

# Diagnosis and treatment of Primary Cardiac Lymphoma: A Case Report

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## Case Report

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# Abstract

**Background:** Primary cardiac lymphoma (PCL) is an extremely rare cardiac tumor and progresses rapidly, which often has short median survival time and poor prognosis unless diagnosed and treated in time.

**Case presentation:** A 61-year-old man was admitted to hospital with repeated chest tightness and pain for 3 days and the right atrial mass by ultrasonography in the other hospital for 1 day. The echocardiography revealed an irregular hyperechoic mass in the right atrium with the size of the mass is about 5.3cm\*4.3cm. Partial resection of right atrial tumor under general anesthesia and cardiopulmonary bypass was performed. After operation, enhanced chest computed tomography (CT) showed a mild enhancement mass in the right atrium. The histopathological examination revealed diffuse large B cell lymphoma. After careful examination, PCL was diagnosed and chemotherapy of low dose R-CHOP for 6 courses were used. After the last course, enhanced CT examination showed that the tumor almost disappeared and the patient's symptoms improved significantly.

**Conclusion:** PCL is an extraordinarily rare disease with poor prognosis, and it is necessary to improve the awareness of this disease. Prompt diagnosis and timely chemotherapy in combination with surgery could lead to excellent clinical outcomes of the patient with PCL.

## 1. Background

Primary cardiac lymphoma (PCL) is an extremely uncommon cardiac tumor, which accounting for only 1.3% of all cardiac tumors and only 0.5% of all extranodal lymphomas [1, 2]. The incidence of PCL is low, but once the disease starts, it will progress rapidly and the median survival time is only about 12 months, so timely diagnosis and treatment are extremely important [3].

## 2. Case Presentation

Herein we present a case of a 61-year-old man who was referred to our hospital. The patient had a history of repeated chest tightness and pain for 3 days and found a right atrial mass by ultrasonography in the other hospital for 1 day. The patient had no complaints of the B symptoms (night sweats, body temperature above 38 degree centigrade or weight loss of more than 10% of body within six month). The echocardiography revealed an irregular hyperechoic mass in the right atrium with the size of the mass is about 5.3cm\*4.3cm, which is connected to the lateral wall of the right atrium without obvious movement (Fig. 1). After admission, partial resection of right atrial tumor under general anesthesia and cardiopulmonary bypass was performed. During the operation, we can see the mass in the right atrium with a diameter of about 6cm, fused with the myocardium and presented invasive growth. The mass has a jelly fish-like texture, protruding from the right atrial and blocking the opening and closing of the tricuspid valve. The mass was partial excised along the edge of the tumor for biopsy, and also can relieve the effect of the mass on the activity of the tricuspid valve. After the surgery, careful examinations were performed, including enhanced CT and positron emission tomography-computed tomography (PET-CT).

Plain CT showed a low-density mass with necrosis that appears irregular in the right atrium, following enhancement CT scan findings of the tumor was slightly enhancing and the necrosis was no enhancing, the size of the tumor was about 6.1cm\*7.7cm, and the right coronary artery can be seen floating on the tumor (Fig. 2). PET-CT showed the radioactivity uptake of right atrium was increased extremely, and the size of the uptake area was about 9.2 cm\*6.3 cm, the standard uptake value (SUV) max was 29.03 (Fig. 3). The histopathological examination revealed diffuse large B cell lymphoma, immunohistochemical results showed CD20 (+), CD79a (+), PAX5 (+), MUM1 (-), BCL-6 (-), CL-2 (+), C-MYC (-), CD30 (-), Eber (-), CD117 (-), KI-67 (60%), Vimentin (+), CD31 (-), Desmin (-), CK (-) (Fig. 4).

The patient was started chemotherapy with low dose of R-CHOP for 6 courses. The symptoms of panic and chest distress gradually disappeared as the chemotherapy treatment continued and the mass gradually smaller. After 7 months, the treatment continued for five cycles and at the last chemotherapy, the patient underwent another enhanced CT examination, which showed the tumor almost disappeared (Fig. 5). The symptoms of patient disappeared completely after 6 courses of chemotherapy.

### 3. Discussion

PCL is defined as extranodal lymphoma that occurs only in the heart and pericardium. On the other hand, when there is extranodal lymphoma in the heart with asymptomatic extracardiac localized lesions can also be diagnosed as PCL [4]. PCL belongs to non-hodgkin's lymphoma, and its main pathological type is diffuse large B cell lymphoma. The most common site of PCL is right atrium, followed by right ventricle, left ventricle, left atrium and atrial septum. Our patient was diagnosed as PCL, because there was no evidence of lymphoma present in any other organ of him and his histological exams consistent with diffuse large B cell lymphomas. Symptoms of PCL are usually nonspecific and associated with the extension of the tumor, differing from lymphoma located in other organs, PCL often causes cardiovascular events, such as heart failure, dyspnea, arrhythmia and pericardial effusion.

Although definitive diagnosis is confirmed by pathology, multimodality imaging optimizes the diagnostic evaluation. Echocardiography is often the preferred imaging method, but CT imaging can provide preferable soft tissue contrast and anatomic information. On CT image, PCL is usually presented as a single or multiple iso-density or low-density mass which infiltrating into the myocardium, with slight enhancement on contrast-enhanced scans. Meanwhile, coronary artery can be seen floating in the mass, which is a typical imaging manifestation of lymphoma called vascular floating sign [5, 6]. Nuclear medicine imaging (PET-CT) may assist in differentiate from other more common cardiac tumors, high uptake is the specific manifestation of PCL [7]. Previous study has shown that SUV values of PCL patients was significantly higher than those of patients with other cardiac malignancies (such as metastatic tumors, sarcomas) and benign tumors [8].

The main treatment of PCL is the combination of surgery and chemotherapy [9]. It is almost impossible to remove the tumor by single surgery, the purpose of surgery is to alleviate clinical symptoms, remove mechanical obstruction when hemodynamics is disturbed, and provide basis for chemotherapy. It also

provides pathology information which could make us determine the pathological type of the tumor by biopsy [10]. Chemotherapy is currently described as the most effective treatment for PCL, and it has been reported in the literature that 61% of patients can get remission from chemotherapy alone [11]. Chemotherapy with CHOP (cyclophosphamide, doxorubicin, vincristine and prednisone) is a classic treatment for Hodgkin's lymphoma and it's also used for the treatment of PCL. In this case, the tumor hindered the normal opening and closing of the tricuspid valve, resulting in the interference of venous blood flow to the pulmonary artery. Therefore, partial tumor resection was performed to remove the effect of the tumor on the tricuspid valve activity and correct hemodynamics. After R-CHOP treatment for 6 courses, the tumor was significantly smaller and the patient had an excellent clinical outcomes.

The prognosis for patients with PCL is usually poor, and the median survival is only about 12 months. Benign tumors such as cardiac myxoma account for a high proportion of primary cardiac tumors, while PCL account for only 1.3% of primary cardiac tumors. Due to the rarity of PCL and atypical radiologic, people usually cannot get a complete recognition of it, which maybe the reason of unable to diagnose in time. PCL is also considered as an acute tumor because of the severe infiltration of the myocardium and the rapid progression of the tumor. Late diagnosis and rapid evolution of the tumor are major factors causing the poor prognosis.

## **4. Conclusion**

PCL is a very rare disease with poor prognosis. This case provides an example of a patient with prompt diagnosis and appropriate chemotherapy in combination with surgery, which lead to excellent clinical outcomes. It is necessary to diagnose PCL as early as possible and it should be kept in mind in the antidiastole of cardiac tumors. At the moment, chemotherapy is an effective treatment and can improve the prognosis.

## **Abbreviations**

PCL: Primary cardiac lymphoma; CT:computed tomography; PET-CT:positron emission tomography-computed tomography; SUV:standard uptake value.

## **Declarations**

### **Ethics approval and consent to participate**

This study was approved by the ethics committee of Nanjing Medical University.

### **Consent for publication**

This manuscript and accompanying images were published with the patient's written informed consent.

## **Availability of data and materials**

**All data generated or analyzed during this study are included in this published article.**

## **Competing interests**

The authors have no conflicts of interest to declare.

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## **Authors' contributions**

ZA and LJ collected the image data. XY made the diagnosis. MP provided the clinical information. ZA and LJ drafted the manuscript. XY and HX participated in manuscript revision. All authors read and approved the final manuscript.

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Not applicable

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## Figures

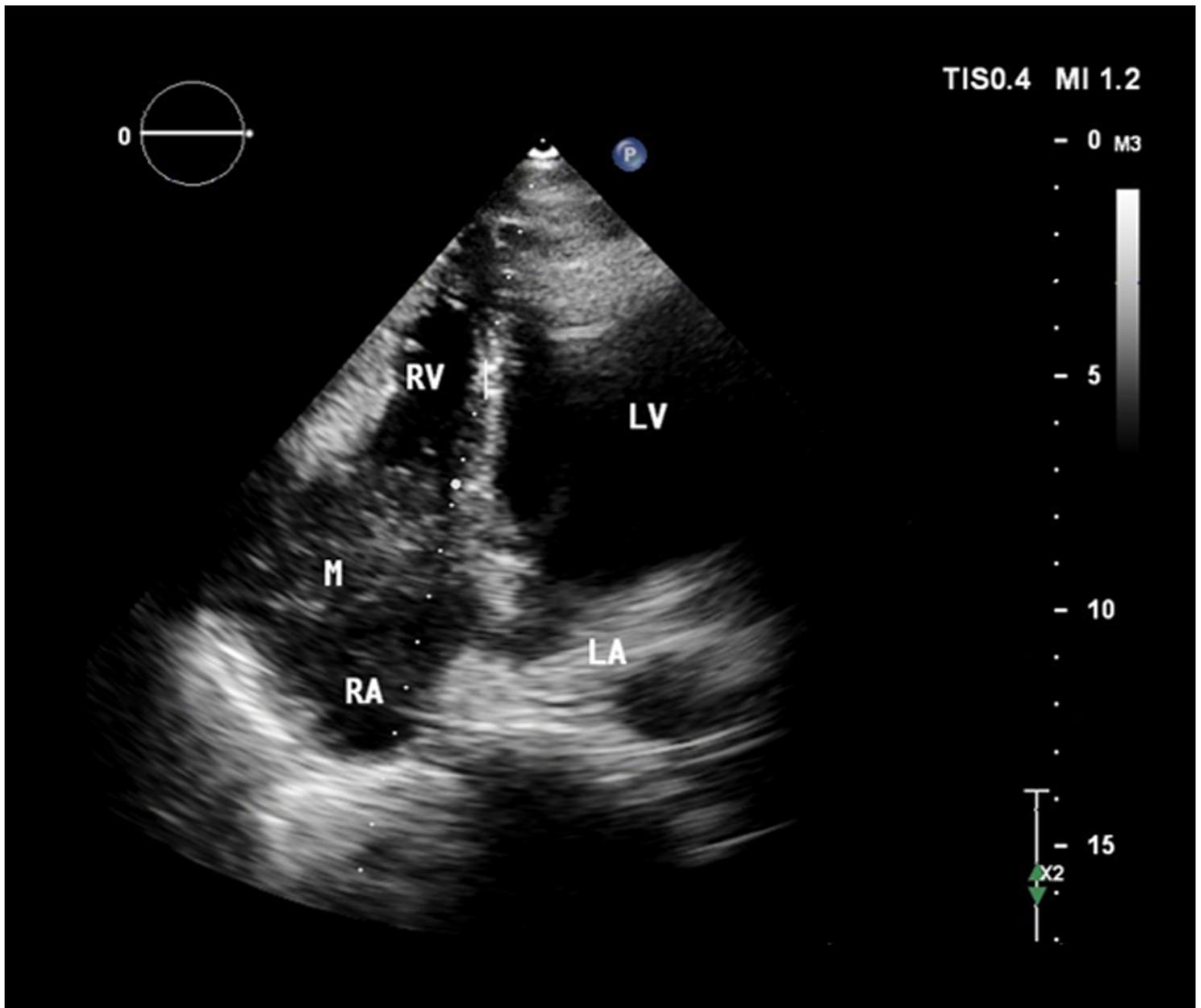
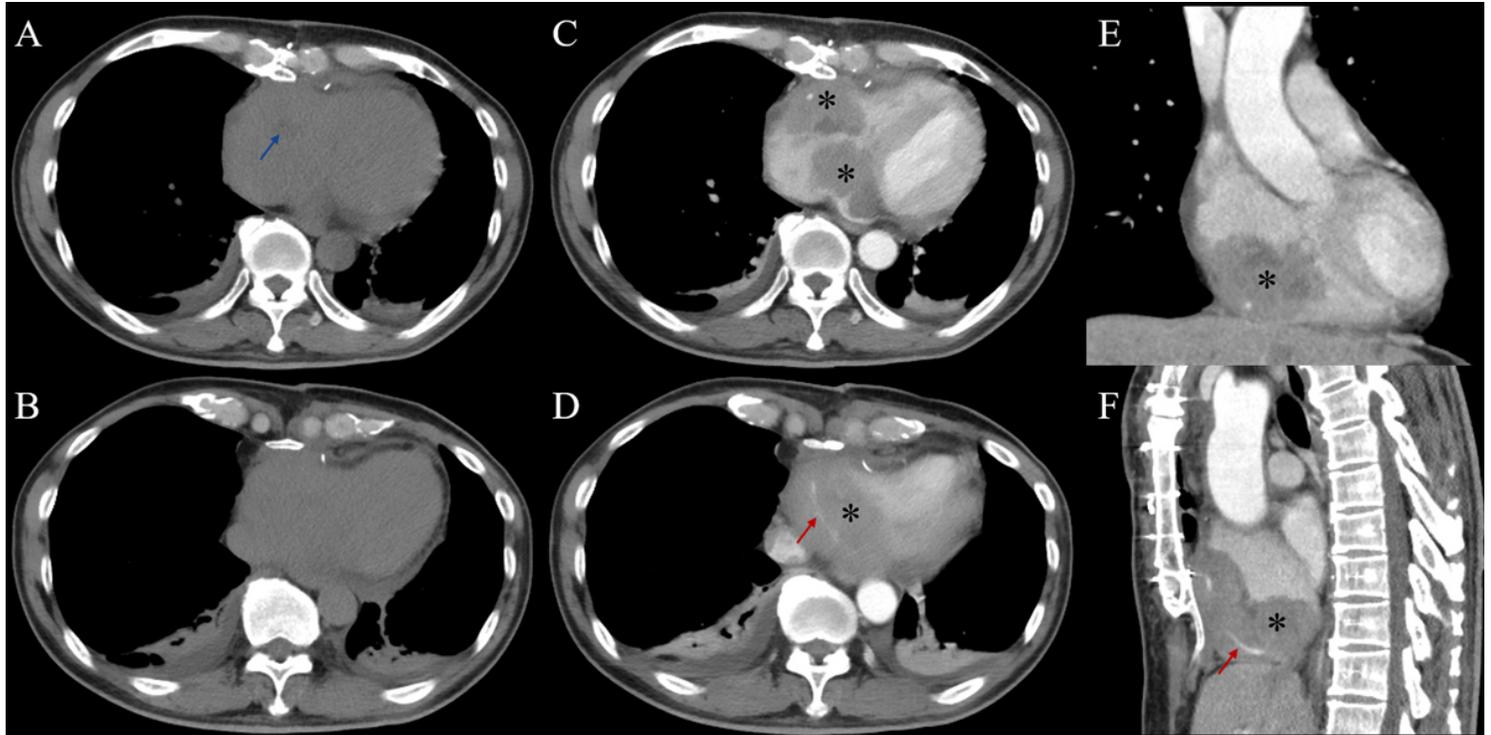


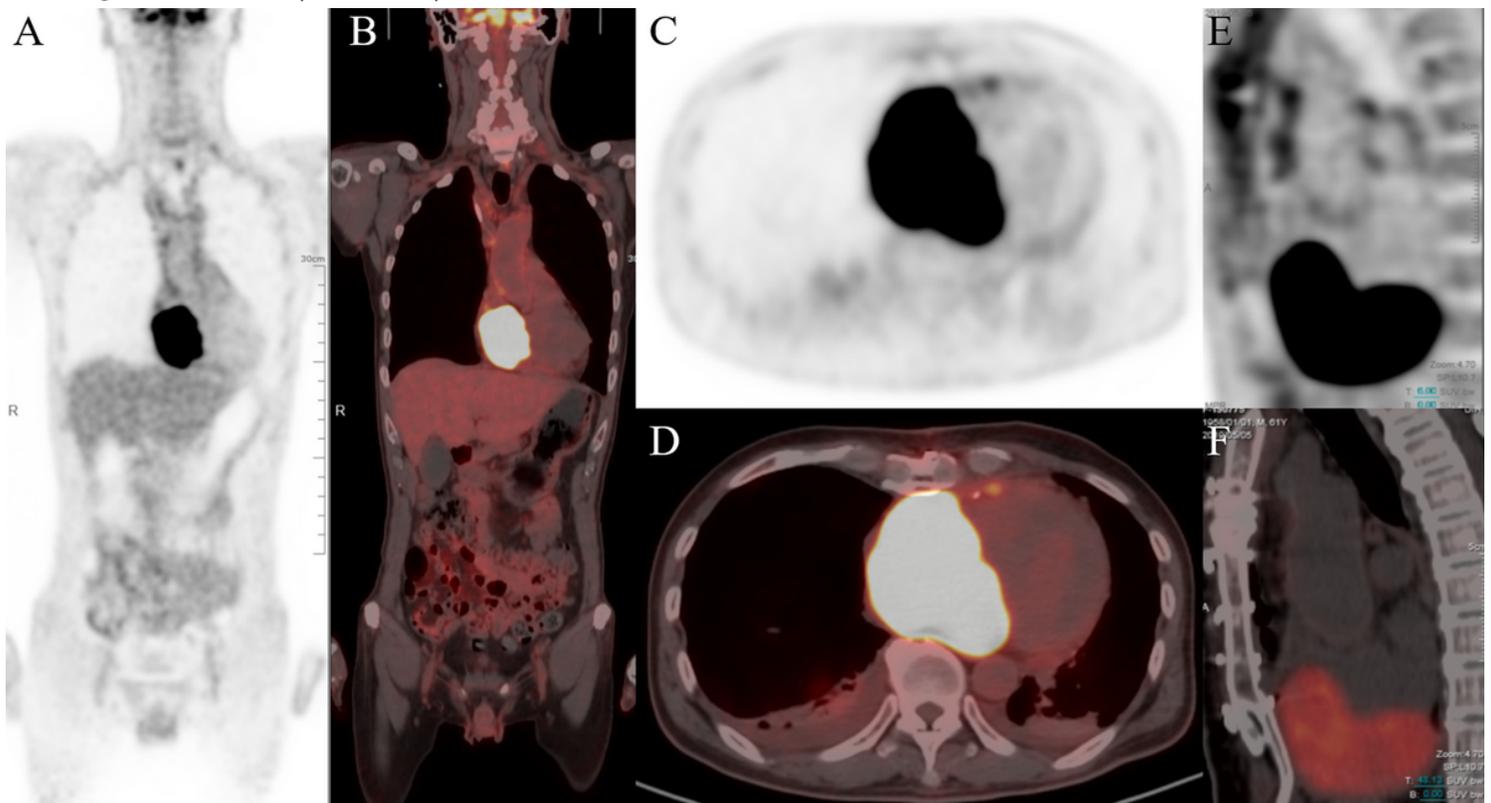
Figure 1

Preoperative echocardiography image shows an irregular hyperechoic mass in the right atrium. RA: right atrium, RV: right ventricular, LA: left atrium, LV: left ventricular, M: mass.



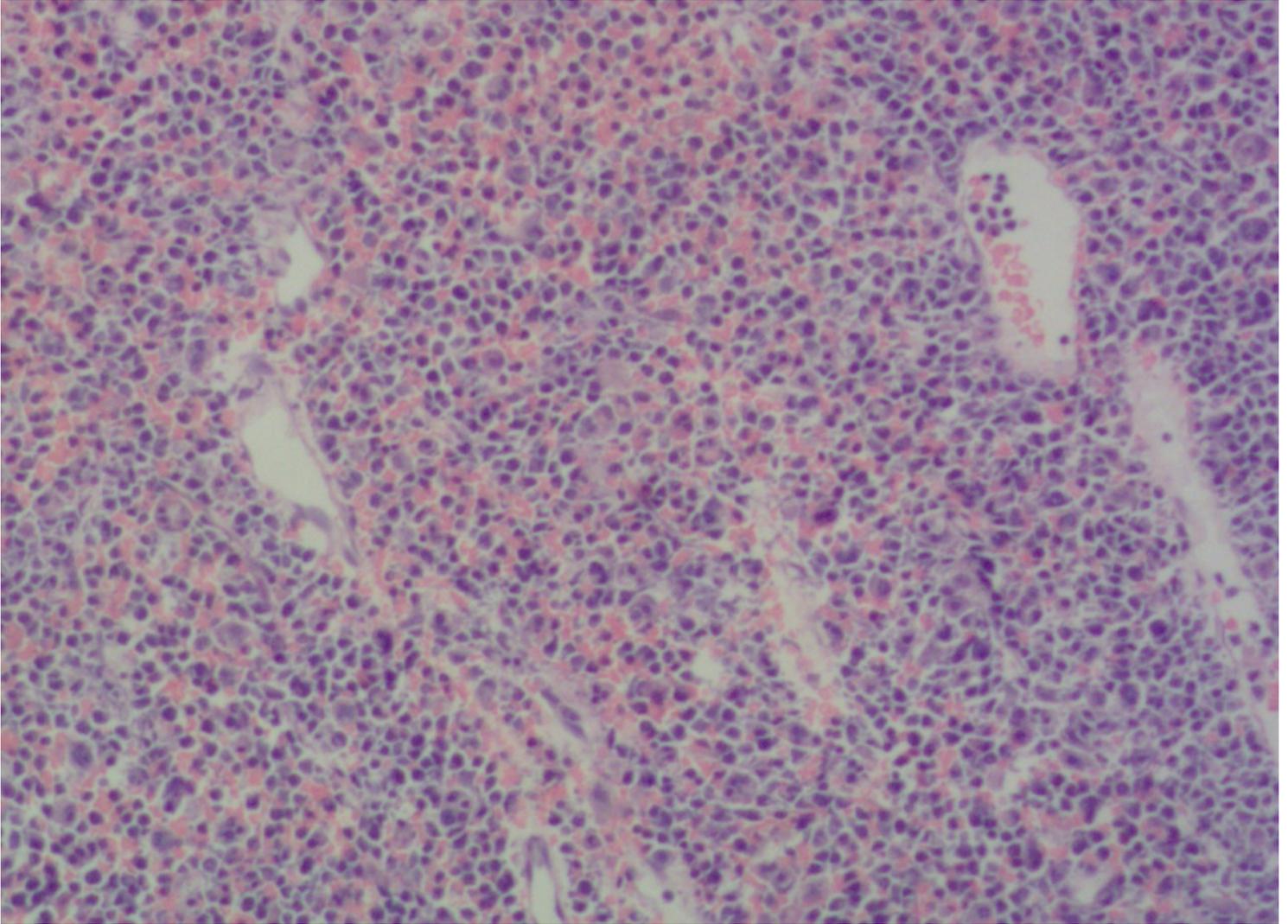
**Figure 2**

Axial plain CT (A, B) shows patchy low density in the right atrium (blue arrow); Enhanced CT (C-F) shows a mild enhancement mass with necrosis in the right atrium (black asterisk) and right coronary artery floating in the mass (red arrow).



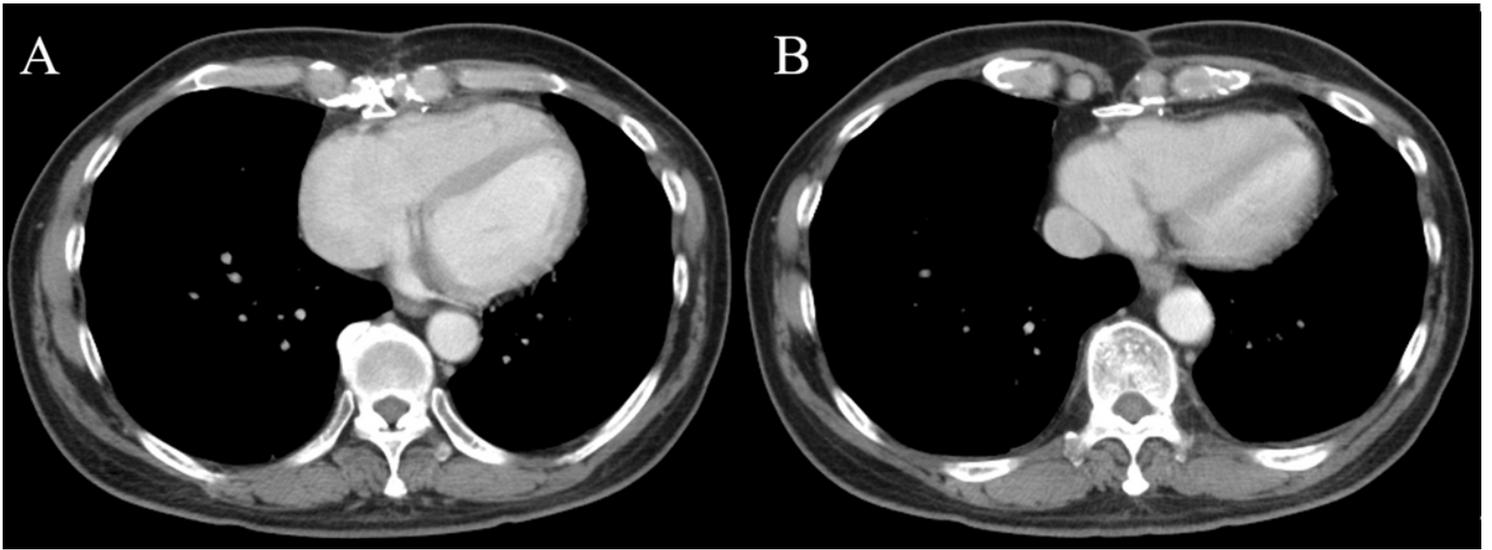
### Figure 3

PET-CT (A and B: coronal images; C and D: axial images; E and F: sagittal images) shows high uptakes in the right atrium (SUV max was 29.03).



### Figure 4

Tumor histology shows diffuse myocardial infiltration with large B cells (Hematoxylin-eosin stain, original magnification  $\times 100$ ).



**Figure 5**

Follow-up enhanced CT after the last chemotherapy shows that the tumor almost disappeared.