 1a] and a chest X-ray showed mild cardiomegaly [cardiothoracic ratio = 57%, Figure 1b]. A bedside echocardiography revealed a big irregular hypoechoic mass in the right atrium. Cardiac magnetic resonance (CMR) images showed a large soft tissue mass within the right atrium measuring about 8.3 cm × 5.2 cm. The mass was homogeneously isointense. Postcontrast T1-weighted imaging revealed moderate enhancement. The right coronary artery traversing through the mass could be observed [Figure 1c and 1d]. Then, a cardiotomy was carried out with cardiopulmonary bypass. A large, smooth, moderate hardness, and lobulated mass was identified at the right atrium impinging the atrioventricular junction. The mass had a pedicle which was attached to the coronary sinus. The final pathological diagnosis was a diffuse large B-cell lymphoma (BCL). The patient subsequently received treatment with CHOP (cyclophosphamide, clarithromycin lactobionate, vindesine, and dexamethasone) therapy. After a course of treatment, the patient showed refractory pleural effusion, in which prolymphocyte was found. Therefore, repeated thoracic drainage was conducted to relieve the oppression symptoms. The patient has been followed up for 6 months and then lost to follow-up.

Another patient, a 27-year-old woman, who had a history of recurrent palpitation and chest distress for 6 months before hospitalization, was admitted on October 19, 2014. The patient had no complaints of fever above 38°C or loss of more than 10% of body weight over a period of 6 months or less. Abnormalities in the laboratory test results included LDH, 273.8 U/L; CRP, 7.35 mg/L; CA72-4, 12.61 U/ml; and cholesterol, 7.22 mmol/L. ECG exhibited atrial fibrillation. Transthoracic echocardiography (TTE) indicated the presence of an irregular right atrial tumor with a little pericardial effusion, yet the left ventricular systolic function was normal. The CMR revealed a tumor in the right atrium, which was isointense on T1-weighted imaging, slightly hyperintense on T2-weighted imaging, and showed obviously homogeneous enhancement on postcontrast T1-weighted imaging. Fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (FDG-PET/CT) imaging demonstrated a hypermetabolic lesion in the right atrium (RA) area, and no other areas of pathological FDG...
uptake were observed in the rest of the body. The patient received a tumor resection surgery in another hospital. A detailed histological examination of the tumor confirmed that it was DLBCL. Immunophenotypic analysis revealed that the tumor was positive for CD20, CD79a, BCL2, BCL6, and MUM1. Then, the patient returned to our hospital and subsequently commenced on R-CHOP chemotherapy, which consisted of rituximab, cyclophosphamide, clarithromycin lactobionate, vindesine, and dexamethasone. After 5 cycles of chemotherapy, the patients achieved complete remission. The follow-up has lasted for 20 months since.

A considerable part of PCLs occurs in immunocompromised patients, especially in individuals with human immunodeficiency virus infections or transplant recipients. Pathologically, the most common type of PCL is of B-cell origin (especially DLBCL). The strict definition of PCL is that the tumor involves only the pericardial space and myocardium, without any other evidence of lymphoma on autopsy. It is reported that PCL presents more common in the elderly age, with a male-to-female ratio of 2(3):1. The most common symptoms include dyspnea, chest pain, arrhythmia, palpitation, weight loss, and fatigue. There are also some cases presenting serious symptoms such as heart failure due to obstruction of blood flow through the cardiac chambers or pericardial effusion. However, all the above symptoms are nonspecific.

Obviously, it is of great significant to choose the appropriate approach among several imaging modalities to detect and evaluate PCL. TTE is useful for detecting the tumor with a mode of real-time imaging. CT and magnetic resonance imaging could not only reveal the infiltrating extent but also provide the intratumoral characteristics. More specifically, CT can be used to clearly show the extracardiac spread rather than the intracardiac spread. By contrast, CMR provides a better alternative to demonstrate the infiltration of the myocardium and pericardium, which is the preferred tomographic method for observing PCL currently. PET/CT is helpful in confirming cardiac isolation and deciding whether the tumor is benign or malignant. However, biopsy is still recommended to obtain a correct pathological diagnosis. The imaging findings of PCL have certain characteristics. First, in contrast to myxoma (the most common type of primary cardiac tumor), the most common original site of PCL is the right atrium and right ventricle. Second, it is often presented as isoattenuating lesion relative to the myocardium on CT, while appears as isointense or hypointense mass on both T1- and T2-weighted images and demonstrates heterogeneous enhancement after administration of gadolinium on CMR. Furthermore, the tumor might encase the adjacent coronary artery without invasion on contrast-enhanced CT, which has been reported as the vessel floating sign. Finally, PCL is less likely to present intratumoral necrosis and rarely involved vessels.

In summary, PCL is more common in the right site of the heart, and it is imperative to employ multiple imaging modalities for evaluating the tumor.
Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient’s guardians have given their consent for their images and other clinical information to be reported in the journal. The patient’s guardians understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
This study was supported by a grant from the Research Innovation Program for College Graduates of Jiangsu Province (No. KYZZ15_0335).

Conflicts of interest
There are no conflicts of interest.

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