

Contrasting fatty involvement of the right ventricle: lipoma versus lipomatous hypertrophy

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ABSTRACT

We report two rare cases of right ventricular lipomatous involvement, and discuss their contrasting diagnostic images and clinical manifestations. Case 1 is a 21-year-old Malay man who presented with recurrent syncope. Transthoracic echocardiography showed a homogeneous discrete mass attached to the right ventricular septum, occupying the right ventricular outflow tract. Cardiac magnetic resonance (MR) imaging showed features consistent with intracardiac lipoma, which was confirmed on histology, upon surgical resection. Case 2, a 73-year-old Caucasian woman, was found to have incidental lipomatous infiltration of the right ventricle on echocardiography and MR imaging. In contrast to the well-defined right ventricular lipoma seen in the first case, lipomatous involvement was manifested as a diffuse homogeneous thickening of the right ventricular free wall. Multimodality imaging allows for a complementary and detailed characterisation of lipomatous masses involving the ventricular septum.

Keywords: cardiac magnetic resonance imaging, echocardiography, intracardiac mass, lipoma, lipomatous hypertrophy, right ventricular fat

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INTRODUCTION

Fatty deposits around the heart are not uncommon. Asymptomatic epicardial fat may be documented on routine transthoracic echocardiography. Similarly, lipomatous hypertrophy of the interatrial septum has been well described. However, besides the relatively uncommon arrhythmogenic right ventricular dysplasia, other forms of lipomatous involvement of the right ventricle are exceedingly rare. Discrete or more homogeneous infiltrations of the right ventricle may or may not present with symptoms and physical signs, depending on their size and the extent of the involvement.

Various imaging modalities can evaluate and provide information on the anatomy, effects on cardiac physiology and nature of these masses. In turn, clinical decisions are then made, particularly if the resection is indicated, taking into consideration the entire clinical scenario, patients' inclination and available surgical expertise. We describe two cases of right ventricular lipomatous involvement, and discuss their contrasting diagnostic images and clinical manifestations.

CASE REPORTS

Case 1

A 21-year-old Malay man with no significant past medical history, presented with a second episode of syncope, unpreceded by prodrome, and which occurred while walking, within a three-month period. There was an absence of seizure. After regaining consciousness, the physical examination findings were essentially normal. There were no cardiac murmurs, abnormal heart sounds or focal neurological deficits. Laboratory tests showed normal blood count, electrolytes and glucose. Electrocardiogram, chest radiograph and computed tomography (CT) of the brain were also normal. Transthoracic echocardiography showed a large mass measuring 57 mm × 20 mm attached to the right ventricular (RV) septum (Figs. 1a–d), and protruding into and occupying most of the RV outflow tract (OT) (Fig. 1b). The right ventricle was mildly dilated with a normal systolic function. The main pulmonary artery was also mildly dilated, but its systolic pressure (33 mmHg), derived from Doppler interrogation of the tricuspid regurgitation jet, was not elevated. The left chambers and the aorta were normal.

Cardiac magnetic resonance (CMR) imaging showed a well-circumscribed and slightly lobulated mass attached to the RV side of the interventricular septum. It arose from the infundibular septum, 15 mm below the pulmonary valve, and extended into the ventricular apex and the RVOT. As a result, the OT was narrowed. However, there was no evidence of overt obstruction at rest. The CMR imaging showed the content of the mass to be consistent with fat, indicating that it was most likely

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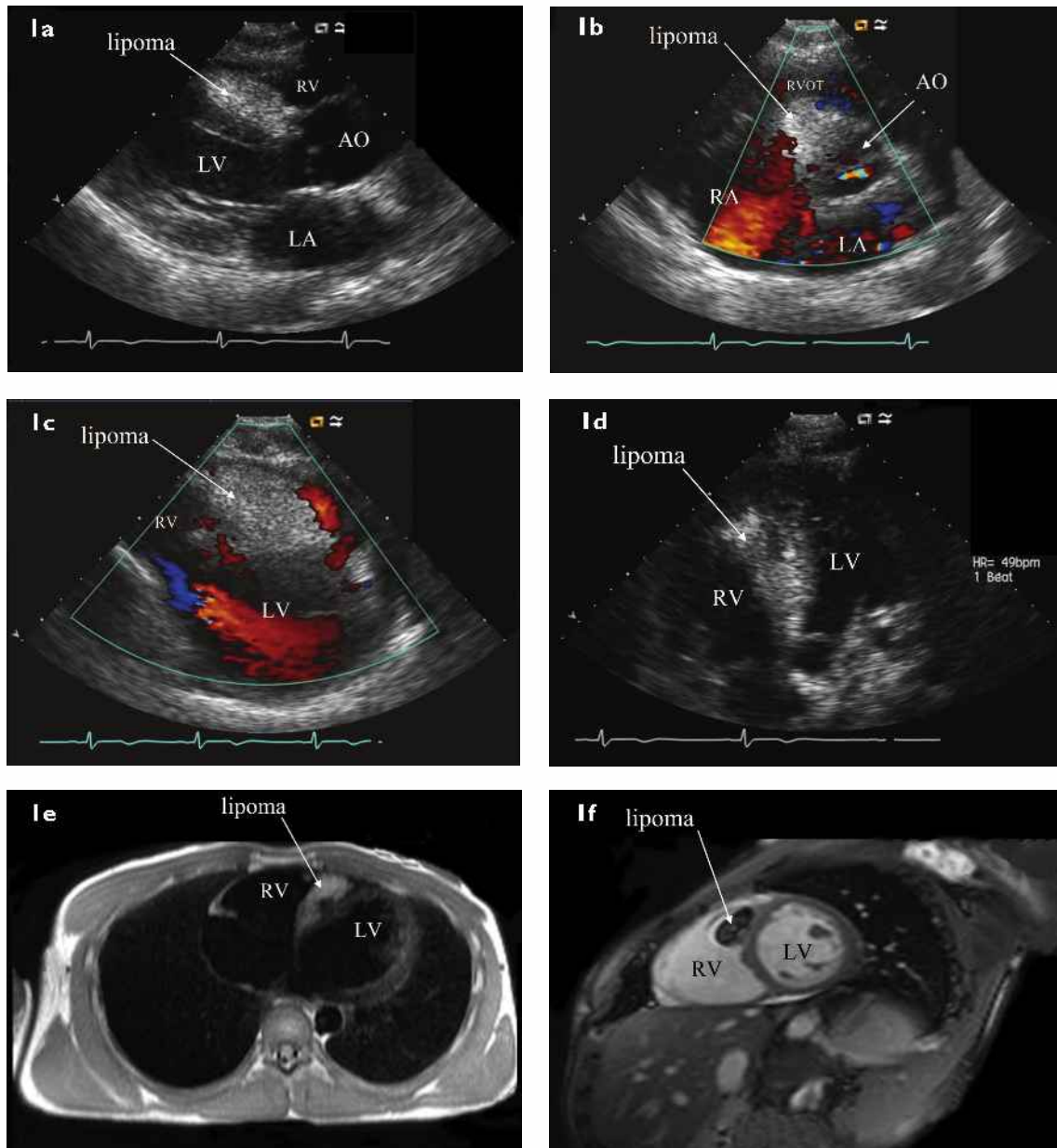


Fig. 1 Transthoracic echocardiography in the (a) parasternal long-axis view shows a right ventricular (RV) lipoma; narrowing of the RV outflow tract (OT), with absence of obstruction on colour Doppler in the (b) short-axis view at the aortic valve level, (c) extent of the RV involvement at the mitral valve level, and (d) extension of lipoma to the RV apex in the apical five-chamber view. Cardiac MR images show the mass in the (e) four-chamber view with high-signal intensity on T1-W spin-echo imaging, and (f) low-signal intensity on fat-suppressed spin-echo images at the short-axis view. AO: aorta; LA: left atrium; LV: left ventricle; RA: right atrium

a lipoma (Figs. 1e & f). Extracardiac lymphadenopathy was not present. Due to recurrent symptoms associated with exertion, the patient underwent a surgical resection. The histology showed a fatty composition of the mass, confirming the diagnosis of intracardiac lipoma. He remained well six months after the resection.

Case 2

A 73-year-old Caucasian woman slipped while attempting to sit on a toilet seat, sustaining a closed fracture of the proximal shaft of her right femur. The

physical examination, inclusive of the cardiovascular system, was otherwise unremarkable. As part of her preoperative assessment, an echocardiography which was performed, showed a marked increase in the thickness and echogenicity of the RV free wall (Figs. 2a & b). There was no other abnormality except for the concomitant presence of lipomatous atrial septal hypertrophy. The CMR imaging showed fat-like features consistent with a RV lipomatous infiltration. As the patient was asymptomatic cardiac-wise and there was no discrete lesion for surgical resection, she was managed conservatively.

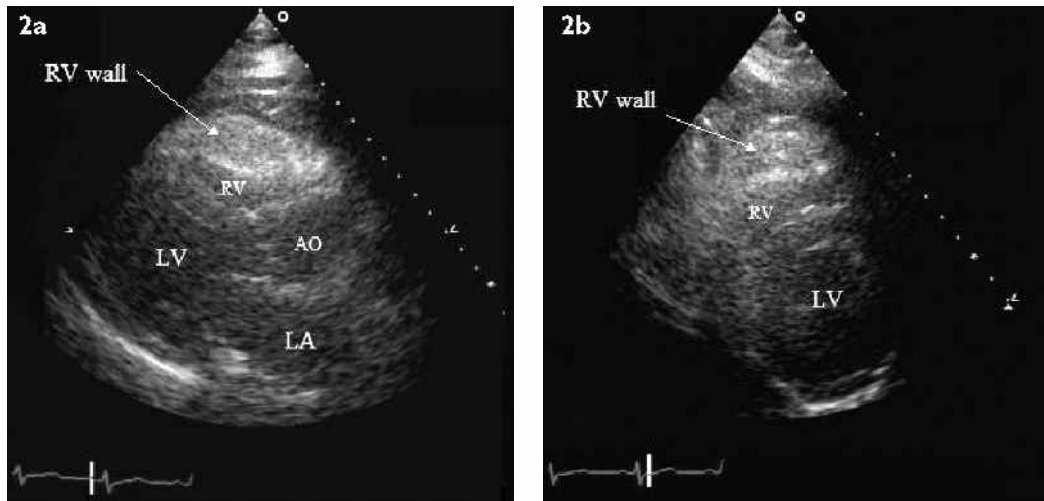


Fig. 2 Transthoracic echocardiography in the (a) parasternal long-axis view, and (b) the short-axis view at the mitral valve level show the lipomatous infiltration of the right ventricular free wall. AO: aorta; LA: left atrium; LV: left ventricle; RV: right ventricular

DISCUSSION

Intracardiac lipomas consist of mature fat cells and are uncommon benign tumours.⁽¹⁾ They may occur at any age with an equal frequency in both genders. They can involve the endocardium, myocardium, pericardium and the emergence of great vessels, or occur within the cardiac chambers. The clinical manifestations of cardiac lipoma depend on its location and the possible resultant compression or obstruction. Those that interfere with cardiac functions, such as a valvular or outflow obstruction, may produce earlier symptoms or audible murmurs. On the other hand, lipomas that involve the epicardium or infiltrate the myocardium may remain asymptomatic, and thus grow to larger dimensions. Lipomas of the right atrium, the interatrial septum and the right ventricle may predispose to arrhythmias or are associated with other pro-arrhythmogenic conditions. For instance, Agadikien et al reported a case of cardiac lipoma in a patient with arrhythmogenic RV dysplasia.⁽²⁾ In both of our cases, arrhythmias associated with the RV lipoma or lipomatous infiltration were not documented. In the first case, exertion-related RV obstruction may have resulted in the syncopal episodes.

Unlike discrete lipoma, lipomatous hypertrophy is not well circumscribed. This may remain asymptomatic for years, as in our second case. Though lipomatous myocardial or intracardiac involvement may result in the appearance of a mass, it does not represent a neoplastic process. Microscopically, the tumour is composed of both mature adipocytes and “brown fat”, similar to a hibernoma. However, sudden death has been attributed to the fatty infiltration of the myocardium.⁽³⁾ More recently, it has been shown that lipomatous infiltration with

increased RV thickness, but without regional or global functional abnormalities of the right ventricle, are distinct from fatty right ventricle associated with arrhythmogenic RV dysplasia.⁽⁴⁾ A more common location for lipomatous hypertrophy is the interatrial septum. The fatty infiltrate is often echogenic and results in a “dumbbell-shaped” appearance on a two-dimensional echocardiography. Prominent crista terminalis may also appear as abnormal masses on echocardiography. Other areas of lipomatous hypertrophy are uncommon, though Izumi et al reported a case of massive lipomatous infiltration to the left ventricle, mimicking a cardiac tumour.⁽⁵⁾

Transthoracic echocardiography is usually the initial diagnostic test for patients with a suspected cardiac mass or myocardial infiltration, due to its portability and availability. The anatomical definition and haemodynamic consequences can often be assessed by this imaging modality. However, its sensitivity for differentiating tissue characteristics is limited. On the other hand, multislice CT allows for a characterisation of the components within the mass. Low-attenuation features with a density similar to fat on CT are pathognomonic for lipoma. Similarly, CMR imaging also shows cardiac lipomas or lipomatous hypertrophy with characteristic low-signal-intensity margins and high-central-signal intensity on “bright-blood” balanced gradient-echo sequences. T1-weighted dark-blood spin-echo sequences demonstrate high-signal intensity within lipomatous masses, and the application of a fat saturation pre-pulse will effectively suppress the signal intensity within the mass, confirming the presence of adipose tissue. In addition, these modalities permit detailed examinations of the intramural components, the degree of extension and the associated pericardial or

extracardiac masses. Gulati et al evaluated the use of these different imaging modalities and found that CMR imaging was advantageous over echocardiography in the detection, and complete morphological and functional assessments of cardiac tumours.⁽⁶⁾ However, transthoracic echocardiography would likely be sufficient and most apt in the serial follow-up of these patients, while transoesophageal echocardiography will be most suitable in providing guidance in the intraoperative resections of cardiac tumours. Multimodality imaging allows for the complementary and comprehensive assessments of lipomatous involvement of the right ventricle.

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