

[CASE REPORT]

Primary Cardiac Diffuse Large B-cell Lymphoma Promptly and Safely Diagnosed with Pericardial Effusion Cytology

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Abstract:

Primary cardiac lymphoma is a rare condition with a poor prognosis, and patients are at risk for sudden cardiac death. A prompt diagnosis and early treatment are therefore essential. A 68-year-old woman was admitted for shortness of breath and peripheral edema. Echocardiograms showed massive pericardial effusion and a mass on the free wall of the right atrium and ventricle. Subsequent pericardial effusion cytology revealed diffuse large B-cell lymphoma. We started chemotherapy with rituximab and achieved a good clinical course. This case is made unique by the use of pericardial effusion cytology, which allowed us to diagnose primary cardiac lymphoma promptly and safely.

Key words: primary cardiac tumor, primary cardiac lymphoma, pericardial effusion cytology, chemotherapy with rituximab

(Intern Med 59: 1997-2001, 2020)

(DOI: 10.2169/internalmedicine.4716-20)

Introduction

Primary cardiac lymphoma is extremely rare, and its prognosis is often poor (1-3). It often shows rapid growth, and its symptoms may vary according to the heart sites involved. Some patients present with cardiac tamponade or systemic obstruction and may progress to sudden death (4). Prompt diagnosis of this disease is therefore essential to reduce the risk of mortality. We report a case in which we were able to make an early diagnosis and initiate successful treatment for primary cardiac lymphoma.

Case Report

A 68-year-old woman was admitted to our hospital because of worsening dyspnea on exertion. She had been well until a few months before, when she noticed peripheral edema. Her conditions of Hashimoto's disease and ulcerative colitis had been stable for years. A physical examination revealed jugular venous distension and bilateral pitting edema of the lower extremities. There was neither superficial lym-

phadenopathy nor hepatosplenomegaly. Her electrocardiogram showed atrial fibrillation and a low voltage, and chest radiography demonstrated moderate cardiomegaly and bilateral pleural effusion (Fig. 1). Laboratory data were mostly unremarkable except for the elevation of lactate dehydrogenase (594 IU/L) and soluble interleukin-2 receptor (4,870 U/mL).

Cardiac echocardiography showed massive pericardial effusion with a collapsed right ventricle. An abnormal mass was seen on the free wall of the right atrium and ventricle, part of which was adjacent to the tricuspid valve and had mobility in the intracardiac space (Fig. 2). We performed pericardiocentesis and collected 700 mL of bloody pericardial effusion. Laboratory data showed elevation of lactate dehydrogenase (5,420 IU/L) and total protein (4.6 g/dL). Both bacterial and mycobacterial culture were negative, which suggested the presence of malignancy. Pericardial effusion cytology revealed diffuse large B-cell lymphoma (Fig. 3). Both a bone marrow examination and cerebrospinal fluid tests showed no infiltration of lymphoma cells. Contrast-enhanced cranial magnetic resonance imaging (MRI) also revealed no evidence of brain metastasis.

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Received: February 25, 2020; Accepted: March 11, 2020; Advance Publication by J-STAGE: April 30, 2020

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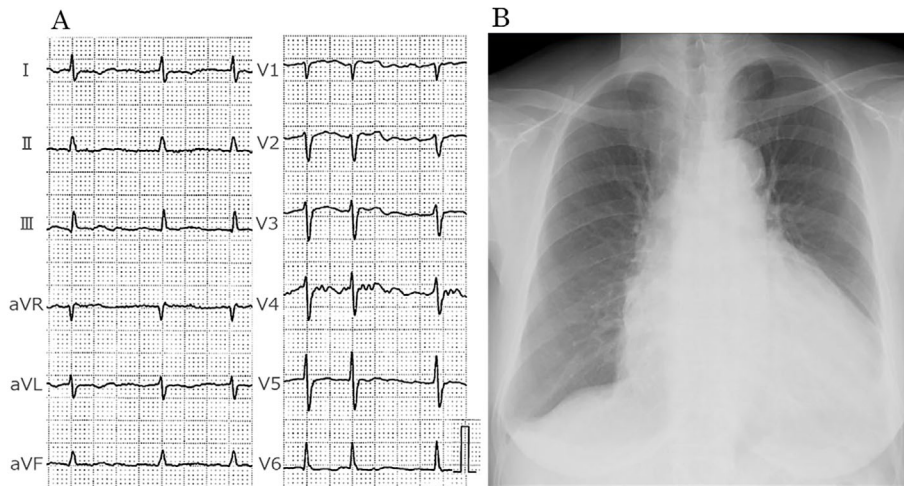


Figure 1. A) An electrocardiogram on admission showed atrial fibrillation and a low voltage. B) Chest radiography revealed moderate cardiomegaly and bilateral pleural effusion.

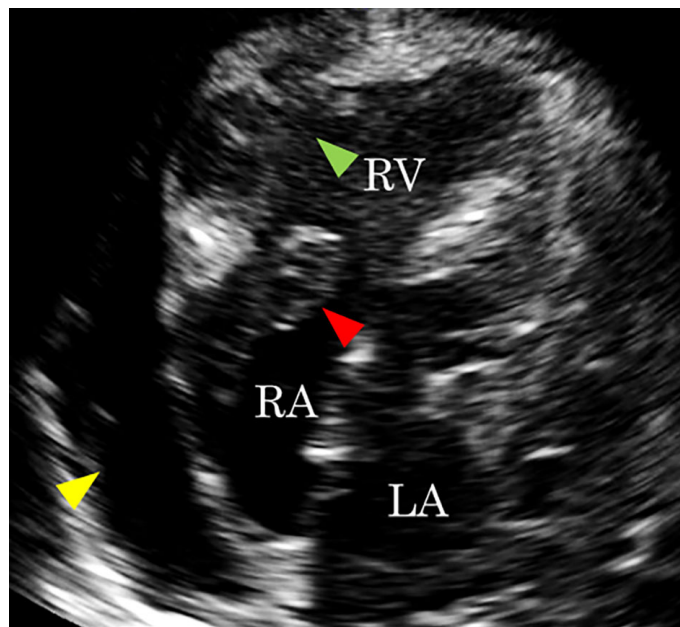


Figure 2. Cardiac echocardiography showed massive pericardial effusion (yellow arrowhead). An abnormal mass was situated on the free wall of the right atrium and ventricle (green arrowhead), part of which was adjacent to the tricuspid valve and had mobility in the intracardiac space (red arrowhead). (LA: left atrium, RA: right atrium, RV: right ventricle)

We diagnosed primary cardiac lymphoma (Ann Arbor stage: IV A, International Prognostic Index: high risk) and treated the patient with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone (R-CHOP) at full dose. After six cycles of chemotherapy, contrast-enhanced computed tomography (CT) and fluorodeoxyglucose-positron emission tomography (FDG-PET)/CT showed a marked reduction in the tumor size (Figs. 4 and 5).

Discussion

A study by Reynen reported that the incidence of primary cardiac tumor was very low at approximately 0.02%, corre-

sponding to 200 tumors per million autopsies (1). Malignant lymphoma is far rarer, comprising approximately 1-2% of those cases (2).

Symptoms of primary cardiac lymphoma are non-specific and may present as cardiac manifestations, including heart failure, pericardial effusion, and arrhythmias such as atrial fibrillation and complete atrioventricular block. However, it may also present with a fever, night sweats, and weight loss (3-5). Some patients present with cardiac tamponade or obstruction of the systemic circulation, which, depending on the tumor site, may cause sudden death (4). Primary cardiac lymphoma progresses far more quickly than benign cardiac tumor or angiosarcoma, which comprises the majority of

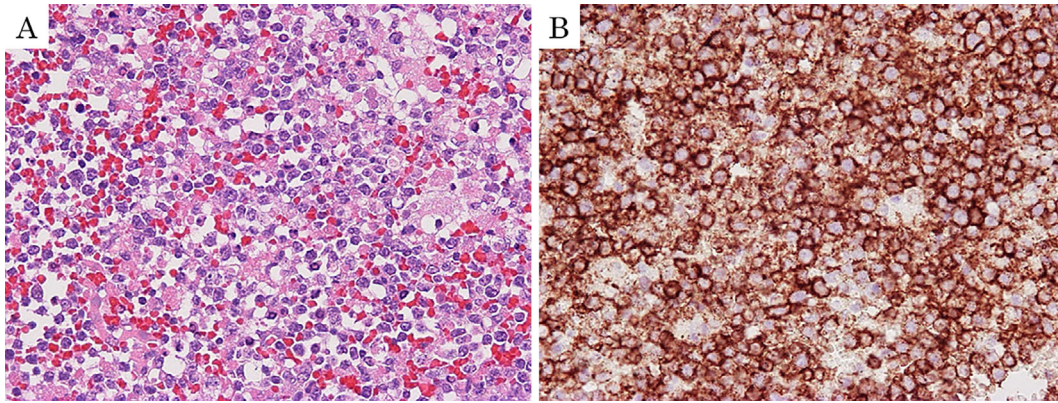


Figure 3. A) Cytology revealed highly pleomorphic large lymphocytes with markedly irregular nuclei on Hematoxylin and Eosin staining ($\times 400$). B) Immunostaining showed that the neoplastic lymphoid cells were diffusely positive for CD20 ($\times 400$).

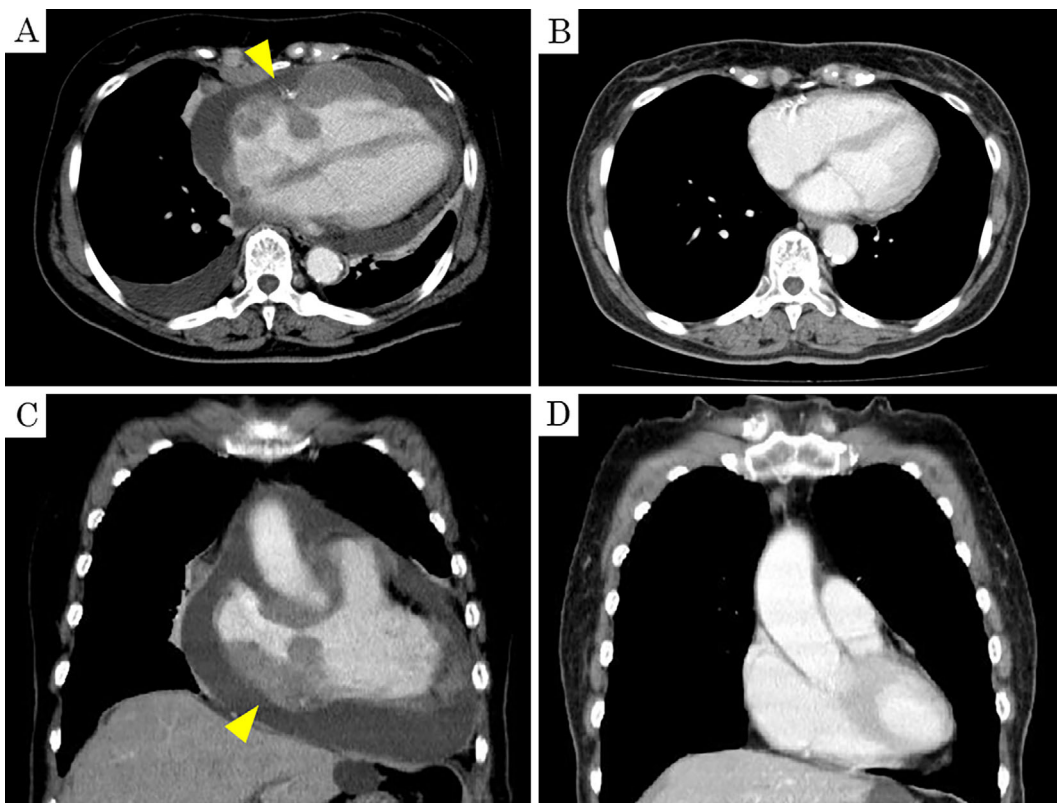


Figure 4. A, C) Contrast-enhanced computed tomography revealed an abnormal mass situated on the free wall of the right atrium and ventricle (yellow arrowhead). B, D) After six cycles of chemotherapy, the tumor showed a marked reduction in size.

malignant cardiac neoplasms (5-7). The diagnosis of primary cardiac tumor has been improved by the use of echocardiography, CT, MRI, and nuclear imaging (4). However, the diversity and non-specificity of its symptoms, as well as its rarity, still make the diagnosis difficult. In our case, the patient also showed non-specific symptoms, such as exertional dyspnea, chest discomfort, and peripheral edema. Therefore, we could not have predicted that she might be at risk for sudden death if we had not requested echocardiography be performed. Pérez et al. reported a case in which the tumor developed on the right side of the heart, eventually progress-

ing to tumoral pulmonary embolism, followed by death (8). It is thus clear that an early diagnosis is essential for preventing such complications.

The diagnosis of lymphoma is ultimately made only after a histological evaluation of the tumor is performed. A transvenous tumor biopsy and open biopsy are often performed. In cases in which the tumor has mobility in the intracardiac space, there is an especially high risk of tumor embolism, and the biopsy itself may lead to death (9-11). What makes this case unusual is our use of pericardial effusion cytology to obtain a histological diagnosis, which allowed us to avoid

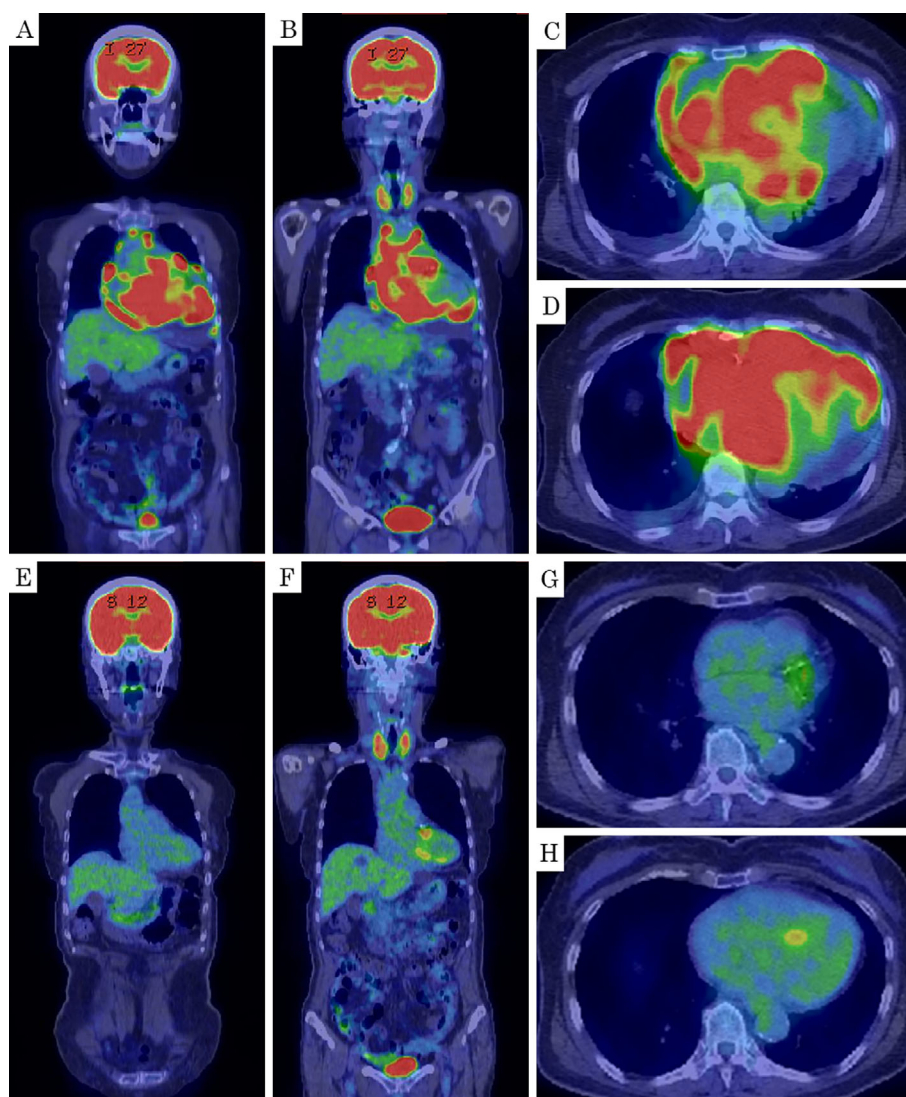


Figure 5. A, B, C, D) An abnormally high accumulation of FDG was detected in the cardiac tumor (SUV_{max} 29.33). E, F, G, H) The abnormal uptake decreased after six cycles of chemotherapy.

the risk of embolism associated with a direct biopsy.

Chemotherapy is commonly used to manage primary cardiac lymphoma. Radiation therapy is sometimes combined with chemotherapy. If cardiovascular system collapse occurs, surgery is sometimes performed as a palliative treatment to reduce the tumor volume (5, 12). In our case, we treated the patient with a standard chemotherapy protocol of six cycles of R-CHOP (3, 4). Five months later, her transthoracic echocardiography and positron emission tomography showed the marked reduction of the tumor and maintenance of remission of the disease.

The 5-year mortality rate is approximately 40% (3). However, in this case, we were able to make the diagnosis through pericardial fluid cytology, which incurs less risk than a transvenous or open biopsy. This allowed us to initiate medical treatment promptly, thus preventing the occurrence of sudden cardiac death in this patient. To improve the chances of a patient survival, the importance of an early diagnosis and implementation of treatment should be emphasized.

The authors state that they have no Conflict of Interest (COI).

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Intern Med 59: 1997-2001, 2020