

CASE REPORT

Swinging heart caused by diffuse large B-cell lymphoma

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Abstract

Pericardial disease is a common complication of solid tumors and occasionally seen in hematologic malignancies. Pericardial effusion, when it occurs, is usually caused by tumor seeding of the pericardium leading to a serous effusion or by mass effect from mediastinal lymphadenopathy blocking drainage of lymphatic ducts. Pericardial disease from non-Hodgkin's lymphoma is uncommon and malignant pericardial effusion is even rarer. Here we present a case of a 31-year-old male with diffuse large B-cell lymphoma who developed cardiac tamponade from a malignant pericardial effusion.

INTRODUCTION

Solid tumor metastasis is a leading cause of pericardial effusion [1]. Its presence is indicative of advanced malignancy and portends a poor prognosis [2, 3]. With large effusions (>500 mL), the heart can appear as swinging from the great vessels, like a pendulum, on echocardiogram [1]. Likewise, the swinging heart is a classic, but uncommon, echocardiographic finding of cardiac tamponade [1]. Diffuse large B cell lymphoma (DLBCL) is the most common of the non-Hodgkin's lymphomas (NHL), which typically presents as a rapidly enlarging mass in the neck, abdomen, or mediastinum [4]. Mediastinal NHL may produce a pericardial effusion, but rarely presents as cardiac tamponade [5]. Herein we report the first case, to our knowledge, of a patient with DLBCL who presented with cardiac tamponade.

CASE REPORT

A 31-year-old male presented to the emergency room for evaluation of progressive cough, dyspnea, pleuritic chest pain and orthopnea, 1 week after diagnosis of DLBCL. Imaging a week ago revealed diffuse lymphadenopathy involving his right neck, right axilla and a right supraclavicular/mediastinal mass encircling the right brachiocephalic vein with severe narrowing of the superior vena cava. Also present were bilateral pleural effusions and a moderate-to-large pericardial effusion. Vital signs were normal and physical exam remarkable only for lymphadenopathy at that time, thus no intervention was performed. The patient was discharged with plans to initiate outpatient treatment for his malignancy which he had not yet begun.

Currently, the patient presented with tachycardia, marginal blood pressure with pulsus paradoxus, muffled heart sounds,

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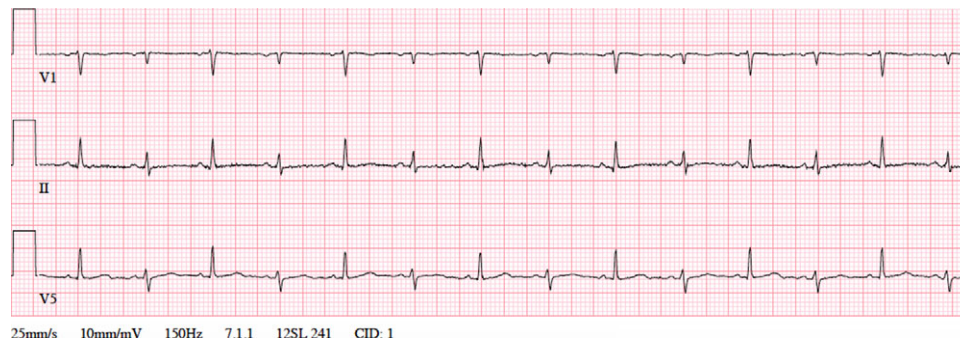


Figure 1: Electrical alternans, with changing QRS complex amplitude on a 12-lead electrocardiogram

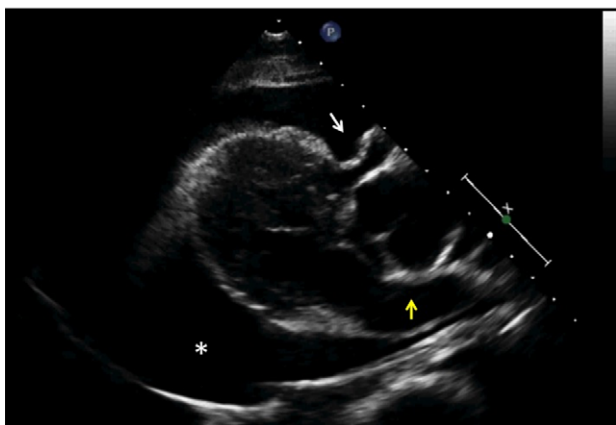


Figure 2: Parasternal long axis view of patient's transthoracic echocardiogram. The heart is seen surrounded by a very large pericardial effusion (asterisk), with features of tamponade including severe collapse of the right ventricle (white arrow) and the left atrium (yellow arrow)

and jugular venous distention, but absent facial and upper extremity edema. His white blood cell count at this time was 7900 cells/mL. Imaging revealed the serosal effusions had increased and the mediastinal mass was now encroaching the right atrium. A 12-lead electrocardiogram showed sinus tachycardia and low-voltage QRS with marked beat-to-beat variation (Fig. 1). A transthoracic echocardiogram confirmed the presence of a very large pericardial effusion (Fig. 2), with multiple features of tamponade including right atrial (RA) and right ventricular (RV) diastolic collapse, inferior vena cava (IVC) plethora, >25% respiratory variation of the mitral valve inflow, and a swinging heart, as shown in Video 1 of supplementary material.

Emergent pericardiocentesis drained 1100 mL of serosanguineous fluid containing 14 000 white blood cells/uL that were 100% lymphocytic. Thoracentesis drained 1100 mL of serous fluid containing reactive mesothelial cells without any significant leukocytosis. The pericardial effusion soon recurred requiring pericardial window placement. After two cycles of rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone (R-CHOP), the effusion was minimal. However, interval imaging after four cycles of chemotherapy demonstrated the pericardial effusion had again recurred. Echocardiogram demonstrated the effusion was borderline significant for tamponade, although clinically, the patient denied any symptoms. There was no metabolic activity within the fluid per positron emission tomography scan. Seven months after completing six cycles of R-CHOP the patient had only a small residual

mediastinal mass and no serosal effusions. He has been lost to follow-up since.

DISCUSSION

Malignant pericardial effusions most often occur in the setting of solid tumors [2, 5]. Hematologic malignant pericardial effusions may also occur, most often with Hodgkin's disease and less commonly with NHL, the incidence of which has been reported to be as low as 0.8% [2]. We presented here the first known reported case of malignant cardiac tamponade secondary to DLBCL.

Malignant pericardial effusion associated with NHL may represent direct malignant infiltration, effusive-constrictive pericarditis secondary to radiation therapy, invasion of the pericardial space with opportunistic pathogens or an inflammatory effusion secondary to chemotherapy [2, 5], as we suspect was the case in our patient after his third effusion recurrence. In two-thirds of cases, pericardial effusion is a benign complication, and not an extension, of malignancy [6]. A lymphocyte-rich effusion may represent a reactive lymphocytosis indistinguishable from malignancy, but this equivocal yield can be overcome by using morphometry with 85% sensitivity and 95% specificity [2]. Combining cytology with flow cytometry may increase diagnostic yield to 100% sensitivity and 94% specificity [2].

Patients with cardiac tamponade may present with Beck's triad of distant heart sounds, distended neck veins, and hypotension, but combined these signs are only 50% sensitive for tamponade [7]. Pulsus paradoxus, low-voltage QRS, and electrical alternans are also commonly associated with tamponade, yet the individual sensitivities for each are only 82, 56 and 42%, respectively [8]. The gold standard for diagnosing cardiac tamponade remains right heart catheterization. Echocardiography may also be used and findings such as RA systolic collapse, early RV diastolic collapse, or IVC plethora are all at least 90% sensitive for tamponade [5, 8, 9], all of which were present in our patient.

Cardiac tamponade is a life-threatening emergency with an estimated 85% mortality, requiring immediate pericardiocentesis [9]. Malignant pericardial effusions recur frequently and guidelines for treating recurrence are lacking. Surgical pericardial window has long been a standard therapy but requires general anesthesia, which many advanced cancer patients may not tolerate. For those too sick for surgery, treatment options include percutaneous balloon pericardiectomy, subxiphoid pericardiectomy and pericardial sclerosis, all with various success rates of >90% preventing recurrence [5, 9]. Finally, treatment of

pericardial effusion with either systemic chemotherapy or localized radiation has also been attempted, but success is highly dependent on the sensitivity of the underlying malignancy to the treatment modality [4]. The standard therapy for DLBCL is R-CHOP, with excellent 5- and 10-year overall survival rates [4]. Our patient, like the majority of patients with neoplastic pericardial effusion, presented a positive pericardial fluid study for malignancy, which is associated with a worse prognosis [3, 10]. However, after six cycles of R-CHOP the patient demonstrated near-total resolution of his lymphadenopathy, with no pericardial effusion.

CONCLUSION

Malignant cardiac tamponade is a rare complication of NHL, but when present, represents a life-threatening emergency. The many classic signs and symptoms of tamponade are insensitive for diagnosis; thus, clinicians should have a heightened vigilance of any malignancy involving the mediastinum. Any pericardial effusion significant for tamponade requires immediate drainage, yet preventing recurrence is problematic. Many potential options for preventing recurrence exist with variable success rates, but the only potential preventive treatment is to address the underlying malignancy.

SUPPLEMENTARY MATERIAL

Supplementary material is available at *Oxford Medical Case Reports* online.

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CONFLICT OF INTEREST STATEMENT

The authors of this publication have no financial or nonfinancial relationships to disclose.

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ETHICAL APPROVAL

Patient has been lost to follow-up. This report has omitted any potential identifier.

GUARANTOR

Neal Olarte, Raúl A. Montañez-Valverde.

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