Primary cardiac lymphoma (PCL) is extremely rare, defined as lymphoma that involves only the heart, pericardium, or both. We report a 73-year-old man who suffered from exertional dyspnea and bilateral lower leg pitting edema for 1 month. Imaging study including cardio-echography, computed tomography, magnetic resonance imaging, and angiography were performed, showing cardiac tumor arising from left ventricular wall associated with pericardial effusion. The patient underwent bilateral partial pericardiectomy and open surgical biopsy. PCL with reactive pericarditis was diagnosed on the basis of imaging features and histopathological findings. We review other reported cases of PCL and summarize the imaging characteristics that help to establish the diagnosis of PCL.

Primary cardiac tumor is a rare entity, with a reported prevalence in autopsy series of between 0.001% and 0.03%. Primary cardiac lymphoma (PCL) accounts for approximately 1.3% of all primary cardiac tumors [1]. In comparison to PCLs, metastatic cardiac lymphomas occur more commonly. Recent advances in imaging and surgical techniques, however, have made the diagnosis of PCL easier. We present a case of PCL which originates from left ventricle and the clinical manifestations and imaging findings.

CASE REPORT

History and Examination
A 73-year-old man suffered from exertional dyspnea and bilateral lower leg pitting edema for 1 month. He had hypertension under medication for years, and smoking for more than 50 years. He was admitted to our cooperating hospital because of suspicious acute cardiogenic shock. In the initial physical examination the patient presented with mild fever and bilateral basal rales in breath sounds. Electrocardiography (ECG) revealed normal sinus rhythm and slight ST-T depression. The results of a hemogram disclosed leukocytosis (white cell count of 11800/μl with 82% segmented neutrophils) and anemia (hemoglobin 11.2 g/dl). The data of cardiac enzymes (CK/CK-MB and GOT/GPT) and tumor markers (α-FP, CEA, β-HCG, CA-199) were within normal.

Imaging Studies
Chest radiography revealed cardiomegaly and bilateral pleural effusion. A series of imaging studies displayed a relatively ill-defined cardiac mass that measured approximately 5 × 3 cm in size and located at the inferior posterior wall of left ventricle, associated with pericardial effusion and pleural effusion. This mass demonstrated hypoechoic and
solid pattern on transthoracic echocardiography and transesophageal echocardiography (TEE) (Fig. 1), and hypodense pattern with heterogeneous enhancement on computed tomography (CT) scans (Hispeed CTi scanner, GE medical systems, Milwaukee, Wis) (Fig. 2a, 2b). Magnetic resonance (MR) imaging (1.5T Signa Cvi, GE medical systems, Milwaukee, Wis) showed an intramural mass that was isointense to myocardium on fast spin-echo T1-weighted (FSE T1W) images (repetition time/echo time [TR/TE] 1379/44.1 ms echo train: 32) (Fig. 3a) and heterogeneous high signal intensity on FSE T2W images (TR/TE 21498/97.7 echo train: 16) (Fig. 3b); there was less central enhancement on postcontrast SE T1W fat saturation images (TR/TE 500/20) (Fig. 3c). The findings of pericardial effusion, mild pericardial thickening, and an obliterated interphase between the tumor and the pericardium suggested the probability of pericardial invasion or reactive pericarditis (Fig. 4a, 4b). Coronary angiography and ventriculography revealed adequate performance of the left ventricle (ejection fraction 65%), regional hypokinesia at the inferior posterior wall of left ventricle, and possible external compression to a posterior interventricular branch of right coronary artery; neither tumor blush nor neovascularity was detected.

No other tumor focus was identified on the staging CT scans. These results led to the impression of primary cardiac tumor. However, according to the above clinical manifestations and imaging findings, it was still difficult to achieve a definite diagnosis. The patient then underwent bilateral partial pericardiectomy and open surgical biopsy.

**Histopathological Examination**

The tumor displayed characteristic microscopic features: large, blue, round, cells with prominent central nucleoli and with thick nuclear membrane distributed between the myocardium cells (Fig. 5). Fibrinous exudates, reactive fibroblasts, fibrosis, and...
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Pericardial wall thickening infiltrated by lymphoplasmocytes were also found; however, malignant cells were not seen in the pericardial tissues. The key results of immunohistochemical staining for the tumor included positive reactions for leukocyte common antigen and L-26, and negative reactions for ubiquitin carboxyl terminal esterase L 1, cytokeratin, and Epstein–Barr virus-encoded small RNA 1 in situ hybridization.

Based on the histopathological results, the final diagnosis was stage I (Ann Arbor Staging Classification) PCL—diffuse large B-cell lymphoma—originating from left ventricular wall accompanied by fibrinous pericarditis.

**Postoperative Course**

Regimens of CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) were admin-

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**Figure 3.** Axial MRI of the heart. a. FSE T1WI (TR/TE 1379/44.1 echo train: 32). b. FSE T2WI (TR/TE 21498/97.7 echo train: 16). c. SE T1WI+Gd+fat-saturation (TR/TE 500/20). An isointense tumor at the left ventricle wall on FSET1W image (white arrows) shows heterogeneously high signal intensity on FSET2W image (black arrows) and slightly peripheral enhancement (white arrows).

**Figure 4.** MRI of the heart. a. end-systole and b. end-diastole precontrast short-axis ECG gated FIESTA (Fast Imaging Employing Steady State Acquisition) images (TR/TE 3.9/1.7 Filp angle: 40°) demonstrates the location and the extent of the tumor more clearly. The associated pericardial effusion, mildly thicken pericardium, and obliterated margin between the tumor and the pericardium can also be seen (black arrows).
istered. The tumor shrank in response to the first course of chemotherapy. Unfortunately within half a year, rapid growth of recurrent tumor was found before the second course of chemotherapy. The patient was thereafter lost to follow up.

**DISCUSSION**

PCL is uncommon, and it accounts for approximately 1.3% of all primary cardiac tumors [1]. In contrast, cardiac spreading of non-Hodgkin’s lymphoma is more common (approximately 16–28% in one autopsy series) [2]. In 1978 McAllister and Fenoglio first defined a PCL as an extranodal type of non-Hodgkin’s lymphoma that involves only the heart, pericardium, or both [1]. Absence of lymphatic vessels and lymph nodes may be reason for the infrequent occurrence of PCLs, which hypothetically arise from primitive multipotential mesenchymal cells [1].

The reported age of patients with PCLs ranges from 13 to 90 years (mean 60 years); there is a slight male predilection and a higher incidence in immunocompromised patients [1]. Immunocompetent patients can also be affected. The presenting symptoms include shortness of breath, superior vena cava obstruction, cardiac tamponade, chest pain, etc. The symptoms varies, depending on the site and extent of tumor involvement [1, 3, 4]. In addition, some patients may present with arrhythmia because of tumor infiltration of myocardium and subsequent interference with the conducting system. Although the nonspecific symptoms may result in misdiagnosis of a PCL, the use of advanced imaging modalities can be helpful in accurate interpretation. Cardiomegaly remains the first choice for initial evaluation of cardiac masses. CT and ECG-gated MRI can provide more information about tumor characteristics, location, and extent [1, 5].

According to the reported cases in the literature, PCLs are typically demonstrated as hypoechoic solid masses on sonography and as hypodense masses on CT scans. Furthermore they seldom show fatty, calcified, necrotic, or hemorrhagic components. The tumors are primarily iso- to hypointense on T1W images and iso- to slightly hyperintense to myocardium on T2W images. Unlike ordinary lymphomas, PCLs always display heterogeneous enhancement after administration of contrast medium. The explanation may be that tumor cells inhomogeneously infiltrate the myocardium [1, 3, 4]. More than 80% of PCLs arise from right heart wall and approximately 56% are associated with pericardial involvement [1], which may be related to tumor spreading or reactive pericarditis. The tumors seldom involve the heart valves or extend into the chambers [1]. They can, however, occur in any chamber. Angiography and gallium-67 scanning play a limited role in the diagnosis of PCL [1].

Compared with PCL, angiosarcoma has the highest incidence among primary cardiac malignancy (approximately 37%) and has a tendency to hemorrhage. Rhabdomyosarcoma, osteosarcoma, liposarcoma, and fibrosarcoma have a reported prevalence between 1% and 9%, and frequently contain large areas of necrosis, calcification or fat. Because of the lack of specific characteristic features, other primary cardiac sarcomas, for example malignant fibrous histiocytoma (MFH) or leiomyosarcoma, may mimic a PCL and become a dilemma for diagnosis. Anyway MFH and leiomyosarcoma are more common (approximately 8-24%) and located more frequently at left atrium [3, 4]. Biopsy sampling for pathological analysis and special immunohistochemical staining is still recommended for the diagnosis of PCL. Routine cytology examination of pericardial effusion has been reported but not reliable [6]. Most PCLs are unresectable. There is a report on a trial of TEE-guided intracardiac biopsy to replace open surgical biopsy [7]. The majority, approximately 80%, of the pathological composition of a PCL is that of aggressive B-cell lymphoma [1]. Staging evaluation including abdominal and pelvic CT scans and bone

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**Figure 5.** Photomicrograph of the tumor tissue discloses large, blue, round cells (arrowheads) with prominent central nucleoli and thick nuclear membrane infiltrating the myocardium (arrow).
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marrow biopsy is necessary to exclude metastatic cardiac lymphomas. To our knowledge, the lack of literature on large surveillance series renders the results of treatment for PCLs controversial. Some recent reports indicate that early diagnosis is still clinically significant because treatment by chemotherapy and radiation may be effective and may prolong survival time [1, 8].

In conclusion, the imaging features of PCL are not specific. In practice, when a cardiac mass with unexplained pericardiac effusion is encountered, PCL should be suspected. A mass in the heart and/or pericardium without evidence of fatty, calcified, necrotic, or hemorrhagic components also suggests the possibility of a PCL.

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原發自左心室之心臟淋巴癌：病例報告

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原發性心臟淋巴癌是一種很罕見的腫瘤，而根據報告統計有百分之八十的原發性心臟淋巴癌發生於右側心臟，原發自左側心室更是少見。我們報告一個七十三歲男性病例，因為呼吸急促和下肢水腫來求診，經過完整的影像檢查，包括心臟超音波、電腦斷層、心臟血管攝影、磁振造影，發現一個左心室腫瘤併發心包膜和肋膜積水。經由開心手術做心臟切片和部分心包膜切除術，診斷為原發自左側心室心臟淋巴癌併發纖維性心包膜炎。我們將對於他的臨床表現和影像做一個簡短的討論。