EJCRIM
European Journal
of Case Reports
in Internal Medicine

SUCCESSFUL CHEMOTHERAPY TREATMENT FOR A TRICUSPID VALVULAR STENOSIS PATIENT DUE TO RIGHT VENTRICULAR LYMPHOMA

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Received: 09/03/2024 **Accepted:** 12/03/2024 **Published:** 25/03/2024

Conflicts of Interests: The Authors declare that there are no competing interests.
Patient Consent: This study was approved by the Institutional Review Board of 108 Central Military Hospital. The patient has given consent to participate as well as consent to publish the data.
Acknowledgements: The authors would like to thank the patient for providing data.

How to cite this article: Chien DV, Quy TQ. Successful chemotherapy treatment for a tricuspid valvular stenosis patient due to right ventricular lymphoma. *EJCRIM* 2024;**11**:doi:10.12890/2024_004451.

ABSTRACT

Background: This study presents a patient diagnosed with tricuspid valvular stenosis due to right ventricular lymphoma, who was treated successfully.

Case presentation: A 66-year-old man with a history of worsening shortness of breath during activity for the last three weeks sought medical attention. The patient later experienced swelling in the extremities, fluid build-up around the lungs and abdominal fluid accumulation, with no reported chest pain, fever, or weight loss. An echocardiogram found a mass in the lateral wall near the tricuspid valve of the right ventricle, leading to moderate tricuspid stenosis. The cardiac magnetic resonance imaging (MRI) revealed a lumpy, poorly defined mass that invaded the heart muscle and displayed varied enhancement after contrast administration. Suspicion arose for a malignant tumour or metastatic lesion due to its features and contrast uptake capability. A percutaneous biopsy was carried out on the mass in the right ventricle to confirm the diagnosis. The pathology report indicated a diagnosis of non-Hodgkin's lymphoma. After being diagnosed, the patient underwent chemotherapy using the R-CHOP regimen. Over time the symptoms improved, and echocardiograms revealed a decrease in the size of the tumour. After undergoing six rounds of chemotherapy, a cardiac MRI four months later showed no signs of a tumour. After that, the patient resumed their regular activities.

Conclusion: Right ventricular tumours are mostly malignant lesions and often have an inferior prognosis. Early diagnosis with imaging techniques and myocardial biopsy is necessary to deliver life-saving treatment quickly.

KEYWORDS

Tricuspid valvular stenosis, right ventricular lymphoma, chemotherapy

LEARNING POINTS

- Right ventricular tumours are mostly malignant lesions and often have an inferior prognosis.
- Early diagnosis with imaging techniques and myocardial biopsy is necessary to deliver life-saving treatment quickly.
- Right ventricular lymphoma is a sporadic tumour, but the prognosis is favourable if detected in time.

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INTRODUCTION

Cardiac tumours, both primary and metastatic, are infrequent with the occurrence of primary cardiac tumours estimated to range from 0.001% to 0.3% based on autopsy findings[1]. The majority of primary tumours of the heart are benign, with malignant cardiac tumours comprising only 10% of all primary cardiac neoplasms. Prior research has indicated that intracardiac masses are predominantly located in the left atrium and are infrequently observed in the right ventricle[2]. A thrombus represents the predominant right cardiac mass, with metastatic lesions following as the next most prevalent type[3].

Primary cardiac lymphoma is a cancerous lymphoma in the heart or the surrounding pericardium. This condition could also include tumour tissue from lymphoma in the heart or the surrounding pericardium, or infiltration of lymphoma into the heart muscle, resulting in cardiac symptoms when first diagnosed[4]. Primary cardiac lymphomas are rare and often found as isolated cases, making them challenging to diagnose. Thus, due to the delayed diagnosis, primary cardiac lymphoma has a dismal outlook, with an average survival time of seven months after diagnosis[5,6].

Primary cardiac lymphoma presents with cardiac symptoms resulting from the infiltration of the myocardium by the lymphoma. Other indicators may be noted, such as enlargement of the mediastinal lymph nodes, presence of pleural exudate and occurrence of pulmonary embolism[7]. Patients with atrioventricular block as their sole presenting symptom are often at risk for misdiagnosis of primary cardiac lymphoma[8]. This report presents a patient diagnosed with tricuspid valvular stenosis due to ventricular lymphoma, who was treated successfully.

CASE DESCRIPTION

The patient was a 66-year-old man experiencing increasing shortness of breath during physical activity for the previous three weeks. The patient had no prior record of cardiovascular issues. Roughly four months prior, the patient underwent a regular medical examination and received a normal echocardiogram result. Three days before being admitted, the patient's condition deteriorated. He was experiencing difficulty breathing while lying down, along with swelling in the lower extremities, abdominal bloating, extreme fatigue and decreased appetite. The patient had no fever, chest pain or recent weight loss. The physical examination showed widespread congestion with increased jugular venous pressure, fluid build-up in the chest cavity, an enlarged liver and abdominal swelling, as well as a slight heart murmur in the left fourth intercostal space. No swollen or enlarged lymph nodes were detected.

The patient's EKG revealed a sinus rhythm of 100 beats per minute and low QRS voltage in the limb leads. The chest X-ray showed a slight fluid build-up on both sides of the lungs without any other signs of abnormalities in the lung area. The patient received a transthoracic echocardiogram (TTE) as a component of a dyspnoea evaluation. The findings revealed

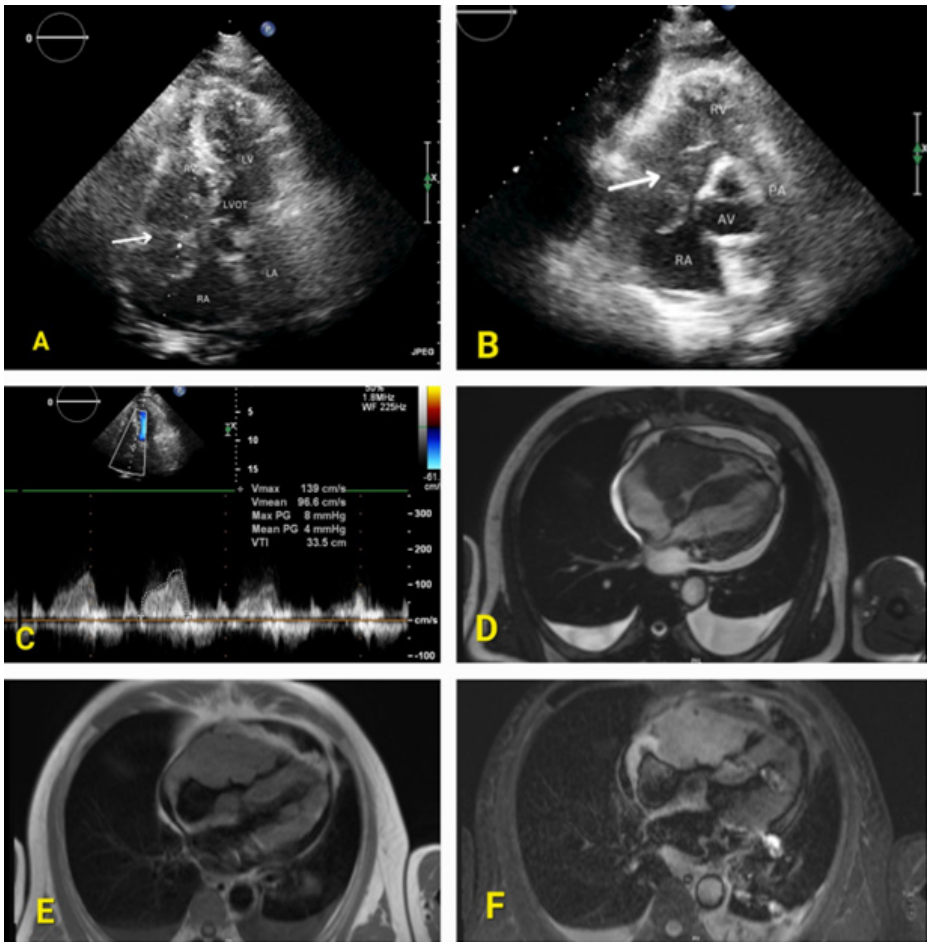

Figure 1. A) RV mass is noted on the five-chamber view of TTE. B) RV mass grew into a tricuspid valve orifice on the parasternal short axis view on TTE. C) Tricuspid transvalvular mean pressure gradient was 4 mmHg. D–F) Cardiac MRI showed RV mass that has a normal signal on the T2-weighted and T1-weighted images and a high signal on the STIR.

Abbreviations: RV, right ventricle; LV, left ventricle; RA, right atrium; LA, left atrium; AV, aortic valve; PA, pulmonary artery; LVOT, left ventricle outflow tract; STIR, short tau inversion recovery.

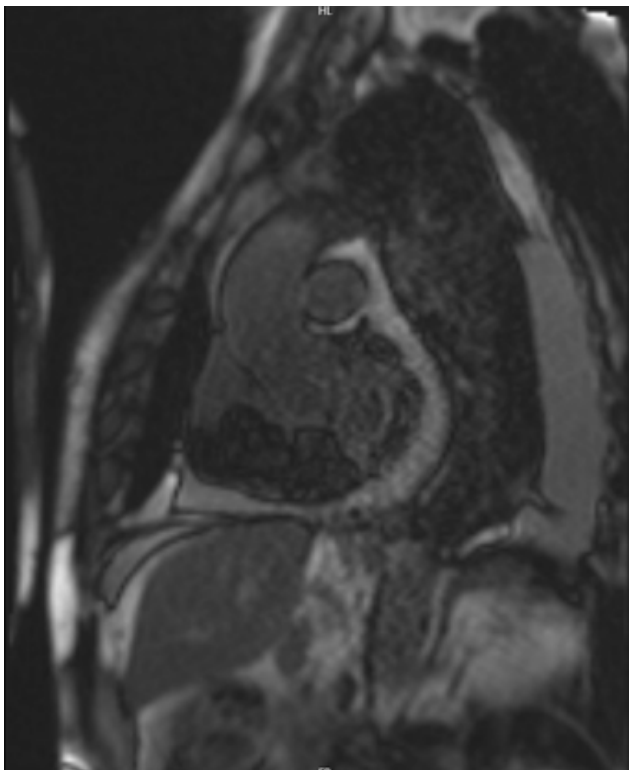

Figure 2. Late gadolinium enhancement of the RV mass on cardiac MRI.

a varied, immobile mass attached to the right ventricle side wall, measuring 69 × 40 mm, with an uneven edge *(Fig. 1A and 1B)*. This mass increased in size and developed into the orifice of the tricuspid valve, leading to a mild tricuspid stenosis, resulting in a transvalvular gradient of 4 mmHg *(Fig. 1C)*. The functioning of the left ventricle was normal, and there was minimal fluid around the pericardium.

Magnetic resonance imaging (MRI) was employed to thoroughly evaluate the attributes of the cardiac mass and its invasion of neighbouring structures, while differentiating between a neoplasm and an intracavitary mural thrombus by administering a contrast agent. The findings indicated the presence of a lobular, poorly demarcated mass that was infiltrating the myocardium. The mass exhibited typical signal characteristics on the T1-weighted and T2-weighted images and appeared hyperintense on the short tau inversion recovery (STIR) images *(Fig. 1D, 1E and 1F)*.

Following the intravenous administration of gadolinium, the MRI images revealed a diverse enhancement pattern within the right ventricular mass *(Fig. 2)*.

The presence of distinct features and contrasting uptake patterns has led to the suspicion of a potentially malignant tumour or metastatic lesion. To establish a conclusive diagnosis, a percutaneous biopsy of the right ventricular mass was executed. Five tumour specimens were collected and forwarded to the pathology department for examination. The pathological investigation yielded a diagnosis of non-Hodgkin's lymphoma *(Fig. 3A–D)*. After the diagnosis, the patient underwent a course of chemotherapy utilising the R-CHOP regimen.

The symptoms showed gradual improvement, and subsequent echocardiographic evaluations demonstrated a reduction in the tumour size, indicating remission. After a period of four months and six cycles of chemotherapy, no evidence of a cancer was detected on TTE and cardiac MRI *(Fig. 4)*. The patient successfully reintegrated into their daily activities and routines.

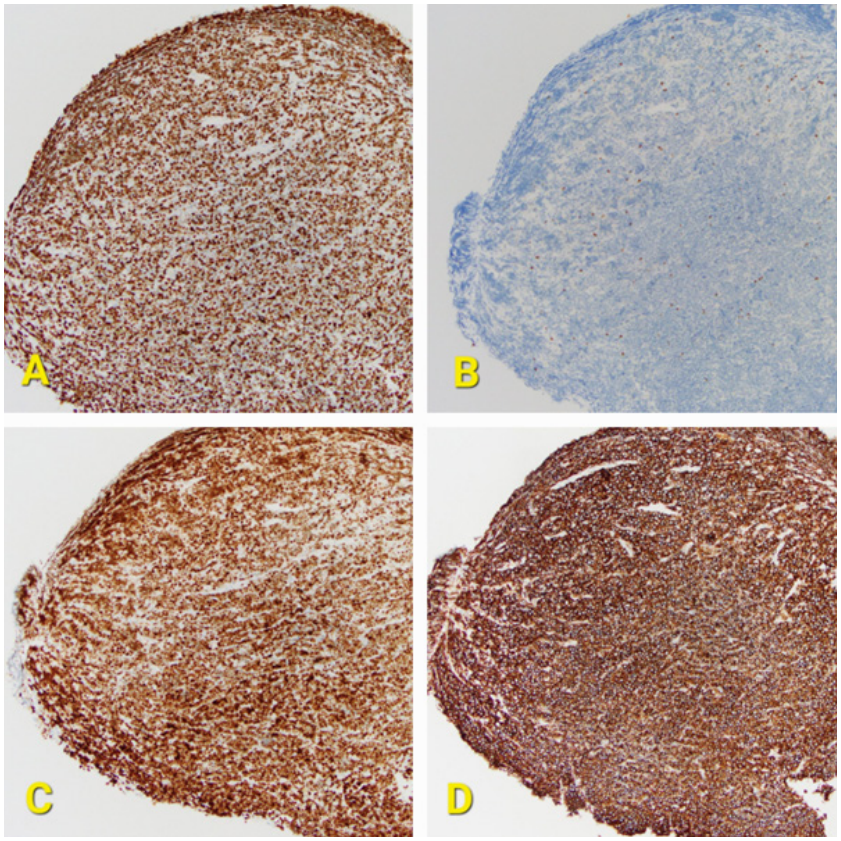

Figure 3. A) Immunohistochemistry shows high percentage of Ki-67-positive cells. B–D) Immunohistochemistry shows the tumour's cells are negative for CD3 and positive for MUM1, CD20.

DISCUSSION

The most interesting finding in this case is that this patient was masked by symptoms of triscupid valve stenosis due to right ventricular lymphoma, and successfully treated with chemotherapy. The cardiac biopsy plays a key role to define the pathology of underlying causes.

Primary cardiac lymphoma is a markedly uncommon condition. This pathology typically affects the right atrium, right ventricle, atrial septum, and inferior vena cava. An anomalous mass located within the right ventricle is commonly attributed to a thrombus or metastatic lesion rather than being diagnosed as a primary cardiac tumour. The prevalence of non-Hodgkin's lymphoma occurring in the right ventricle is exceedingly uncommon, representing one of the most infrequent forms of cardiac tumours, accounting for less than 1% of all primary cardiac tumours[9]. The neoplasm is typically identified in the right cardiac chambers (both atrium and ventricle), although rare occurrences in the left ventricle have also been documented[10,11].

The diagnosis of primary cardiac lymphoma poses a challenge due to the presence of non-specific symptoms. The manifestation of symptoms is often attributed to the presence of tumours that impede cardiac blood flow, invade neighbouring tissues or cause tumour embolisation. The patient was admitted to the hospital presenting symptoms consistent with right heart failure attributed to tricuspid stenosis, including dyspnoea, pleural effusion, hepatomegaly and oedema, without the presence of B symptoms such as fever, night sweats and weight loss. There was no evidence of adenopathy or tumours identified in any other organ of the patient. Furthermore, it is essential to consider that in conjunction with the manifestations of heart failure exhibited by the patient, cardiac lymphoma has the potential to induce arrhythmias, intracardiac conduction blockage, pericardial effusion or stroke[12,13].

The wide range of varied and non-specific symptoms necessitates a comprehensive examination of patients with suspected cardiac lymphoma. Furthermore, it is recommended that more advanced imaging techniques, such as echocardiography, heart computed tomography or MRI, be employed alongside the physical examination. TTE serves as the principal diagnostic modality for identifying anomalous cardiac masses and is widely used in clinical practice.

Transoesophageal echocardiography (TEE) is known for its superior image quality and potential utility in transvenous biopsy procedures[14,15]. CT scans and MRI have commonly utilised imaging modalities for distinguishing between intracardiac thrombus and tumours in clinical practice. While both methods exhibit superior contrast resolution compared to echocardiography and offer a precise diagnosis for particular types of tumours, the conclusive diagnosis of lymphoma can only be established following a histological assessment of the tumour[16]. Although TEE can be utilised to obtain specimens[15], in this case the patient underwent a successful endomyocardial biopsy combining fluoroscopic imaging and TTE due to the elevated risk of respiratory failure associated with TEE.

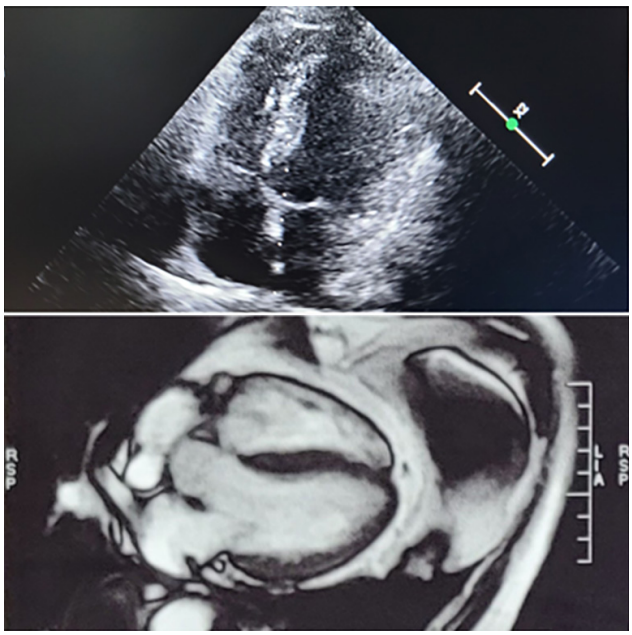

Figure 4. TTE and cardiac MRI images showed no sign of tumour after six cycles of chemotherapy.

The primary therapeutic approach for primary cardiac lymphoma consists of chemotherapy, associated with a relatively high rate of complete response, reaching up to 59%[10]. In specific clinical scenarios surgical intervention may be warranted, such as when a tumour exerts a sizeable presence, leading to partial or complete obstruction of the right ventricular outflow tract. Nonetheless, it is essential to note that this approach does not significantly impact the overall prognosis of the patient[11].

To conclude, this report delineates an uncommon occurrence of cardiac lymphoma, which was proficiently managed and resulted in a complete remission. The majority of right ventricular tumours are malignant in nature, and their prognosis is typically unfavourable. Timely identification through imaging techniques and myocardial biopsy is essential to promptly administer life-saving interventions. Right ventricular lymphoma is an exceedingly uncommon neoplasm; however, the prognosis is favourable if diagnosed promptly.

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