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CHEST

A 70-Year-Old Man With Relapsed CNS Lymphoma Has Incidental Finding of Right Atrial Mass

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CASE PRESENTATION: A 70-year-old man was admitted to the hospital for planned chemotherapy for recently diagnosed CNS lymphoma. His medical history included follicular lymphoma (achieved remission 1 year prior with chemotherapy) and tonic-clonic seizure 1 month prior to admission, which led to his eventual biopsy-confirmed diagnosis of CNS lymphoma. Physical examination revealed temperature 36.4 °C, heart rate of 60 beats/min, BP of 160/81 mm Hg, and 98% oxygen saturation on room air. Neurologic condition, including mental status examination, was normal. His cardiac examination revealed regular rate and rhythm with normal first and second heart sounds without murmurs, rubs, or gallops. The remainder of the examination was unremarkable. Review of systems noted progressive and intermittent confusion prior to his seizure. He denied any shortness of breath, dyspnea on exertion, orthopnea, lower extremity edema, palpitations, or syncope. Laboratory data were unremarkable.

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Before beginning chemotherapy for his relapsed lymphoma with CNS involvement, he underwent CT scans of the chest, abdomen, and pelvis for evaluation of systemic disease. Chest radiograph revealed a normal cardiac silhouette and no signs of acute cardiopulmonary processes. CT chest scan with contrast demonstrated a large hypodense homogenous mass, with no contrast uptake, occupying the right atrium (RA), that measured 3.6 × 2.7 × 3.2 cm (Fig 1). This was an interval change compared with CT imaging 1 month earlier, which was obtained during initial diagnosis of CNS lymphoma. The mass did not extend into the superior vena cava, and there was no evidence of thrombotic or embolic events in the lung. There was no lymphadenopathy or other evidence to suggest malignancy found elsewhere in the imaged chest, abdomen, or pelvis.

Transthoracic echocardiography (TTE) demonstrated preserved left ventricular ejection fraction without wall motion abnormalities, a 3- × 3-cm rounded pedunculated mass in the RA. The mass appeared to be attached to the posterior free wall, near the tricuspid annulus (Video 1). There was no apparent flow obstruction through the RA and no significant tricuspid regurgitation present. The RA was of normal size and function.

Cardiac MRI with contrast demonstrated a mildly mobile mass with irregular and lobulated contour in the RA that was attached to the posterior medial and superior wall of

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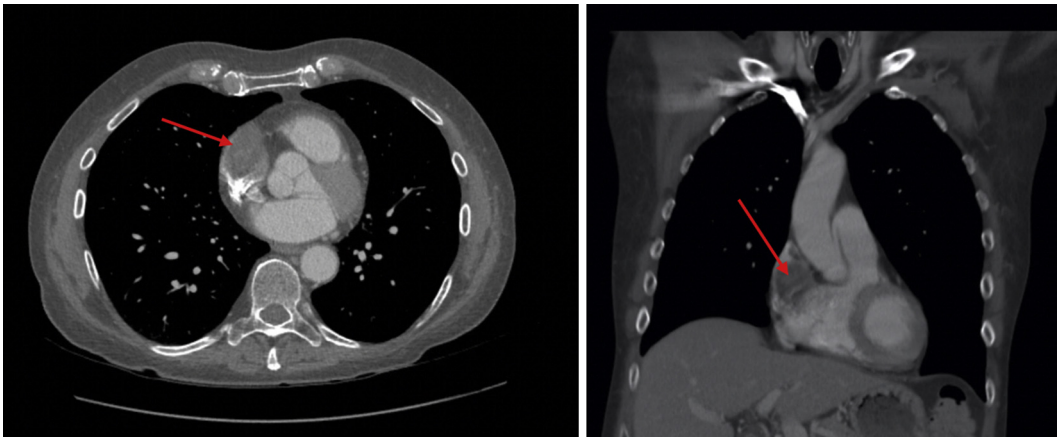

Figure 1 – *Chest CT scan, with axial orientation on the left and coronal on the right, demonstrating a hypodense homogenous mass in the right atrium (red arrows).*

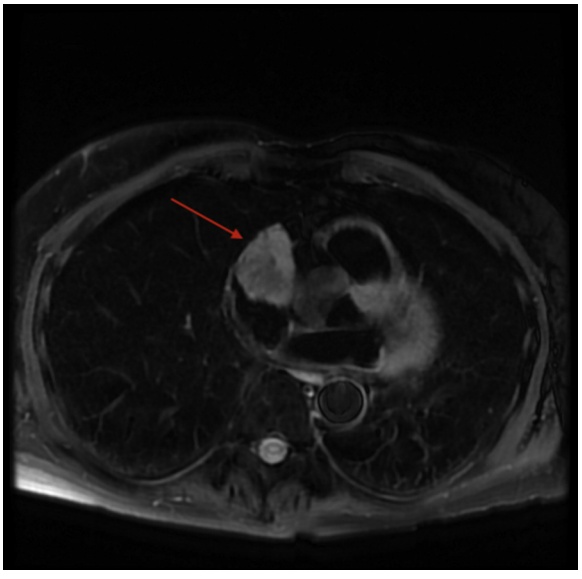

Figure 2 – *Cardiac MRI T1 sequence, in axial orientation, with indeterminate diffuse enhancement (red arrow).*

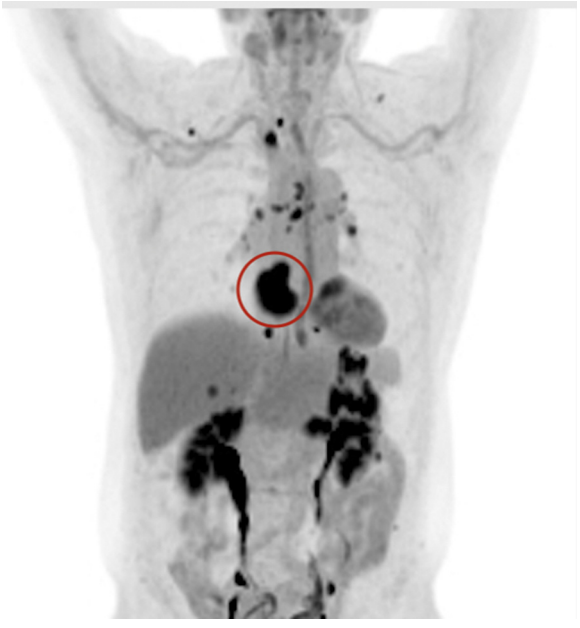

Figure 3 – *PET/CT scan demonstrating avid fluorodeoxyglucose (FDG) uptake in right atrium (red circle), with multiple small areas of uptake in the superior and retrosternal mediastinum, bilateral hilar and pre-carinal nodes.*

the RA (Fig 2). There was no invasion into the adjacent walls visualized. T1 and T2 sequences had indeterminant signal intensity, with mild-to-low central enhancement. On delayed enhancement, there was diffuse enhancement in the central portion of the mass with some peripheral segments not enhancing. PET scanning demonstrated lesions with intense fludeoxyglucose uptake in the brain, mediastinum, and cardiac RA (Fig 3).

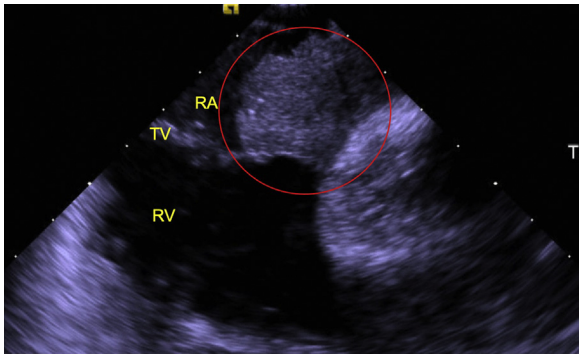

Figure 4 – *Intracardiac echocardiography (ICE) demonstrating the mass (red circle) within the right atrium (RA). RV, right ventricle; TV, tricuspid valve.*

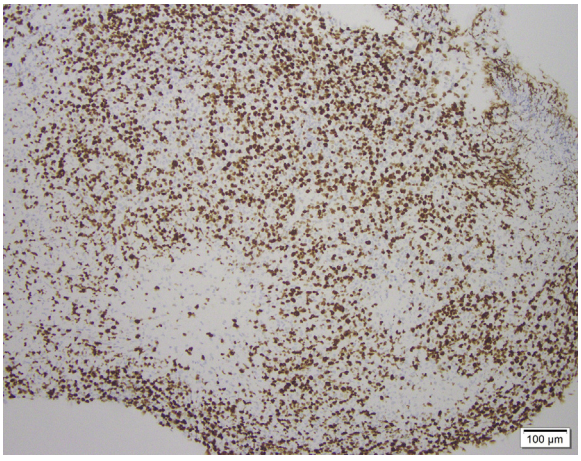

Figure 6 – *Staining for ki67 proliferation index, high at 80-90%, consistent with aggressive lymphoma.*

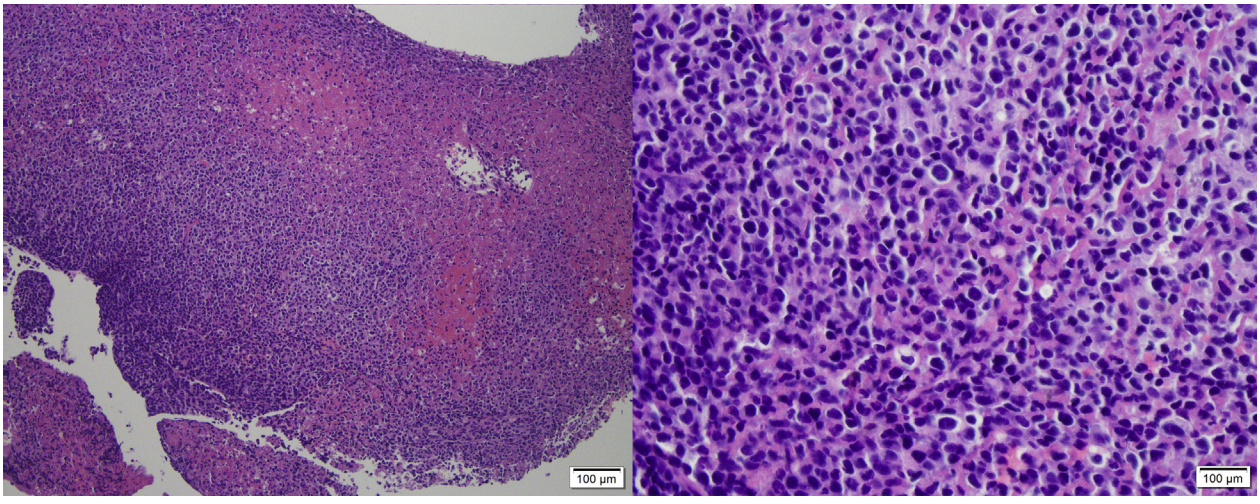

Figure 5 – *Hematoxylin and eosin (H&E) staining at 10× (left) and 40× (right) magnification demonstrating large neoplastic lymphocytes with extensive necrosis and karyorrhectic debris typical of large B-cell lymphoma.*

The mass was biopsied via transvenous access to the RA under intracardiac echocardiography guidance (Fig 4). A limited coronary angiography was performed during the biopsy procedure that demonstrated multiple feeder vessels supplying the mass coming from the right coronary artery (Video 2). Specimens demonstrated several fragments of soft tissue that was infiltrated with large neoplastic lymphocytes, which were positive for CD20, PAX5, CD10, BCL2, and BCL6 (Figs 5, 6).

What is the diagnosis?

Diagnosis: Large B-cell lymphoma with cardiac mass

Discussion

Clinical Discussion

Cardiac lymphoma is a rare cause of cardiac mass, and presentation as a pedunculated mass is very uncommon. Cardiac tumors remain very rare, with an autopsy study finding an incidence of < 1.5%.[1] When cardiac tumors are identified, there is a limited differential, with benign primary cardiac tumors, such as myxoma, being the most common.[2] Secondary malignant tumors of the heart can be from either distant metastasis or local extension, with lung and breast cancer, lymphoma and melanoma being the most common.[3] Cardiac manifestations are seldom the presenting symptom of lymphoma; however, autopsy studies have shown cardiac involvement in up to 20% of patients with terminal disseminated lymphoma.[2] Although primary cardiac lymphoma is much more rare than secondary, both present with similar clinical and radiographic findings.

Cardiac lymphomas, like other cardiac tumors, often present with nonspecific symptoms, such as dyspnea, palpitations, or chest pain. Asymptomatic presentation is not uncommon, with metastatic cardiac tumors identified as an incidental finding in approximately 45% of cases.[4] New arrhythmias can be a common presenting symptom and can include atrial fibrillation, ventricular tachycardia, and even sudden cardiac death.[5] Cardiac lymphomas most often take the form of ill-defined infiltrative masses within the myocardium, often with an associated pericardial effusion, which causes patients to experience heart failure symptoms.[6]

Both primary and secondary cardiac lymphomas have a poor prognosis, attributed to the initial nonspecific symptoms that can lead to delayed diagnosis. Arrhythmia has been shown to be a protective prognostic factor, prompting earlier diagnosis.[7] The mainstay of treatment in both primary and metastatic cardiac lymphoma is chemotherapy, which differs from other benign and malignant cardiac tumors, for which surgical resection often is required. Diagnosis can be made with transesophageal or intracardiac

TABLE 1] Typical Characteristics of Common Cardiac Masses

Mass	Location	Cardiac CT Scanning	MRI
Benign			
Myxoma	Left atrium > right atrium; ventricles	Pedunculated, mobile; low attenuation; may be calcified	Isointense to hypointense on T1; hyperintense on T2; heterogenous enhancement with contrast
Lipoma	Varies	Smooth, encapsulated; no enhancement; fat attenuation	Hyperintense on T1 and T2; reduced intensity with fat suppression technique; no enhancement with contrast
Thrombus	Left atrium, left ventricle	Low attenuation, high sensitivity to rule out thrombus of left atrial appendage	Hypointense on T1 and T2 (hyperintense if recent); no enhancement with contrast
Malignant			
Angiosarcoma	Right atrium > right ventricle; pericardium	Broad base, irregular, heterogeneous; low attenuation; infiltrative; often with pericardial effusion	Heterogenous in T1 and T2; heterogenous contrast enhancement
Metastatic solid cancers	Pericardium (65%-70%) > epicardium (25%-35%); myocardium (30%)	Pericardial thickening or effusion; myocardial thickening or nodularity	Hypointense on T1; hyperintense on T2; heterogenous enhancement with contrast
Lymphoma	Varies	Infiltrating epicardial or myocardial mass; iso to hypoattenuation compared with myocardium	Isointense on T1 and T2; no or variable enhancement with contrast

echocardiography-guided endomyocardial or intracardiac biopsy, avoiding the need for thoracic surgery in these patients.

Imaging Discussion

TTE is the recommended first imaging modality in cases of suspected cardiac masses. Ability of TTE to detect cardiac lymphoma is only 55%; cardiac CT scanning and MRI are positive in 71% and 92% of cases, respectively.[8] The typical imaging findings of common cardiac masses are presented in Table 1.

Although cardiac myxomas often present as discrete pedunculated masses, cardiac lymphomas most commonly affect the myocardium and pericardium, presenting as infiltrative masses with associated pericardial effusion.[7] In the patient, TTE imaging was most consistent with myxoma, which classically presents as a pedunculated mass. The mass's attachment to the posterior free wall, without contact to the nearby tricuspid valve, made thrombus or vegetation less likely. However, thrombus remained on the differential because the malignancy increased the risk of hypercoagulability and interval development of mass over 1 month. Echocardiographic contrast, which was not used in this case, can help differentiate between vascular tumors, which have greater contrast enhancement than myocardium, and nonvascular masses.[9] Lymphomas and other malignant tumors are highly vascular, whereas myxomas have little blood supply and thrombi are avascular.

The findings seen in cardiac MRI again suggested that myxoma was the most likely diagnosis. Lack of restricted diffusion made aggressive metastatic solid cancers and angiosarcoma less likely. MRI results also suggested against lymphoma because of the lack of invasion into adjacent walls, which is often characteristic of cardiac lymphoma. Although both TTE and MRI suggested myxoma, there was still high clinical suspicion for lymphoma, given this patient's active CNS lymphoma. As a result, further imaging was obtained with PET/CT scans. PET/CT scanning with fludeoxyglucose allows for differentiation between benign and malignant tumors via measurement of metabolic activity, which ultimately led to correct diagnosis in this patient. There have been cases in which PET/CT scanning has been instrumental in the identification of malignant cardiac tumors that appear to mimic benign myxoma.[10]

Pathologic Discussion

Biopsy of the cardiac mass was achieved with minimally invasive fluoroscopic and intracardiac echocardiography guidance. The specimen showed several fragments of soft tissue infiltrated with medium-to-large neoplastic lymphocytes with extensive necrosis and karyorrhectic debris in the background. Immunophenotype staining was consistent with B-cell lymphoma, and rearrangement of BCL2/BCL6 with high Ki-67 value suggests that his current large B-cell lymphoma was a transformation from his prior follicular lymphoma. Despite the lymphomatous mass being supplied by branches off the right coronary artery, necrotic segments were evidence of the mass outgrowing its blood supply.

The morphologic condition and immunophenotype of the atrial biopsy specimen were consistent with pathologic evidence that was obtained from a brain biopsy 1 month earlier. The presence of systemic disease in addition to CNS relapse made treatment options significantly more limited. This subgroup of patients is traditionally one with a very poor prognostic status at the time of diagnosis, and treatment goals consist of eventual progression to autologous stem cell transplantation.

Conclusions

This case highlights the challenges of diagnosing cardiac masses, despite the use of multimodal imaging. The imaging findings were atypical of lymphoma and more characteristic of myxoma, requiring PET/CT scanning and ultimately cardiac biopsy to confirm lymphoma as the final diagnosis.

Clinical suspicion was high for lymphoma because the patient's history, which prompted caution in the diagnosis of benign myxoma despite the characteristic imaging findings. Although cardiac imaging can be very useful in the identification of cardiac tumors, biopsy remains the gold standard for diagnosis. Furthermore, because cardiac metastasis can be present in up to 20% of patients with metastatic lymphoma, there are currently no guidelines for dedicated cardiac imaging in patients with lymphoma, despite the poor prognosis of cardiac involvement and need for early detection.

Acknowledgments

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Additional information: Videos for this case are available under "Supplementary Data."

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