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Case Report

An uncommon cardiac lymphangioma in hypertrophic cardiomyopathy ☆,☆☆

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ABSTRACT

Cardiac lymphangiomas represent one of the rarest types of cardiac tumors. We describe the case of a 28-year-old male with cystic lymphangioma and hypertrophic cardiomyopathy. There have been only a few cases reported and to the best of our knowledge this is the first documented case associating a cardiac lymphangioma with cardiomyopathy.

Introduction

Primary cardiac tumors are exceptionally rare, with an autopsy incidence ranging from 0.001 % to 0.03 %. Among these, cardiac lymphangiomas are even rarer, characterized as benign malformations of the lymphatic system forming mass-like structures. These tumors can originate from various cardiac regions and are often asymptomatic. Symptomatic cases may present with non-specific manifestations such as chest pain, arrhythmias, or signs of right heart failure, depending on the tumor's size and location. Hypertrophic cardiomyopathy (HCM), on the other hand, is a relatively common genetic cardiac disorder. HCM is associated with left ventricular hypertrophy and variable degrees of outflow tract obstruction, arrhythmias, and sudden cardiac death. We report the case of a 28-year-old male presenting with intermittent chest pain and a history of syncopal episodes, who was found to have both obstructive HCM and an intrapericardial cystic lymphangioma. This case highlights the critical role of multimodality imaging in diagnosing and managing rare cardiac tumors and underscores the surgical challenges posed by such tumors. To date, the coexistence of HCM and cardiac lymphangioma

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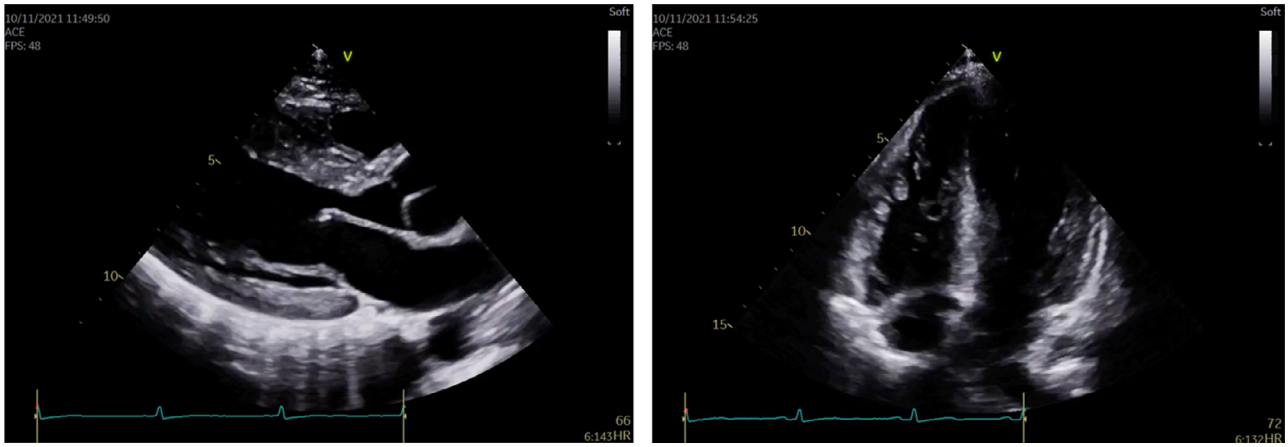

Fig. 1 – Transthoracic echocardiography revealing LV hypertrophy (septal thickness 17 mm) and normal systolic function. Systolic anterior motion of the mitral valve and right ventricle hypertrophy (free wall thickness of 11 mm).

has not been documented in the literature, making this case uniquely significant.

Case report

A previously healthy 28-year-old male presented to the Emergency Department with intermittent chest pain exacerbated by deep inspiration, beginning 12 h prior. He denied dyspnea or palpitations but reported 2 apparent neurocardiogenic syncopal episodes in the previous year. On examination he was hemodynamically stable, with no abnormal findings except for a grade III/VI high-pitched midsystolic murmur heard best at the left lower sternal border. Family history revealed that the patient's mother had hypertrophic myocardiopathy (HCM), although she declined genetic testing or family screening.

The electrocardiogram showed sinus rhythm and was suggestive of an early repolarization pattern. All laboratory results were within normal limits, except for elevated D-dimers (587 ng/mL; normal range: <500 ng/mL). Computed tomography (CT) angiography revealed a space-occupying lesion adjacent to the right border of the ascending aorta, molding the right atrium, extending paracardially, within which the coronary artery trajectory was identified, ruling out pulmonary embolism. Transthoracic echocardiography (Fig. 1) revealed left ventricular (LV) hypertrophy, normal systolic function, and a resting LV outflow tract peak gradient of 59 mmHg which increased to 95 mmHg during the Valsalva maneuver. Systolic anterior motion of the mitral valve and right ventricle hypertrophy were noted. No mass was visible on standard imaging planes. Thoraco-abdominopelvic CT confirmed an intrapericardial lesion encasing the proximal and middle right coronary artery (RCA), molding the right atrium (Fig. 2). No other lesions or adenopathy were found. Cardiac magnetic resonance imaging (MRI) (Fig. 3) showed LV hypertrophy (maximal wall thickness of 26 mm in the basal septum), preserved ejection fraction and diffuse late gadolinium enhancement (LGE) in hypertrophied regions. There was systolic anterior motion of the mitral valve leaflets, apical displacement of the posterior papillary muscle and LV cavity obliteration in the middle segment during systole. It confirmed the presence of an intrapericardial mass (9 × 3-4 cm) (Fig. 4), isointense on T1 and hyperintense on T2 imaging. The mass extended from the aortopulmonary junction along the right side of superior vena cava. It overlapped the anterior and right lateral walls of the aorta, not infiltrating it, with inferior extension along the right side of the right atrium and the right atrioventricular groove, surrounding the proximal and middle RCA. The first-pass perfusion sequence showed an avascular mass with RCA perfusion visible inside it

In the case of obstructive symptomatic HCM and a cardiac mass requiring histological diagnosis and possible complete excision, a surgical approach was taken. A septal myomectomy was performed without intervention on the mitral valve or subvalvular apparatus. Tumor resection was performed with a good cleavage point up to the right atrioventricular groove allowing surgical separation of the upper part of the mass (Fig. 5). Further resection was impeded by the presence of the RCA within the mass with no possibility of dissecting it without damaging the RCA wall. A bypass graft was placed to the distal RCA, considering the possibility of future arterial compression.

Histopathological examination revealed a collection of vessels that stained positive for podoplanin, CD31, and CD34, confirming the lymphatic nature of the mass. The tumor also contained other cellular elements, including adipocytes, smooth muscle cells, and fibrocytes, with no significant collagen deposition. The findings were most consistent with a cystic lymphoma. Histopathological examination of the myectomy specimen revealed features consistent with HCM including myocyte disarray, cardiac sarcomeres with large nuclei, and subendocardial and interstitial fibrosis. The findings were supportive of HCM.

In short-term follow-up, the patient is asymptomatic and further imaging is contemplated at one year of follow-up.

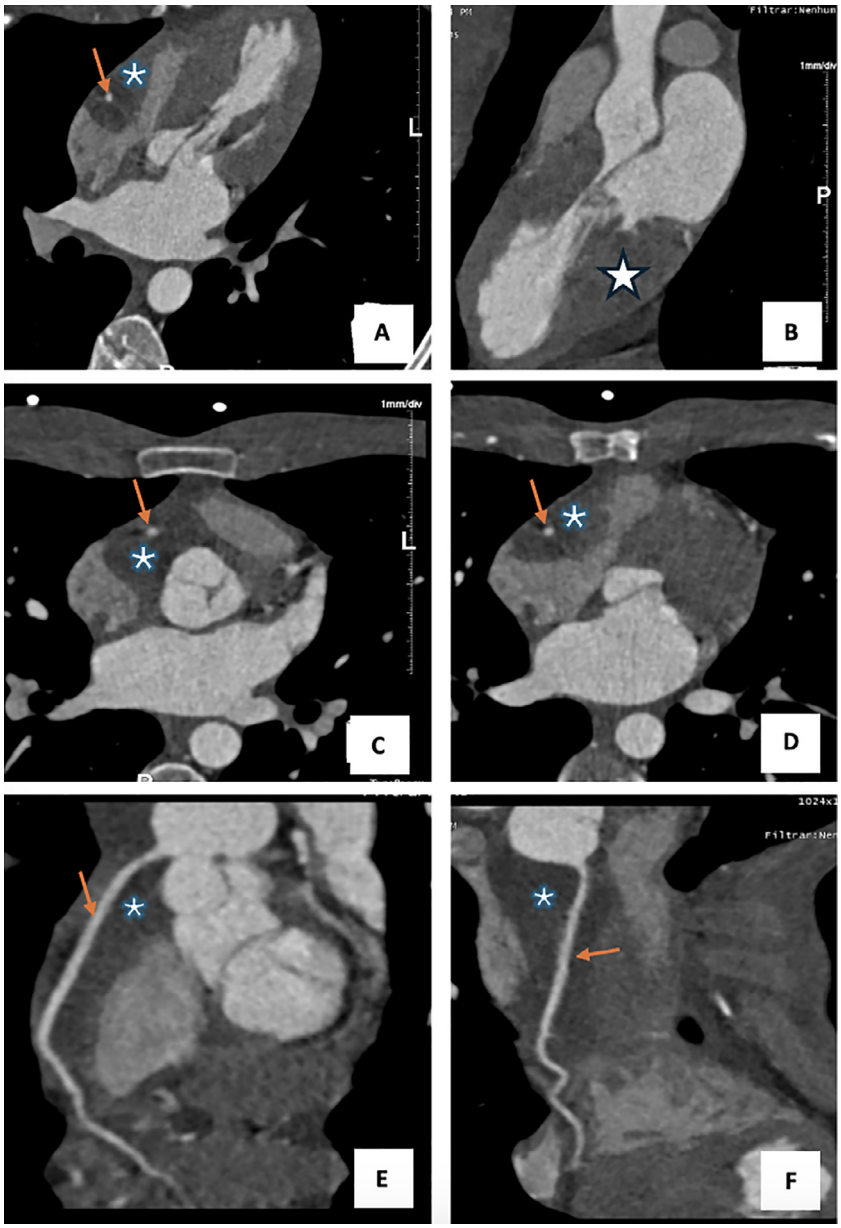

Fig. 2 – Multiplanar reconstructed coronary CT showing a mass (*) and systolic anterior motion of the mitral valve (A and B); intrapericardial lesion (*) with molding effect on the right atrium (C and D), surrounding the RCA (→) (E and F). LV hypertrophy (star).

Discussion

Primary cardiac tumors are exceedingly rare, with an incidence of 0.001%-0.030 %, based on autopsy reports [1]. Among these, lymphangiomas, which are benign masses arising from abnormal collections of lymphatic vessels, are extremely uncommon [2]. Primary cardiac lymphangioma have only been reported in a few cases in the literature [3].

Cardiac tumors cover a wide spectrum of benign and malignant pathologies, including thrombi, myxomas, lipomas, and fibromas, as well as more aggressive entities like angiosarcomas, lymphomas, and metastatic diseases [3,4]. Cardiac MRI plays a critical role in differentiating between these masses. Its ability to provide detailed imaging across multiple sequences allows for a reasonably conclusive diagnosis in certain types of tumors, such as lipomas, fibromas, pericardial cysts, and thrombi, which have characteristic imaging features [4]. Additionally, combining clinical information with imaging findings can help in the identification of other tumors like myxomas, rhabdomyomas, right atrial angiosarcomas, paragangliomas and secondary deposits from primary extracardiac neoplastic disease [4]. The specific location of a cardiac tumor can also provide diagnostic clues. The region around the right atrium and the atrioventricular sulcus is particularly home to right atrial angiosarcomas, primary cardiac lymphoma, paragangliomas and hamartomas, the last of which are abnormal non-neoplastic collections of

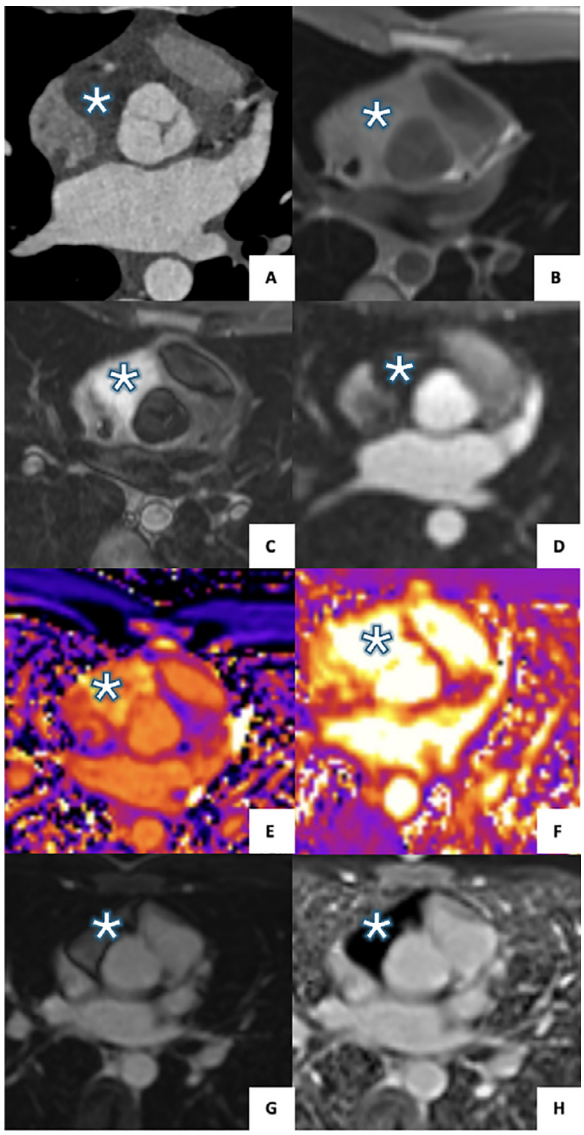

Fig. 3 – Multimodality mass (*) characterization. CT revealed a mass molding the right atrium, with right paracardiac extension, within which the coronary artery path was identified (A). Cardiac MRI showed: isointense mass on T1 spin echo with encasement of the superior vena cava without compression (B); hyperintense signal on T2 STIR, suggesting a watery or liquid content (C); non-vascularized mass on resting perfusion (D); hyperintense signal on T1 (E) and T2 mapping (F), suggestive of edema or a liquid content (F); possible liquid content on magnitude-only inversion recovery (MAG) LGE (G) and confirmed liquid content on phase-sensitive inversion recovery reconstruction (PSIR) LGE (H).

non-proliferating normal tissue. Conversely, thrombi tend to develop within the heart chambers and are frequently associated with underlying structural heart diseases, particularly following myocardial infarctions [4].

In terms of cardiac lymphangiomas, they can arise from different cardiac regions, including the atrial septum, myocardium, atrioventricular node, and even the heart valves [3]. However, literature review showed that the most common anatomic location was the pericardium, most often the visceral layer. Infrequently, lymphangiomas may be found within the myocardial wall of the interatrial septum or ventricles. Despite their potentially diverse origins, cardiac lymphangiomas are often asymptomatic, with non-specific symptoms, such as dyspnea, arrhythmias, angina, signs of right heart failure, pericardial effusions and thromboembolic events, appearing only when the tumor reaches a significant size [3]. Most cardiac lymphangiomas are incidentally discovered during imaging studies [4]. In our case, multimodality imaging, particularly cardiac MRI, played a major role in diagnosing the tumor, delineating its extent, and evaluating its relationship with surrounding structures such as the coronary arteries, which was crucial for surgical planning. On MRI, lymphangiomas typically exhibit high signal intensity on both T1- and T2-weighted images, reflecting their proteinaceous content in the lymph within the stroma and cystic liquid content, respectively. The mass exhibited no contrast enhancement on first-pass perfusion sequences, suggesting an avascular lesion. This finding aligns with the diagnosis of a lymphatic tumor, which is typically low in vascularity. However, it is important to note that lymphatic lesions may exhibit markedly delayed contrast enhancement due to the slow transit of lymphatic fluid through the lesion. This is a known limitation in detecting contrast uptake in lymphatic tumors, where the washout effect from surrounding tissue could further obscure any subtle signal enhancement.

The absence of aggressive imaging features helped rule out more malignant entities like a right atrial angiosarcoma or a primary cardiac lymphoma [4]. Further imaging findings were inconsistent with other potential diagnoses. In the absence of systemic symptoms of a catecholaminergic syndrome, a paraganglioma was unlikely. The tumor characteristics on T2-weighted images, namely the presence of signal on T2-weighted despite fat suppression, excluded lipomas and liposarcomas, whereas the absence of LGE made fibromas unlikely [4,5]. Although extracardiac myxomas involving the right atrium with entrapment of the RCA artery have been described in the literature, they were ultimately not considered a likely diagnosis in this case. Workup for primary extracardiac neoplastic disease was also negative in this patient.

Distinguishing between benign from malignant masses through noninvasive imaging alone is challenging. Although cardiac MRI is highly effective, histopathological examination remains the gold standard for definitive diagnosis [6]. Surgical biopsy, although risky due to the vascular nature of these tumors, is often the safest approach for obtaining tissue samples. In most cases, complete surgical resection of cardiac lymphangiomas is feasible, though the proximity to coronary arteries and the tumor's anatomical location present significant surgical challenges.

While hypertrophic cardiomyopathy is a well-known genetic disorder affecting 1 in 200 to 500 individuals [7], the co-occurrence of HCM with a rare benign tumor like cardiac lymphangioma is unusual. Of note, the histopathological examination of the myectomy specimen was supportive of HCM, particularly in the context of the patient's clinical presentation and positive family history and by the clear presence of asymmetric septal hypertrophy, but it is not definitively

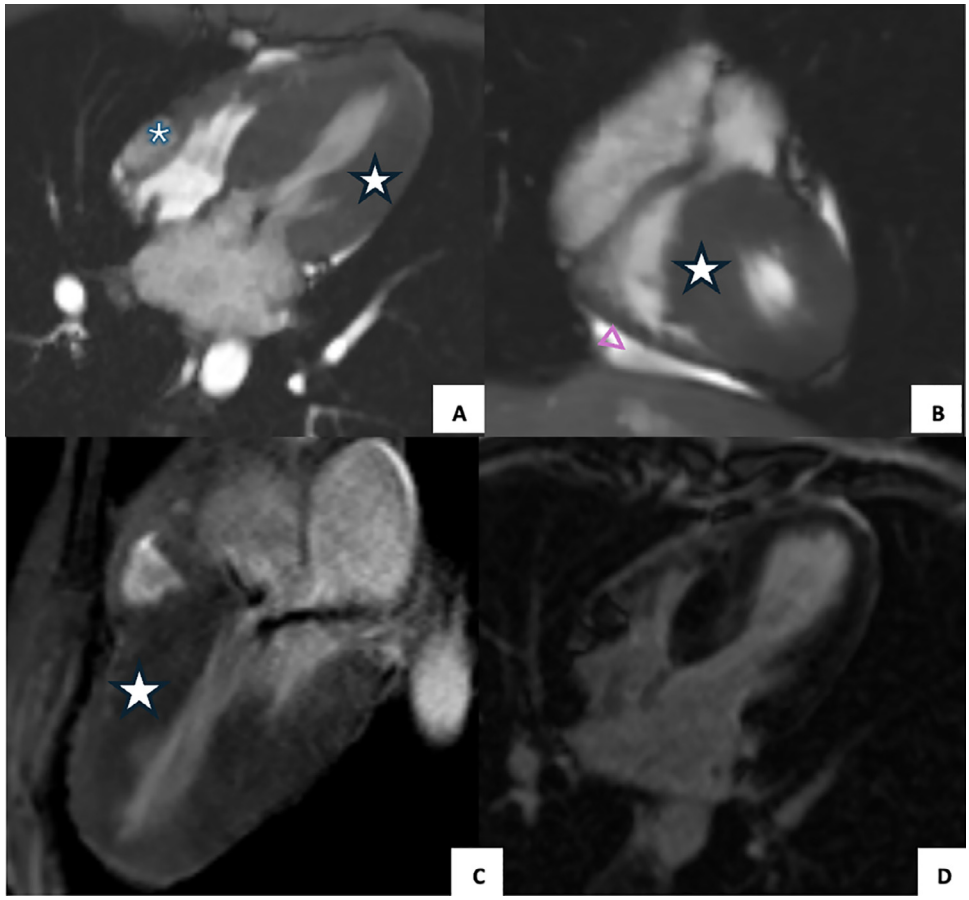

Fig. 4 – Cardiac MRI showing an intrapericardial mass on the right ventricular groove (A, *), mild pericardial effusion (B; triangle), systolic anterior motion (C) and no LGE in the LV (D).

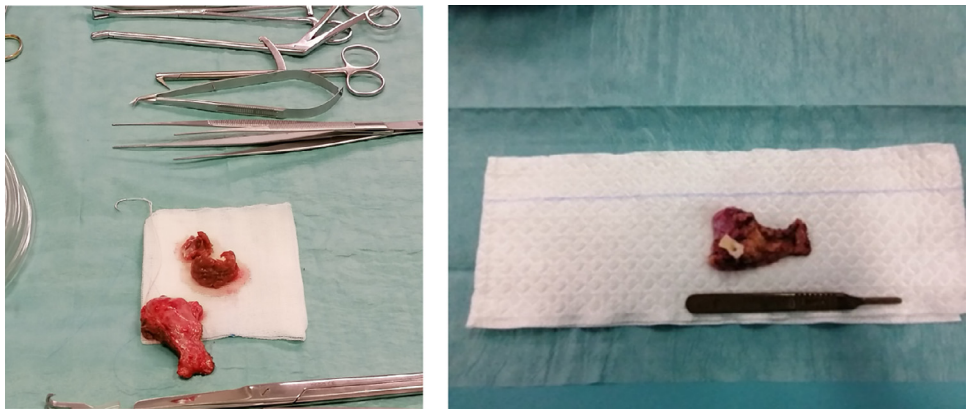

Fig. 5 – Tumor's macroscopic view.

diagnostic, as myocyte disarray found in superficial myectomy sections can also observed in non-HCM conditions.

Our case highlights not only the importance of imaging in diagnosing these rare cardiac masses but also the complexity of surgical management when such tumors are located near critical structures like the coronary arteries.

Author contributions

All authors listed have made substantial contributions to the conception, design, analysis, or interpretation of the case report and have approved the final manuscript.

Patient consent

Informed consent was obtained from the patient for his data and images to be used in this study and published in this journal. The patients understood that their anonymity would be preserved, and no identifiable information would be disclosed.

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