Lipomatosis of interventricular septum and both ventricles: an extremely rare pattern

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Abstract

Fatty masses of the heart are relatively uncommon. This report is about an extremely rare case of cardiac lipomatosis that implicates both ventricles and inter-atrial septum. The patient was a 27-year-old female who was accidentally diagnosed during a routine physical examination. Trans-thoracic echocardiography (TTE) discovered a large homogenous bi-lobed, non-encapsulated echogenic mass. Magnetic resonance imaging (MRI) showed soft tissue abnormal enhancement in the anterior left base of the heart and sub-aortic outflow tract. A trans-jugular echomyocardial biopsy was done because of a lack of a definite diagnosis by non-invasive techniques. Pathological study showed benign fatty infiltration suggestive for benign lipomatous hypertrophy.

Keywords: Cardiac tumor; Left ventricle; Lipomatosis

1. Introduction

Fatty masses of the heart are relatively uncommon which include cardiac lipomas, lipomatous hypertrophy of the inter-atrial septum (LHIS), lipomatous infiltration of the right ventricle and arrhythmogenic right ventricular dysplasia (ARVD) [1]. LHIS is a rare but benign condition characterized by fat accumulation in the inter-atrial septum and a septal wall thickness of more than 2 cm [2]. More than 200 cases with cardiac lipomatous hypertrophy have been reported so far, but there has been only one report of LHIS involving the right ventricle up to 2007 [3]. The lesion is confined to the right atrium and the masses project only into the right side of the heart; however, extension into the free wall of the left atrium has been documented [4]. No absolute diagnostic criteria have been established but specific echocardiographic and magnetic resonance imaging (MRI) findings could be suggestive for benign lipomatous hypertrophy.

Here, we report a benign lipomatous hypertrophy of the heart having an extremely rare involvement pattern which implicated inter-atrial septal wall, proximal third of interventricular septum, aortic and pulmonary outflow tract.

2. Case report

The patient was a 27-year-old female with no previous history of any health issues. A systolic murmur was revealed in routine physical examinations so she was referred to cardiologist without any symptoms. In primary trans-thoracic echocardiography (TTE) evaluation, a huge infiltrative homogenous mass in the anterior aspect of aorta, proximal third of inter-ventricular septum and postero-lateral left ventricular wall was found. Further assessments included thoracic high-resolution computed tomography (HRCT), MRI and follow-up echocardiography. The thoracic HRCT revealed no significant lesion except a soft tissue density next to the right border of the aorta. MRI showed a soft tissue abnormal enhancement in the anterior left base of the heart and sub-aortic outflow tract with smooth borders and a mildly different signal from normal myocardium (Fig. 1). The nine-month follow-up TTE discovered a large nearly homogenous bi-lobed, non-encapsulated echogenic mass encasing the aorta and infiltrating into septum, encroachment of the right ventricular outflow tract (RVOT) and flow acceleration in RVOT with pulmonary gradient being 15 mmHg which was a new finding since the last examination, atrial septal wall thickened to about 22 mm and left main coronary artery embracement by the mass. The new TTE was suggestive for a malignant sarcomatous nature of the mass, most likely a primary pleomorphic liposarcoma. We decided to perform a more aggressive approach towards diagnosis and treatment due to the suspected malignancy and the newly founded RVOT obstruction, so a trans-jugular endomyocardial biopsy from the right ventricular wall was obtained. Pathological study of the specimen showed benign lipomatous hypertrophy. No further surgical intervention seemed to be necessary. As a result, the patient was discharged from the hospital and is now without any cardiac symptoms and under echocardiographic follow-up every six months.
4. Conclusion

According to the above-mentioned evaluative and usually helpful tools in diagnosing cardiac lipomatosis, MRI, CTs and echocardiography could not make a definite diagnosis in such a case. As a result, aggressive approaches like endomyocardial biopsy were inevitable. After pathological confirmation of the benign nature of the lipomatous lesion, no further surgical intervention was necessary as there was no indication for surgical removal. To our knowledge, this is the first reported case of benign cardiac lipomatosis which involves the left ventricle.

References


eComment: Lipomatous hypertrophy of the atrial septum: clinical approach and surgical indications

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We read with great interest this case report, concerning cardiac lipomatosis involving the inter-atrial septum and both ventricles [1]. Considering that most patients with lipomatous hypertrophy of the inter-atrial septum (LHIS) are asymptomatic and over 60 years of age, the young age of the patient is also rare. In our opinion, a new trans-thoracic echocardiography (TTE) should have been performed much earlier than 9 months, in view of the patient’s age and the lack of accurate diagnosis following high-resolution computed tomography (CT) and magnetic resonance imaging (MRI), so as to prevent potential complications (arrhythmias or even sudden cardiac death) of rapid mass development. In particular, the expanding use of non-invasive imaging techniques in recent years has led to an increase in the reported incidence of LHIS up to 8% [2]. Histologically, LHIS is characterized by proliferation of adipocytes and interspersed hypertrophied cardiomyocytes, while massive fat deposition in the atrial wall results in a thickness of the atrial septum of >20 mm up to 60 mm, associated with a higher incidence of atrial arrhythmias [3]. Cardiac irritability may be a result of atrophy of the myocardium with fibrosis that accompanies deposition of large amounts of fat tissue, in contrast to the majority of true cardiac lipomas that present with symptoms proportional to their size and localization [4].

Surgical management is usually not necessary and should be limited to cases in which LHIS is complicated by severe rhythm disorders, superior vena cava syndrome, right atrial obstruction or altered hemodynamic cardiac function leading to congestive heart failure [4]. In the case of a small tumor the septum may be excised and closed primarily by suture, although after large LHIS removal including atrial septum, the replacement of the septum with a dacron or autologous pericardial patch may be necessary. Of note, LHIS is usually situated in the area of at least two described interatrial conduction pathways (anterior and middle interomodal pathways), and their interruption could be the major reason for rhythm disorders in these patients [5]. As a consequence, partial or total resection of the interatrial septum probably will not eliminate the rhythm disturbances, especially when the
atrioventricular node region is involved. In that case, the patient may end up with a pacemaker [4, 5].

References


