Lipomatous Hypertrophy of the Interatrial Septum in a Patient with Chronic Dyspnea and Episodes of Paroxysmal Atrial Fibrillation: A Case Report

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What's Known

• Lipomatous hypertrophy of the interatrial septum can be detected as a tumor-like lesion that spares the fossa ovalis typically and needs differentiation from other cardiac neoplasms.

• Although it is usually an asymptomatic condition, it can cause several cardiac rhythm abnormalities.

What's New

• Although surgery is generally considered unnecessary in patients with persistent cardiac symptomatology or patients without a definite diagnosis, prompt resection is a useful option.

Abstract

Lipomatous Hypertrophy of the Interatrial Septum (LHIS) is an unusual condition usually found as an incidental finding on echocardiography. It can create a mass-like bulge typically sparing the fossa ovalis. We describe a 73-year-old overweight Cypriot woman, who visited our clinic in November 2018 and was complaining of dyspnea on exertion, chest tightness, and two episodes of paroxysmal atrial fibrillation for the last eight months. A big sessile mass was detected in the interatrial septum measuring 3.7×4.7 cm during transoesophageal echocardiography. The patient underwent resection of the mass and pericardial patch reconstruction of the defect with significant improvement of her clinical status during the follow-up period. The histopathological analysis suggested LHIS. In patients with persistent cardiac symptoms, a prompt resection is a useful option, although surgery is generally considered unnecessary in this clinical setting.

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Keywords • Atrial septum • Cardiac tumor • Atrial fibrillation • Dyspnea

Introduction

Lipomatous Hypertrophy of the Interatrial Septum (LHIS) is an unusual condition that is characterized by non-encapsulated accumulation of adipose tissue first described in 1964 by Prior.¹ The prevalence of LHIS is reported to be between 1-8%, depending on the diagnostic modality applied, with most data described in autopsy studies.² Interatrial Septum (IS) thickness >2 cm suggests the diagnosis of LHIS.³ However, it can also be detected as a tumor-like lesion that spares the fossa ovalis typically and represents approximately 0.6% of benign cardiac tumors.⁴ It should be differentiated with other cardiac neoplasms that can arise in the atrial septum (lipoma, myxoma, rhabdomyoma, fibroma, fibroelastoma, mesothelioma), cardiac sarcomas (rhabdomyosarcoma and liposarcoma), and distal metastases.^{3,4} Moreover, LHIS may coexist with other intracardiac malignancies, for which necessary medical interventions should be made.⁵ We describe a 73-year-old overweight woman with chronic dyspnea on exertion and two past episodes of paroxysmal atrial fibrillation having a large mass in IS as a result of LHIS. The patient underwent the resection of the mass and pericardial patch reconstruction of the defect with significant improvement of her clinical status during the follow-up period.

Case Presentation

A 73-year-old Cypriot woman presented in our clinic in November 2018 complaining of dyspnoea on exertion [New York Heart Association class II] and chest tightness for the last eight months.6 She also experienced two episodes of paroxysmal atrial fibrillation (AF) with a rapid ventricular response and hemodynamic instability treated with amiodarone. She was overweight [body mass index: 28 Kg/m²] with a past medical history of hypertension, hypercholesterolemia, and severe claustrophobia. Her treatment at the time included ramipril (Sanofi-Aventis, United Kingdom) 5 mg daily, bisoprolol (Merk Global, Germany) 5 mg daily, furosemide (Sanofi-Aventis, United States) 40 mg daily, atorvastatin (Pfizer, Australia) 10 mg daily, apixaban (Bristol Myers, United States) 5 mg every 12 hours, escitalopram (H. Lundbeck A/S, Denmark) 20 mg daily, and alprazolam (Upjohn UK Limited, United Kingdom) 0.25 mg daily. She had never smoked.

In physical examination, the patient appeared to be in good condition having 12 breaths per minute, a temperature of 36.5 °C, blood pressure of 110/70 mm Hg, and heart rate of 75 beats per minute. The rest of the clinical examination was unremarkable. Laboratory investigations were within the normal limits, including liver and renal function indices, electrolytes, and thyroid profile. Her electrocardiogram demonstrated sinus rhythm, and the chest radiography disclosed a normal cardiac silhouette. A chest computed tomography (CT) scan disclosed a big lipomatous lesion arising from the right cardiac chambers (figure 1). The CT pulmonary angiography was normal. The transthoracic echocardiogram revealed normal left and right ventricle dimensions and contractility with a left ventricular ejection fraction of approximately 65%. A mass in the base of the right atrium originating from IS was noted (figure 2). Transoesophageal echocardiography better clarified the nature of this large sessile mass in IS measuring 3.7×4.7 cm, which spared the fossa ovalis (figure 3). Fatty infiltration of IS was also noted. Cardiac magnetic resonance imaging (MRI) was not performed due to the patient's



Figure 1: Chest computed tomography scan illustrates a big lipomatous lesion arising from the right chambers (red arrow). RT.A.: Right atrium; RT.V.: Right ventricle; LT.A.: Left atrium; LT.V.: Left ventricle; Ao: Aorta; M: Mass



Figure 2: Transthoracic echocardiogram depicts normal left and right ventricle dimensions and contractility. A mass in the base of the right atrium originated from the interatrial septum is observed (red arrow). RA: Right atrium; RV: Right ventricle; LA: Left atrium; LV: Left ventricle



Figure 3: Transesophageal echocardiography discloses a large sessile mass in the interatrial septum measuring 3.7×4.7 cm, sparing the fossa ovalis (red arrow).

medical history of claustrophobia. Although she was suggested general anesthesia during her cardiac MRI, she was completely negative to cooperate. Non-significant (30%) left anterior descending artery stenosis was found, after the coronary angiography was performed.

Due to the unexplained dyspnea of our patient, the recent serious arrhythmia episodes

with hemodynamic instability, and the lack of MRI data for further clarification of tumor origin, the patient was referred to a cardiac surgeon. She underwent resection of the mass in IS and pericardial patch reconstruction of the defect in December 2018. The histopathological analysis showed LHIS. Microscopically, mature adipose tissue was the major cellular component found and was interspersed through cardiac muscle fibers (figure 4). Encapsulated fat cells were not found. From December 2018 until today, a significant improvement in her clinical status was observed. Written informed consent was obtained from the patient for the publication of all the data in this case report on the condition of anonymity.



Discussion

LHIS is usually found as an incidental finding in echocardiography and is more common in older obese people.^{2, 4} It has been associated with long-term corticosteroid use and obstructive lung disease.⁷ Although it is usually a benign lesion, it can induce several complications, namely superior vena cava obstruction, altered P wave configuration, atrial arrhythmia, atrioventricular block, recurrent pericardial effusions, and sudden death.^{2, 4, 7, 8} Interestingly, in a series of 32 autopsy cases with LHIS, the cause of death was directly related to this tumor in 28% of the population enrolled.9 Histologically, a combination of mature adipose tissue interspersed by fetal fat cells, fibrous tissue, and hypertrophied myocytes is found. The absence of encapsulated fat cells and the presence of fetal fat cells distinguish LHIS from cardiac lipoma. Fibrous fibers can affect the myocardial fibers and eventually lead to electric perturbation, cardiac arrhythmias, and myocardial atrophy. Moreover, LHIS is usually located in two proposed interatrial conduction pathways (middle and anterior internodal pathways). The interruption of these pathways could also be an important reason for rhythm defects.⁹ Although it is difficult to argue for the true causality of the dyspnea and AF in this elderly overweight hypertensive woman, the fact that she was treated with the same medication before and after the surgery, and that she experienced a significant improvement of her clinical status two years following the surgery (during her follow-up) favors this possibility.

Indications for surgical resection of LHIS are mainly superior vena cava obstruction and intractable rhythm disturbances. However, it was suggested that in patients referred for surgical therapy of other cardiac pathology, concomitant treatment for LHIS should be considered even in the absence of these indications.^{8, 10, 11} In large LHIS, after tumor resection, including the atrial septum, the replacement of the septum with a Dacron or autologous pericardial patch can be necessary.¹⁰ Surgical removal usually has excellent short- and long-term results similar to our patient.^{10, 11} The limitation of this report was the lack of cardiac MRI data of the patient.

Conclusion

LHIS is an unusual cardiac lesion, which needs to be differentiated from other cardiac neoplasms. It can present with a variety of morphologic features, such as an adherent mass. It is not clinically apparent in most cases. However, it can cause several cardiac rhythm abnormalities. Although surgery is generally considered unnecessary in patients with persistent cardiac symptomatology or patients without a definite diagnosis, prompt resection is a useful treatment option.

Authors' Contribution

G. S. P: Study concept and design, Acquisition, and interpretation of data, Drafting or critical revision of the manuscript; A. D. A Study concept and design, Acquisition, and interpretation of data, Drafting or critical revision of the manuscript; S. A. K. Study concept and design, Acquisition, and interpretation of data, Drafting or critical revision of the manuscript; All authors have read and approved the final manuscript and agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Conflict of Interest: None declared.

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