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CASE REPORT

ADVANCED

CLINICAL CASE

Primary Epicardial Malt Lymphoma

A New Physiopathologic Entity Mimicking a Pericardial Compressive Syndrome

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ABSTRACT

MALT lymphoma is a non-Hodgkin lymphoma developing from B cells and is a type of marginal zone lymphoma. It can develop in any organs, but no case of primary cardiac location has yet been reported. We report the first observation of a primary epicardial MALT lymphoma mimicking a compressive pericardial syndrome. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:1711–1715) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

An 83-year-old man presented with acute dyspnea. Physical examination showed multiple subcutaneous lipomas (**Figure 1A**), edema of the lower extremities, and recent weight gain. Electrocardiogram (ECG) identified atrial fibrillation (**Figure 1B**). Chest x-ray revealed cardiomegaly (**Figure 1C**). Echocardiography disclosed a 4-cm-wide × 10-cm-long solid mass compressing the right atrium (RA) and the right ventricle (RV) (**Figure 1D,** Video 1), which suggested a chronic hematoma or a lipoma because of an encapsulated-like structure. The left ventricular ejection fraction was 45%, with inferior wall hypokinesia. Computed tomography (CT) revealed a prominent epicardial mass, which suggested an organized hematoma or a lipomatous hypertrophy of the epicardium (**Figure 1E**). The result of coronary angiography was normal (**Figures 1F and 1G**).

LEARNING OBJECTIVES

- To report an exceptional case of primary cardiac MALT lymphoma.
- To discuss differential diagnoses.
- To review imaging and immunohistochemical characteristics.
- To discuss the therapeutic management.

MEDICAL HISTORY

Two years earlier, similar symptoms were reported. and the result of magnetic resonance imaging (MRI) gave rise to a suspicion of transmural infarction with either an epicardial hematoma or a lipoma (**Figure 2A,** Video 2). Owing to the lack of late gadolinium enhancement, liposarcoma had been ruled out. Unfortunately, the patient was lost to follow-up.

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ABBREVIATIONS AND ACRONYMS

CT = computed tomography

ECG = electrocardiogram

FDG PET = fluorodeoxyglucose positron emission tomography

MALT = mucosa-associated lymphoid tissue

MCL = mantle cell lymphoma

MRI = magnetic resonance imaging

MZ = marginal zone

RA = right atrium

RV = right ventricle

SCAD = spontaneous coronary artery dissection

DIFFERENTIAL DIAGNOSIS

Considering imaging stability over the previous 2 years with no clinical alteration, cardiac malignancy was initially discarded, as well as a pericardial cyst, on the basis of the echocardiographic and MRI characteristics.

Therefore, several diagnoses were hypothesized:

1. A compressive hematic mass resulting from either of these conditions:
   a. A contained free-wall rupture complicated by either a thrombosed pseudoaneurysm or a chronic hematoma. This cause was assumed by echocardiography and previous MRI showing a moderate hypokinesia of the inferior wall and characteristics of transmural infarction, respectively. Nevertheless, the mass was not consistent with a pseudoaneurysm because the usual echocardiographic shape of a pseudoaneurysm generally has a narrow neck with an abrupt disfiguration of the ventricle (1).
   b. A spontaneous coronary artery dissection (SCAD) with a large epicardial hematoma. However, SCAD is rare, accounts for <1% of myocardial infarctions, and generally occurs in women between the ages of 47 and 53 years (2).
2. A compressive cardiac lipoma, mostly based on echocardiographic findings.

MANAGEMENT

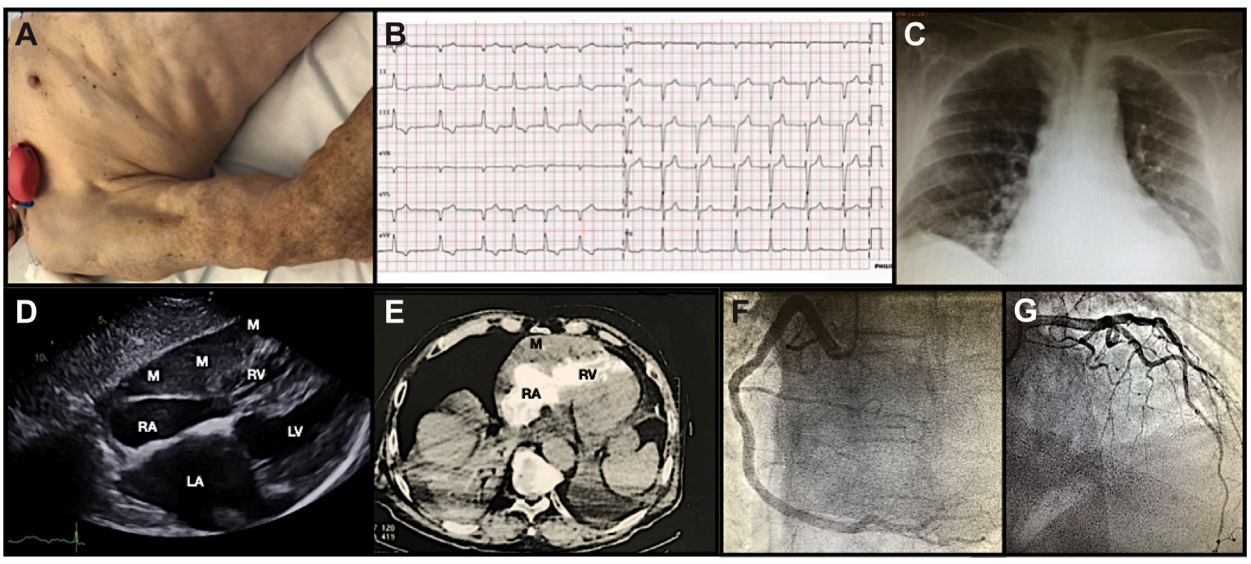
Regarding the size of the mass, a decision for surgery was made to relieve compression of the right cavities. Immediately after anesthetic induction, the systolic arterial blood pressure decreased from 100 mmHg to 70 mmHg, and the diastolic pressure was 55 mmHg. The cardiac index (CI) was 2.4 L/min/m^2, and the pulmonary artery pressure ranged between 65 and 70 mmHg. Therefore, inotropic support with vasopressors was infused.

After median sternotomy and pericardiotomy, instant hemodynamic relief was observed (CI: 3.2 L/min/m^2).

Neither pericardial adherence nor a hematoma was found. Nonetheless, we noticed an unusual thickened salmon-pink epicardial fatty layer of the RV (**Figure 2B**) contrasting with the usual yellow fatty layer of the left ventricle (Video 3) and consistent with a cardiac lipomatosis supported by the presence of multiple subcutaneous lipomas as previously reported (3). Biopsies of the epicardium were made for a comprehensive analysis.

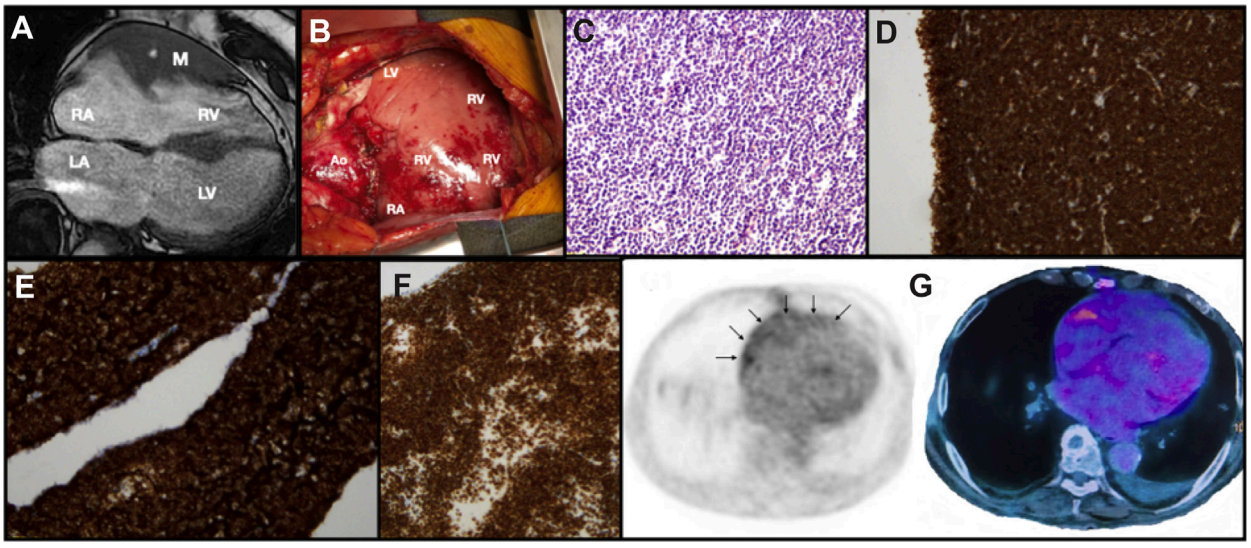
To relieve the risks of intrapericardial crowding with pericardial compression, we performed a pericardiectomy with resection of the anterolateral

FIGURE 1 Preoperative Findings



(A) Subcutaneous lipomas. **(B)** Electrocardiogram shows atrial fibrillation. **(C)** Chest X ray reveals cardiomegaly. **(D)** Echography shows a heterogenous mass (M) compressing the right ventricle (RV) and the right atrium (RA). **(E)** Computed tomography-scan shows a mass compressing the right ventricle. **(F, G)** Normal result of coronary angiography.

FIGURE 2 Operative and Postoperative Findings



(A) Magnetic resonance image of cardiac chambers during diastole. The right ventricular (RV) filling is severely jeopardized by the mass (M), whereas the left ventricular (LV) filling is preserved. **(B)** Operative view showing the RV covered with a thickened salmon-pink epicardial fatty layer with no visible myocardium. The LV displays a normal yellow color of the epicardial fatty layer. **(C)** Hematoxylin and eosin staining (×20) highlights the diffuse lymphoproliferation. The chromatin appears irregular, with patterns evoking lymphoplasmocytic cells. **(D)** Positive staining of IgM antibody suggesting a lymphoplasmocitary differentiation (×20). **(E)** Evidence of B lymphocyte lineage characterized by positive cytoplasmic staining of CD20 antibody (×20). **(F)** Tumoral lymphoproliferation characterized by positive staining of BCL2 antibody (×20). **(G)** Fluorodeoxyglucose positron emission tomographic view at 3 months showing moderate metabolic activity **(black arrows)**. Ao = aorta; LA = left atrium; RA = right atrium.

pericardium between the 2 phrenic nerves and the pericardium from the superior vena cava-RA junction to the inferior vena cava-RA junction.

The patient was weaned from inotropic support in the operative room. His postoperative outcome was uneventful, and he was discharged on postoperative day 10.

PATHOLOGIC RESULTS

Although we expected evidences of cardiac lipomatosis, histopathologic examination revealed a B-cell lymphoproliferation with small monomorphic lymphocytes associated with large immunoblastic-like cells consistent with a marginal zone (MZ) B-cell lymphoma of the mucosa-associated lymphoid tissue (MALT) of epicardium (Figures 2C to 2F).

IMMUNOHISTOCHEMICAL AND CYTOGENIC CHARACTERIZATION

Cytogenic characterization confirmed no BCL2 and BCL6 rearrangement, with a monoclonal population of B lymphocytes detected by κ/λ rearrangement analysis. These patterns suggested a low-grade B-cell lymphoma. Additionally, a diagnosis of follicular lymphoma was discarded by fluorescent in situ hybridization for BCL2 and BCL6.

DISCUSSION

MALT lymphoma belongs to the group of non-Hodgkin lymphomas called MZ lymphomas. MZ lymphomas have several subgroups according to the location where they develop:

1. The nodal MZ lymphoma (10%).
2. The splenic MZ lymphoma (20%) involving the spleen and the bone marrow.
3. The extranodal MZ lymphoma (70%) involving the MALT.

The MZ is an anatomic interface between the nonlymphoid area and the lymphoid area, as described in the spleen and in other lymphoid organs such as palatine tonsils and Peyer plaques. The MZ receives large amounts of blood from the general circulation. Its major role is to trap particulate antigen from the circulation and present the antigen to the lymphocytes.

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The mucosa is the moist tissue that lines some organs and body cavities, including the nose, mouth, lungs, and digestive tract such as the stomach. Hence, MALT lymphoma starts in the body organs, not in the lymph nodes.

MALT lymphoma is a low-grade disease most often diagnosed in the stomach and hence called gastric MALT. This latter form is frequently associated with chronic inflammation as a result of the presence of *Helicobacter pylori* and the action of *H. pylori* virulence factors such as CagA (4). Inasmuch as virtually any mucosal site can be involved, MALT lymphoma can also develop in the lung, thyroid (association with Hashimoto thyroiditis), salivary glands (association with Sjögren syndrome), ocular adnexa (association with *Chlamydia psitaci* or autoimmunity), skin, bladder, breast, bowel (association with *Campylobacter* species), or soft tissues (therefore called non-gastric MALT).

This observation is the first reported case of a primary epicardial MALT lymphoma.

Another case of cardiac MALT lymphoma was recently reported in a 67-year-old woman with a history of MALT lymphoma of the right orbit and treated with surgical resection (5). Nonetheless, that observation is considered a secondary metastatic cardiac location associated with a specific inflammatory condition, not as a primary cardiac MALT lymphoma.

MALT lymphoma may be difficult to distinguish from reactive infiltrates, especially in specific organ locations such as stomach (6), thyroid (Hashimoto thyroiditis) and salivary glands (Sjögren syndrome), so that multiple biopsies are needed to enable a confident diagnosis.

In our case, histologic examination disclosed a B-cell lymphoproliferation with small monomorphic lymphocytes consistent with either a MALT lymphoma, a small lymphocytic lymphoma, or a mantle cell lymphoma (MCL), which is a rare subtype of non-Hodgkin lymphoma.

A confident diagnosis was definitely ascertained with immunohistochemistry that showed expression of CD20, CD79a, BCL2, and IgM with a negative staining for CD5, CD23, and Cyclin D1. By contrast, if a small lymphocytic lymphoma and an MCL are negative for CD5, they can be distinguished with CD23, positive in lymphocytic lymphoma (7) and cyclin D1 positive in MCL (8), respectively. In contrast to small lymphocytic lymphoma and MCL, staining for CD5 is usually negative in MALT lymphoma except in a small proportion of MALT lymphomas, particularly those in the ocular adnexa (9).

Usually, MALT lymphoma is not associated with BCL1, BCL2, or BCL (6,9,10). In our case, these latter characteristics were also confirmed.

MALT lymphoma usually displays an indolent course even when the lymphoma is quite widespread. Several therapeutic options are available, including surgery, radiotherapy, and rituximab alone or in combination with chemotherapy. Additionally, antibiotic therapy may be an effective option in gastric MALT lymphoma because *H. pylori* is known to be frequently associated with MALT (4). Among these options, the watch-and-wait strategy may be justified, subject to regular clinical and imaging follow-up of patients (11).

Because resection of the mass was impossible, resection of an inelastic pericardium was the adequate option to relieve restriction of the RV diastolic expansion.

Additionally, F-18 fluorodeoxyglucose positron emission tomography (FDG PET) can be useful for optimizing diagnosis, posttherapy evaluation, and survival of B-cell MZ lymphoma (12).

FOLLOW-UP

Owing to an indolent evolution, immunohistochemical characteristics, and age, a decision was made for a regular clinical and imaging follow-up without any chemotherapy. Three months after surgery, F-18 FDG PET showed a moderate metabolic activity, which suggested a slow evolution (**Figure 2G**). At the 10-month follow-up visit, the patient was symptomatically much better, with neither echocardiographic compression (Video 4) nor progression of the epicardial mass.

CONCLUSIONS

We report the first case of a primary epicardial MALT lymphoma. This observation highlights the limitations of diagnostic imaging and the importance of pathologic and immunohistochemical characterization. Prognostic and therapeutic strategies need to be further investigated.

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APPENDIX **For supplemental videos, please see the online version of this paper.**