Lipomatous hypertrophy of the interatrial septum: report of two cases where histological examination and surgical intervention were unavoidable

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Received 27 March 2009; accepted after revision 16 May 2009; online publish-ahead-of-print 13 June 2009

Lipomatous hypertrophy of the interatrial septum (LHIS) is an increasingly recognized heart condition characterized by fatty deposits in the interatrial septum with sparing of the fossa ovalis. Its distinctive characteristic features by imaging techniques, benign nature, and the fact that most patients remain asymptomatic, has limited the need for histological confirmation and operative intervention in most cases. In this report, we describe two cases of LHIS where cardiac surgical intervention was indispensable: in the first patient, due to the presence of an additional left atrial tumour found out as mixoma and in the second, to relief a superior vena cava obstruction together with bypass grafts for severe coronary artery disease. Histological samples of the interatrial septal lesion were obtained in both cases either because of uncertainty of the diagnosis (Case 1) or to confirm the diagnosis (Case 2).

Keywords
Lipomatous hypertrophy of the interatrial septum; Cardiac tumours; Transesophageal echocardiography; Vena cava obstruction

Background
Lipomatous hypertrophy of the interatrial septum (LHIS) is a benign cardiac mass characterized by massive fatty deposits in the interatrial septum (IAS).1–4 With a still unknown aetiology and once described as a relatively rare condition, the expanding use of non-invasive imaging techniques in recent years leaded to an increase in the reported incidence up to 8%.5,6 The common imaging tool to evaluate LHIS is transthoracic and transoesophageal echocardiography, where the IAS appears unusually thick sparing the membrane of the fossa ovalis, giving the mass the typical dumbbell shape.7 However, its distinction from physiological thickening of IAS is not always easy and circumstances exist where differential diagnosis with neoplastic infiltration of the septum have to be considered.1 The ability of other non-invasive imaging techniques (CT scan and MRI) to recognize the entity of LHIS, including tissular characterization as fatty nature, makes them the preferred image tools to confidently recognize this benign disorder, avoiding the need for tissue biopsy.5,6,8 Despite the well-defined entity of LHIS, it remains under-recognized and can be easily taken for a malignant tumour.8 The differential diagnosis of LHIS might be particularly difficult when it appears associated with other intra-atrial cardiac masses. Moreover, the choice of treatment of LHIS is sometimes controversial, and truly surgical indications are few and rare.9

Case reports
Case 1
A 73-year-old man with worsening exertional dyspnea for the previous 2 months was referred for echocardiographic evaluation. He had several cardiovascular risk factors (hypertension, hyperlipidaemia, cigarette smoking, and obesity) and atrial fibrillation diagnosed since 2 years, when warfarin was started. On the transthoracic echocardiogram, a cardiac mass was incidentally found in the left atrium and multiplane transoesophageal echocardiography was performed to better delineate the nature and extension of the mass. An ovoid (40 × 30 mm) echodense and mobile mass was observed in the left atrium, attached to the IAS. The mass displaced towards the mitral valve orifice in diastole, still not causing valve obstruction (Figure 1A and B). Originating from the entire length of the IAS, a second large echogenic and immobile mass was identified with a
typical dumbbell shape (Figure 2). Cardiac surgery was undertaken to remove the mobile left intra-atrial mass. Despite the morphological characteristics of the IAS suggested LHIS, its close contiguity with the other atrial mass raised doubts about its histological nature. At surgery, a tumour was attached to the IAS between the mitral valve and fossa ovalis, measuring 50 × 40 mm. It was with regular contours, heavy consistency, and some areas of intratumoral necrosis. There was also an impressive increased thickness of the septum secundum and primum which was biopsied but not excised.

Histological examination of the left intracavitary mass disclosed a mixoma (Figure 3) and the septal lesion confirmed as LHIS (Figure 4). The patient was discharged asymptomatic a few days later.

Case 2

A 66-year-old woman with angina pectoris for the last 2 years was admitted with NYHA class III dyspnea. This obese patient had a history of hypertension, diabetes, cigarette smoking, and chronic obstructive pulmonary disease. Results of the chest and cardiac examinations and electrocardiogram were unremarkable. Transthoracic echocardiography detected a thickened IAS, not well delineated (Figure 5A). On transoesophageal echocardiography, a bulky septal mass (4.6 × 3.4 cm) protruded into the right atrium and the superior vena cava (SVC), causing some degree of upper right atrial inflow obstruction (Figure 5B).

The interatrial septal lesion as well as the SVC narrowing was further confirmed by multi-slice computed tomography (Figure 6), cardiac magnetic resonance (Figure 7), and superior caval venography (Figure 8A).

Coronary angiography showed severe calcified two-vessel disease. Reconstructive enlargement of the SVC with autologous pericardium in combination with coronary artery bypass grafting (CABG) was planned. During surgery, a tissue sample of the interatrial mass was taken for histological examination which confirmed LHIS. The post-operative course was complicated by transient supraventricular tachycardia. The patient was discharged without dyspnea a few days later, after repeated superior caval venography (Figure 8B).

Discussion

We report two cases of LHIS with associated cardiac pathology where cardiac surgical intervention was unavoidable. In Case 1, the indication for surgical treatment was the presence of the left atrial myxoma. In this case, LHIS was asymptomatic and not causing any obstruction to flow. In Case 2, however, surgical correction of lipomatous hypertrophy was needed to relieve a certain degree of SVC obstruction.
Figure 4  Histology of the IAS: vesicular brown fat (→). Entrapped enlarged myocytes (**) and normal fat cells (Δ). Absence of lipoblasts.

Figure 5  Transthoracic (A) and transoesophageal echocardiography (B). (A) Apical four-chamber view: thickening of the IAS (white arrow); (B) mid-esophageal view at 0°: volumous interatrial septal mass (*)

Figure 6  Multislice computed tomography (MSCT) depicting heavy thickening of the upper segment of the IAS and narrowing of the distal part of the superior vena cava (SVC).

Figure 7  Cardiac magnetic resonance. Arrow points out the thickened IAS, protruding into the right atrial (RA) cavity.
Uncertainty in the diagnosis, connected to the narrow proximity with the left atrial tumour (Case 1), and the will to achieve a definitive diagnosis excluding a proliferative disease (Case 2), justified tissue biopsy of the interatrial septal lesion in both patients.

Lipomatous hypertrophy of the IAS is a benign cardiac mass that should be considered in the differential diagnosis of any atrial cardiac mass involving the IAS. Because it can be mistaken with a malignant tumour leading to unwarranted surgical removal, recognition of this lesion by different imaging techniques is of paramount importance. In Case 1, the patient presented with a left atrial mass besides the typical atrial septal lesion. This fact gave place to doubts about the diagnosis, namely for the possibility of a link between the two lesions, precluding the histological evaluation performed during the surgical procedure to remove the left atrial tumour. In Case 2, although no diagnostic doubts remained after the diversity of non-invasive imaging techniques performed before the operation, the surgeon intra-operatively decided for a partial resection of the tumour. Pericardial patch reconstruction of the SVC to relief upper right atrial inflow obstruction, in combination with CABG was carried out as planned. In this particular case, the demand for a sureness diagnosis dictated tissue sampling. Lipomatous hypertrophy of the IAS usually remains asymptomatic and presents as an incidental finding of echocardiography or intra-operative. In most cases, it does not require any specific treatment. Surgical management of LHIS is usually not necessary and should be limited to patients with intractable severe rhythm disorders, haemodynamic instability, symptoms of SVC syndrome or right atrium obstruction.

The two cases reported represents unusual examples of this entity, where contrary to the current practice dictating a conservative approach, an interventionist attitude including biopsy and surgical intervention was justifiably undertaken. Finally, we also highlight the importance of histological examination on definitive diagnosis of cardiac tumours.

References