Turmoil of Symptoms: The Devastating Impact of Cardiac Lymphoma – Multiple Clinical Manifestations and the Lethal Causes of Cardiac Involvement

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Abstract

Primary cardiac lymphoma (PCL) is a rare and highly lethal neoplasm, whose nonspecific clinical presentation makes early diagnosis difficult. This article reports the case of an elderly patient, previously healthy, who experienced dizziness and syncope for four months before the discovery of a large tumor mass in the heart. Imaging exams, such as echocardiography and magnetic resonance imaging, were crucial for tumor identification, and the biopsy confirmed it to be a B-cell lymphoma with high replication. However, given the severity of the condition and the family's refusal to proceed with treatment, the patient progressed to sudden death two weeks after diagnosis. Primary cardiac tumors (PCTs) are underdiagnosed, with varied symptoms that may include vascular obstructions, arrhythmias, and heart failure. PCLs account for only 1% to 2% of primary cardiac masses and are more common in elderly men. The diagnosis is challenging due to the absence of specific signs, making imaging exams and biopsies essential for disease confirmation. This case illustrates the complexity of managing PCLs, whose rapid progression limits therapeutic options. The lack of specific symptoms and silent progression highlight the importance of early diagnosis. The high mortality rate reinforces the need for greater medical attention to this condition, which frequently leads to fatal outcomes before an effective intervention can be implemented.

Introduction

Primary cardiac tumors (PCTs) are rare entities in clinical practice and can be classified as benign lesions (myxomas) or malignant tumors (sarcomas and lymphomas).[1,2] With the increasing life expectancy of the population, a higher incidence of these tumors is expected, highlighting the need for greater attention to their possible symptoms, especially considering their varied clinical manifestations.[2,3]

Considering this, it is important to emphasize that PCTs are often underdiagnosed, which may lead to fatal outcomes before an effective therapeutic approach can be implemented.[2] This article presents a case of primary myocardial lymphoma in an elderly patient, previously healthy, who experienced sudden death two weeks after diagnosis.

Thus, the objective of this study is to discuss the clinical and diagnostic characteristics of PCTs.

Case report

A 78-year-old man, previously healthy, presented with dizziness and syncope worsening over four months. The patient had been seen at other emergency care units, but his symptoms progressively worsened.

On physical examination, no significant findings were noted. The electrocardiogram showed sinus rhythm with diffuse ventricular repolarization abnormalities. A chest X-ray revealed a cardiac silhouette at the upper limit of normal and bilateral costophrenic angle blunting. A transthoracic echocardiogram detected a large tumor mass infiltrating the interventricular septum and the inferolateral wall of the right ventricle, extending into the right atrial and ventricular cavities, as well as the left ventricular cavity adjacent to the septum (Figure 1).

Given this, the patient was referred for a magnetic resonance imaging scan for a more detailed assessment of the neoplasm, which confirmed the echocardiographic findings (Figure 2). An endomyocardial biopsy was performed, guided by transthoracic echocardiography and fluoroscopy, conducted simultaneously in the hemodynamics room. The histopathological study and immunohistochemical analysis concluded that it was an invasive B-cell lymphoma with intense cellular replication (Figure 3).

After a detailed explanation of this complex and severe clinical case to the patient and his family, they refused the implementation of any form of treatment and requested discharge to his home, where he remained asymptomatic for approximately two more weeks, before experiencing sudden death.

Keywords

Heart Neoplasms; Echocardiography; Cardiac Arrhythmias; Biopsy

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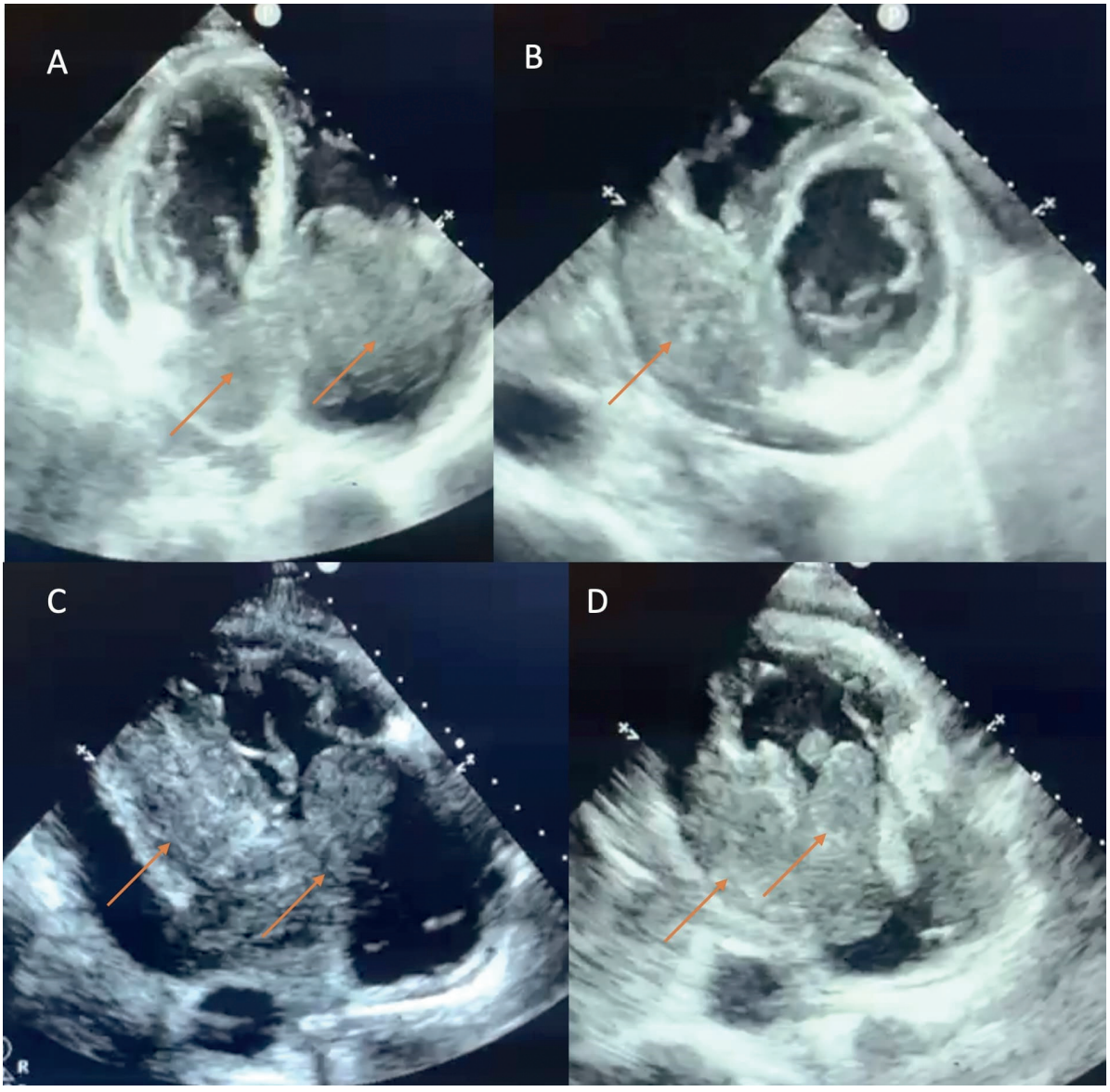

Figure 1 – *Transthoracic echocardiogram showing a tumor mass infiltrating the atrioventricular valves, the ventricular myocardium at the septal level, and the anterior and inferior walls of both ventricles. The tumor occupies the atrial and right ventricular cavities (arrows); pericardial effusion.*

Discussion

Although reported since the 16th century, PCTs are rare in clinical practice, with an incidence ranging from 0.0017% to 0.03% in autopsy studies.[1-3] PCTs can primarily manifest as benign lesions, with an incidence rate of 75%, or as malignant tumors, accounting for 25% of cases.[3-5]

On the other hand, primary cardiac lymphomas (PCLs) are classified as malignant neoplasms, representing 1% to 2% of all primary cardiac masses.[2-5]

Although PCTs generally affect more women, PCLs are more common in men over 60 years old, which is well represented by the patient described in this case report.[4-6]

PCLs often present nonspecific and multifaceted clinical manifestations.[14] Initially, the patients may be asymptomatic or oligosymptomatic, but as the disease progresses, they develop various signs and symptoms due to direct and/or indirect involvement of multiple organs and systems through heterogeneous, simultaneous, and diverse mechanisms. These include recurrent vascular obstructive phenomena caused by

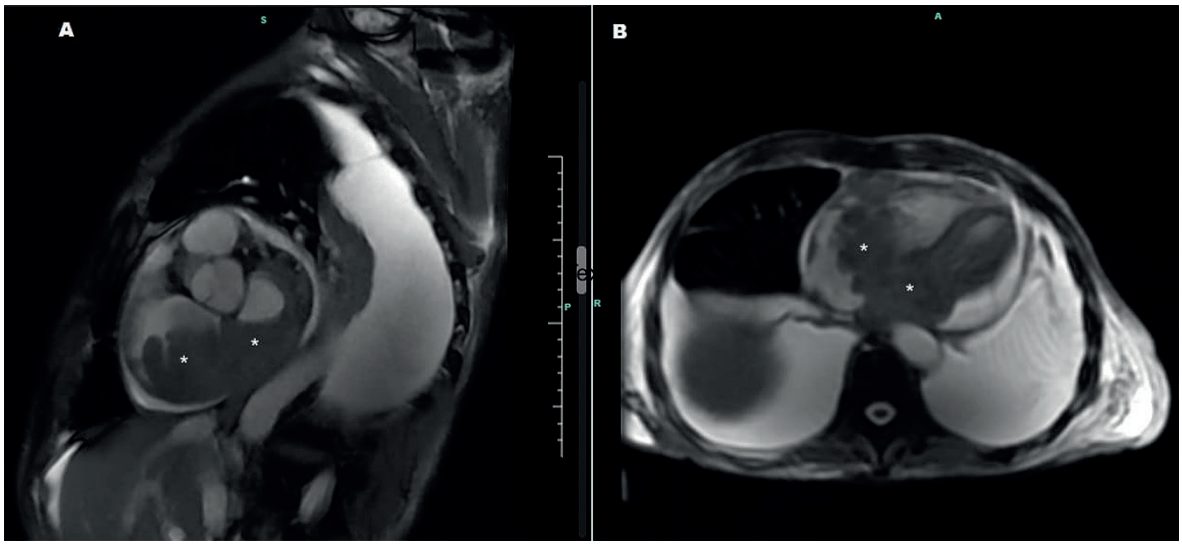

Figure 2 – *Magnetic resonance imaging showing the extent of cardiac involvement by lymphoma (*) demonstrated in the sagittal (A) and transverse (B) planes, along with pericardial and bilateral pleural effusion.*

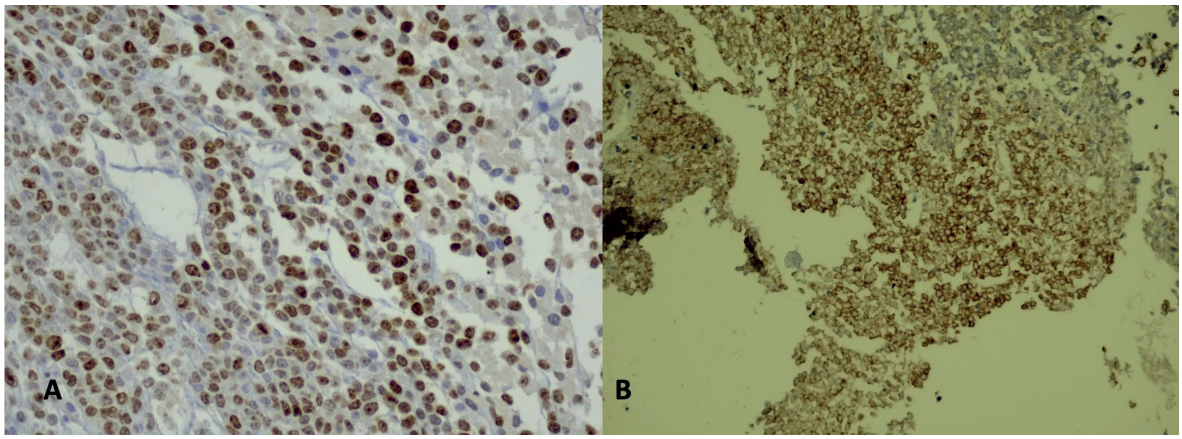

Figure 3 – *Histopathological and immunohistochemical findings demonstrating B-cell lymphoma with intense cellular proliferation in Ki-67 (A), and B lymphocytes marked in CD20 (B).*

thrombi and/or tumor fragments, leading to stroke, peripheral and visceral arterial embolism, and pulmonary thromboembolism.[4-7]

The tumor itself may cause fixed or transient intracardiac blood flow obstruction, depending on its size and morphology (intramural or pedunculated intracavitary), potentially compromising cardiac output.[2,4] When the myocardial electrical conduction system is affected, arrhythmia may occur, sometimes unexpectedly fatal, as possibly seen in the case presented here.[5-7]

Thus, in cases of cardiac tumors, it is undeniable that, due to their nonspecific symptoms, establishing a timely diagnosis to apply effective therapy is a challenge for the healthcare team, as demonstrated in this report.[6,7] Furthermore, it is essential to highlight the importance of imaging exams, such as echocardiography and cardiac magnetic resonance imaging, in the detection of these tumor masses and in performing endomyocardial biopsies to establish a histopathological diagnosis.[8-12]

Conclusion

PCL represents a rare but extremely severe condition, whose nonspecific clinical presentation and silent progression make its diagnosis a major challenge.

As discussed in this report, its multiple manifestations can mask the true extent of the disease, resulting in a discouraging prognosis. Cardiac involvement by lymphoma not only compromises organ function but also exposes the patient to a high risk of lethal events, such as fatal arrhythmias and critical vascular obstructions.

Given this scenario, the importance of early and thorough investigation using imaging exams, histopathological aspects, and immunohistochemical analysis for diagnostic definition is fundamental. However, as evidenced in the case described, even with the diagnosis established, therapeutic options may be limited due to the rapid progression of the disease and the patient's own decisions.

Thus, the devastating impact of cardiac lymphoma extends beyond its physiological repercussions, encompassing therapeutic limitations and abrupt outcomes, emphasizing the need for greater medical-scientific attention and understanding for future approaches to this unusual and fatal entity.

Author Contributions

Conception and design of the research: Carvalho G; acquisition of data: Carvalho G, Carvalho MFM, Magalhães T, Hayashida MR, Staszko KF; analysis and interpretation of the data: Carvalho G, Carvalho MFM, Varela AM; writing of the manuscript: Carvalho G, Carvalho MFM; critical revision of the manuscript for intellectual content: Carvalho G, Carvalho MFM, Magalhães T, Hayashida MR, Staszko KF, Varela AM; review of references: Carvalho MFM; magnetic resonance imaging: Magalhães T; pathological anatomy analysis: Hayashida MR; echocardiogram images: Staszko KF.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

This study was approved by the Ethics Committee of the Hospital De Clínicas Da Universidade Federal Do Paraná under the protocol number 90118225.4.0000.0096. All the procedures in this study were in accordance with the 1975 Helsinki Declaration, updated in 2013. The Free and Informed Consent Form was waived.

Use of Artificial Intelligence

The authors did not use any artificial intelligence tools in the development of this work.

Availability of Research Data

The underlying content of the research text is contained within the manuscript.

References

1. Leja MJ, Shah DJ, Reardon MJ. Primary Cardiac Tumors. Tex Heart Inst J. 2011;38(3):261-2.
2. Tyebally S, Chen D, Bhattacharyya S, Mughrabi A, Hussain Z, Manisty C, et al. Cardiac Tumors: JACC CardioOncology State-of-the-Art Review. JACC CardioOncol. 2020;2(2):293-311. doi: 10.1016/j.jaccao.2020.05.009.
3. Cresti A, Chiavarelli M, Glauber M, Tanganelli P, Scalese M, Cesareo F, et al. Incidence Rate of Primary Cardiac Tumors: A 14-Year Population Study. J Cardiovasc Med. 2016;17(1):37-43. doi: 10.2459/JCM.0000000000000059.
4. Carras S, Berger F, Chalabreysse L, Callet-Bauchut E, Cordier JF, Salles G, et al. Primary Cardiac Lymphoma: Diagnosis, Treatment and Outcome in a Modern Series. Hematol Oncol. 2017;35(4):510-9. doi: 10.1002/hon.2301.
5. Sarjeant JM, Butany J, Cusimano RJ. Cancer of the Heart: Epidemiology and Management of Primary Tumors and Metastases. Am J Cardiovasc Drugs. 2003;3(6):407-21. doi: 10.2165/00129784-200303060-00004.
6. Butany J, Nair V, Naseemuddin A, Nair GM, Catton C, Yau T. Cardiac Tumours: Diagnosis and Management. Lancet Oncol. 2005;6(4):219-28. doi: 10.1016/S1470-2045(05)70093-0.
7. Bruce CJ. Cardiac Tumours: Diagnosis and Management. Heart. 2011;97(2):151-60. doi: 10.1136/hrt.2009.186320.
8. Biersmith MA, Tong MS, Guha A, Simonetti OP, Addison D. Multimodality Cardiac Imaging in the Era of Emerging Cancer Therapies. J Am Heart Assoc. 2020;9(2):e013755. doi: 10.1161/JAHA.119.013755.
9. Zaragoza-Macias E, Chen MA, Gill EA. Real Time Three-Dimensional Echocardiography Evaluation of Intracardiac Masses. Echocardiography. 2012;29(2):207-19. doi: 10.1111/j.1540-8175.2011.01627.x.
10. Motwani M, Kidambi A, Herzog BA, Uddin A, Greenwood JP, Plein S. MR Imaging of Cardiac Tumors and Masses: A Review of Methods and Clinical Applications. Radiology. 2013;268(1):26-43. doi: 10.1148/radiol.13121239.
11. Jeong D, Patel A, Francois CJ, Gage KL, Fradley MG. Cardiac Magnetic Resonance Imaging in Oncology. Cancer Control. 2017;24(2):147-60. doi: 10.1177/107327481702400207.
12. Esposito A, De Cobelli F, Ironi G, Marra P, Canu T, Mellone R, et al. CMR in the Assessment of Cardiac Masses: Primary Malignant Tumors. JACC Cardiovasc Imaging. 2014;7(10):1057-61. doi: 10.1016/j.jcmg.2014.08.002.