Case report

Lipomatous hypertrophy of the interatrial septum and upper right atrial inflow obstruction

Martin Breuer*, Jens Wippermann, Ulrich Franke, Thorsten Wahlers

Department of Cardiothoracic and Vascular Surgery, Friedrich-Schiller-University, Bachstrasse 18, 07743 Jena, Germany

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Abstract

Lipomatous hypertrophy of the heart is a benign rare abnormality characterized by large fatty tissue deposits in the interatrial septum. An increased incidence of atrial arrhythmias is described in these patients, significant blood flow obstruction however is not the rule. We report a case of lipomatous hypertrophy of the interatrial septum, detected by transeosophageal echocardiography (TEE). The tumour mass protruded into the right atrium and the superior caval vein (SCV), thus causing upper right atrial inflow obstruction. Partial resection of the tumour and pericardial patch-reconstruction of the SCV were performed in combination with coronary artery bypass grafting due to coronary artery disease (CAD). The diagnostic and therapeutic management is discussed and a review of the literature performed. © 2002 Elsevier Science B.V. All rights reserved.

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1. Case report

A 69-year-old female patient was admitted to a regional hospital for diagnostic evaluation and therapy of minor stroke. Computed tomography of the chest and transthoracic echocardiography (TTE) detected a large intracardiac tumour. Therefore, after recovering she was referred to our hospital for further evaluation and treatment. Medical history showed a rheumatic arthritis, a nephrotic syndrome and arterial embolic occlusion of one finger. History for atrial arrhythmia was uneventful. Physical examination revealed a normal pulse of the jugular veins, upper venous congestion was not visible. X-ray of the chest demonstrated a normal heart size and slightly emphysematous lungs. In electrocardiogram (ECG) a normal sinus rhythm was found. Transeosophageal echocardiography (TEE) examination detected a bilobed echogenic hypertrophy of the interatrial septum (58 × 35 mm), sparing the foramen ovale. A large mass extending into the superior and posterior parts of the right atrium was described, narrowing the superior caval vein (SCV) and the SCV-atrial junction (Figs. 1 and 2). Chest computed tomography showed a lipomatous tumour starting from the posterior wall of the right atrium, almost completely compressing the SCV. Magnetic resonance imaging confirmed these results. Coronary angiography demonstrated coronary artery disease (CAD) with 70% stenosis of the main stem, 80% stenosis of the left anterior descended artery (LAD) and a significant stenosis of the 2nd diagonal branch.

Operative approach: following median sternotomie ascending aortic cannulation with bicaval venous return was applied. The heart was vented but not arrested. Larger parts of the fatty tissue mass were removed in particular out of the septal portion of the SVC-atrial junction for urgent histologic evaluation. Due to invasion of the tumour into the skeleton of the heart and a non-malignant histology, no attempt was made to remove the tumour in total. After inspection of the SVC-atrial junction it became obvious that, even after sharp decompression, a significant obstruction of the right atrial inflow with a diameter of only 12 mm was remaining. Therefore additionally a reconstructive enlargement of the SVC and the SVC-atrial junction with pericardium was performed. Finally, after cardioplegic arrest of the heart, coronary artery bypass grafting was performed. The patient was successfully weaned from cardiopulmonary bypass with a slow junctional rhythm making ventricular pacing necessary. On the 3rd postoperative day the heart converted into a stable sinus rhythm.

Pathohistologic examination of the resected tissue showed the typical pattern of lipomatous hypertrophy, that
means predominantly adult fat cells with a background of interspersed hypertrophic cardiac muscle cells.

2. Comment and review of the literature

Lipomatous hypertrophy of the interatrial septum is a rare but pathohistologic and morphologic well described cardiac tumour [1]. The clinical importance is not defined because most of the data was derived from autopic studies. Its diagnosis during life is a rarity mostly seen incidental.

Lipomatous hypertrophy of the interatrial septum as a different entity was first described by Prior in 1964 [2]. He detected that lipomatous hypertrophy is a non-encapsulated hypertrophic alteration of normal structures of the interatrial septum. It must be differentiated from the lipoma and in particular from the liposarcoma of the heart. In the literature there are meanwhile approximately 200 cases described [3,4]. The first in vivo diagnosis of this kind of tumour is from the year 1982 [5].

Histologically, lipomatosis of the heart is characterized by mostly adult lipocytes with interspersed hypertrophic cardiac muscle fibers. Occasionally, multivacular fat cells similar to fetal fat cells can be found [6]. Macroscopically it cannot be differentiated from epicardial fatty tissue. Moreover it seems that in patients with lipomatous hypertrophy additionally an enormous increase of the epicardial fatty tissue can be observed [7]. Clinical symptoms of the tumour are not specific or absent, but most of the reported cases occur in obese patients and most commonly in women [4]. Only the incidence of supraventricular arrhythmias seems to be significantly increased. A septal wall thickness of more than 3 cm was combined with an incidence of atrial arrhythmias of more than 60% [7]. Hemodynamical significant obstructions of the atrial inflow or outflow tract usually are not observed. Only few cases with clinical relevant obstruction of the SVC can be found in the literature [5,8]. In our patient at the time of surgery no clinical signs of upper inflow obstruction were seen. TEE examination and in particular intraoperative findings however revealed a dramatic obstruction of the SCV-atrial junction.

Surgical therapy of lipomatous hypertrophy of the interatrial septum is reserved to patients having SCV obstruction or an intractable rhythm disturbance [8,10]. If complete excision of the tumour is planned, reconstruction of the interatrial septum, with either autologous pericardium or Dacron must be performed [10]. On the other hand, lipomatous hypertrophy of the interatrial septum has no tendency towards a rapid increase. So, in fact, there is no real need for complete excision. Particularly as partial or total resection of the interatrial septum also will not relieve the patient from rhythm disorders in any case. In our patient we diag-

Fig. 1. Lipomatous hypertrophy of the interatrial septum (arrow) detected by magnetic resonance imaging. Moreover, a remarkably increased epicardial fatty tissue layer can be seen.

Fig. 2. Lipomatous hypertrophy of the interatrial septum: computer tomographic detection of caval vein obstruction. The diameter in the area of the junction between SCV and atrium is reduced to one third (arrow).
nosed a significant obstruction of the SCV. Due to the benign character and the central intracardiac position of the tumour we decided against a complete resection. Narrowing of the SCV could be easily repaired by insertion of a pericardial patch.

In conclusion, lipomatous hypertrophy is a rare, acquired, benign deposition of fatty tissue within the interatrial septum, mostly diagnosed incidental. TEE and computed tomography are the diagnostic tools of choice. In case of hemodynamic alteration of the great vessels or severe rhythm disorders a surgical correction must be considered.

References