

Tamponade with recurrent chylopericardium in patient with non-Hodgkin's lymphoma: chemotherapy is the key management after pericardiocentesis

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SUMMARY

Cardiac tamponade, the accumulation of fluid in the pericardial space, leads to impaired venous return, loss of left ventricular preload and haemodynamic collapse. **Chylopericardium** is an unusual cause of the pericardial effusion. This is often secondary to malignancy. Non-Hodgkin's Lymphoma is a primary malignancy from the lymph node. It can be produced by B lymphocytes, T lymphocytes or natural killer cells. The term chylopericardium refers to a pericardial effusion containing milky fluid within the intrapericardial space. We present a case of a **42-year-old** male patient who came with dyspnoea as a result of cardiac tamponade caused by a **massive milky pericardial effusion** (chylopericardium) secondary to mediastinal non-Hodgkin's lymphoma.

BACKGROUND

Pericardial effusions can occur due to malignancy, inflammation, infection, connective tissue disorders, uremia, pleural effusion with extension or trauma, or they can occur spontaneously.^{1,2} Chylopericardium is a **rare condition** in which fluid-containing chylomicrons accumulates within the pericardial space.^{1,3} The accumulation of chyle can also cause tamponade and cardiac tamponade must be treated promptly to avoid fatality.^{4,5} Though concurrent accumulation of chyle in all serous cavities is uncommon, it can be associated with non-traumatic causes. By far, malignancy (particularly, lymphoma) is frequently being the underlying cause. The management strategy consists of pericardiocentesis in massive pericardial effusion, a strict dietary fat restriction, surgical intervention if indicated, octreotide therapy (recommendation class IIb) and treating the underlying cause.^{1,6,7}

CASE PRESENTATION

A man in his 40s presented to the emergency department with progressive dyspnoea for months, worsening dyspnoea and shortness of breath for the last 3 days. He denied any other associated symptoms such as chest pain, fever, nausea or vomiting.

His medical history was significant for non-Hodgkin's lymphoma, discovered a few months ago. He was found to have **B-cells non-Hodgkin's lymphoma**. The mass was biopsied and revealed **anaplastic variant diffuse large B cell lymphoma**. He was treated with **R-EPOCH chemotherapy**, which

includes etoposide, epirubicin, vincristine, rituximab and cyclophosphamide, during the second cycle. The patient had previously been hospitalised with a chylothorax diagnosis and had pleural fluid evacuated via thoracentesis. Thoracosentesis with pigtail catheter placement was done and there was 8 L of pleural fluid drained in 4 days. The analysis of pleural fluid showed triglycerides raise of **755 mg/dL**, which was consistent with chylothorax. At that time, the patient continued chemotherapy and discharged with stable condition.

Then a month later, he came with dyspnoea in the emergency department. Pulsus paradoxus and jugular venous distention were noted on physical examination. **Cardiovascular examination** revealed muffled heart sounds that were not accompanied by murmurs and there was no evidence of pericardial friction rub. His lungs were examined, and it was discovered that he had slight diminished breath sounds in his right lower lobe. Examinations of the abdomen were unremarkable.

INVESTIGATIONS

Routine blood investigations and coagulation profiles were within normal limits. Chest radiograph was suggestive of cardiomegaly (**figure 1**). Electrocardiography showed low-voltage complexes (**figure 2**). An echocardiogram revealed extensive anterior, posterior, left lateral and right lateral pericardial effusion, with evidence of cardiac tamponade those were **swinging of the heart**, the early diastolic collapse of the right ventricle, the late diastolic collapse of the right atrium and exaggerated **respiratory variability (>25%)** in mitral inflow velocity (**video 1** and **figure 3**).

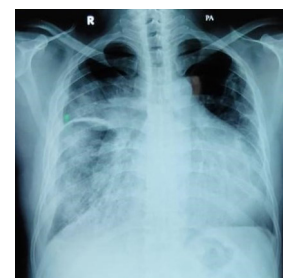


Figure 1 Chest radiograph demonstrated an apparent enlargement of cardiac silhouette.



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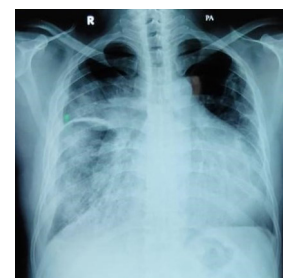


Figure 1 Chest radiograph demonstrated an apparent enlargement of cardiac silhouette.



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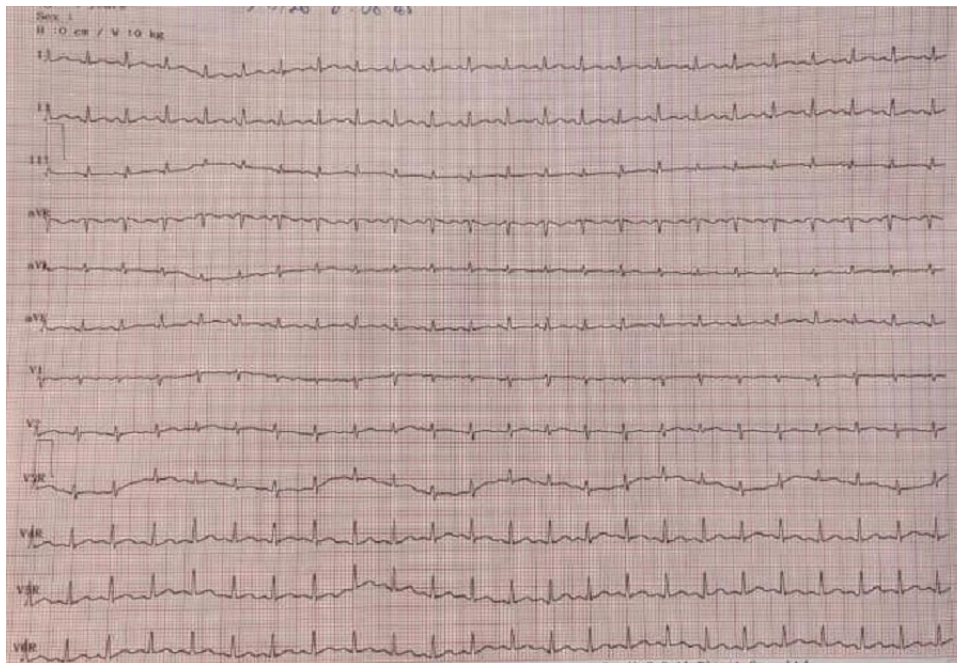


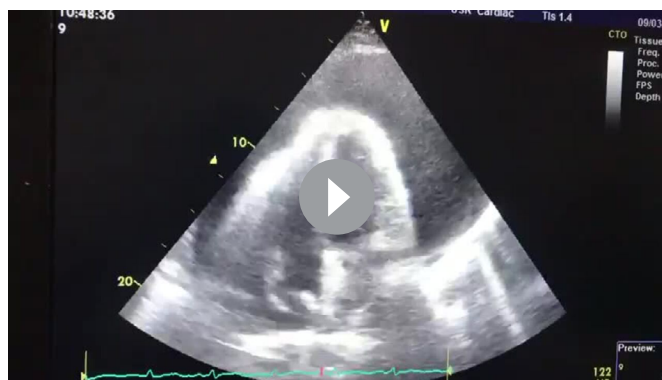
Figure 2 ECG showed sinus tachycardia 150 bpm with low voltage.

DIFFERENTIAL DIAGNOSIS

Chylopericardium, cholesterol pericarditis or purulent pericardial effusion.

TREATMENT

The patient underwent **pericardiocentesis**, and 1L of milky fluid was removed (**figure 4**). Pericardial fluid differential showed a total white cell count of $0.513 \times 10^9/L$, which contained monocytes of 87.4% and polymorphonuclear cells of 12.6%. Analysis of pericardial fluid showed protein 3.9 mg/dL, glucose 221 mg/dL, cholesterol 79 mg/dL, LDL cholesterol 15 mg/dL and also elevated triglycerides of 2678 mg/dL consistent with chylopericardium. A **pericardial drain** was left in place for 15 days, and it drained a total of **4L of chylous fluid**. Then **bleomycin** intra-pericardial was given with the expectation of a decrease in the production of pericardial effusion to less than 25 mL/day. But the production of pericardial effusion was **still about 100 mL/day**. After discussing with **haematology consultant**, we decided to continue the R-EPOCH chemotherapy.



Video 1 Swinging of the heart (cardiac tamponade)

OUTCOME AND FOLLOW-UP

Following the **fourth cycle of chemotherapy**, the production of pericardial effusion was diminished to less than 25 mL/day. We followed up on the patient's condition, and he reported decreased dyspnoea and improvement in his shortness of breath. He was discharged in good health. Prior to discharge, the pigtail catheter was removed. Echocardiography was done prior to discharge, it showed only minimal pericardial effusion in basal and no pericardial effusion in lateral (**video 2** and **figure 5**). Following completion of chemotherapy, the patient experienced significant symptomatic improvement.

DISCUSSION

Chylopericardium is a pericardial effusion composed of chyle, the natural fluid that fills lymphatic vessels. Chylopericardium can be caused by various factors, including trauma, surgery (particularly for congenital heart disease), congenital lymphangiomas, radiotherapy, subclavian vein thrombosis, infection (eg, tuberculosis), mediastinal neoplasms and acute pancreatitis.^{1,3,6} Chylopericardium is frequently associated with chylothorax. In a systematic PubMed and Wangfang database search for English and Chinese studies reporting **idiopathic chylopericardium**

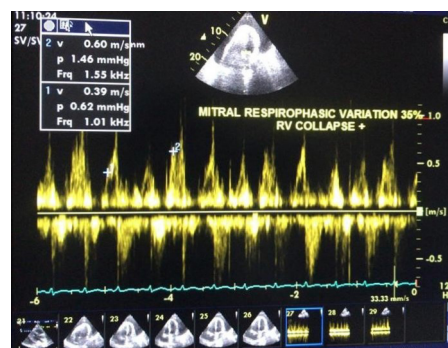


Figure 3 Mitral respiratory variation 35% (right ventricle collapse).



Figure 4 Pericardial milky fluid.

during the period of 1950–2015, 104 cases were found and less than 50% came with cardiac tamponades.^{8–10} Any disruption or dysfunction of the flow of chyle through the thoracic duct can cause chylothorax and also chylopericardium. The component of chyle causes its distinctive colour of effusion. Mediastinal non-Hodgkin's lymphoma is the leading cause (11%–37%) of non-traumatic chylothorax and chylopericardium.^{11 12} In this case, the patient had previously experienced chylothorax and had pleural fluid evacuated.^{6 13}

Chylopericardium is diagnosed by the presence of a milky opalescent pericardial effusion.^{1 6} The triglyceride content in the effusion fluid is >500 mg/dL. The cholesterol:triglyceride ratio of the pericardial effusion fluid is <1 with a predominant lymphocyte count in the hundreds.¹ This is in accordance with what was found in this case, namely, milky fluid, was obtained. Pericardial fluid analysis showed that the total cholesterol was 79 mg/dL, triglyceride was 2678 mg/dL and cholesterol:triglyceride ratio was <1. In addition, there were also predominant lymphocytes with a cell count of 513. Therefore, it can be concluded that this patient has chylopericardium.



Video 2 Minimal pericardial effusion

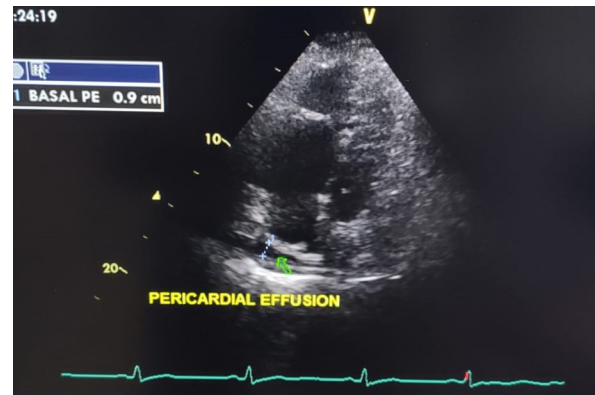


Figure 5 Minimal pericardial effusion at basal (0.9 cm).

The management of pericardial effusion with tamponade or in selected large effusion without tamponade is pericardiocentesis. Furthermore, treating the underlying cause is of foremost importance.¹ The treatment of chylopericardium-induced cardiac tamponade in patient with non-Hodgkin's lymphoma, as is the case, is identical to that of massive pericardial effusion in general, namely, pericardiocentesis. Pericardial drainage must be performed immediately to avoid the patient developing unstable haemodynamics and alleviate the symptoms.^{1 2} Chemotherapy is still the mainstay of treatment. Pericardiocentesis is just life saving. In this case, the pericardial effusion was still productive even after the administration of bleomycin intracardiac. Then, complete chemotherapy to treat the underlying cause of malignancy was performed. The chemotherapy solved the fluid accumulation.

Learning points

- ▶ Chylopericardium is rare, might also cause tamponade. Diagnosis of chylopericardium is established by direct analysis of fluid from pericardial effusion. The fluid contains a milky opalescent pericardial effusion, with a triglyceride level of >500 mg/dL, cholesterol:triglyceride ratio of <1, negative cultures and lymphocyte predominance.
- ▶ Chylopericardium, distinguished from cholesterol pericarditis in which cholesterol pericarditis showed clearer fluid and frequently associated with tuberculous pericarditis, rheumatoid pericarditis and trauma. On the other hand, chylopericardium can be caused by various factors, which in this case was caused by mediastinal neoplasms.
- ▶ Pericardiocentesis is warranted in cardiac tamponade for live saving. The treatment of chylopericardium that results in cardiac tamponade is to perform pericardiocentesis immediately to evacuate pericardial fluid, preventing the patient's haemodynamics from deteriorating and alleviating the patient's symptoms. Chemotherapy, which includes systemic therapy—not only local therapy (intrapericardium bleomycin), itself is the mainstay of treatment. In this case, the primary treatment for the underlying cause is mediastinal non-Hodgkin's lymphoma, must be carried out by completing the chemotherapy regimen.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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