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DOI:10.14744/AnatolJCardiol.2020.59507

An unusual case of cardiac lymphoma diagnosed using computed tomography-guided percutaneous transthoracic biopsy

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Introduction

Cardiac tumors are the extremely rare and least investigated tumors in oncology. The most common type of tumor originating primarily in the heart is myxoma, while other types are sarcoma, lipoma, fibroelastoma, teratoma, lymphoma, and mesothelioma. Only 10% of cardiac tumors are malignant, and 95% of which are sarcomas and remaining 5% are lymphomas and mesotheliomas (1).

Primary cardiac lymphoma (PCL) is a type of non-Hodgkin lymphoma, which originates from myocardium or pericardium and those are the only sites of involvement at the time of diagnosis. The patients are mostly, but not always, admitted with cardiac manifestations (2). PCL is very rare and fatal unless it is

diagnosed and treated on time. Patients with PCL usually die few months after diagnosis (3, 4).

In this article, we present a challenging case of PCL diagnosed using computed tomography (CT)-guided percutaneous transthoracic biopsy of the cardiac mass encircling the atrioventricular septum.

Case Report

A 63-year-old female patient was admitted to the cardiology clinic with the complaints of dyspnea, weight loss, edema on the legs, and prominent fatigue. According to her definition, she has lost more than 8 kg in the last 2 months, involuntarily. She has a history of diabetes and was on metformin for the last 6 years with excellent glycemic control. She did not report any microvascular complications. She did not have any history of cardiac or pulmonary diseases. In the physical examination, it was found that she had clear S1 and S2, her pulse was regular, and no murmur or gallop was heard. There was prominent pretibial edema on both legs. The laboratory results revealed a high sedimentation rate of 85 mm/h, normal complete blood count except mild iron deficiency anemia (Hb: 11 mg/dL, MCV: 72, and ferritin: 3), elevated C-reactive protein (22 mg/dL), and three-fold increase in lactate dehydrogenase (650 u/L, normal range: 110-206 u/L). ECG and routine biochemistry were normal. As the patient described that dyspnea was worsening during exercise or emotional stress, echocardiogram and chest CT were ordered. The transthoracic echocardiogram which was performed under suboptimal conditions revealed normal ejection fraction and no thrombus, vegetation, or intraventricular mass. In the chest CT, a



Figure 1. Cardiac mass encircling the atrioventricular sulcus and atrium wall

mass sized 2.7 cm was detected, which was encircling the right atrium and atrioventricular septum and compressing the inferior vena cava (Fig. 1). There was pericardial effusion, of which the thickest diameter was 16 mm, and accompanying bilateral pleural effusion. Positron emission tomography (PET) imaging was performed with 18F-fluorodeoxyglucose (FDG) to understand if the lesion is benign or malignant and there was any extracardiac involvement. The procedure was performed after a ketogenic high-fat diet that was given to the patient 2 days before the procedure to enhance the decreased physiologic glucose utilization of the heart during imaging. PET showed a focal uptake of glucose in the cardiac mass with standardized uptake value of 8. The cardiologists tried to obtain a percutaneous transvenous endomyocardial biopsy of the mass, but it was unsuccessful due



Figure 2. CT image of the needle penetrance during percutaneous biopsy

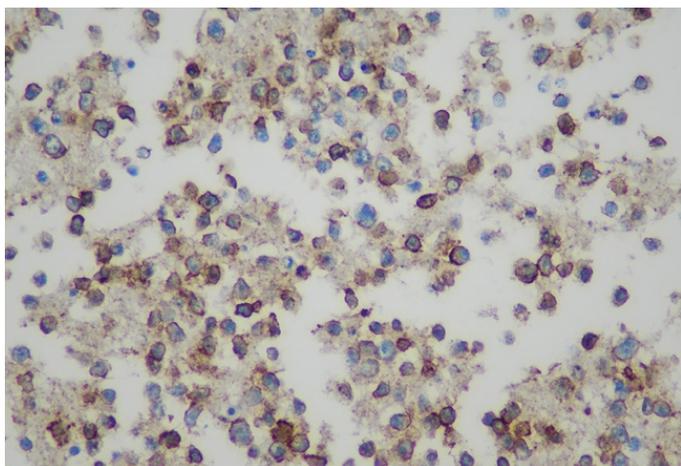


Figure 3. CD20 positive B lymphocyte infiltration in the immunohistochemistry staining of the biopsy specimen taken from the cardiac mass

to the lack of enough specimens to reveal the possible diagnosis. In addition, the patient had arrhythmia and hypotension during the procedure, which limited the efficacy. While waiting for the diagnosis, patient's general condition deteriorated and the degree of dyspnea increased. Mini thoracotomy for repeat biopsy was planned, but anesthetists considered the procedure as high risk and did not approve general anesthesia. The patient was then referred to the interventional radiology clinic and a CT-guided percutaneous transthoracic biopsy was performed under local anesthesia (Fig. 2). No serious adverse events occurred after the intervention. The pathologic evaluation detected atypical lymphoid cell infiltration, and the immunohistochemistry was positive for CD20 positive B cells diagnosing "diffuse large B-cell lymphoma" (Fig. 3). Chemotherapy was started immediately after the biopsy.

Discussion

There are no predefined criteria to call a cardiac tumor as "PCL" Zaharia and Gill (5) state that PCL can be diagnosed if the tumor involves only myocardium or pericardium (5). However, some authors oppose the definition and suggest that PCL can be diagnosed only if the tumor is limited to the heart and there is no extracardiac involvement that is proven by autopsy (6). In our case, the tumor was limited within the myocardium, and there was no extra involvement in the other body sites as proved by CT and PET imaging.

PCL is a rare disease, which composes 1.3% of all cardiac tumors, and is usually seen in the elderly (7, 8). The clinical symptoms of PCL are nonspecific, including the complaints related to heart failure, pericardial effusion, or malignant arrhythmia (9). Our patient was admitted with the symptoms of inferior vena cava compression and heart failure, including pretibial edema, and severe dyspnea.

An echocardiogram may detect the tumor location and measure the size when possible, and a color Doppler may be useful in detecting the blood flow disturbances within the tumor (10). In our case, the echocardiogram was unsuccessful for tumor visualization, and the tumor was detected using chest CT. Also, the inferior vena cava compression was prominent, and the tumor was encircling the right atrium with unclear boundary with endocardium. Other imaging methods that can be used in cardiac mass evaluation are cardiac MRI and PET/CT. Unlike other malignancies, such as sarcomas, lymphomas are reported to lack areas of central necrosis and hemorrhage on MRI. They are typically homogeneous and isointense on T1- and T2-weighted imaging with minimal contrast uptake at late gadolinium enhancement (11). A PET with FDG is useful in differentiating malignant lesions from benign ones. In our case, there was an increased focal uptake of FDG in the cardiac mass without any extra organ or lymph node involvement. In a previous report, FDG-PET was successful in diagnosing mesothelioma originating from pericar-

dium in a 54-year-old male patient who was admitted to the cardiology clinic with severe dyspnea and pericardial effusion (12).

There are several techniques to obtain a biopsy specimen from a cardiac mass. The biopsy of the mass can require open chest procedure, which is very difficult both for the patient and the clinician, and may not be possible if the patient's performance status is poor or anesthesia is risky. Another method is transvenous endomyocardial biopsy, which carries a risk of tumor embolism. It can be performed with the guidance of fluoroscopy or transesophageal echocardiogram (13). Herein, we presented a case of cardiac lymphoma who was diagnosed using percutaneous transthoracic biopsy. To our knowledge, there are very few studies conducted on this method in the literature. In a previous report, a 70-year-old female patient was diagnosed with PCL in the right ventricle wall using a percutaneous transthoracic biopsy with an 18G needle similar to our case (14). In another case, cardiac angiosarcoma was diagnosed using a percutaneous transthoracic biopsy in a 44-year-old man who was admitted for a large serosanguineous pericardial effusion and a tumor invading the right atrium (15). Shah et al. (16) reported a case of B-cell lymphoma in a 72-year-old immunocompetent male patient who presented with paroxysmal atrial fibrillation. A CT-guided trans-sternal biopsy was successfully performed for the histopathological diagnosis in that case.

Conclusion

In conclusion, PCL is a life-threatening malignancy, which should prompt immediate diagnosis and treatment. A tumor sampling is required for the diagnosis of PCL. It can be done using various methods even if the patient's condition is unstable and inappropriate for general anesthesia or sedation for long durations.

Informed consent: Informed consent was taken from the patient for publication of her data.

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DOI:10.14744/AnatolJCardiol.2020.95079

