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Case Report

Mediastinal Mass Invading the Right Ventricle

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ABSTRACT

Lymphoma with cardiac involvement is rare; however, there can be very serious consequences. It is usually revealed in autopsy studies. Cardiac invasion by lymphoma may occur through retrograde lymphatic flow, hematogenous spread, or direct invasion from neighboring structures. Direct invasion is the most common, and has the most destructive results. The clinical signs and symptoms are nonspecific. Presently described is the case of a patient who was initially examined in a cardiology polyclinic due to exertional dyspnea and cardiomegaly seen in a chest X-ray. Echocardiographic examination revealed a suspected mass and the patient was referred to our clinic. A mass lesion filling the mediastinum and invading the right ventricle was detected in a computed tomography image of the chest. On the same day, a tru-cut biopsy with thoracic ultrasound guidance was performed by the pulmonologist. Three days after the biopsy, the patient was referred to the hematology clinic for treatment of pathological lymphoma. This case was presented to draw attention to the importance of prompt diagnosis and treatment of thoracic and mediastinal mass lesions with a multidisciplinary approach and to emphasize that a biopsy can be performed quickly and safely in patients with a mediastinal mass with the guidance of ultrasound, even when there is cardiac involvement.

INTRODUCTION

Cardiac involvement of systemic lymphoma is most often revealed in autopsy studies. Manifestations of lymphomatous involvement of the heart identified at autopsy have a reported incidence of 9% to 24%. Cardiac involvement is believed to take place via 3 different pathways: retrograde lymphatic flow, hematogenous spread, and direct invasion from neighboring structures. Direct invasion is the most frequently seen, and the most destructive.[1] In studies investigating cardiac involvement of lymphoma, a greater incidence has been reported in cases of non-Hodgkin lymphoma. Since, most often, diagnosis is made after death, antemortem diagnosis and prompt treatment using aggressive diagnostic methods are very important.[2]

Ultrasound (US)-guided transthoracic biopsy has the advantages of not exposing the patient or the physician to harmful radiation, the use of Doppler mode may protect the patient from vascular injuries, and the physician can manipulate the needle within the lesion in real-time. Transthoracic biopsy with thoracic US guidance may be preferred for large, mediastinal tumor invasion of the anterior chest wall.[3,4]

This case of a patient who presented at the outpatient cardiology clinic with exertional dyspnea and in whom a mediastinal mass was detected in the vicinity of the heart during echocardiographic examination which necessitated US-guided tru-cut biopsy was presented to draw attention to the importance of rapid diagnosis and treatment, and to share available literature information.

CASE REPORT

The patient had presented at 2 outpatient cardiology clinics with the complaints of chest pain, dyspnea, and orthopnea. Coronary angiography was performed at the first clinic with the suspicion of stable angina pectoris and ischemic heart disease, and a fistula from the left anterior descending artery to the pulmonary artery was revealed. Some time later, the patient consulted another medical center with the same complaints. An echocardiogram revealed a mass lesion 3.5x5.5 cm in size in the vicinity of the right ventricle. There were no symptoms of compression or pericardial effusion leading to cardiac tamponade, so the patient was referred to the outpatient clinic of chest diseases.

Mediastinal enlargement was observed on a chest radiogram (Fig. 1). A thoracic computed tomography (CT) image indicated the presence of a mass lesion extending bilaterally from the upper mediastinal area and surrounding the major vascular structures, the pulmonary artery, and ascending aorta. This mass lesion, 101x96x181 mm at the widest point, compressed the right atrium and right ventricle on interposing planes, which raised the suspicion of invasion (Fig. 2). The physical examination and examination of other systems were unremarkable. The hemogram, biochemical parameters, and coagulation profile of the patient were within normal limits. On the day of admission to the outpatient clinic of chest diseases, the patient underwent US-guided tru-cut biopsy (Fig. 3). The histopathology result reported 3 days later was diffuse large B-cell lymphoma (Fig. 4). Positron emission tomography/computed tomography (PET-CT) indicated diffusely increased fluorodeoxyglucose (F18-FDG) uptake (SUD max 51.6) in conglomerated lymphadenopathies, and a mass lesion 101x96x181 mm in size at the maximal diameter. The mass extended from the upper mediastinal region to occupy the superoinferior paratracheal region, enclose the vascular structures, aortic arch, major brachiocephalic vascular structures, both pulmonary arteries, and the ascending aorta on planes between the right atrium and the right ventricle. The mass lesion also completely enclosed vena cava superior and surrounded the vena cava inferior from the medial aspect. The patient was referred to the hematology polyclinic (Fig. 5). The patient underwent 6 cycles of CHOP treatment (cyclophosphamide, doxorubicin, vincristine, and prednisone). At the third month control visit, a PET-CT displayed dramatic disease regression and

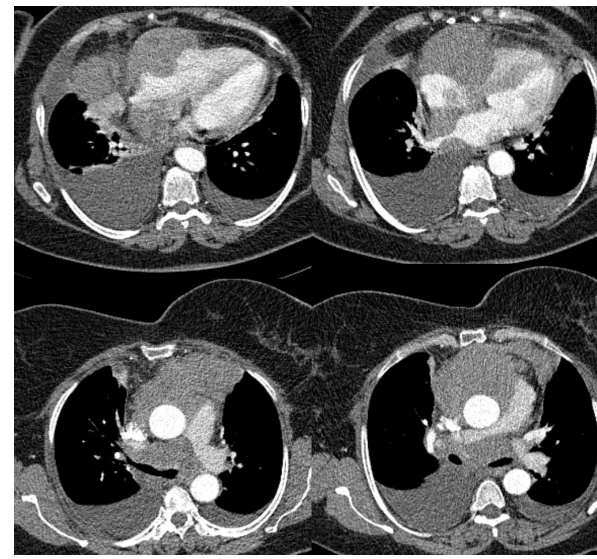

Figure 2. Thoracic computed tomography revealed a mass lesion with maximal dimension of 101x96x181 mm. It extends from the mediastinal area and compresses the right ventricle, which suggested the presence of invasion.

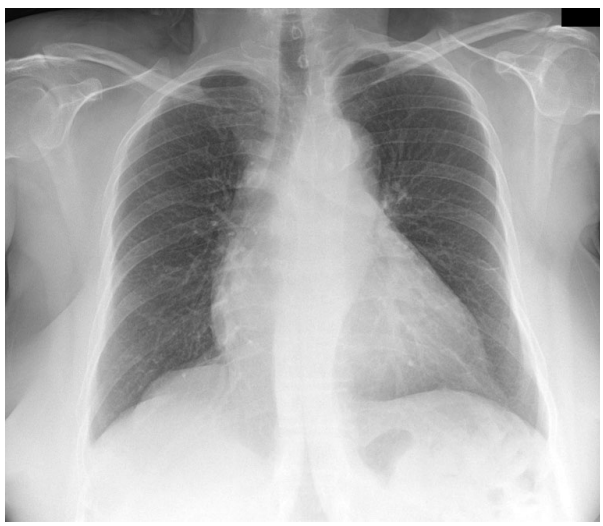

Figure 1. Mediastinal enlargement and an increase in the cardiothoracic index suggesting cardiomegaly seen on an anteroposterior chest X-ray.

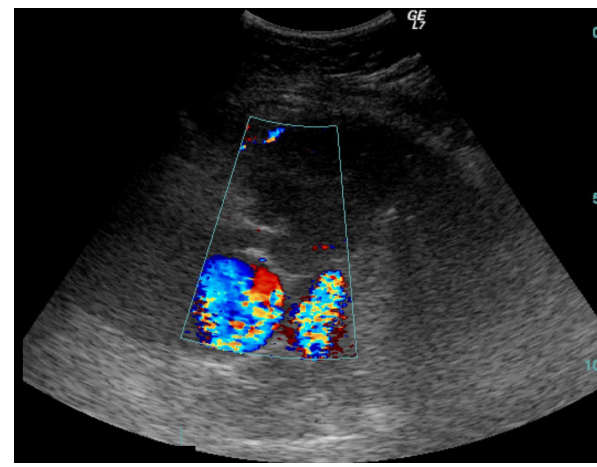

Figure 3. A hypoechoic mass lesion measuring 6x5 cm with irregular contours is seen on thoracic ultrasound in the anterior mediastinal region from the parasternal approach, as well as the color Doppler flow in the vascular structures adjacent to the lesion.

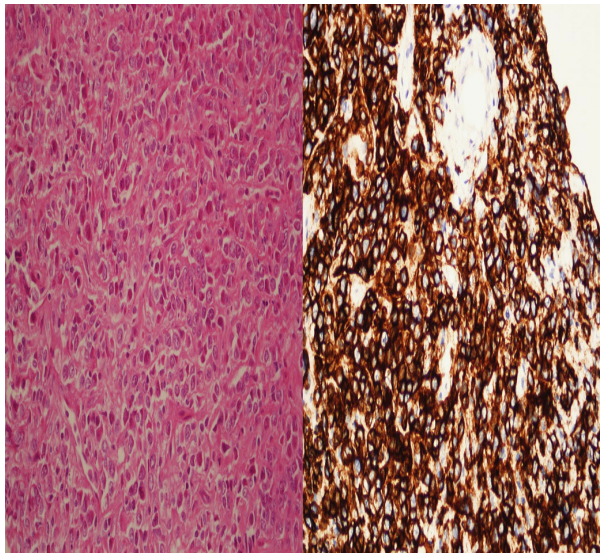

Figure 4. Moderate to large-sized lymphoid cell proliferation and CD20-positive lymphocytic cells (CD20 x400/H&E x400).

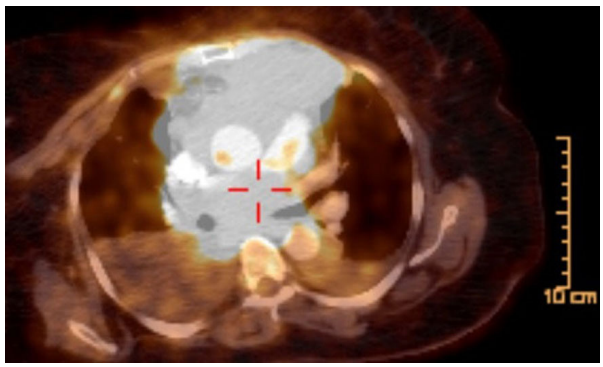

Figure 5. Diffuse, increased fluorodeoxyglucose uptake (SUD max 51.6) in conglomerated lymphadenopathies and a mass lesion with maximal diameter of 101x96x181 mm is seen. This mass lesion extends from the upper mediastinal region and occupies the superoinferior paratracheal region, surrounds the vascular structures, aortic arch, major brachiocephalic vascular structures, both pulmonary arteries, ascending aorta, and planes between the right atrium and right ventricle (invasion?). The mass lesion also completely encloses the vena cava superior and vena cava inferior the from medial aspect.

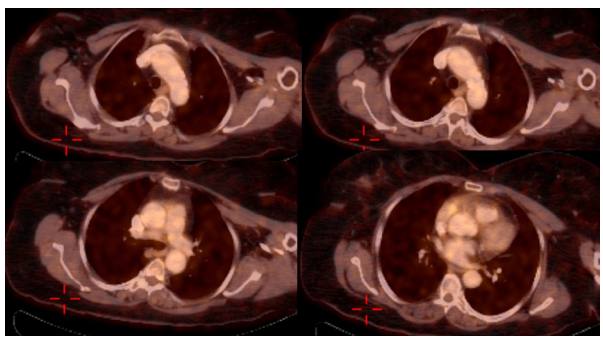

Figure 6. Abnormal focus of fluorodeoxyglucose uptake is observed in the parenchymal tissue of both lungs and the mediastinum.

pathological foci demonstrating F18-FDG uptake was not observed in the parenchymal tissue of the lungs or the mediastinum (Fig. 6).

DISCUSSION

Involvement of the heart in secondary malignancies is uncommon. The clinical signs and symptoms of secondary cardiac involvement are nonspecific. The consequences, however, can be serious. Therefore, early diagnosis is lifesaving. Our patient was examined due to the presence of exertional dyspnea and cardiomegaly seen on a chest X-ray. The patient was referred to our clinic with the suspicion of a mass lesion observed on an echocardiogram. Thoracic CT revealed the presence of a mass completely occupying mediastinum that also radiologically suggested invasion into the right ventricular wall. A pulmonologist performed a US-guided tru-cut biopsy, and 3 days later the patient was referred to the hematology polyclinic for treatment based on the histopathological diagnosis. All of the patient's complaints had regressed by the third week of treatment. This article was presented to emphasize the importance of rapid diagnosis and treatment using a multidisciplinary approach.

Secondary cardiac malignancies (metastases) are seen more frequently than primary cardiac malignancies. The most frequently seen primary benign cardiac tumor is myxoma, while the primary malignant cardiac tumor most often seen is angiosarcoma. Among metastatic malignant tumors of the heart, lymphomas rank third after lung and breast tumors. Primary tumors of the heart may involve the endocardium, myocardium, and epicardium, separately or in combination. Metastatic cardiac tumors are most often localized in the pericardium.[5]

Lymphoma rarely invades the heart, but when it does, serious cardiac complications can ensue. The symptoms and signs of cardiac involvement may be nonspecific. Early diagnosis is critical, and prompt treatment should be administered, as cardiac involvement has a poor prognosis, with a very high mortality and morbidity rate. Clinically, the patient may present with symptoms and signs of heart failure, pericardial effusion, hypotension, cardiac tamponade, tachycardia, or arrhythmia.[6] McDonnell et al.[7] detected 13 cases with cardiac involvement in 150 patients with lymphoma. Nine of the 13 patients had non-Hodgkin lymphoma. The authors noted that these cases with cardiac involvement in lymphoma had received an antemortem diagnosis due to nonspecific signs and symptoms. The authors attributed the frequent inability of clinicians to make an early diagnosis to factors such as the rarity of cardiac metastasis, the lack of clinical suspicion, the use of noninvasive methods with low sensitivity such as electrocardiography, and the difficulty of performing cardiac biopsy and demonstrating cardiac involvement with conventional imaging modalities.

Early diagnosis and treatment is required in cases of involvement of vital organs, such as the heart and the major vessels. The high sensitivity and specificity of echocardiography and magnetic resonance imaging, as well as multidetector CT, are helpful in making the diagnosis.[8,9] It is not always possible to differentiate between benign and malignant lesions based on a histopathological examination of specimens obtained through a fine-needle aspiration biopsy. The tissue sample must be of adequate size, and in addition to a classic histological examination, cytogenetic, immune analysis, and immunophenotypic examination should be also performed.[10] Diagnosis of lymphoma made using US-guided transbronchial fine-needle aspiration biopsy (TBFNAB) is now a controversial issue.[11] Since specimens obtained by endobronchial US-guided TBFNAB are not always appropriate for immunohistochemical and flow cytometric examinations, diagnosis of lymphoma subtypes can be difficult.[12] However, recent studies have demonstrated that diagnostic accuracy has significantly improved as a result rapid on-site pathological evaluation.[13]

Chemotherapy (CHOP) is the standard treatment for lymphoma. Particularly in cases with cardiac or major vessel invasion, bulky lymphadenopathy (>10 cm), or a mass lesion with a diameter greater than one-third the transverse diameter of the chest, radiotherapy is typically combined with standard therapy.[14]

Consistent with the literature, our case was detected using echocardiography and thoracic CT. Tru-cut biopsy was preferred because of presumptive cardiac involvement. Combined CT and radiotherapy was administered, given the presence of vital organ involvement, and the patient was followed up accordingly.

Informed Consent

Consent was obtained from the patient who participated in this study.

Peer-review

Internally peer-reviewed.

Authorship Contributions

Concept: C.D.; Design: T.S.G.; Data collection &/or processing: N.Ö.B.; Analysis and/or interpretation: S.Ş.C.; Literature search: T.S.G.; Writing: C.D.; Critical review: G.Y.

Conflict of Interest

None declared.

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Sağ Ventriküle İnvaze Mediastinal Kitle

Lenfomaların kalp tutulumu nadirdir ve çoğunlukla otopsi çalışmaları ile ortaya konulur. Buna karşın kalp tutulumunun son derece ciddi sonuçları vardır. Lenfomaların kalp tutulumu retrograd lenfatik, hematojen ve doğrudan komşuluk yoluyla direkt invazyon şeklinde olur. Direkt invazyon en sık görüleni ve en destrüktif bulgulara yol açanıdır. Klinik bulgu ve belirtileri nonspesifiktir. Bu yüzden erken tanı ve tedavi hayat kurtarır. Olgumuz efor dispnesi ve akciğer grafisinde görülen kardiyomegali nedeni ile uzun süre kardiyoloji polikliniğinde tetkik edilmiş, ekokardiografik incelemede mediastinal kitleden şüphelenilmesi üzerine kliniğimize refere edilmiştir. Çekilen toraksın bilgisayarlı tomografik incelemesinde mediasteni dolduran, sağ ventrikül duvarına radyolojik olarak invazyon düşündüren kitle saptanan hastaya aynı gün göğüs hastalıkları uzmanı tarafından toraks ultrasonografisi rehberliğinde trucut biyopsi yapılmış, biyopsiden üç gün sonra lenfoma patolojik tanısı ile beraber hasta tedavi için hematoloji polikliniğine yönlendirilmiştir. Tedavinin üçüncü haftasında ise hastanın tüm yakınmaları gerilemiştir. Bu makale kalp tutulumu şüphesi olan olgularda multidisipliner yaklaşımla hızlı tanı ve tedavinin önemine ve toraks ultrasonografi ile mediastinal kitle lezyona hızlı ve güvenli yapılabilen biyopsi işlemine dikkat çekmek için sunulmuştur.

Anahtar Sözcükler: Lenfoma; mediastinal kitle; ultrasonografi.