Case Report

Atypical Presentation of a Cardiac Lipoma

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Abstract

Cardiac masses can have atypical presentations and diagnosis can be difficult because of limitations in imaging modalities. We report the case of a 69-year-old man with a history of hypertension and hyperlipidemia who presented with several months of dyspnea with exertion. There was a soft systolic murmur on exam. Transthoracic echocardiogram showed a large right atrial mass that appeared to be adherent to the interatrial septum. Considering the size and apparent attachment to the interatrial septum the suspicion was a myxoma. After further imaging however, including transesophageal echocardiogram, cardiac computed tomography (CT) and cardiac magnetic resonance imaging (MRI) the diagnosis changed to a lipoma. The patient had surgical resection of the mass which confirmed the diagnosis. A multi-imaging approach may be helpful and required to accurately diagnose cardiac masses.

Keywords
cardiac tumor; lipoma; atrial myxoma; lipomatous hypertrophy; echocardiography; MRI; cardiac CT

Introduction

Primary cardiac tumors are rare, with an incidence of 0.001 to 0.19% of which 90% are benign [1–4]. Cardiac lipomas are the second most common cardiac tumor, representing 10–19% of the total [2,3]. They vary in size depending on location. Pericardial lipomas can be large in size (median 10 cm) while intracavitary locations tend to be smaller (median 3.5 cm). They are broad based with no stalk and appear homogenous on echocardiograms and cardiovascular magnetic resonance imaging (CMR) [1]. Cardiac myxomas are the most common primary tumor, representing 45% of the total. They are most commonly found in the left atrium attached to the interatrial septum (IAS) by a discrete stalk. They can be of varying sizes and appear heterogeneous [1]. Lipomatous hypertrophy is a benign condition in which non-encapsulated fatty tissue infiltrates the interatrial septum [5]. It has a characteristic dumbbell appearance of the interatrial septum with sparing of the fossa ovalis. It can be mistaken for a mass resulting in unnecessary surgery [6]. Other cardiac masses typically considered in the differential include: benign tumors such as papillary fibroelastomas, rhabdomyomas, and cardiac fibromas. Malignant masses include: angiosarcomas, liposarcomas and leiomyosarcomas with non-neoplastic masses characterized by vegetations and thrombus. We report the case of a cardiac lipoma that was initially thought to be a myxoma based on the size and unclear attachment site on the transthoracic echocardiogram (TTE). Multiple imaging modalities including transesophageal echocardiogram (TEE), CMR and cardiac computed tomography (CCT) were utilized to clarify the attachment site, tissue characterization and diagnosis.

Case Presentation

A 69-year-old male with a history of hypertension and hyperlipidemia presented with several months of dyspnea with exertion. His physical examination was notable for a soft systolic ejection murmur and trace lower extremity edema. Electrocardiogram showed left ventricular hypertrophy with repolarization abnormalities. Chest x-ray was unremarkable. A TTE showed normal ejection fraction with a large mass in the right atrium measuring 3.5 × 2.5 cm which appeared adherent to the interatrial septum. Doppler imaging showed no clear hemodynamic effect. There was no tricuspid stenosis and only trace tricuspid regurgitation. The inferior vena cava (IVC) was dilated however, suggesting increased right atrial pressure. A TEE was then performed to better evaluate the mass and attachment site. It showed a large, pedunculated, well-circumscribed mass measuring 4.4 × 3.9 cm. In some views it appeared the mass was either pressing on or was attached to the IAS (Fig. 1A,B). In other images, the attachment seemed to be the right atrial wall along the insertion of the inferior vena cava (IVC) (Fig. 1C). Considering the large size of the mass and questionable attachment to the IAS, the initial diagnosis was myxoma. He was seen by cardiothoracic surgery and had a CCT which showed a focal mass with smooth borders. There were some technical difficulties with mix-
Fig. 1. Transesophageal echocardiogram (TEE) showing right atrial mass. (A) Mid esophageal four chamber view, showing a right atrial mass (red arrow) compressing the interatrial septum (IAS) (blue arrow). (B) TEE, mid esophageal short axis view at the level of aortic valve also showing the mass (red arrow) with possible attachment to the IAS (blue arrow). (C) TEE, mid esophageal bicaval view suggesting the mass (red arrow) is attached to the right atrium (RA) along the insertion of the inferior vena cava (IVC) or IAS (blue arrow).

Fig. 2. Cardiac computed tomography (CT) showing right atrial mass. (A) Axial view shows fat density mass (~83 Hounsfield Units) adjacent to coronary sinus and IVC (arrow). (B) Sagittal view shows the thin rim of the right atrial wall (arrow), indicating the mass is intra-atrial. (C) Right ventricle three chamber view shows the tricuspid valve (dashed arrow) and lipoma (solid arrow). No soft tissue densities within the mass are present.

There was concern the mass could be extracardiac compressing the right and left atria (Fig. 2). Finally, a cardiac magnetic resonance imaging (MRI) was recommended and showed the mass had features consistent with lipoma but severe lipomatous hypertrophy could not be excluded (Fig. 3). Considering the size of the mass and persistent dyspnea, the patient underwent surgical exploration and excision of the mass. The surgery was performed with a midline sternotomy approach. After opening the right atrium, the lesion was exposed and appeared lipomatous macroscopically. It was within the right atrium attached to the atrium in the space between the IVC cross and the coronary sinus measuring 4.5 cm. The mass was sharply resected in its entirety. Frozen section confirmed lipoma. Patch closure with bovine pericardium was required for the posterior wall of the right atrium. Considering the patient’s intolerance to opiates, an incisional VAC dressing was placed to minimize pain and help stabilize the sternum (Fig. 4). The patient had an uneventful post operative course and was discharged home on post op day #4. Post operative TTE showed no residual right atrial (RA) mass with no dilation of the IVC suggesting normal RA pressure. The CARE checklist was used when writing this case report (Supplementary Table 1).

Discussion

Cardiac lipomas are second to myxomas as the most common primary cardiac tumor. They are more common in
middle-aged or older adults, and primarily originate from the subendocardial layer [7]. They are encapsulated by a thin layer of fibrous tissue and are predominantly composed of mature white adipose tissue although fetal brown fat has also been reported [7,8]. The etiology of the tumor is unknown. There are no clear genetic correlations and the presentations can vary [7]. They tend to be small in size and can be located in any of the cardiac chambers or in the pericardium. They can also grow very large in size with or without infiltration of the myocardium [7]. Often time there are no symptoms but patients can present with dyspnea, chest pain, syncope, palpitations, stroke-like symptoms and sudden cardiac death [1,2,9]. Mechanisms for the symptoms include: obstruction of intracardiac and inferior and superior vena cava blood flow, cardiac valve dysfunction, phrenic nerve stimulation and embolic events [2]. In
Intraoperative photographs: Yellow fatty mass seen after entry into right atrium (top left). Mass is identified with forceps arising from the wall of the right atrium (middle). Mass is completely resected (right). The black arrows in the intraoperative images are pointing to the right atrial mass.

In our case, the patient’s only symptom was dyspnea with suggestion of elevated RA pressure but no hemodynamic compromise of the tricuspid valve.

By echocardiogram imaging, lipomas appear homogeneous, well circumscribed, and sessile with a broad-based attachment. Unlike myxomas, they tend to not have a discrete stalk. Cardiac lipoma should not be confused with lipomatous hypertrophy of the interatrial septum which is non-encapsulated fatty tissue that can appear asymmetric at times and can lead to unnecessary surgical resection [6].

Computed tomography (CT) and MRI can aid with tissue differentiation. CT imaging shows homogeneous, hypodense, encapsulated masses with or without septa. They have the same appearance as subcutaneous fat. High resolution CT can identify the wall and boundaries of the mass to help localize the attachment [7]. Cardiac MRI images similarly have features consistent with subcutaneous fat. There is signal loss of the mass on fat suppression sequences [7].

Due to the rare occurrence of cardiac lipomas there are no guidelines for treatment [2]. Surgical resection is recommended for masses that are causing symptoms, including hemodynamic compromise [7]. Cure rates approach 95% and prognosis tends to be good [2]. Asymptomatic masses can potentially be monitored for rate and size or growth [2].

The diagnostic approach in evaluating cardiac masses can be complicated. Factors considered in the evaluation include: age of the patient, clinical presentation, location of the tumor and noninvasive tissue characteristics [1]. In our case, several of the masses on the differential were excluded. Fibroelastomas and vegetations tend to be associated with cardiac valves. Rhabdomyomas and fibromas occur in younger patients. Sarcomas appear more invasive and infiltrative to the surrounding tissue and thrombus seemed unlikely considering the location, clinical presentation and MRI findings [1].

The various imaging modalities available have limitations. Echocardiography can be challenging depending on the imaging plane and is unable to make a determination of tissue characterization. MRI has lower temporal resolution than echocardiography and thus cannot adequately visualize small lesions [10]. CT disadvantages include: radiation exposure, contrast induced nephropathy, and limited soft tissue and temporal resolution compared to MRI [1]. The most challenging aspects of this case were the size and location of the lipoma. Despite the multiple imaging modalities utilized in the case, there was some confusion about the location and attachment site. In this case, by TTE and TEE, the mass occupied a significant portion of the right atrium and appeared to either compress or be attached to the IAS. The other possible attachment site appeared to be along the RA free wall near the IVC. Depending on the imaging plane and view, it can be challenging distinguishing this. The use of X-plane imaging or three dimensional (3D) may have helped but were not utilized in this case. The CT images had some technical limitations which made border recognition difficult. The CT density of the mass was ~83 Hounsfield. There were areas of streak artifact from iodinated contrast located in the right atrium which extended into the mass, making evaluation of the borders and internal density somewhat challenging. The MRI images questioned whether the mass was extracardiac or represented asymmetric lipomatous hypertrophy. The MRI limitations included: (1) the
large size of the mass made border distinctions difficult, (2) the very low signal intracardiac border of the mass appeared to connect with the very low signal outer border of the atrium and (3) the faint low signal thin wall of the atrium to which the lipoma was attached is underappreciated on MRI (slice thickness of 8 mm) compared to the higher resolution CT (slice thickness of 2 mm). MRI did show complete loss of signal on fat suppressed images confirming fat composition without internal soft tissue.

Despite the limitations of each of the imaging modalities, the multimodality imaging approach was vital in this case. The echocardiogram findings made it clear the mass was intracardiac and not consistent with lipomatous hypertrophy. Although the large size and possible attachment to the IAS made myxoma a possibility, the addition of the CT and MRI findings confirmed this was a lipoma.

During surgery, it became clear the attachment site was the right atrial wall between the coronary sinus and inferior vena cava. Post operatively, the patient did well and was discharged in stable condition. The symptom of dyspnea improved during follow up visits but has not completely resolved as of 1 year following excision of the mass. Workup for alternative causes of the persistent dyspnea is ongoing. Follow up echocardiograms 1 month and 6 months following surgery have shown no evidence of recurrence and there were no changes with respect to doppler hemodynamics and RA pressure remained normal.

Conclusions

Cardiac lipomas are rare tumors that can become large in size and cause symptoms requiring surgical excision. Lipomas can be challenging because of their variable presentations including: sizes of the masses, multiple locations, differing symptoms and rare occurrence with no established guidelines regarding management [2]. A multimodality imaging approach including CT/MRI and echocardiography can help determine the type of mass and location, which can aid with surgical and long-term management.

Availability of Data and Materials

Data sharing is not applicable to this article, as no datasets were generated or analyzed during the current study.

Author Contributions

PL, KL, ADK, MG and EJK contributed to the conceptualization. ADK, PL, KL and EJK drafted the work. MG was involved with revising it critically for important intellectual content. EJK, MG and PL were involved with editing and review. ADK, KL and MG contributed to the images used in the work. All authors contributed to editorial changes in the manuscript. All authors read and approved the final manuscript. All authors 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.

Ethics Approval and Consent to Participate

Ethical approval to report this case was not obtained by the IRB at Abington hospital given no patient identifier information included and that article is a retrospective chart review. Written consent was obtained from the patient.

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Conflict of Interest

The authors declare no conflict of interest.

Supplementary Material

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References