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A case of Exuberant Lipomatous Hypertrophy of Interatrial septum in the setting of Narrow complex Tachyarrhythmia with diagnostic and management challenges.

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Abstract:

Background

Exuberant Lipomatous growth of Interatrial septum is an interesting but rare and challenging case. It can mimic the diagnostic dilemma of Cardiac malignancy leading to nonessential resection. A sound understanding of its radiological features and ability to define it based on the non-invasive modality is of utmost importance in the clinical decision making

Case Summary

We present an interesting case in a 49 year old Caucasian lady which was picked up as incidental finding on TTE. Clinical disease progression was further complicated with the new onset of narrow complex tachycardia i.e. Atrial Fibrillation. Multiple imaging modalities including TTE, TOE, MDCT , CMR and PET scan was used to assess the property of mass where the features were suggestive of the benign nature of Exuberant lipomatous hypertrophy of Interatrial septum. A multidisciplinary meeting was organised for the consensual opinion regarding the surgical Vs medical management where on the balance medical management with rate control strategy was implemented. Surveillance with the serial imaging is ongoing to monitor the changes

Discussion

Cardiac masses are rare but an important cause of different presentations like narrow complex tachyarrhythmia, tamponade and pericardial effusion. With the frequent use of non-invasive modalities like CT, CMR and PET its incidence frequency is on rise. Exuberant lipomatous growth is due to the fat deposition in embryonic aberrancy of septum secundum leading to septal thickness which can be readily detected on imaging modalities however its close resemblance to infiltrative process like sarcoidosis or neoplasia can lead to unnecessary surgical resection.

Keywords: Exuberant Lipomatous growth, Interatrial septum, Narrow complex tachyarrhythmia

Issue Section Cardiovascular imaging

Learning Points

Features of different cardiac masses and its aetiology, MRI appearance of exuberant lipomatous growth of interatrial septum, Narrow complex tachyarrhythmia in the setting of cardiac mass and its medical management with rate control vs need of surgical interventions

Introduction

We present an interesting case of a large intracardiac mass in the setting of narrow complex atrial tachyarrhythmia where imaging features were suggestive of exuberant lipomatous growth of interatrial septum. The management was challenging given the multiple co-morbidity of obesity, suspected sleep apnoea and poor respiratory reserve. Initial rate control strategy for atrial arrhythmia was successful, and patient was conservatively managed with long term rate control measures.

Due to ongoing intermittent symptomatic atrial tachycardia an ablation was considered however, not attempted yet particularly due to anaesthetic concerns. Surveillance Echo and CT chest were considered most appropriate for the follow up of the intracardiac mass

Timeline

June 2018 Case introduced to local cardiology services for a Troponin leak and T wave inversion inpatient investigation with a TTE revealed an echogenic mass adjacent to right atrium

June 2018 Outpatient Computed tomographic and PET scans organised for the assessment of mass.

March 2019 Represented with new onset of Atrial Fibrillation with Rapid ventricular response

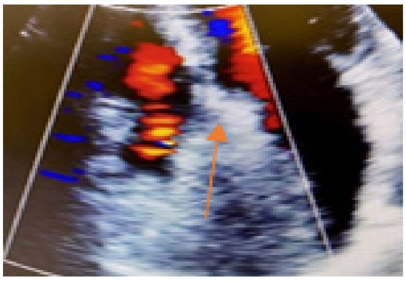
March 2019 Multiple imaging modalities including CT chest, Cardiac MRI, TOE, PET scan showing features of Exuberant lipomatous growth of interatrial septum

July 2019 A consensual decision made for medical management and serial imaging for follow ups after discussion with Cardiothoracic surgeons, Electrophysiologist and Imaging Cardiologists.

Case Presentation

49-year old Caucasian lady has been notified to our cardiology services approximately two years ago for an opinion and management of troponin leak of 0.68(<0.04) and TWI(T wave inversion) in the inferior leads. This was in the setting of cutaneous sepsis in the postoperative period for an incarcerated hernia requiring urgent surgical bowel resection and hernia repair. There was no suggestion of acute coronary syndrome based on clinical history. As a preliminary investigation an inpatient transthoracic echocardiogram(TTE) was organised revealing an echogenic mass adjacent to right atrium with no regional wall motion abnormalities(Fig 1.1) There was no hemodynamic compromise and she was planned for serial echocardiogram and outpatient computed Tomographic(CT) / PET (Positron Emission Tomographic)scans for further clarification of this mass.

Fig 1.1: TTE colour doppler images from apical 4 chamber focused on the inteatrial septum showing the mass with no colour communication



Within a few months, she re-presented with atrial fibrillation and rapid ventricular response requiring amiodarone infusion in addition to beta blocker therapy. A CT chest was performed which demonstrated a prominent fat density, posterior and lateral to the right atrium, measuring 9.2x4.5x3.6cm in maximum dimension, resulting in a significant attenuation of the SVC just proximal to its entry in to the right atrium. There was no evidence of collateral formation or of extrinsic compression of right subclavian/axillary veins at the level of the thoracic inlet. A new moderate volume pericardial effusion measuring 2.1 cm maximally inferiorly, was noted with evidence of SVC compromise and significant effacement of the right atrium. (Fig 2.1 and 2.2). These findings prompted revaluation of the known mass with TOE and PET scan.

Fig 2.1: Contrast enhanced axial images of chest through the interatrial septum showing a large mass compressing the right atrium with fatty density and no contrast enhancement

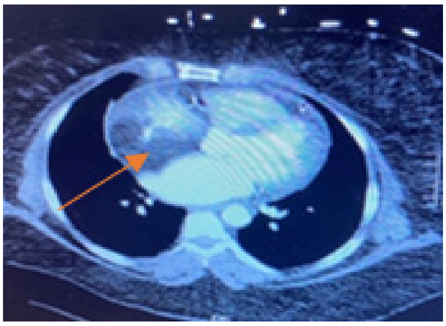


Fig 2.2: Small pericardial effusion is noted which prompted further evaluation of the mass

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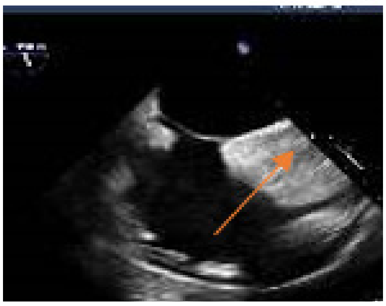
The TOE images confirm evidence of prominent lipomatous hypertrophy but no obvious compression of the superior vena cava. There was a small pericardial effusion with no evidence of tamponade physiology. (Fig 1.2 and 1.3)

Fig 1.2: Exuberant liomatpous growth seen on TOE - Bicaval image focused on the mass 3D.

Fig 1.3: Exuberant lipomatous growth on TOE (2D bi-caval image)



A 24 hour Holter monitoring in out-patient setting was performed to assess the rate control which revealed a predominantly sinus rhythm with no sustained atrial arrhythmia reassuring current medical therapy.

Cardiac MRI (3T Siemens Verio) confirmed a homogenous fatty dense lesion in relation to right atrium with no late gadolinium enhancement. Overall appearance was highly suggestive of exuberant lipomatous hypertrophy of interatrial septum. There was no evidence of late gadolinium uptake in the post contrast images. (Fig 3.1 – 3.6)

Fig 3.1: Exuberant growth spreading beyoind the interatial septum (SSFP retrospectively gated 4 Chamber view showing the mass)

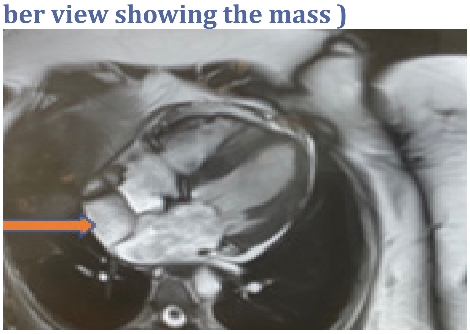


Fig 3.2: Black blood axial HASTE images immediately below the pulmonary bifurcation. The arrow shows narrowed SVC from the mass.

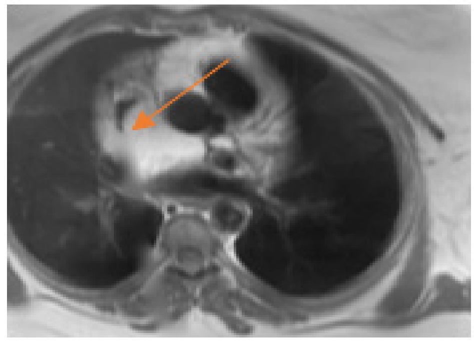


Fig 3.3: TSE T2 weighted images showing the mass with a smiilar density to subcutaneous fat

Fig 3.4: MRI first pass study showing no perfusion in the mass.

Fig 3.5: Nonenhancing lesion on LGE (3Ch , T1 PSIR image)

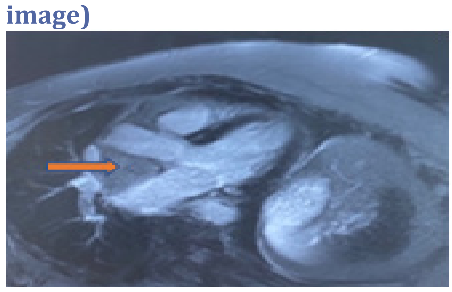
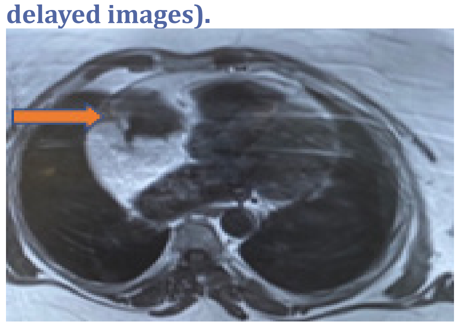


Fig 3.6: Nonenhacing lesion on LGE (Axial stack T1 delayed images).



The FDG -PET study did not reveal any tracer avidity favouring a benign pathology.

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Management options were discussed with the involvement of Cardiothoracic surgeons, Electrophysiologist and Imaging Cardiologist. The consensus was to continue to monitor the 'cardiac mass' given that there was no indication of surgical intervention. Biopsy was also deemed unnecessary or high risk given the patient's co-morbidities. Atrial tachycardia ablation for long term management of rhythm was considered. However, this had not been attempted due to fewer symptoms and patient's high anaesthetic risk.

DISCUSSION

Cardiac masses are rare but important cause of different pathology including arrhythmia, tamponade, pericardial constriction and effusion. Clinical imaging by echocardiography, ECG gated CT/MRI and PET scan has been on rise and frequently characterizing the location of masses and its relation to vital structures. Up to some extent these may be able to predict the tissue property, however histopathology is still the gold standard.(1, 2). Primary cardiac tumours are uncommon and most of them are benign tumours with the frequency(in descending order) of myxoma, Papillary fibroelastoma, Rhabdomyoma and Fibroma.(3) Malignant tumours are predominantly Sarcomas.(4) Lipomatous hypertrophy of interatrial septum is not a neoplasm rather a mass forming process in the atrial septum resulting from developmental aberrancy with entrapment of fat in the embryonic septum secundum. Its typically detected in 50s and more common in women(5). This is usually asymptomatic but may cause atrial arrhythmias and in extreme cases SVC obstruction. Although Septal thickening can be easily detected in Cardiac imaging it can be easily misinterpreted as infiltrative process like Sarcoidosis or Neoplasia. Surgical resection is usually not necessary but its close resemblance to neoplastic appearance has led to resection in few cases. (2)

Macroscopically the mass formed in Lipomatous hypertrophy is more prominent on Right atrium with the limbus portion of fossa ovalis most affected. The gross appearance is same as fatty tissue elsewhere. Its bright yellow in appearance and Atrial septal thickness is noted to be >1.5-2 cm in most cases. (5)Microscopically there is no capsule and mature/ brown fat cells are seen. Lack of encapsulation and presence of both mature and brown fat helps differentiating it from Lipoma.(6)

Transthoracic Echocardiogram is widely available and hence the first line investigation for the evaluation of cardiac masses. Moreover many of the cardiac masses are detected during the routine echocardiogram procedures. In many cases Echocardiogram can pick up the location, anatomical property and structural details of the masses which can lead to the differentials. With the evolution of the three dimensional echocardiography its role has been more pronounced.(7) The obvious limitation with this technique would be a limited field of view in large body habitus and underlying lung condition patients. Limited view of Right heart, mediastinum and extra cardiac masses are other limitations.

Transesophageal echocardiogram is used when in diagnostic dilemma with the addition of different planes for further assessment of lesion though it's an invasive procedure. Some masses do reveal specific echogenic property but the details of tissue characteristic is beyond the scope of TTE unless the advanced research technique such as ultrasonic backscatter or strain rate analysis is used. (8, 9). The strain rate analysis is a mathematical calculation where a velocity gradient is used to calculate the myocardial deformation. This test permits the differentiation of passive motion due to tethering from active deformation in contractile myocardium.(9)

Cardiac computed tomography is second most commonly used modality for the assessment of cardiac masses.(10, 11)Increased use of CT to assess coronary artery disease(CTCA) is leading to incidental findings of Cardiac masses.(12, 13). Advancement in technologies and Electrocardiographic gated scans are leading to better image qualities and frequent identification of cardiac masses(14, 15).CT can also provide information about vascularity by means of contrast enhancement, presence of calcification and presence of fat. Its known limitation would include exposure to radiation, and lower resolution as compared to Echo and MRI.(16)

Cardiac MRI is so far the most useful technique to study the property and characteristic of cardiac masses(17). It can be used to predict the likely malignancy of a cardiac mass(18, 19). Hoffman et al has derived a Cardiac MRI protocol to assess the different morphology, location, infiltrative pattern and contrast enhancement in patients(18). As with CT scan , MRI imaging also needs ECG gating and hence in the presence of significant arrhythmia the artefacts and image degradation are significant challenge(20). Most cardiac MR sequence needs a breath holding spell which can be particularly challenging in sick patients, however with the development of hardware and pulse sequences faster acquisition of images are getting obtained now. Respiratory navigator tracking methods are other alternative where free breathing is allowed during the acquisition of images. With the advancements in pacemaker and defibrillator manufacturing the MRI compatibility is on the rise in last few years(21).

CONCLUSION

Exuberant lipomatous growth of inter atrial septum is unique and interesting aetiology mostly present in middle aged women who are overweight or obese. They may predispose to atrial arrhythmia. The diagnosis is usually made with the use of Echocardiography, CT. In cases where this mimics tumour cardiac MRI would be useful for further anatomical location delineation and tissue characterisation. In extreme cases like ours FDG -PET study has been further useful particularly when the suspicion of malignancy is raised. Our case highlights the unique presentation of cardiac mass complicated with atrial fibrillation and pericardial effusion.

Conflict of interest

The Author has no conflict of interest regarding the publication of this report.

Consent

The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Abbreviation

CMR- Cardiac Magnetic resonance im aing
MDCT- Multi Detector Computed Tomo raphy
FDG-PET- Fluoro deoxy glucose, Posi tron Emission Tomography
TTE- Transthoracic Echocardiogram
TOE- Transesophageal Echocardiogram
SVC- Superior Vena cava
CTCA – CT Coronary Angiogram
LGE – Late Gadolinium Enhancement
SSFP- Steady State Free Precession
HASTE- Half fourier Single -shot Turbo spin-Echo

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