



Palpitations as manifestation of lipomatous hyperplasia interatrial septum: case report and review of the literature

Palpitaciones como manifestación de hiperplasia lipomatosa del tabique interauricular: reporte de caso y revisión de literatura

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Keywords:

lipomatous hyperplasia, palpitations, atrial septum, cardiac resonance imaging.

Palabras clave:

hiperplasia lipomatosa, palpitaciones, tabique interauricular, resonancia cardiaca.

ABSTRACT

Lipomatous hypertrophy of the interatrial septum is increasingly recognized and should be considered as part of the differential diagnosis of any cardiac tumor. We present the case of a 70-year-old patient who presented to the outpatient service for palpitations. Transthoracic echocardiogram (TTE) detected thickening of the interatrial septum with proximal predominance with respect to the fossa ovalis. The diagnosis was confirmed by cardiac magnetic resonance imaging. In such cases, the opinion of a cardiologist with expertise in cardiac imaging would help to avoid misdiagnosis and unnecessary intervention. This condition is more common than initially thought and remains under-recognized by most physicians.

RESUMEN

La hipertrofia lipomatosa del tabique interauricular se reconoce cada vez más y debe considerarse como parte del diagnóstico diferencial de cualquier tumor cardiaco. Presentamos el caso de un paciente de 70 años que acudió al servicio ambulatorio por palpitaciones. El ecocardiograma transtorácico (ETT) detectó engrosamiento del septo interauricular de predominio proximal con respecto de la fosa ovalis. El diagnóstico fue confirmado por resonancia magnética cardiaca. En tales casos, la opinión de un cardiólogo experto en imagen cardiaca ayudaría a evitar un diagnóstico erróneo y una intervención innecesaria. Esta condición es más común de lo que se pensó inicialmente y sigue siendo poco reconocida por la mayoría de los médicos.

INTRODUCTION

Lipomatous hypertrophy of the interatrial septum (LASH) is a benign lesion characterized by an excessive accumulation of adipose tissue in the interatrial septum exceeding 2 cm in thickness and forms part of the differential diagnosis between malignant and benign tumors of the atrium.¹ As most patients with this condition remain asymptomatic, cases are usually detected as an incidental finding at the time of cardiac imaging, surgery or necropsy.² Consequently, this condition remains under-recognized by most physicians and therefore can easily be

mistaken for a malignant lesion, subsequently leading to unwarranted surgical removal.

We present a case of a patient with persistent palpitations as a manifestation of an LASH, discuss the main features of this lesion and the importance of the diagnostic role of noninvasive imaging modalities, and briefly review the current literature.

DESCRIPTION OF THE CASE

A 70-year-old woman with a history of hypertension and dyslipidemia. The patient consulted for a long-standing clinical picture of occasional irregular palpitations, without

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syncope, attended cardiology consultation where Holter was taken with evidence of frequent supraventricular ectopias with arrhythmic load of 1%, echocardiogram with image suggestive of lipomatous hyperplasia (Figure 1) cardiac MRI was performed with evidence of left ventricle with normal function and size confirming the diagnosis without evidence of late enhancement (Figures 2 and 3), beta-blockers were added and the patient is currently under follow-up with improvement of the initial symptoms.

DISCUSSION AND LITERATURE REVIEW

Lipomatous hypertrophy of the cardiac interatrial septum is a rare but increasingly recognized benign anomaly of the heart.³ It was first described in 1964 following postmortem examination.⁴ Since then, this condition has been extensively documented in various case reports and original articles. The lipomatous lesion derives mainly from the superior and/or inferior part of the interatrial septum, typically respecting the fossa ovalis, giving a characteristic, considered by some to be pathognomonic, hourglass-shaped image, with a tendency to stand out in the right atrium, which may be related to a thickening of the terminal ridge (Figures 1 and 2).

Lipomatous hypertrophy of the interatrial septum is a frequent finding on echocardiography

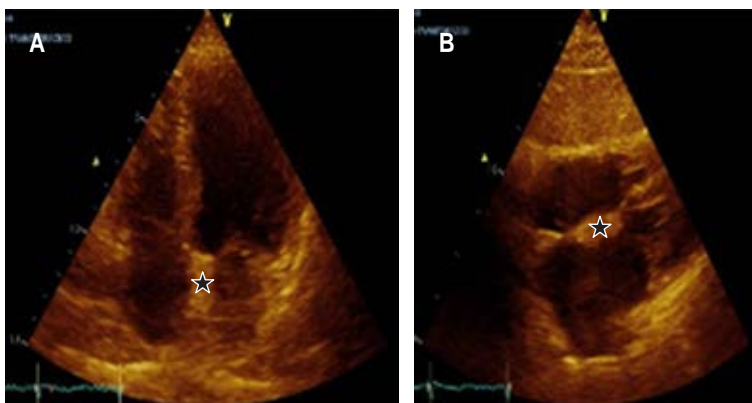


Figure 1: A) Four-chamber view shows increased thickness of the interatrial septum. **B)** Subcostal view, thickening of the interatrial septum with proximal predominance with respect to the fossa ovalis («hourglass sign»).

★ = interatrial septum.

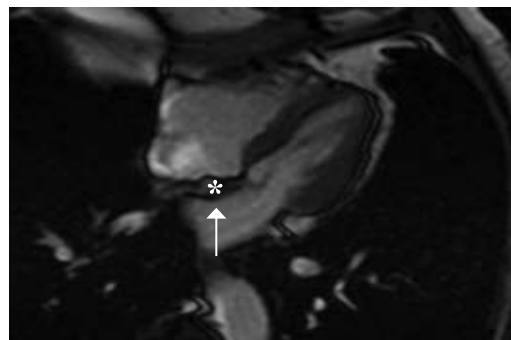


Figure 2: 4-chamber cine. Lesion in the interatrial septum with high signal intensity in relation to the myocardium (asterisk). The lesion presents artifact in India ink (low intensity line surrounding the lesion), indicating the presence of fat (arrow).

with a reported incidence of 1 to 8% of the general population.⁵ Cases previously described in the literature were based on incidental autopsy, surgical, and clinical imaging findings or were associated with the symptomatic course of the disease.⁶ Risk factors for LASH include emphysematous pattern with use of steroid therapy in higher frequency for elderly women, in which predisposition to mediastinal and intracardiac deposition of fatty tissue, cerebrotendinous xanthomatosis, mediastinal-abdominal lipomatosis and long-term parenteral nutrition are observed.⁷ Lipomas account for about 10% of cardiac neoplasms and are discrete accumulations of tissue grade that are usually extramyocardial and present as a defined rounded mass.⁸ While similar in tissue characterization to lipomas, LASH on TTE respects the boundaries of the atrial septum and usually respects the fossa ovalis.⁹ On transesophageal echocardiogram, LASH appears as an echodense globular thickening of the interatrial septum that is best seen in the bicave projection.¹⁰

Although it is a benign tumor that remains asymptomatic in most people, it may be associated with atrial arrhythmias that often require antiarrhythmic drugs. The arrhythmia, rarely associated with LASH, was first observed in 1969 by Kluge.¹¹ The mechanism of its occurrence has not been well explained, however, it appears to be related to infiltration of fatty tissue that interferes with the structure of atrial myocytes, thus disrupting normal

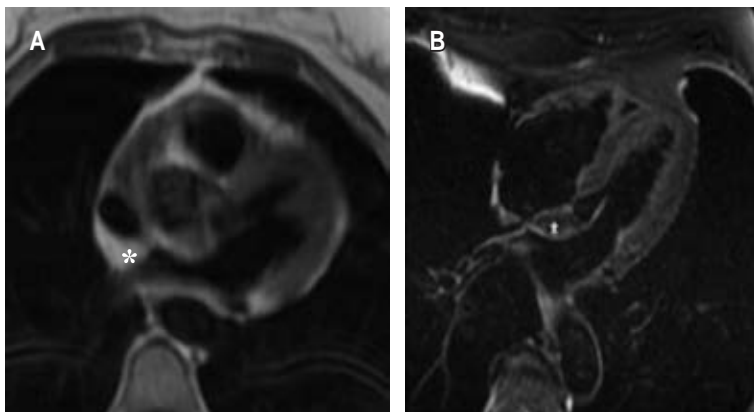


Figure 3: Axial T1 FSE (A) and 4-chamber STIR (B). Lesion in the interatrial septum (asterisk), of «dumbbell» or «hourglass» morphology, with high signal intensity in T1 (A), with signal drop in fat saturation sequence (B), suggesting the presence of fat. The morphology of the lesion and the presence of fat suggest the diagnosis of lipomatous hypertrophy of the interatrial septum.

conduction pathways.³ The arrhythmia manifests mainly in atrial fibrillation, atrial extrasystoles, supraventricular arrhythmias, ectopic and junctional rhythms. The incidence of atrial arrhythmia is also presumed to be related to septal thickness.¹²

This case highlights that while most LASH has the standard «dumbbell» appearance on TTE, there are cases where it may appear more like an adherent mass causing a wider differential. Noncontrast cardiac computed tomography (CT) can be used to confirm LASH by presenting low attenuation values for tissue (-80 to -120 HU) in the area of the atrial septum.⁷ Alternatively, CMR (cardiac magnetic resonance imaging) can be used for tissue characterization and confirmation of LASH. Steady-state pre-contrast free precession T2/T1-weighted cine CMR images show high signal

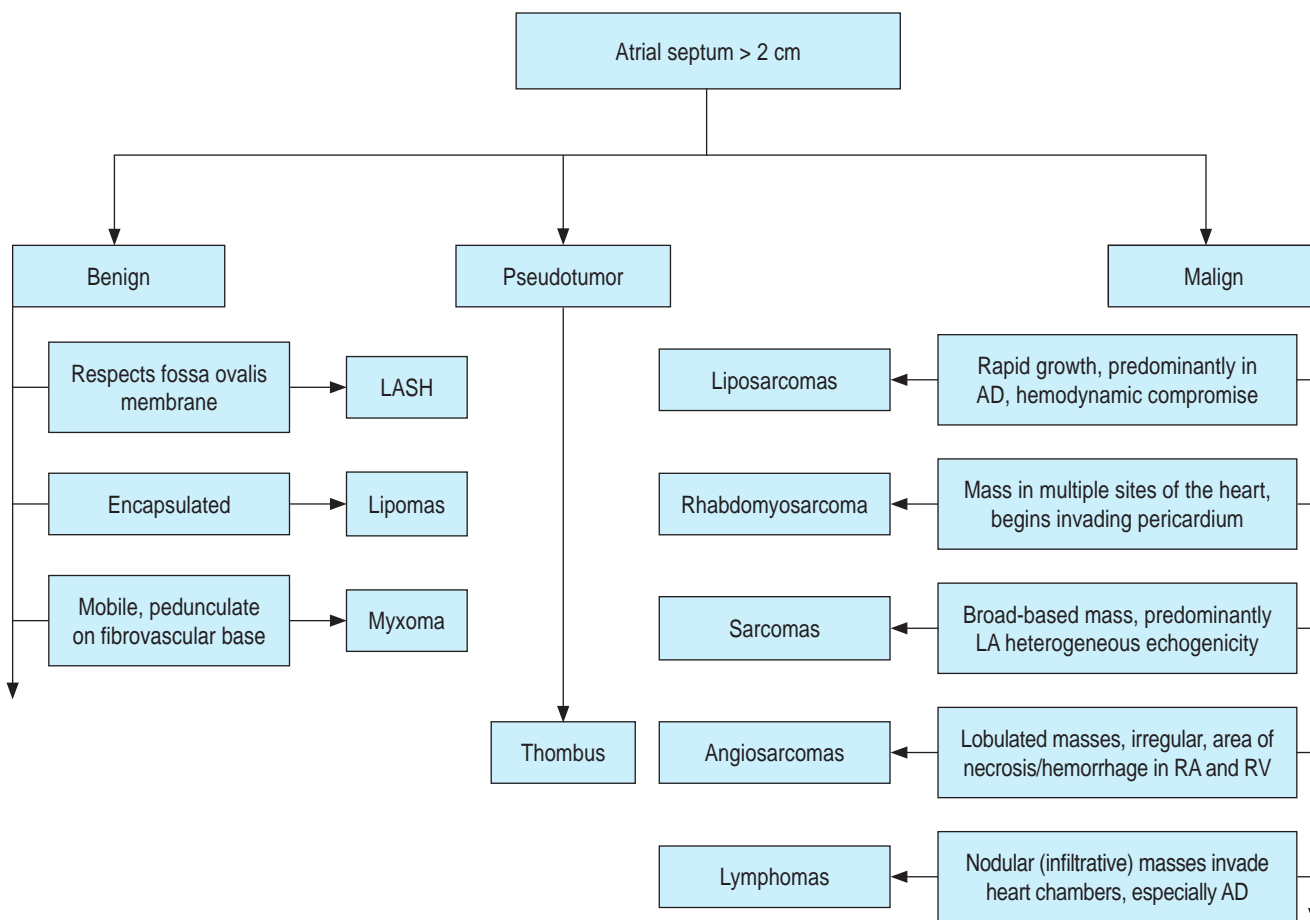


Figure 4: Atrial septal thickening approach.

intensity in the area of LASH and elicit a black limiting/hypointense effect between fat and myocardium (Figures 2 and 3).¹³ Because the structural features of this lesion are so distinct from any other intracardiac mass, definitive diagnosis without the need for tissue biopsy is now widely accepted.

Making the differential diagnosis of LASH from other cardiac neoplasms based on conventional imaging findings can be difficult. Myxomas are the most common primary cardiac tumors and account for 30% to 50% of all cases.¹⁴ Most myxomas are solitary and are located in the atria. They arise from the interatrial septum in the vicinity of the foramen ovale, whereas the foramen ovale is always spared in LASH. In addition, most myxomas are pedunculated on a fibrovascular stalk, which differentiates them. Cardiac lipoma is a true neoplasm occurring in younger patients. Lipomas are encapsulated, which is never seen in LASH.¹⁴ Rhabdomyomas and fibromas are common cardiac tumors in infants and children, and usually occur in the ventricles. Cardiac liposarcoma, a rare entity occurring predominantly in the right atrium, is a rapidly growing tumor with early signs of local invasion and hemodynamic compromise.¹⁵ Intraseptal cardiac liposarcomas have never been described. Its incidence is high in malignant melanoma, lymphoma and leukemia, where it occurs in the context of extensive disease.¹⁶

In our case presented with the «typical» hourglass appearance seen on TTE, a multimodality approach was used to narrow the differential diagnosis and arrive at a definitive diagnosis of LASH without requiring tissue biopsy. Lipomatous hypertrophy of the interatrial septum is a benign condition in most cases. Rarely, if severe interatrial hypertrophy is present, patients may develop right atrial filling obstruction (marginal obstruction of the superior vena cava or right atrium), shortness of breath and/or symptoms of heart failure. LASH can coexist with other intracardiac malignancies, which would warrant resection of the lesion with simultaneous interatrial septal plasty.¹⁷ Our case does not present these conditions and therefore was not a candidate for surgical management.

Considering the increase in life expectancy of the general population, the evolution of non-invasive imaging techniques and the increase in the prevalence of obesity, the probability of identifying this lesion will increase. We suggest a diagnostic algorithm for atrial septal hypertrophies in order to have a timely diagnosis and an adequate approach (Figure 4).

CONCLUSIONS

LASH is a benign lesion of the atrial septum, often asymptomatic. The role of multimodality imaging techniques in the diagnosis of LASH is essential. Making a correct and timely diagnosis prevents the patient from undergoing futile examinations with probable economic, social and psychological consequences, since in most cases of LASH, management consists of rapid diagnosis, reassurance and periodic follow-up.

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