

Taponamiento cardiaco como forma de presentación inicial de un linfoma linfoblástico de células T

Cardiac tamponade as the initial presentation of a T-cell lymphoblastic lymphoma

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ABSTRACT

We report a 27-year-old man who presented with cardiac tamponade and was eventually diagnosed of T-cell lymphoblastic lymphoma (T-LBL) by the flow cytometric analysis of pericardial fluid. Despite pericardiocentesis and institution of standard chemotherapy, the patient developed malignant arrhythmia with hemodynamic instability, and died soon after admission. Cardiac tamponade is rarely the first manifestation of a T-LBL.

Keywords: T-cell lymphoblastic lymphoma, Pericardial effusion, Cardiac tamponade.

INTRODUCTION

T-cell lymphoblastic lymphoma (T-LBL) has an annual incidence of 0.3 cases per 100,000¹. Although pericardial involvement has been previously reported, cardiac tamponade as the initial presentation of the disease is extremely rare.

CASE REPORT

A 27-year-old male was admitted to the hospital for a 3-week history of fever and chest pain and, during the last 3 days, dyspnea on exertion. Medical history was unremarkable.

On physical examination, temperature was 37,5°C, blood pressure 100/80 mmHg with pulsus paradoxus (12 mmHg), pulse rate 100/min, respiratory rate 21/min, and oxygen saturation 96%. Jugular venous distension was present, heart sounds were distant, and there was no pericardial friction rub. Pulmonary examination was normal. Generalized lymphadenopathy (painless, firm, and movable lymph nodes), and a red plaque on the left shoulder were detected (Fig. 1A). Laboratory studies revealed a hemoglobin level of 13 g/dL, platelet count of 184x10⁹/L, and total leukocyte count of 4.68x10⁹/L (13.5% lymphocytes, 77% polymorphonuclear leukocytes, and 5.1% monocytes without atypical cells). Serum C-reactive protein was of 36.1 mg/L, and lactate dehydrogenase (LDH) level was 1822 U/L.

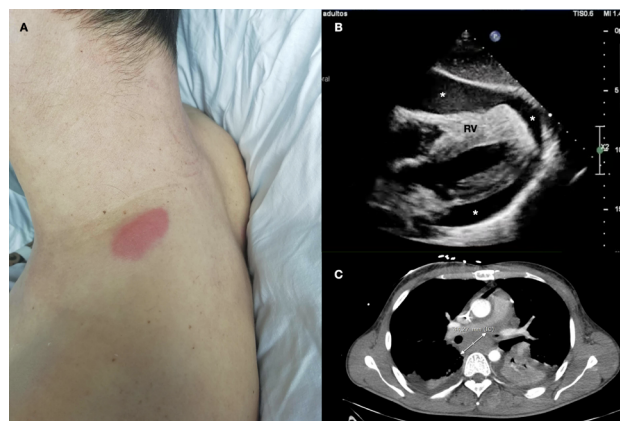
Chest X-ray showed an enlarged cardiac contour with a possible widening of the mediastinum, while the electrocardiogram demonstrated low voltages and electric alternance. The echocardiography revealed a severe pericardial effusion with right-sided chambers' collapse (Fig. 1B), dilated and plethoric inferior vena cava, septal wall thickness (19 mm), and heterogenous granular

RESUMEN

Describimos el caso de un varón de 27 años que se presentó con un taponamiento cardiaco y fue diagnosticado de linfoma linfoblástico de células (T-LBL) mediante citometría de flujo del líquido pericárdico. A pesar de una pericardiocentesis y de la instauración de quimioterapia estándar, el paciente desarrolló una arritmia maligna con inestabilidad hemodinámica y falleció poco después de su ingreso. El taponamiento cardiaco es rara vez la primera manifestación de un T-LBL.

Palabras clave: T-cell lymphoblastic lymphoma, Pericardial effusion, Cardiac tamponade

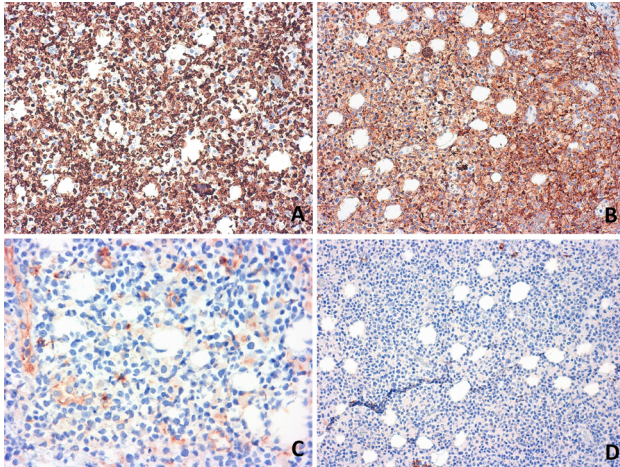
Figure 1. A) Red infiltrative plaque on the left shoulder; B) Subcostal echocardiographic image showing pericardial effusion (*) and right ventricular (RV) collapse; C) Chest CT showing a retrocardiac mass.



echocardiographic texture which suggested an infiltrative disease. A chest and abdominal computed tomography showed cervical, axillary, mediastinal, abdominal and inguinal lymphadenopathies of 2-2.5 cm², bibasal lung collapse/consolidations, small bilateral pleural effusions, isolated ill-defined lung nodules, peribronchial thickening, and a 3x5x3 cm retrocardiac mass (Fig. 1C).

Drainage of 850 mL of a serosanguineous fluid was accomplished with pericardiocentesis. The cytological examination of pericardial fluid was negative, but its flow cytometric analysis was consistent with T-LBL. A skin biopsy showed an infiltration of T lymphocytes

Figure 2. Skin biopsy. T-cell lymphoma was illustrated by diffuse positivity of CD3 (A), and diffuse positivity for CD4 (B), without a precursor T-cell phenotype as demonstrated by negativity for TdT (C) and CD1a (D).



in the dermis and subcutaneous tissue. Immunohistochemistry revealed positivity for CD3+, CD4+ and CD5+, and negative B-cell markers as well as TdT (terminal deoxynucleotidyltransferase) and CD1a; all of which was consistent with a peripheral T-cell lymphoma (Fig. 2). A bone marrow aspirate showed increased cellularity, with cells in all stages of maturation, decreased fatty tissue, and 3% of lymphoblasts. A cytogenetic study reported a karyotype with multiple chromosomal breakages and non-clonal alterations suggesting chromosomal instability.

Two days after admission, the patient experienced cardiac arrest due to ventricular fibrillation and recovered in 30 minutes after advanced cardiopulmonary resuscitation. He was admitted to the intensive care unit with multiorgan failure and cardiogenic shock. Five days after pericardiocentesis, a bedside echocardiography showed a moderate quantity of pericardial effusion, with no signs of cardiac tamponade. Treatment with high-dose corticosteroids (methylprednisolone 250 mg IV per day), and prophylaxis of the tumor lysis syndrome (rasburicase 15 mg IV per day) was started, followed by chemotherapy with cyclophosphamide 1g IV 1 day and cytarabine 3 g IV every 12 hours for 2 days. On the second day of chemotherapy the patient had another cardiac arrest and died. A necropsy study was not performed.

DISCUSSION

Both T-cell acute lymphoblastic leukemia (T-ALL) and T-LBL are clonal expansions of lymphoid blasts. The disease is categorized as T-LBL when the process is confined to lymph nodes or a mass lesion, with minimal or absent blood and bone marrow involvement. With extensive bone marrow and blood involvement, the term T-ALL is preferred. Generally, the diagnosis of T-ALL should be avoided when there is less than 20% of lymphoblasts in the bone marrow. T-LBL is less common than T-ALL and it appears mostly in childhood and young adults, with a 2:1 male predominance¹. In our case, clinical evaluation and laboratory results revealed a T-LBL, as there were just 3% of lymphoblasts in the bone marrow.

T-LBL is often diagnosed in advanced stages (70% in the Ann Arbor stages III-IV), with B symptoms being present in most of the cases. About 50-75% of the patients exhibit a mediastinal mass,

which is usually anterior, bulky, and often associated with pleural effusions¹.

The presentation of T-LBL with cardiac tamponade is extremely rare. A literature search in the PubMed database (keywords "pericardial effusion" AND (T-cell acute lymphoblastic leukemia OR T-cell lymphoblastic lymphoma), time period 1982-2021, and no language restrictions), yielded 36 publications on disease-associated pericardial effusions. Fourteen of them were excluded due to lack of clinical information on the development of cardiac tamponade, and one due to the existence of an alternative diagnosis to T-ALL/T-LBL. The remaining 21 publications comprised a total of 22 patients with pericardial effusions in the context of T-ALL or T-LBL, of whom only 8 had cardiac tamponade. One case of cardiac tamponade was deemed to be secondary to the oncological treatment, in particular methotrexate². This leaves only 7 reported cases of cardiac tamponade attributable to the underlying lymphoma (Table 1)³⁻⁹. Patients were young and predominantly male, and most had a mediastinal mass. In all cases, cardiac tamponade was the initial presentation of T-LBL.

Effusion cytology aims to achieve an early and quick diagnosis of malignancy. In our patient, cytological studies of the pericardial fluid were negative. The diagnosis in such cases requires correlation of various cytomorphologic features, immunocytochemistry (ICC) and patient's clinical details. In a retrospective study, 51 positive cytological fluid samples from patients with lymphoreticular malignancies that included 30 (58.8%) pleural, 18 (35.3%) peritoneal, and 3 (5.9%) pericardial, were also analyzed by flow cytometry (FCM) and/or ICC¹⁰. Forty-eight samples were diagnostically confirmed by using these techniques, but 3 (5.9%) were considered to be false positives since FCM and/or ICC did not support a lymphoreticular malignancy. Currently, effusion cytology should be combined with FCM and/or ICC studies if lymphoma is suspected.

Our paper highlights cardiac tamponade as a life-threatening complication and rare clinical presentation of T-LBL. Moreover, the role of pericardial fluid FCM analyses to obtain a prompt diagnosis of lymphoma is stressed.

Table 1. Cases of cardiac tamponade in T-LBL and T-ALL patients

Publication	Patient	Presence of mediastinal mass	Other manifestations	Follow up
Sögüt et al 1999 ³	3-year-old female	-	Pericardial infiltration	-
Basu et al 2009 ⁴	27-year-old male	No	Pancytopenia, Left pleural effusion, Mild hepatomegaly and ascites, 90% lymphoblasts in the bone marrow	Pericardiocentesis was performed. The patient deteriorated and died on the fifth day of chemotherapy.
Erdoğan et al 2012 ⁵	20-year-old male	Yes	Chylopericardium, Bilateral moderate pleural effusions	Pericardiocentesis was performed. The patient received chemotherapy, radiotherapy, and an allogenic stem cell transplantation.
Kapur et al 2014 ⁶	25-year-old male	Yes	Massive left-sided and moderate right-sided pleural effusions, Pericardial infiltration	Subxiphoid pericardial window was done, and bilateral chest tubes were inserted, which improved patient's condition. Combination chemotherapy and intrathecal methotrexate was administered.
Ozdemir et al 2015 ⁷	7-year-old male	No	Anemia, Large amounts of blastic cells in pericardial liquid	Pericardiocentesis was performed. After initiation of chemotherapy, pericardial effusion completely disappeared within 2 weeks.
Alimi et al 2020 ⁸	18-year-old male	No	Pulmonary stenosis Pericardial infiltration	Pericardiocentesis was performed. After six courses of chemotherapy and one-year follow up, the patient remained asymptomatic.
Sablak et al 2021 ⁹	13-year-old male	Yes	Large left-sided pleural effusion	Small pericardial effusion with probable compressive effect from the pleural effusion, with hemodynamic and respiratory improvements after therapeutic thoracentesis. Chemotherapy was instituted.
Current case	27-year-old male	Yes	Lymphadenopathy, Small pleural effusions, Skin lesion	Pericardiocentesis was performed. On the second day of chemotherapy the patient died because of a cardiac arrest.

CONCLUSION

Cardiac tamponade is a rare initial presentation of T-cell lymphoblastic lymphoma and can be life-threatening. Flow cytometry of pericardial fluid may be a useful tool to provide the diagnosis of T-LBL.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interests.

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