

Hodgkin's Lymphoma Presenting with Myocardial Metastasis and Heart Failure

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ABSTRACT

Although involvement of the heart by malignant lymphoma is relatively common, it is unusual to be detected premortem. We report a case of Hodgkin's lymphoma who presented with systemic signs and symptoms including abdominal distension, weakness, pallor, chills and fever, hepatosplenomegaly and generalized lymphadenopathy as well as signs of heart failure. Echocardiography revealed lucent myocardial lesions, pericardial effusion, and left ventricular hypertrophy. Right cervical lymph node and bone marrow biopsies established the diagnosis of nodular sclerosis Hodgkin's lymphoma with bone marrow involvement. After 14 periods of chemotherapy, systemic and cardiac abnormalities improved. To the best of our knowledge, Hodgkin's lymphoma primarily presenting with myocardial metastasis and heart failure is not previously reported.

Key words: Hodgkin's lymphoma, Nodular sclerosis, heart failure

Introduction

ardiac involvement in malignant lymphoma is one of the least investigated subjects in oncology (1). Cardiac metastases are found in 20-25% of patients with lymphoma (2, 3) and 9% of all metastatic cardiac tumors are due to lymphoma (3, 4). Several authors have described primary cardiac lymphomas presenting with pericardial effusion (5), arrhythmias and heart failure (6). Patterns of cardiac involvement vary by the types of lymphoma, suggesting that

different pathologic types of lymphoma may have different mechanisms of metastatis to the heart. Diffuse myocardial infiltration documented by echocardiography has rarely been described as a presenting feature of this condition (7, 8), but has been more commonly found at postmortem examinations (9). Primary presentation with heart failure in Hodgkin's lymphoma has not been reported before. We report a case of Hodgkin's lymphoma that presented with heart failure and had patchy myocardial involvement.

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Case Report

An 8-year-old girl from Afghanistan presented with a 2-month history of edema, abdominal distension, weakness, pallor, chills, fever, anorexia, and weight loss. Her past medical history was unremarkable. Physical examination showed severe pallor, generalized edema and abdominal distension. She also had non tender mobile lymph nodes in right neck (5x5cm), bilateral inguinal areas (0.5x0.5cm) and left axillary (0.7x0.7cm), marked hepatosplenomegaly and ascitis with shifting dullness. A systolic murmur (II/III) was heard; lungs were clear. Laboratory findings included severe anemia with moderate anisopoikilocytosis, hemoglobin 3.2 (NI: 12-16gr/dl), hematocrit 12 (N1:37-47%), MCV 105.3 (N1:81-90 FL), reticulocyte count 8.5% (NL:1-2%), iron 34 (N1:60-150mg/dl), TIBC 414 (N1:260-400 mg/dl), ESR 50 (N1<15 mm/hr) and positive C-reactive protein. Polymerase chain reaction (PCR) for tuberculosis, blood culture, urine culture, hydatid antibody, coomb's wright and 2ME, direct coomb's test, bone marrow culture and blood smear for malaria and borrelia were negative; G6PD level was normal.

Chest X-ray showed cardiomegaly; lungs were clear. CT scan of chest revealed multiple lymphadenopathy in paratracheal and subcarnial area. In abdominal sonography, marked hepatosplenomegaly (liver span 17cm) and two round hypoechoic areas in hepatic portal space due to adenopathy were seen. In abdominal CT scan (with and without contrast), severe hepatosplenomegaly, a hypodense area in liver, para aortic lymphadenopathy and dilated small bowel loops with thickened wall were reported.

Echocardiography revealed pericardial effusion with mild mitral regurgitation, mitral valve vegetations, left ventricular hypertrophy and multiple lucencies in myocardium ranging between 0.5 and 1 cm (Figure 1). On histologic examination of cervical lymph node (Figure 2) a mixture of histiocytes, eosinophils, plasma cells and few lymphocytes with scattered Hodgkin cells

and Reed-Sternberg cells within severely sclerotic stroma were seen. Bone marrow biopsy showed involvement by Hodgkin's lymphoma (Figure 3). The patient underwent medical treatment for bacterial endocarditis and took 14 courses of chemotherapy with diagnosis of nodular sclerosis Hodgkin's lymphoma, stage IV. Echocardiograpy after four weeks showed resolution of vegetations and mitral regurgitation. By the end of chemotherapy, all systemic, cardiac and laboratory abnormalities improved and myocardial lesions disappeared (Figure 4). She has had no evidence of recurrence in 3 year follow up.



Figures 1: Echocardiography, long axis: Increased thickness of left ventricle with myocardial lucencies and pericardial effusion.

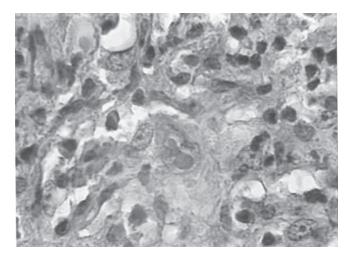


Figure 2: Histologic examination of lymph node showing mixture of inflammatory cells and a Reed Sternberg cell with prominent nucleoli (H&E x 400).

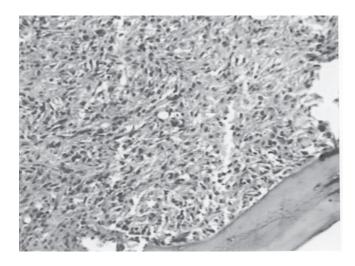


Figure 3:Histologic examination of bone marrow revealing involvement by lymphoma(H&E, x 250).

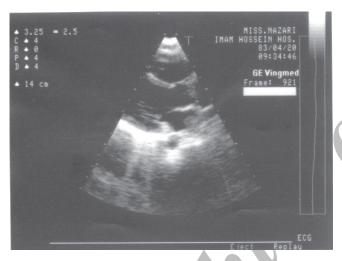


Figure 4: Echocardiography, long axis: Normal left ventricle with no lucencies, no effusion after treatment.

Discussion

Malignant lymphoma constitutes over 9% of the total metastatses by malignant neoplasms to the heart (10). Up to 20% of the patients who died from lymphomas have been found to have cardiac involvement (11). Although cardiac infiltration by lymphoma may sometimes be obvious, clinical sings and symptoms of cardiac metastasis of lymphoma may be typically absent or non-specific. In most cases cardiac involvement in lymphoma remains undetected prior to patients' death.

Echocardiography is a non-invasive, safe, relatively inexpensive and sensitive approach without known side effects for the diagnosis of cardiac involvement in lymphoma. There are three patterns of cardiac involvement in lymphoma: pericardial, epicardial-adventitial, and diffuse intrestitial-perivascular. Each of these patterns may be correlated with one of the three proposed mechanism by which lymphomas, in general, may spread to the heart: via direct extension, retrograde flow through cardiac lymphatic vessels, and hematogenous spread, respectively (12, 13).

Most authors believe that direct extension and / or retrograde lymphatic spread are that most common means by which carcinomas metastasize to the heart (14). In contrast, the hematogenous route has been proposed as the most common mechanism by which lymphomas involve the heart (15). The most common location of metastasis is pericardium (66.7%) followed by myocardium (50%) and coronary arteries (33.3%).

Echocardiographic findings in these patients showed that pericardial effusion is the most common abnormality of cardiac metastasis (1).

identification of metastatic cardiac involvement can benefit lymphoma patients through careful monitoring of these patients, and managing morbidity and minimizing mortality from this complication. Studies have shown that lymphoma patients with cardiac involvement could be treated successfully (16-18).

Although involvement of the heart by malignant lymphoma is relatively common, it is unusual to be detected premortem. The majority of reported cases of heart involvement by Hodgkin's lymphoma are post chemo and/or radiotherapy, mostly in nodular sclerosis type (3, 4, 9). We report an 8-year-old girl from Afghanistan with Hodgkin's lymphoma who presented with signs and symptoms of heart failure and had cardiac metastatic lesions. Primary presentation with myocardial metastasis and heart failure in Hodgkin's lymphoma has not been reported before.

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