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Multimodality Imaging Can Help to Doubt, Diagnose and Follow-Up Cardiac Mass

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ABSTRACT

Primary cardiac lymphoma is a very rare form of lymphoma primarily or mainly involving the heart, as in the two cases presented in this report. Various imaging modalities, including coronary computed tomography angiography, cardiac magnetic resonance imaging and positron emission tomography were useful for the characterization and diagnosis of cardiac mass. Pathologic confirmation was successful with endomyocardial biopsy under echocardiographic guidance, intra- and extracardiacally. In primary cardiac lymphoma, diagnosis using multiple modalities may be useful for mass characterization, and for response monitoring after chemotherapy. (**Korean Circ J 2011;41:555-558**)

KEY WORDS: Lymphoma; Heart, neoplasm; Echocardiography; Magnetic resonance imaging; Positron emission tomography.

Introduction

Primary cardiac lymphoma is a form of non-Hodgkin's lymphoma confined to or mainly located in the heart,¹⁾ which is extremely rare in immunocompetent hosts. Here, we report two cases of primary cardiac lymphoma, upon which coronary computed tomography angiography (CCTA), cardiac magnetic resonance (CMR) imaging and positron emission tomography (PET) were useful to establish diagnosis. The mass was confirmed histologically by endomyocardial biopsy under the guidance transthoracic echocardiography (TTE) and intracardiac echocardiography (ICE).

Cases

Case 1

A 26-year-old, previous healthy man was referred to our emergency room for further treatment of complete atrioventricular block (C-AVB) associated with a one-week history of exertional dyspnea, chest discomfort and dizziness. Twelve-lead electrocardiogram (ECG) showed C-AVB without abnormal findings on chest radiograph. Before implanting permanent cardiac pacemaker, he underwent investigations for reversible causes of C-AVB. Coronary angiography showed intact left and right coronary artery (Fig. 1A). However, CCTA revealed a large infiltrative hypo-attenuated mass involving the atrioventricular (AV) groove. The infiltrative mass circumferentially encased the right coronary artery but did not compromise its lumen (Fig. 1B), which was strongly suggestive of cardiac lymphoma. The mass was demonstrated on TTE to be protruding into the right atrium (RA) and right ventricular cavity (Fig. 1C). A heterogeneously enhancing infiltrative mass involving the myo- and epicardium of both ventricular inferior wall along the AV groove was seen in CMR images (Fig. 2A). Transvenous endomyocardial biopsy (EMB) was performed under TTE guidance (Supplementary Figure Online A). Histology showed many mononuclear cells infiltrating the myocardium positive for CD3, CD4, negative for CD20 and highly positive for Ki-67, confirming the diagno-

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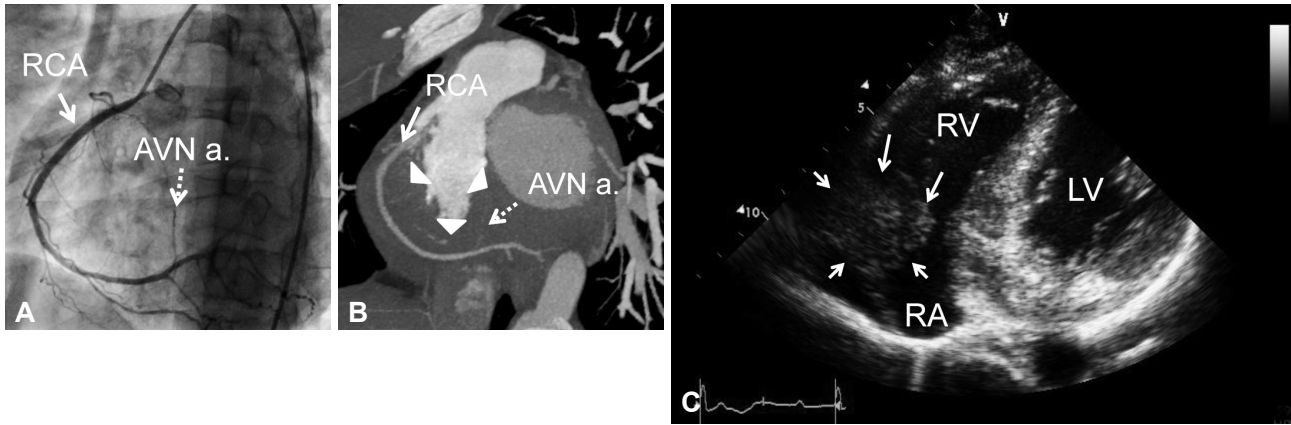


Fig. 1. Angiographic and transthoracic echocardiographic findings of cardiac lymphoma at presentation in case 1. A: coronary angiography shows intact right coronary artery (arrows) and AV nodal artery (dotted arrows) without significant stenosis. B: coronary CT angiography shows preservation of the right coronary artery lumen in spite of the mass at the posterior surface and AV groove of the heart (arrowheads). The AV nodal artery was not compromised in both images. C: modified three chamber view by transthoracic echocardiography shows a mass protruding into the right atrium and right ventricle cavity (arrows), which was the target for endomyocardial biopsy. RCA: right coronary artery, AVN a.: atrioventricular nodal artery, RA: right atrium, RV: right ventricle, LV: left ventricle.

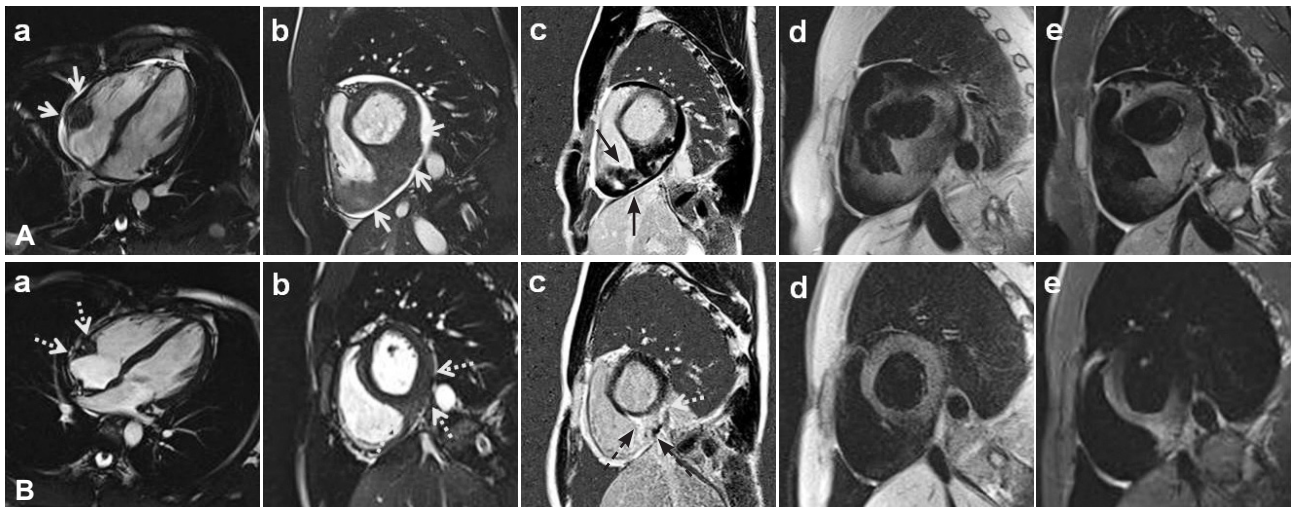


Fig. 2. Cardiac magnetic resonance (CMR) findings of the cardiac lymphoma at presentation (A, upper row) and at follow-up after one cycle of chemotherapy (B, lower row) in case 1. A: iso to slightly high signal intensity mass (arrows) infiltrating the myocardium at the AV groove by steady state free precession (SSFP) image of cardiac MRI (Aa and Ab). Ten-minute delayed MR images with phase-sensitive inversion recovery technique after administration of gadopentetate dimeglumine demonstrated the infiltrative mass (arrows) clearly with heterogeneous enhancement (Ac). B: the mass (dotted arrows) size was markedly reduced after one cycle of chemotherapy in both SSFP (Ba and Bb) and delayed enhancement images with phase-sensitive inversion recovery (Bc). T1 (Ad and Bd) and T2 (Ae and Be) weight images also demonstrate significantly decreased intracardiac mass after chemotherapy.

sis of T lymphoblastic lymphoma (T-LBL). Whole-body PET showed hypermetabolic lesions at the cardiac base (Supplementary Figure Online Ba). Chemotherapy comprised of cyclophosphamide, doxorubicin, vincristine, L-asparaginase (Stanford regimen) commenced after the diagnosis was confirmed, and appropriate staging procedures were conducted without delay, as the patient did not undergo sternotomy for pathologic diagnosis. During chemotherapy, the patient's heart rate was maintained using isoproterenol continuous infusion (1-2 $\mu\text{g}/\text{min}$). Follow-up CMR imaging, including steady-state free precession, delayed enhanced MR (Fig. 2B) and whole-body PET imaging (Supplementary Figure Online Bb) confirmed near complete remission of C-AVB after one cycle of chemotherapy. The patient has been in complete re-

mission for 6 months.

Case 2

A 54-year-old, previous healthy woman was referred to our emergency room with persistent nausea and chest discomfort. She had experienced 8 kg of weight loss in the past one year. Her initial systolic blood pressure (SBP) was 60 mmHg and cardiomegaly and pericardial effusion was respectively noted on chest radiograph and bedside echocardiography. Emergency pericardiocentesis was performed under the presumptive diagnosis of cardiac tamponade, and 700 mL of hemopericardium was drained immediately with recovery of SBP to normal soon after. A mass protruding from the posterior surface of the RA into the atrial cavity was subsequently

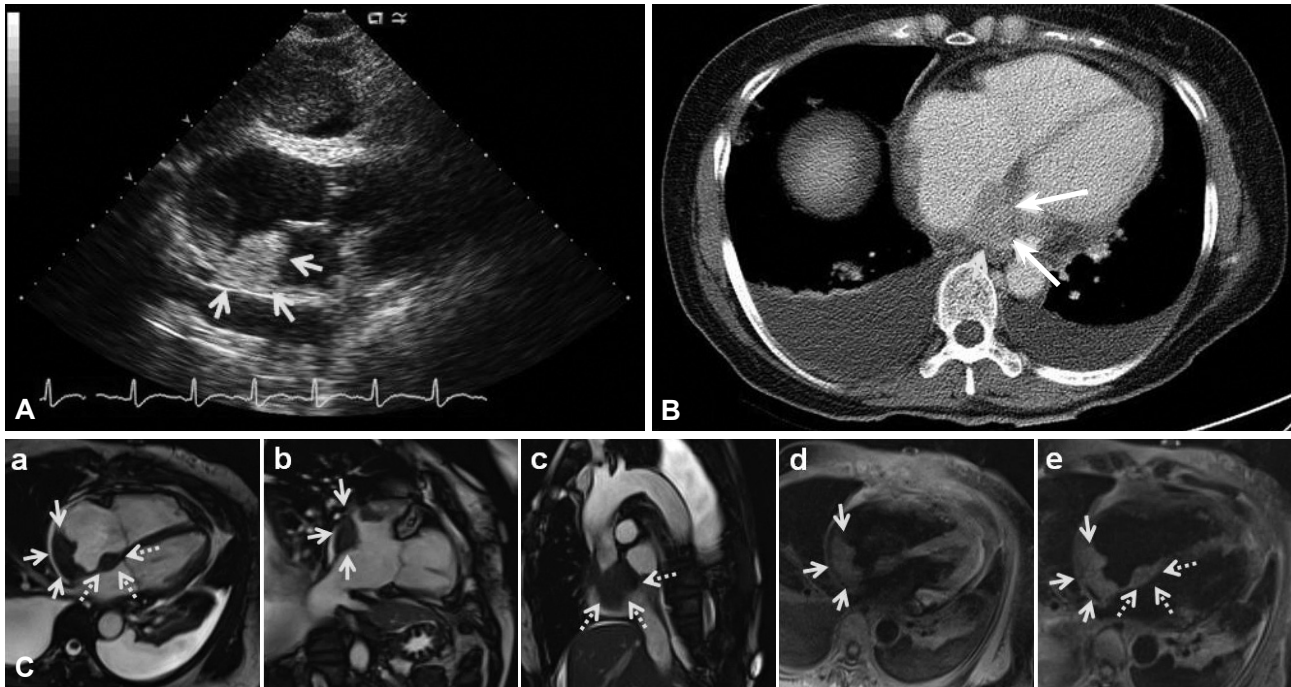


Fig. 3. Transthoracic echocardiography, chest CT and cardiac magnetic resonance findings of the cardiac lymphoma in case 2. A: subcostal view showing a mass in the right atrium (arrows). B: chest CT image demonstrating a homogeneously attenuated mass in the right atrium (arrows). C: mass infiltrating the posterior surface of the right atrium (arrows) and interatrial septum (dotted arrows) is demonstrated in steady state free precession (a, b and c), T1 (d) and T2 (e) images.

demonstrated on TTE (Fig. 3A), and confirmed by chest CT (Fig. 3B). CMR images demonstrated an infiltrative mass involving the posterior surface of RA and interatrial septum (Fig. 3C). Transvenous EMB was performed under ICE guidance (Supplementary Figure Online C). Histological analysis revealed many mononuclear cells infiltrating the myocardium, positive for Igλ and highly positive for Ki-67, but negative for CD3 and CD20, suggestive of diffuse large B cell lymphoma, immunoblastic variant. Whole-body PET showed hypermetabolic lesions at the cardiac base (Supplementary Figure Online Da). Chemotherapy comprised of cyclophosphamide, doxorubicin, vincristine and rituximab commenced after the diagnosis was confirmed, and follow-up whole-body PET images performed 7 weeks later showed reduction of the mass after two cycles (Supplementary Figure Online Db).

Discussion

Primary cardiac lymphoma is an extremely type of lymphoma, the majority of which are of the B cell phenotype,² as seen in the second case. Although there have been case reports and small case series of B cell lymphomas presenting as AV block,^{3,4} the first case is one of the first to demonstrate that T cell lymphomas can also present as such. T-LBL differs from other types of B cell lineage lymphomas with its distinct clinical and biological features, such as high male predominance, and particularly, preponderance of mediastinal involvement and lower chance of complete remission des-

pite adequate chemotherapy.⁵ Therefore our first case is unique in providing an infrequent presentation (C-AVB) of a rare disease entity (T-LBL) occurring at a very unusual site (heart) with favorable outcome, in contrast to two previous case reports of T-LBL in the heart, the results of which were unfavorable in the short-term (<1 month).^{6,7}

Among various diagnostic tools, TTE may be recommended as the initial modality because it readily assesses not only the hemodynamic status, but also the morphological abnormality within the heart. TTE is also useful in guiding the EMB procedure,⁸ which may be superior because it does not delay chemotherapy initiation. Others have also suggested the use of ICE to guide biopsy,⁹ which was the method that we adopted for the second case. Although transvenous biopsy imposes the risk of pulmonary embolism, the mass in our case did not look friable in all imaging modalities (TTE, CT and MRI) and also, firmly infiltrating the myocardium.

In contrast to the characteristic, operator-dependent performance of ultrasonography, CMR may facilitate objective measurement of the mass and provides information on the characteristics of the mass, i.e., vascularity, homogeneity and necrosis. Similar to previous literature,¹⁰ iso-intense but heterogeneous signal and diffusely infiltrating pattern of the mass without evidence of necrosis is a hallmark of cardiac lymphoma. The dramatic reduction of the lymphoma mass in our patient also shows that CMR may be helpful not only for diagnosis but for follow-up of cardiac lymphoma.¹¹ In addition, owing to the diffuse infiltrative pattern of the lympho-

ma, it may be hard to demonstrate whether the lymphoma has genuinely regressed using other modalities (TTE or CT), which are generally poor for soft tissue characterization. Although PET is considered a useful and even a standard modality for lymphoma follow-up recently, the poor spatial resolution of PET, complexity and the dynamicity of the heart structure and the high baseline metabolic rate of the heart renders PET a poor option for follow-up of cardiac lymphoma. In contrast, CMR with the help of late gadolinium enhancement as seen in the first case, may provide useful information to differentiate the mass and the myocardium.

In conclusion, we report the usefulness of various imaging techniques in making a doubt, diagnosis and follow-up of rare primary cardiac lymphoma. As such, multimodality imaging may be helpful not only for proper diagnosis with less invasiveness but also for follow-up in such patients.

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