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Case Report

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Primary pericardial lymphoma with hemodynamic disorder apropos of a case

Geovedy Martínez García[1]✉, MD, MSc; Yanitsy Chipi Rodríguez[1], MD; Annia M. Carrero Vázquez[1], MD; Yaydy González Miguélez[1], MD; Natalia Reynosa Paneque[1], MD; Rosa M. Martínez Peró[1], MD; Lissette Rodríguez Phinhevy[2], MD; Anisleidys Ledesma Duardo[2], MD; and Arturo Martínez García[3], MD

[1] Department of Cardiology, Hospital General Docente Enrique Cabrera Cossío. Havana, Cuba.
[2] Department of Geriatrics, Hospital General Docente Enrique Cabrera Cossío. Havana, Cuba.
[3] Department of General Surgery, Hospital General Docente Enrique Cabrera Cossío. Havana, Cuba.

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Abbreviations
MRI: magnetic resonance imaging
PCL: Primary cardiac lymphoma
TTE: Transthoracic echocardiography

ABSTRACT

Primary cardiac lymphoma is an extremely rare and fatal entity of the heart. It is defined as a lymphoma that compromises the heart as well as the pericardium. It has a poor prognosis because of the difficulty in its diagnosis, due to the inaccessibility of its location. Here is presented the case of an 84-year-old man with healthy history that came to our hospital with symptoms and signs of heart failure. The first suspicion was dilated cardiomyopathy, but in the transthoracic echocardiography was observed a large pericardial effusion with a mass attached to the side wall of the right chambers, without chamber dilation. A pericardial window was performed to drain the effusion, without being able to study the extracted fluid. Finally, a magnetic resonance imaging was performed resulting in a cardiac lymphoma diagnosis that, taking into account the absence of other changes in the rest of the body, was defined as primary. No histological diagnosis could be developed due to refusal of the patient and his relatives.
Keywords: Pericardium, Lymphoma, Cardiac tumors, Diagnosis, Echocardiography, Magnetic resonance imaging

Linfoma pericárdico primario con compromiso hemodinámico a propósito de un caso

RESUMEN

El linfoma cardíaco primario es una enfermedad extremadamente rara y fatal del corazón. Es definido como un linfoma que compromete el corazón y el pericardio. Tiene un pronóstico pobre debido a la dificultad en su diagnóstico por lo inaccesible de su localización. Se presenta el caso de un hombre de 84 años de edad con antecedentes de salud que acudió con síntomas y signos de insuficiencia cardíaca. Se sospechó cardiopatía dilatada, pero en el ecocardiograma transtorácico se observó derrame pericárdico severo con masa adherida a la pared lateral de las cavidades derechas, sin dilatación de cavidades. Se realizó ventana pericárdica para drenar el derrame, sin poder estudiar el líquido extraído. Finalmente se realizó resonancia magnética nuclear que diagnosticó linfoma cardíaco, y teniendo en cuenta la ausencia de otras alteraciones en el resto del cuerpo, se definió como primario. No se pudo realizar diagnóstico histológico por negativa del paciente y

✉ *G Martínez García*
Anita 936 e/ Gertrudis y Lagueruela, Sevillano, Diez de Octubre.
La Habana, Cuba.
E-mail address:
geovedymg@infomed.sld.cu

familiares.
Palabras clave: Pericardio, Linfoma, Tumores cardíacos, Diagnóstico, Ecocardiografía, Resonancia magnética nuclear

INTRODUCTION

The cardiac and pericardial affectation of a malignant lymphoma is very rare, and it represents about 1% of cardiac tumors and 0.5% of extranodal non-Hodgkin lymphomas, and the most common type is the diffuse large B-cell lymphoma[1].

The initial symptoms are given by cardiac failure, syncope, arrhythmias and pericardial effusion of great amount, and the positive diagnosis is made with cardiac imaging techniques and the histological study[2]. Despite these techniques, diagnosis –in most cases– is difficult, due to the inaccessible location of the tumor.

The treatment for a primary cardiac lymphoma (PCL) is variable because the optimal strategy has not yet been established; however, previous studies have shown that chemotherapy is the most effective treatment[3]. It has a poor prognosis, since patients die in the first months after diagnosis, mainly due to systemic damage[4].

We report the case of a patient who came to our institution with symptoms of cardiac failure globally; a pericardial lymphoma without other systemic locations was diagnosed through imaging techniques.

CASE REPORT

An 84-year-old man, with healthy history and three days of weakness, dyspnea on slight effort (class III of the New York Heart Association) and edema in both lower limbs, reasons why he went to the doctor of his health area, which indicated amoxicillin and digoxin; but when continuing with the same symptoms the patient went to the emergency department of our hospital.

Positive data from the physical examination

- Wet and hypo-colored mucous membranes.
- Lower limbs infiltrated with soft, cold, painless edemas, with presence of godet palpation, reaching both knees. Change of skin coloration in the distal region of the lower left limb.
- Vesicular murmur diminished globally, with bibasilar crackles and respiratory rate of 22 breaths per minute.
- Arrhythmic cardiac noises, low intensity, no murmurs, no friction. Heart rate of 106 beats per minute and blood pressure 100/60 mmHg.
- Soft and depressible abdomen, painful on palpation in the right hypochondrium, where painful hepatomegaly is found approximately 3 cm below the edge of the ribs. Presence of hepatojugular reflux.

Table. Blood tests performed.

Additional blood tests	Result	Reference values
Hematocrit	0.39	0.42 - 0.52
Leukogram	10.5 x 10^9/L	4.5 - 10.5 x 10^9/L
Erythrosedimentation	12 mm/h	< 30 mm/h
Alkaline phosphatase	116 U/L	100 - 290 U/L
GPT	28 U/L	0 - 49 U/L
GOT	34 U/L	0 - 46 U/L
GGT	81 U/L	5 - 45 U/L
Cholesterol	2.92 mmol/L	3.87 - 6.20 mmol/L
Triglycerides	1.13 mmol/L	0.46 - 1.88 mmol/L
Creatinine	62 µmol/L	49.0 - 104.0 µmol/L
Uric acid	382 µmol/L	155 - 428 µmol/L
Urea	6.1 mmol/L	3.30 - 8.30 mmol/L
Total proteins	79 g/L	60.0 - 80.0 g/L
Albumin	38 g/L	38 - 54 g/L
Glycemia	6.6 mol	4.20 - 6.11 mmol/L
PSA	0.38 ng/L	0.4 ng/L

GGT, gamma-glutamyl transpeptidase enzyme; GOT, glutamic oxalacetic transaminase enzyme; GPT, glutamic pyruvic transaminase enzyme; PSA, prostate-specific antigen.

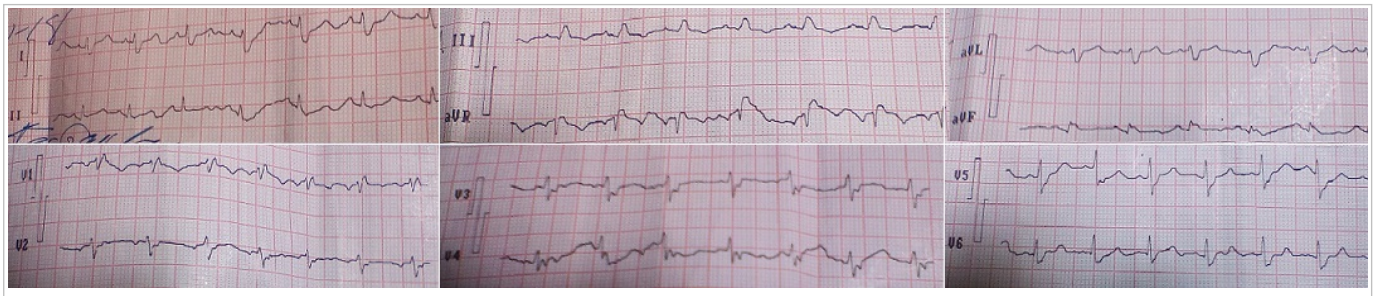

Figure 1. Electrocardiogram of the patient upon arrival at the hospital.

Tests

- Additional blood tests: they are described in the **table**.
- Electrocardiogram: normal electrical axis (△QRS +80º). Pattern of right bundle branch block and left anterior fascicular block (**Figure 1**).
- Posteroanterior thoracic radiography: increase in the transverse diameter of the cardiac silhouette. Dilated aorta. Pleural reaction in both bases, more pronounced on the right side and signs of bilateral hilium and basal pulmonary congestion.
- Abdominal ultrasound: normal-sized liver, homogeneous, conserved echogenicity, without focal lesions. Gallbladder of normal size, with reactive walls. No dilation of bile ducts. Pancreas of normal size, homogeneous, with discrete increase in its echogenicity. Normal caliber aorta, with fibrocalcic deposits and absence of aneurysmal lesions. Kidneys of normal size and position, prominent pyramids, poor corticomedullary relationship, irregular contours, with marked increase in parenchymal echogenicity, not ectasia. Bilateral pleural effusion of moderate amount, presence of perihepatic fluid and small amount in hypogastrium.
- Simple computed tomography. Thorax: bilateral pleural effusion of moderate amount involving fissures, signs of pulmonary fibrosis and emphysema, presence of pneumomediastinum and pericardial effusion, small mediastinal adenopathies smaller than 1 cm. Abdomen: hepatomegaly at the expense of the left lobe, absence of intraabdominal fluid, right convex scoliosis with narrowing of the intervertebral orifices.
- Transthoracic echocardiogram (TTE): Cardiac cavities of normal diameters and structures, preserved systolic function. Pericardial effusion of great amount, with no signs of cardiac tamponade (Witzman index = 54 mm). Hyperdense image, homogeneous, 46 × 61 mm, anchored to the lateral wall of the right cavities that it compresses, in particular, to the atrium where it produces systolic collapse (**Figure 2, A y B**). Dilated inferior vena cava, with inspiratory collapse less than 50%.

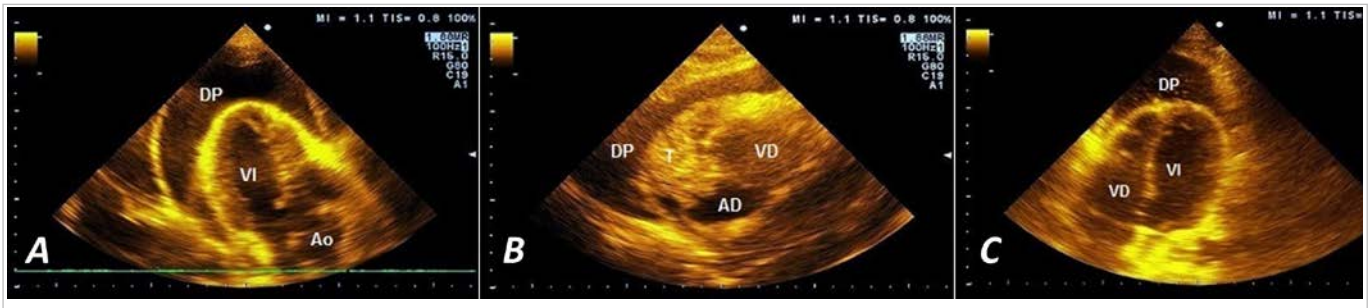

Figure 2. Transthoracic echocardiography. **A.** Long axis parasternal view. A large pericardial effusion is observed that surrounds the cardiac apex. **B.** Image of the pericardial tumor attached to the lateral walls of the right cavities (subcostal view). **C.** Study conducted three days after the pericardial window. The effusion can only be seen around the apex, with abundant remains of fibrin. Abbreviations in Spanish: Ao, aorta; VI, left ventricle; AD, right atrium; DP, pericardial effusion; VD, right ventricle; T, tumor.

With the result of the TTE and the clinical picture of the patient, it was decided –by the collective of Internal Medicine, Cardiology and General Surgery– to make a pericardial window surgery in order to drain the large pericardial effusion, relief the symptoms of the patient and to study, histologically, the liquid obtained. The procedure was performed by via transperitoneal, under local anesthesia, and 600 mL of dark serous liquid were extracted, which was sent for study. Unfortunately, the liquid coagulated before the histological study.

The patient had an improvement on its general condition after the procedure and the dyspnea and edema in the lower limbs disappeared. The TTE was repeated and a reduction of the pericardial effusion was observed, which was concentrated in the apical region of the left ventricle with abundant fibrin bands (**Figure 2C**). The image described above on the right side wall was maintained with the same characteristics.

In order to clarify the diagnosis of pericardial tumor, a magnetic resonance image (MRI) was performed, in which a tumor image was observed at the expense of the pericardium compressing the right cavities, mainly the right atrium, with diameters of 52.2 × 72.7 mm (**Figure 3**). With the studies of tissue characterization, it was concluded as an alleged pericardial lymphoma, but the histological diagnosis could not be made because the patient and his relatives refused to perform the biopsy and requested hospital discharge against medical recommendations.

COMMENTS

The pericardial primary lymphoma is an extremely rare disease and it represents only 0.5% of extranodal lymphomas. It is defined as an extranodal lymphoma limited to the heart or pericardium[1,2]. Histologically, the diffuse large B-cell non-Hodgkin lymphoma is the most common of the PCL (58%), followed by the T-cell (16%), Burkitt (9) and small-cell lymphocytic lymphoma (6%)[2]. In the bibliography, most PCL were diagnosed as a mass anchored to the walls of the cardiac cavities, primarily the right atrium, right ventricle and left atrium, in order of frequency[5].

Patients usually present nonspecific symptoms such as dyspnea, arrhythmias, pericardial effusion and congestive heart failure, as observed in our case[3,5]. Imaging studies such as echocardiography, computed tomography and the MRI are useful for diagnosing a PCL; nevertheless, the histopathological study is the only way to obtain a definitive diagnosis[6]. Chemotherapy is effective for the treatment of some subtypes of this lymphoma. Previous reports have demonstrated that rituximab with the CHOP regime (cyclophosphamide, doxorubicin, vincristine and prednisolone) has had excellent results in the diffuse large B-cell lymphoma, with survival of 30 months after the diagnosis[3]. However, the optimal treatment strategy has not yet been established.

In the case presented, the patient had symptoms of global heart failure, without suspicion of neoplastic origin. The TTE showed pericardial effusion and a mass adhering to the rights cavities, but being

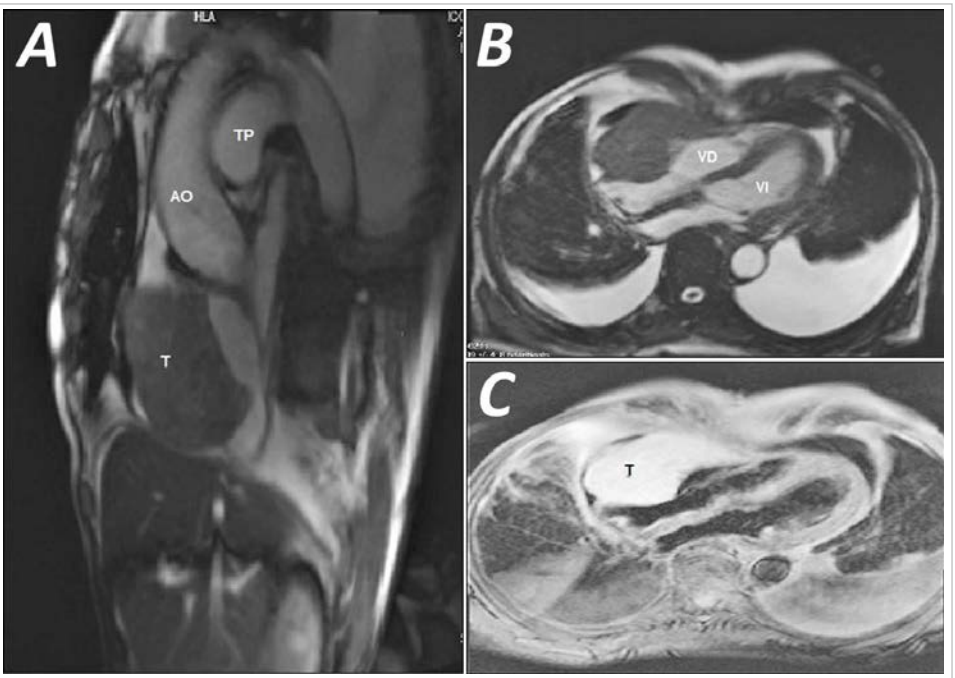

Figure 3. Contrasted cardiac magnetic resonance imaging. **A.** Sagittal cut. **B** and **C.** Transversal cut. A pericardial lymphoma is observed compressing right cavities. Abbreviations in Spanish: AO, aorta; VI, left ventricle; TP, pulmonary artery trunk; VD, right ventricle; T, tumor.

unable to study the pericardial fluid and not finding other tumor locations in the rest of the body, the MRI helped establishing the diagnosis of PCL. There are currently numerous scientific evidences that show the usefulness of MRI in the evaluation of cardiac masses, especially in the morphology, location, movement, vascularization and characterization of different tissues, comparing the intensity of images of T1 and T2 and the proton density width[7-10]. That is why it is currently the most widely used non-invasive diagnostic and characterization resource for cardiac tumors.

In spite of the interesting thing about the case, our presentation has an important limitation, and it is that the histological diagnosis of the tumor could never be corroborated. Nonetheless, we decided to publish it due to: the infrequency of its occurrence, which makes almost impossible its early diagnosis; poor accessibility of its location; and the benefits of MRI for guiding the diagnosis.

REFERENCES

1. Li S, Desai P, Lin P, Yin CC, Tang G, Wang XJ, *et al*. MYC/BCL6 double-hit lymphoma (DHL): a tumor associated with an aggressive clinical course and poor prognosis. Histopathology. 2016;68(7): 1090-8.
2. Gordon MJ, Danilova O, Spurgeon S, Danilov AV. Cardiac non-Hodgkin's lymphoma: clinical characteristics and trends in survival. Eur J Haematol. 2016;97(5):445-52.
3. Petrich A, Cho SI, Billett H. Primary cardiac lymphoma: an analysis of presentation, treatment, and outcome patterns. Cancer. 2011;117(3):581-9.
4. Habertheuer A, Ehrlich M, Wiedemann D, Mora B, Rath C, Kocher A. A rare case of primary cardiac B cell lymphoma. J Cardiothorac Surg [Internet]. 2014 [cited 14 Ene 2019];9:14. Available at: https://cardiothoracicsurgery.biomedcentral.com/track/pdf/10.1186/1749-8090-9-14
5. Miguel CE, Bestetti RB. Primary cardiac lymphoma. Int J Cardiol. 2011;149(3):358-63.
6. Kawamura T, Sakaguchi T, Nishi H, Miyagawa S, Yoshikawa Y, Yamauchi T, *et al*. Successful treatment of a large primary cardiac lymphoma by surgical resection combined with chemotherapy: report of a case. Surg Today. 2013;43(9):1066-70.
7. Li B, Li Q, Nie W, Liu S. Diagnostic value of whole-body diffusion-weighted magnetic resonance imaging for detection of primary and metastatic malignancies: a meta-analysis. Eur J Radiol. 2014; 83(2):338-44.
8. Zhu D, Yin S, Cheng W, Luo Y, Yang D, Lin K, *et al*. Cardiac MRI-based multi-modality imaging in clinical decision-making: Preliminary assessment of a management algorithm for patients with suspected cardiac mass. Int J Cardiol. 2016;203:474-81.
9. Nensa F, Tezgah E, Poeppel TD, Jensen CJ, Schelhorn J, Köhler J, *et al*. Integrated 18F-FDG PET/MR imaging in the assessment of cardiac masses: a pilot study. J Nucl Med. 2015;56(2):255-60.
10. Fathala A, Abouzied M, AlSugair AA. Cardiac and pericardial tumors: A potential application of positron emission tomography-magnetic resonance imaging. World J Cardiol. 2017;9(7):600-8.