



Case Report

Chylopericardium with Chylothorax as an Unusual Presentation of Hodgkin Lymphoma

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Abstract: *Introduction:* Chylous effusions are rare and their etiologies are multiple and diverse. Chylothorax is the most common; however, chylopericardium is exceptional. The aim of this work is to report the epidemiological, diagnostic and therapeutic aspects of Hodgkin lymphoma revealed by chylopericardium and bilateral chylothorax with review of the literature. *Case presentation:* It's about a 34 years old man, with no particular pathological history, admitted in emergency, with a cardiac tamponade associated with a bilateral pleural effusion. The patient underwent a subxiphoid pericardiostomy and pleural drainage, removing milky pericardial (800cc) and pleural (1850cc) fluid. Fluid analysis confirmed a chylous effusion with no malignant cells. The patient was started on a low dietary fat diet which allowed a decrease in the chyle output after 10 days. A mediastinal mass biopsy was performed, through an anterior right mediastinotomy, revealing a Hodgkin lymphoma. The patient was referred to an Oncology center and underwent a full course of chemotherapy. The patient's response to chemotherapy was favorable with a good clinical outcome and complete regression of the superior vena cava syndrome, the edema and the respiratory distress. *Conclusion:* It's important to keep in mind some uncommon presentations of mediastinal tumors in general, and specifically Hodgkin Lymphoma; as well as the role of the 3 cornerstones (low-fat diet, surgery, and treatment of the underlying cause) in the management of this condition.

Keywords: Chylopericardium, Chylothorax, Lymphoma, Mediastinal Tumor

1. Introduction

Chylopericardium is a rare entity defined by the accumulation of the chyle in the pericardial cavity. Diagnosis always requires pericardiocentesis allowing the macroscopic study of the fluid (as a white, milky liquid), triglyceride content, cytologic examination and negative culture. The causes include trauma (e. g. surgery), malignant diseases of

lymphatic system, vein thromboses... or idiopathic [1, 2].

A chylothorax is the accumulation of the chyle in the pleural cavity. It can be idiopathic or caused by various conditions, traumatic and non-traumatic. There are two main categories of traumatic causes, iatrogenic/surgical and the other being non-iatrogenic [3].

Non-traumatic causes mainly include malignant tumors (lymphomas), in 70% of cases, but also congenital disorders [3, 4].

In the largest study about chylothoraces, only 1.4% had Hodgkin lymphoma, which makes it a rare cause of pleural chylous fluid effusion [5].

This case report describes a case of a 34-year-old (yo) man with chylopericardium presenting as a cardiac tamponade and bilateral chylothorax, which revealed a Hodgkin lymphoma. The aim of this study is to help practitioners think of Hodgkin lymphoma when they face similar presentations.

2. Case Presentation

A 34yo man was admitted for a 5-months history of upper body swelling and face puffiness associated with thoracic pain, shortness of breath gradually worsening and dysphonia associated with nocturnal fever, anorexia and loss of body weight and condition. He had undergone, prior to admission in our department, many right and left thoracenteses; then a pleural drainage for a massive right pleural effusion that removed a milky pleural fluid. In view of further investigation and management he was referred to our department.

Admission examinations have revealed a 90% saturation at room air, a mediastinal syndrome with dysphonia and superior vena cava (SVC) syndrome, diminished heart sounds and diminished breath sounds in lung fields bilaterally with lower-extremities pitting edema. The patient was hemodynamically stable. (Figure 1).



Figure 1. Photo of our Patient Showing a Superior Vena Cava Syndrome and Pitting Lower-Extremity Edema.

Chest x-ray showed a right chest tube, a left pleural effusion and a cardiothoracic ratio (CTR) of 0.67. On the chest computed tomography (CT) scan the SVC, the innominate vein and the arch of the azygos vein were not opacified and there was a thrombosis of the mediastinal segments of the subclavian vein. The subcarinal part of the Azygos vein was dilated with a development of collateral veins; it also revealed an encysted right pleural effusion, and an important free left pleural effusion with bilateral pulmonary atelectasis, and a circumferential moderate pericardial effusion.

The echocardiography found a cardiac tamponade pulmonary arterial hypertension. CT scan of the abdomen and pelvis revealed hepatomegaly and splenomegaly without lymphadenopathy. Laboratory tests summarized in (Table 1) showed a thrombocytosis and a lymphopenia with a positive hepatitis B surface antigen. Fluid cultures were negative.

Table 1. Laboratory Tests Processed Before Surgery.

Laboratory tests	Results
White blood cells (/mm ³)	6850
Neutrophils (/mm ³)	4590
Lymphocytes (/mm ³)	940
Monocytes (/mm ³)	770
Basophils (/mm ³)	100
Eosinophils (/mm ³)	450
Hemoglobin (g/dL)	13.2
Platelets count (/mm ³)	1.045.000
Blood type	O+
GOT (UI/L)	47.08
GPT (UI/L)	25.94
Total serum protein (g/L)	59.34
AgHbs	Positive

Given the urgency (Cardiac Tamponade + large pleural effusion), the patient underwent a subxiphoid pericardiostomy and a left pleural drainage, removing milky pericardial (800cc) and pleural (1850cc) fluid (Figure 2 and Figure 3).



Figure 2. Pericardial and Pleural Fluid Extracted During the Surgery and the Pericardial Biopsy.

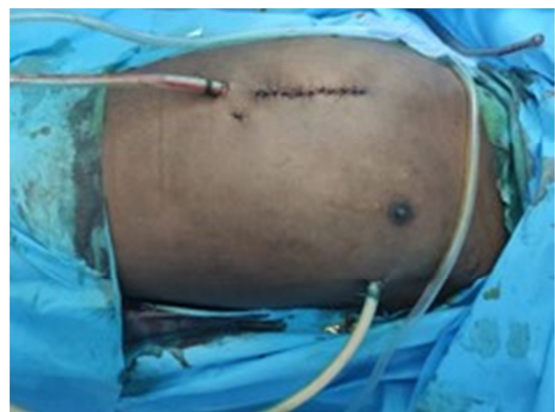


Figure 3. Immediate Post-Operative Photo Showing Pericardial and Pleural Drains Placement.

Fluid analysis confirmed a chylous effusion with no malignant cells (Table 2). The pericardial biopsy was inconclusive. The patient was started on a low dietary fat diet which allowed a decrease in the chyle output after 10 days.

Table 2. Pericardial and Pleural Fluid Analyses.

Pericardial and Pleural fluid analyses	Results
Macroscopic aspect (Fluid color)	milky
Biochemistry Analysis	
Triglycerides (g/L)	6.95 g/L Or 7.92 mmol/L
Protein rate (g/L)	32.11 g/L
Cytologic Analysis	
White blood cells (/mm ³)	247
Neutrophils	5%
Eosinophils	95%
Red blood cells (/mm ³)	1
Malignant cells	Absent
Bacterial detection	
Culture	Negative
Mycobacterium tuberculosis bovis	Negative

Given the worsening of the respiratory distress despite the drainage and oxygen therapy, the patient undertook a new Chest CT-scan (Figure 3) revealing bilateral pleural effusion, a non-tension left pneumothorax and an anterior mediastinal mass with multiple lymphadenopathies previously hidden by the massive effusions. An anterior pleural drainage removed the pneumothorax and a mediastinal mass biopsy was performed through an anterior right mediastinotomy. Thereafter, an intravenous corticotherapy has been started (Methylprednisolone) with an adjuvant treatment.

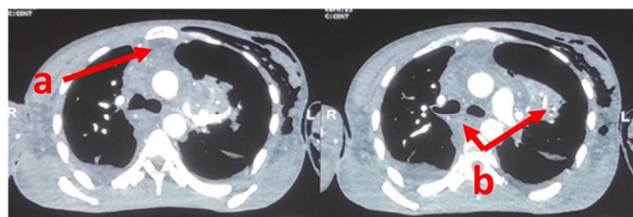


Figure 4. Chest CT-Scan After Pericardial and left Pleural Drainage. (a) Anterior Mediastinal mass, (b) Mediastinal Adenopathies.

Pathology of the mediastinal mass revealed a nodular sclerosing Hodgkin's lymphoma.

The patient was referred to an Oncology center and underwent a full course of chemotherapy (8 cycles of 15days). Hydrocortisone was given the 1st day, “Adriablastine + Bléomycine + Vinblastine + Dacarbazine + Hyperhydratation” on the 1st and 15th day and Allopurinol every day.

The patient's response to chemotherapy was favorable with a good clinical outcome and complete regression of the SVC syndrome, the edema and the respiratory distress. The oncologist transferred those medical records.

3. Discussion

The Chylopericardium is a rare entity. The etiology includes thoracic surgery, mediastinal neoplasm (like lymphomas), thrombosis of the left subclavian vein and more [1, 5, 6].

Moreover, chylothoraces are mainly caused by lymphoma, bronchogenic carcinoma and trauma (including surgery) [5].

Clinical presentation of Hodgkin lymphoma as a cardiac tamponade with bilateral chylothorax is very rare, and to the

best of our knowledge, it has been reported only once before by John O'Donnel and al. in 2019 [7]. Sanjeev Kumar Verma and al. also reported a case of a nodular sclerosis Hodgkin lymphoma revealed by a chylothorax [8].

The age of our patient was 34 years, which is consistent with the literature. Chylopericardium can occur in people across various age ranges, including adults between 18 and 70 years old, and affecting both males and females equally [9].

Treatment of chylopericardium and chylothorax relies mostly on 3 cornerstones: treatment of underlying cause, low fat diet and surgery if required [10, 11].

Our patient required the 3 of them. He underwent the first surgery that relieved the life-threatening condition and allowed the diagnosis of chylous pericardial and pleural effusion. Then, a second surgery was carried out allowing the diagnosis of “Hodgkin Lymphoma”. It should be noted here that the patient had two conditions contributing to the chylous effusion. Firstly, the compression of the thoracic duct by the mediastinal mass and lymphadenopathies, and secondly, the left subclavian vein thrombosis obstructing the venous drainage of the thoracic duct. Low fat diet has allowed a decrease in the chylous production flow and the patient was referred to a specialized department to start a Hodgkin lymphoma management.

Multiple studies have affirmed that chemotherapy is the primary treatment option for chylothorax in patients with Hodgkin lymphoma [12, 13].

Despite the fact that there is no standardised treatment, some authors have proposed an algorithm for interventional lymphatic treatment guided by Dynamic contrast-enhanced magnetic resonance lymphangiography to treat non-traumatic chylothorax and chylopericardium. This new approach is producing promising results [14-16].

4. Conclusion

This case report showed that Hodgkin Lymphoma could be revealed by chylothorax with chylopericardium presenting as a tamponade. After dealing with the surgical emergency, the overall management of our patient's condition was achieved based on 3 cornerstones: low fat diet, surgery and treatment of the cause.

Patient Consent

The patient gave his consent to use his data.

Conflict of Interest Statement

The authors declare that they have no competing interests.

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