

An unusual presentation of lymphoma: Chylotamponade

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ABSTRACT

Chylopericardium is an uncommon but potentially life-threatening clinic entity. Here we reported a case with chylopericardium causing tamponade and shock as an unusual presentation of lymphoma. The patient was managed by immediate pericardiocentesis. Further analysis of the pericardial fluid revealed immature T-cells compatible with precursor T-lymphoblastic lymphoma.

Isolated chylopericardium was first described by Hasebrock in 1888.¹ This is a rare clinical entity in which chyle accumulates in the pericardial cavity. It usually results from the injury of the thoracic duct by chest trauma or surgery, congenital mediastinal lymphangiectasy, Gorham syndrome, mediastinal neoplasms, subclavian vein thrombosis, tuberculosis, radiotherapy, infection, or Behçet syndrome. The pathophysiology of all these conditions seems to be the obstruction of the thoracic duct without development of collateral drainage.² Chylous effusion rarely causes acute pericardial tamponade characterized with the triad of decreased arterial blood pressure, increased venous pressure, and a small quiet heart. We describe a case of secondary chylopericardium with cardiac tamponade due to lymphoma.

CASE REPORT

A 20-year-old man admitted to the emergency department at BezmiÂlem Foundation University presented with fatigue and shortness of breath that had started 3 hours previously. He was orthopneic, tachycardiac (130

beats/min), and hypotensive (85/50 mm Hg) at the time of admission. His physical examination revealed decreased heart sounds, pulsus paradoxus, and jugular venous distention. He had sinus tachycardia, low voltage, and electrical alternans on surface electrocardiography (Figure 1). A chest x-ray showed bilateral pleural effusion, mediastinal enlargement, and cardiomegaly (Figure 2). He underwent urgent thorax computed tomography that revealed an anterior mediastinal mass and pleural and pericardial effusion (Figure 3). Emergency transthoracic echocardiography confirmed severe pericardial effusion with a swinging heart (Figure 4). Pericardiocentesis via subxyphoid approach was performed immediately under intensive care unit monitoring, and 1200 mL of chylous fluid was drained from the patient (Figure 5). Samples from the pericardial fluid were prepared for biochemical, microbiologic, and pathologic examinations. After pericardiocentesis, arterial blood pressure began to increase, and dyspnea subsided. The results of the pericardial fluid were white blood cells > 1000/ μ L (lymphocyte 89.9%), triglyceride 918 mg/dL, protein 4.7 g/dL, and albumin 3.6 g/dL. Biochemical analysis showed chylous pericardial effusion. Acid-fast staining of pericardial fluid was negative. Cytology

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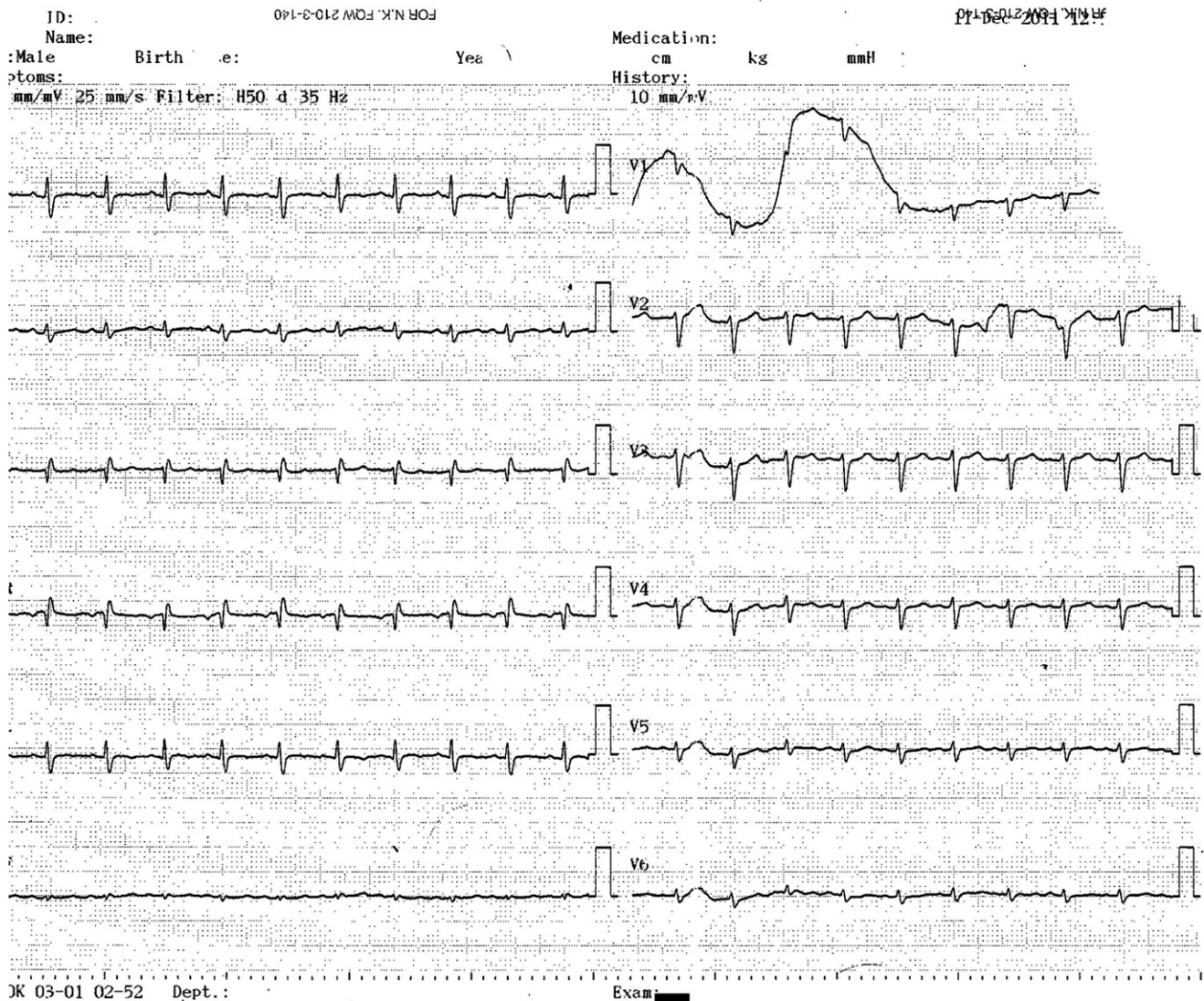


Figure 1 – Baseline electrocardiogram. Sinus tachycardia with electrical alternans.

confirmed the presence of immature T cells. Flow cytometric and immunocytochemical findings were as follows: CD3(+), CD5(+), CD10 focal (+), CD4(+), CD8(+), BCL-2(+), BCL-6(-), TdT+, cyclin D1 (-), CD20(-). Histology, immunocytochemistry, and chromosome analysis revealed precursor T-lymphoblastic lymphoma.

Total parenteral nutrition support was maintained after pericardiocentesis, and repeat echocardiography revealed only a small amount of pericardial effusion. The patient was transferred to the oncology clinic for chemotherapy, radiotherapy, and an allogenic stem cell transplantation.

DISCUSSION

Chylopericardium, characterized by the accumulation of chylous fluid in the pericardial sac, is an

uncommon clinical entity. It was first described by Hasebrock.¹ It may derive from extravasation of chyle from the lymphatic duct into the pericardial space as the result of abnormal communication between the pericardium and the thoracic duct or obstruction of the vessels draining pericardial lymph. Parietal pericardial lymphatic drainage is mainly to the anterior and posterior mediastinal nodes, whereas visceral pericardial lymphatic drainage is via the tracheal and bronchial mediastinal lymph nodes and finally into the thoracic duct.³ Blockage of the thoracic duct does not by itself cause chylopericardium because lymph can also drain via the diaphragmatic and intercostal routes.⁴ There are multiple causes of chylopericardium, including primary (or idiopathic) trauma, mediastinal neoplasm, subclavian vein thrombosis, tuberculosis, radiotherapy, infection, thoracic surgery, Behçet

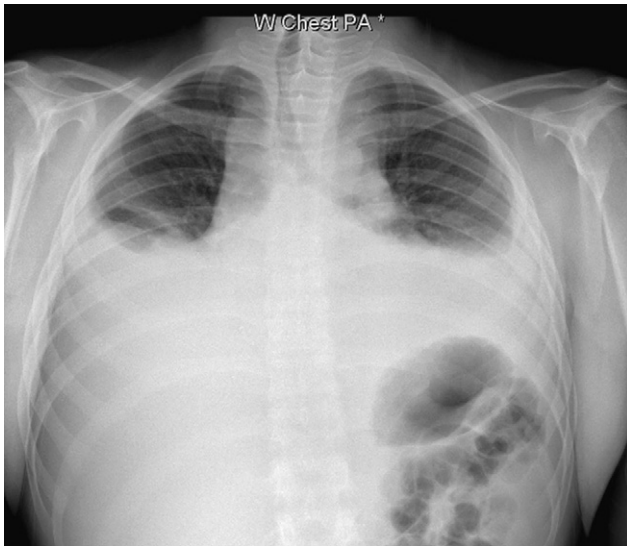


Figure 2 – Chest x-ray shows cardiomegaly and bilateral pleural effusion.

syndrome, or Gorham syndrome.⁴ Dyspnea is the most common presentation for patients with chylopericardium.⁴ Some patients have chest pain or discomfort, cough, palpitations, and syncope. Tamponade may occur if the accumulation of chyle in the pericardial space causes the intrapericardial pressure to be higher than the intracardiac pressure. Up to 17% of patients may present with cardiac tamponade.⁵

In the presence of pericardial tamponade, pericardiocentesis followed by cytology, chemistry, and culture is necessary to make the diagnosis. Analysis of the fluid usually shows a milky yellowish aspirate with a triglyceride level greater than 500 mg/dL, a cholesterol/triglyceride ratio less than 1, high protein content, a predominance of lymphocytes, and the presence of fat globules by Sudan III staining. Blood cultures are universally negative. Aspiration of fluid is continued until there is clear clinical and hemodynamic improvement. Enhanced computed tomography with lymphangiography could be used as a diagnostic tool for identifying abnormalities of the thoracic duct and its lymphatic channels connecting to the pericardium. Lymphoscintigraphy with radioactive oral (131)I-triolein and observation of Sudan III dye distribution into the pericardial cavity after oral intake of Sudan III dye are other described diagnostic modalities.⁶

Although the optimal treatment of chylopericardium is not clear, pericardial drainage in the case of hemodynamic compromise should be performed. For patients who have chylopericardium refractory to pericardiocentesis, surgical options such as pericardiostomy, creation of a pericardial window, thoracic duct ligation via a right open thoracotomy, or thoroscopic direct clipping of the thoracic

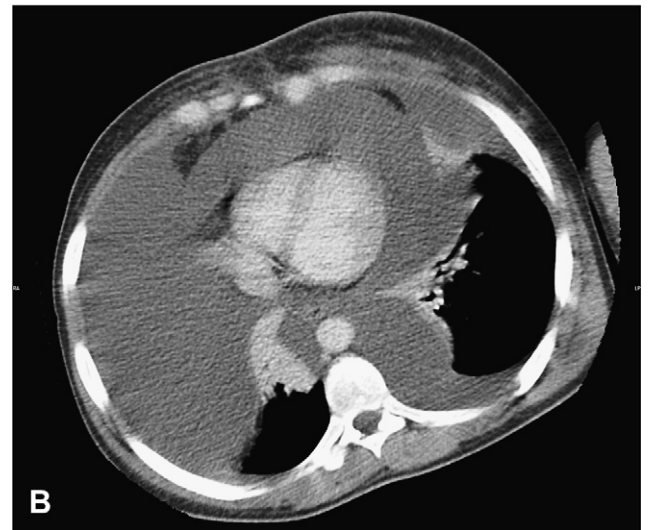
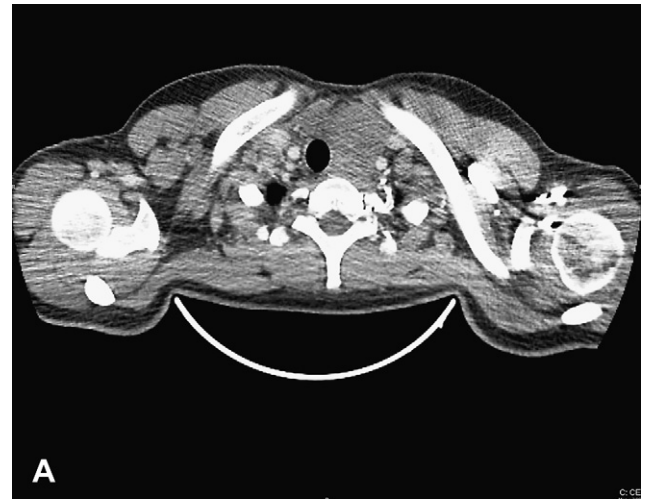


Figure 3 – Computed tomography shows an anterior mediastinal mass compressing the trachea (A) and massive pericardial effusion (B).

duct by video-assisted thoracic surgery should be taken in account. Conservative treatment includes low-fat diet with increased medium-chain triglycerides, total parenteral nutrition, and subcutaneous octreotide.

Precursor T-lymphoblastic lymphoma is a type of non-Hodgkin lymphoma in which immature T lymphocytes are involved in the lymph nodes and spleen.⁷ It is most common in young men. Although patients usually present with painless lymphadenopathy, 50% to 75% of patients have mediastinal mass. It may cause dyspnea and chest pain, which were thought to be symptoms of the heart failure, or it may progress to life-threatening compression of the superior vena cava or tracheobronchial tree.⁸ Chylothorax, chylopericardium, or chylous ascites can be present if major obstruction to lymphatic drainage has occurred.

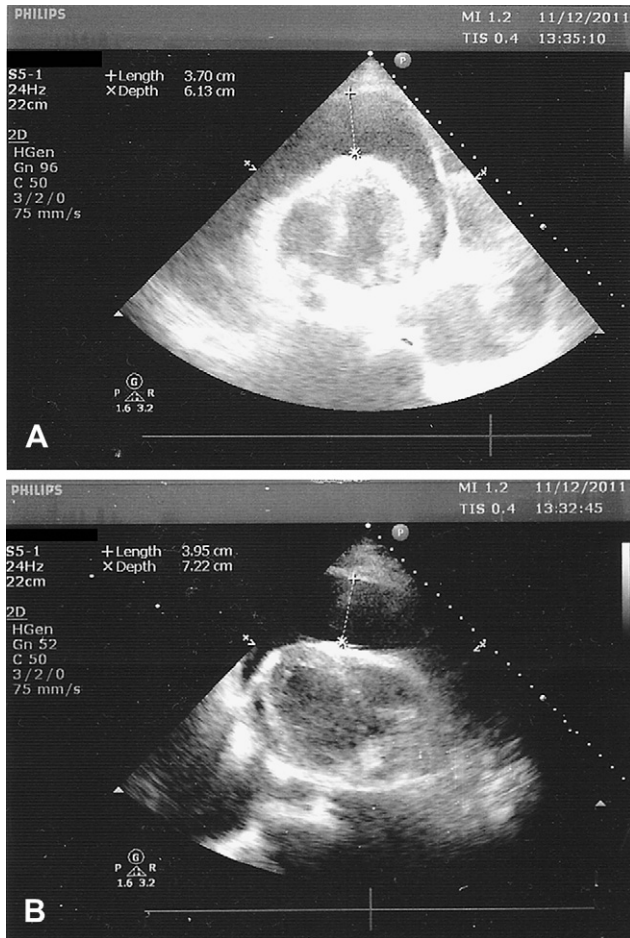


Figure 4 – Transthoracic echocardiogram demonstrating massive pericardial effusion.

CONCLUSIONS

We presented a case with chylopericardium causing tamponade and shock as an unusual presentation of lymphoma. These cases are primarily diagnosed and treated by hematologists or oncologists. In case of progressive dyspnea and hemodynamic compromise during the course of the disease, physicians should take into account chylotamponade and immediately reconsider the case with a cardiologist.



Figure 5 – Macroscopic view of the pericardial fluid.

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