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Cardiac tamponade as the initial presentation of Hodgkin's lymphoma in a young female

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ABSTRACT

Introduction: Hodgkin's lymphoma can affect the heart both as primary and as a metastatic condition. Pericardial compromise is a common finding over the course of the disease, and the manifestations can range from pericarditis to pericardial tamponade. Pericardial tamponade is a life-threatening condition that requires immediate treatment, and is rarely the initial presentation of Hodgkin's lymphoma. We present a case of a young female with Hodgkin's lymphoma initially presenting as acute pericardial tamponade.

Case Report: A 20-year-old female presented with acute shortness of breath. At presentation she was hypotensive, tachycardic, and tachypneic. Chest X-ray showed an enlarged mediastinum, without enlarged cardiac silhouette. Bedside transthoracic echocardiography demonstrated massive pericardial effusion with cardiac tamponade. The patient was in critical condition, and a pericardial window was performed. After the procedure she was hemodynamically stable. Positron emission tomography scan and computed tomography evidenced a ganglionar conglomerate in the mediastinum and supraclavicular area. An excisional biopsy of a neck lymph node confirmed classical Hodgkin's lymphoma. She was started with standard chemotherapy induction. At the time of the report the patient is doing well with favorable response to the treatment, and without any clinical evidence of systemic or cardiac recurrence.

Conclusion: Pericardial involvement is frequent over the course of Hodgkin lymphoma. Pericardial tamponade is rarely the initial presentation of Hodgkin's lymphoma. A team approach is necessary for favorable outcome. Treatment consists of stabilizing the patient by relieving the tamponade, followed by systemic chemotherapy treatment for the underlying malignancy and to prevent recurrence.

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Introduction: Hodgkin's lymphoma can affect the heart both as primary and as a metastatic condition. Pericardial compromise is a common finding over the course of the disease, and the manifestations can range from pericarditis to pericardial tamponade. Pericardial tamponade is a life-threatening condition that requires immediate treatment, and is rarely the initial presentation of Hodgkin's lymphoma. We present a case of a young female with Hodgkin's lymphoma initially presenting as acute pericardial tamponade. Case Report: A 20-year-old female presented with acute shortness of breath. At presentation she was hypotensive, tachycardic, and tachypneic. Chest X-ray showed an enlarged mediastinum, without enlarged cardiac silhouette. Bedside transthoracic echocardiography demonstrated massive pericardial effusion with cardiac tamponade. The patient was in critical condition, and a pericardial window was performed. After the procedure she was hemodynamically stable. Positron emission tomography scan and computed tomography evidenced a ganglionar conglomerate in the mediastinum and supraclavicular area. An excisional biopsy of a neck lymph node confirmed classical Hodgkin's lymphoma. She was started with standard chemotherapy induction. At the time of the report the patient is doing well with favorable response to the treatment, and without any clinical evidence of systemic or cardiac recurrence. Conclusion: Pericardial involvement is frequent over the course of Hodgkin's lymphoma. Pericardial tamponade is rarely the initial presentation of Hodgkin's lymphoma. A team approach is necessary for favorable outcome. Treatment consists of stabilizing the patient by relieving the tamponade, followed by systemic chemotherapy treatment for the underlying malignancy and to prevent recurrence.

Keywords: Cardiac tamponade, Hodgkin's lymphoma, Malignant pericardial effusion, Pericardial window

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INTRODUCTION

Hodgkin's lymphoma (HL) is a hematologic malignancy that arises from germinal center or post-germinal center B cells. It accounts for approximately 10% of all lymphomas and approximately 0.6% of all cancers diagnosed in the developed world annually

[1]. Commonly, the initial clinical presentation is with painless localized peripheral lymphadenopathy, typically involving the cervical region. Over the course of disease Hodgkin's lymphoma can have different types of pericardial involvement. Manifestations can range from an asymptomatic mild effusion to acute pericardial tamponade [2, 3]. Pericardial tamponade is an acute condition that requires immediate treatment; and is rarely the initial presentation of Hodgkin's lymphoma [4–7]. Herein, we present a case of a young female with Hodgkin's lymphoma initially presenting as acute pericardial tamponade.

CASE REPORT

A previously healthy 20-year-old female presented to the emergency department with progressive shortness of breath. On clinical examination she was conscious, had fatigue, cough, swelling of the neck and evident respiratory distress. At presentation she was hypotensive (blood pressure 90/60 mmHg), tachycardic (pulse 125 beats/minute), and tachypneic (respiratory rate 27 respirations/minute), and temperature was 37°C. Electrocardiogram showed normal sinus rhythm with sinus tachycardia. Chest X-ray (Figure 1) showed an enlarged mediastinum, encompassing 60% of the internal transverse diameter of the thorax, without enlarged cardiac silhouette. Bedside transthoracic echocardiography demonstrated massive pericardial effusion with cardiac tamponade. The patient was in critical condition and was shifted emergently to the operating room where she was evaluated by cardiovascular surgery. A pericardial window was performed via subxiphoid incision. The pericardial sac was tense and the pericardium was opened anterior to the phrenic nerve. About 400 mL of bloody fluid was gradually suctioned from the pericardial cavity, and 300 mL from the right pleural space. The pericardium had normal appearance and no cardiac injuries could be identified. The pericardium was inspected and a large swelling was found in the middle mediastinum compressing the heart, the fluid was drained, fluid studies and a biopsy were performed and the bleeding was controlled by applying an absorbable hemostat. Pericardiopleural window was created posterior to the phrenic nerve and the chest was closed after inserting a thoracic drain. After the procedure she was hemodynamically stable. Chest computed-tomography scan evidenced a great ganglionar conglomerate in the anterior and medium mediastinum with a mediastinal mass of 150x100x130 mm with secondary stenosis of the superior vena cava. The cardiac biopsy and the fluid studies were negative for malignancy.

An excisional biopsy of an easily accessible neck lymph node was taken and sent for pathology and immunologic studies. The biopsy confirmed a lymphoproliferative disease consistent with classical Hodgkin's lymphoma of the nodular sclerosing subtype. Cytology evidenced the presence of large mononuclear cells and the classical Reed-Sternberg cells which were CD 30+. A staging positron emission tomography/computer tomography (PET/CT) scan (Figure 2) was performed and the patient was staged as a Hodgkin's lymphoma, stage III-2 (Both sides of diaphragm), A (No B symptoms), X (presence of bulky disease larger than 10 cm) according to Ann Arbor classification. She had compromise of retroperitoneal lymph nodes but without hepatic, splenic, or bone marrow compromise. The patient had three negative prognostic factors based on the International Prognostic Score (IPS); with a calculated 60% freedom from progression and 78% overall survival at five years. She was started with standard induction chemotherapy with adryamicin, bleomycin, vinblastine, and dacarbazine (ABVD). At the time of the report the patient has received the six cycles of chemotherapy and is doing well with favorable response

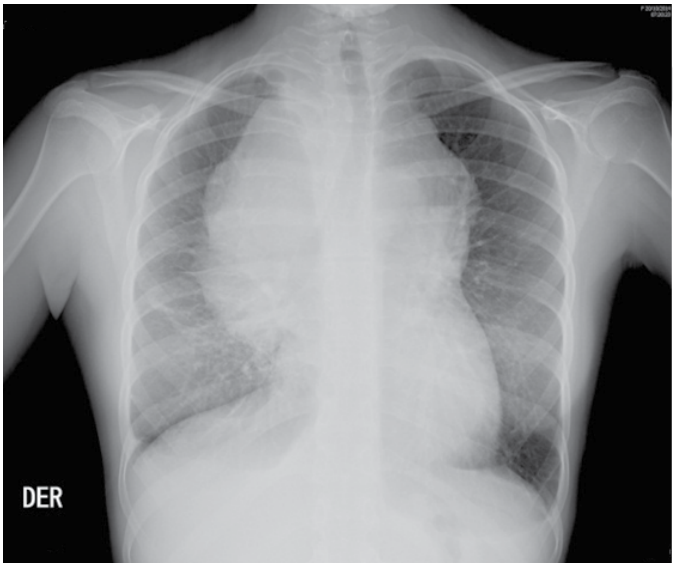

Figure 1: Chest X-ray with widened mediastinum at the initial presentation.

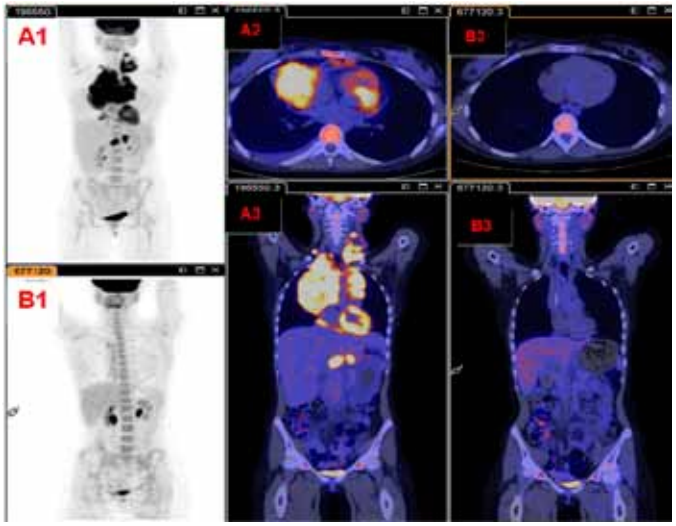

Figure 2: Comparison of initial staging PET/CT (Panels A1, A2, A3), scan showing non-FDG-avid pericardial effusion and the mediastinal mass, with a final PET/CT after six cycles of chemotherapy (Panels B1, B2, B3).

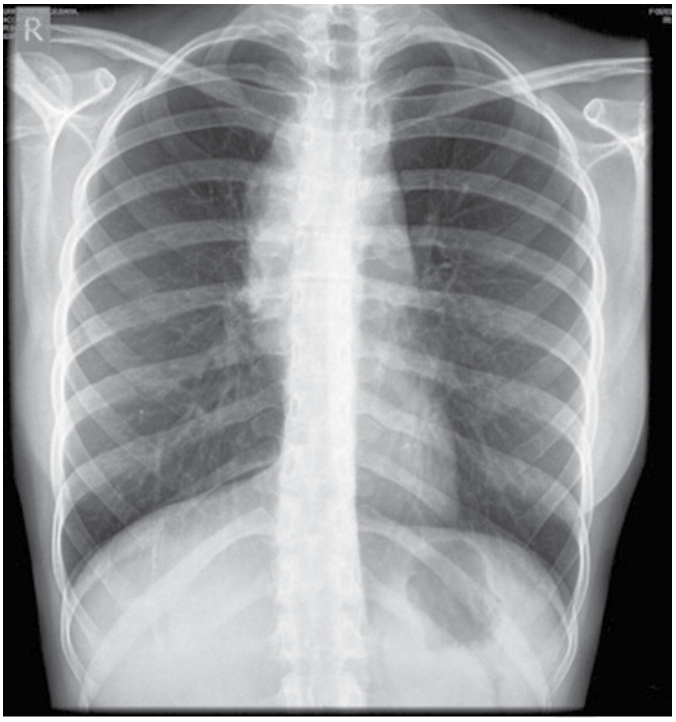

Figure 3: Chest X-ray after three cycles of chemotherapy.

to the treatment (Figures 2 and 3), and without any clinical or imagenological evidence of systemic or cardiac recurrence.

DISCUSSION

Hodgkin's lymphoma is a hematological malignancy with an incidence of 2–4 per 100,000 per year [1]. The most common presentation consists of a painless mass in >70% of the cases. Pericardial involvement is a common finding over the course of the disease, and it can present either as a primary or as a metastatic condition. Pericardial compromise can be asymptomatic or present with manifestations that range from pericarditis to pericardial tamponade [5, 7]. The clinical presentation of pericardial effusions depends upon whether the accumulation is acute or subacute.

Acute pericardial tamponade is a life-threatening condition that requires immediate diagnosis and treatment and is rarely the first presenting sign of Hodgkin's lymphoma [6, 8, 9]. The clinical presentation of acute pericardial tamponade includes: sudden dyspnea, chest pain, tachycardia, softening of heart sounds and echocardiographic signs of right heart compromise secondary to the acute rise in the pericardial pressure limiting the diastolic filling and leading to hemodynamic instability.

The diagnostic approach of pericardial tamponade includes a combination of electrocardiography, radiologic studies, and diagnostic/therapeutic pericardiocentesis. Chest X-ray can show an enlarged cardiac silhouette with clear lung fields. Echocardiography is the primary imaging tool to establish the diagnosis and quantify the amount of pericardial effusion and the hemodynamic impact on the patient. Imaging with computed tomography scan and positron emission tomography scan can provide additional information and can be useful in the evaluation of patients. Pericardial biopsy and examination of the pericardial fluid should be done to perform cytology studies and biomarkers and to rule out other diagnosis [2, 10].

Hodgkin's lymphoma has a good prognosis when treated promptly with chemotherapy. In patients presenting with acute symptoms of pericardial tamponade the mainstay of treatment is the removal of the fluid to obtain normal hemodynamic status and stabilize the patient by relieving the tamponade. A team approach including cardiologist, surgeon, anesthetist, radiologist and oncologist is necessary for favorable outcome. Following the stabilization of the hemodynamic parameters, staging should be completed and induction chemotherapy treatment initiated as soon as possible to treat the underlying disease and prevent reaccumulation of pericardial fluid. Our patient received appropriate acute management of the tamponade and received induction chemotherapy without recurrence of the pericardial fluid.

CONCLUSION

Pericardial involvement is a frequent finding in Hodgkin's lymphoma but is rarely the initial presentation. Given its rarity it is hard to give a specific recommendations regarding diagnosis, but clinical suspicion should remain high in an appropriate clinical context. Diagnosis and treatment should be directed depending on the presentation; with a team approach necessary for favorable outcomes. This atypical presentation should be suspected and promptly confirmed with imaging. Initial treatment consists of stabilizing the patient by relieving the tamponade, followed by systemic induction chemotherapy treatment for the underlying malignancy and to prevent fluid reaccumulation.

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Author Contributions

Miguel Gonzalez – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Amado Karduss-Urueta – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Laura Gutiérrez – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Juan Alejo Jimenez – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Rosendo Perez – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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