CLINICAL CASE - TEST YOURSELF | Thoracic & Cardiovascular Imaging

Myocardial infiltration as an incidental finding in a 64-year-old woman

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PART A

A 64-year-old female patient was referred by cardiology for further investigation, on account of abnormal findings on a routine cardiac ultrasound (US). Her medical history was unremarkable. She underwent Computerized Tomography (CT) (Fig.1) and Magnetic Resonance Imaging (MRI) (Fig. 2 a, b, c), which both confirmed the suspected US findings.

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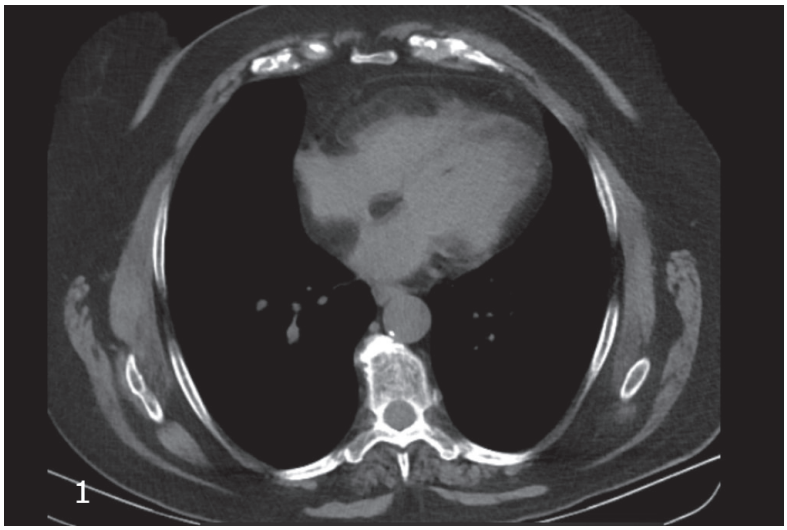
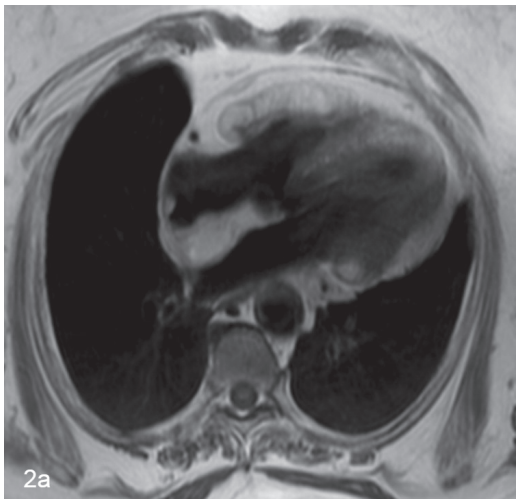


Fig 1. *Axial non enhanced CT.*



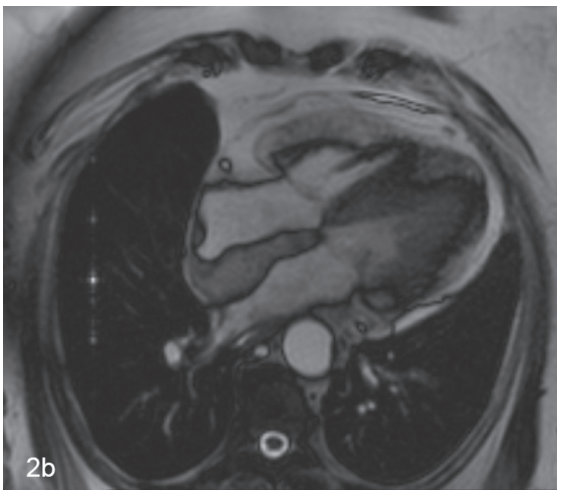


Fig 2. *Four chambers view MRI a.T1W b. T2W TFE c. STIR.*

PART B

Diagnosis: Lipomatous Hypertrophy of the Interatrial Septum (LHIS), with associated fatty replacement of the ventricular myocardium.

Lipomatous Hypertrophy of the Interatrial Septum (LHIS) is a rare benign condition, first described in 1964. It is more frequent in elderly, obese women and has an incidence report ranging from 1-8%, [3,5]. It is most often an asymptomatic condition, except in rare cases when it can cause arrhythmias or obstruction to nearby structures, such as the Superior Vena Cava (SVC). The cause of the arrhythmogenic function, is not fully understood, but it is thought to be due to interruption of conduction, leading to atrial fibrillation, atrial premature complexes, supraventricular arrhythmias, ectopic and junctional rhythm. A correlation between the thickness of the atrial septum and prevalence of arrhythmias has been reported. [4,6]. LHIS is characterized by the presence of excessive, unencapsulated fat depositions in the septum secundum, sparing the fossa ovalis (Fig. 1,2) [3]. The typical radiological characteristics of the interatrial septum are: a) dumbbell or hourglass appearance b) increased thickness (more than 20mm) c) low attenuation identical to fat in non contrast CT of the heart (Fig. 1) d) and in CMR hyperintensity on T1W images (Fig 2a), hypointensity on fat suppression images (Fig. 2c) and hypointense boundary between fat and myocardium (Fig. 2b) on steady state free precession images [1,2,3]. The above characteristics help differentiate LHIS from other causes, deeming the histological confirmation redundant [4]. The differential diagnosis includes cardiac tumours such as myocardial myxomas- rhabdomyomas (which lack significant amounts of fat), and lipomas [9]. Cardiac rhabdomyomas are more prevalent in children, with a history of Tuberous Sclerosis [9]. Myocardial lipomas are encapsulated collections of adipose tissue, arising most commonly on the sub-endocardium of the right atrium and the Left Ventricle (LV) [9, 10].

Fatty replacement of the ventricular myocardium can be a normal aging process or it can be attributed to other causes, such as Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC), post-myocardial infarction lipomatous metaplasia and post-corticosteroid treatment. It is important to differentiate between various etiologies, as they may change overall management. One can determine this by analyzing the anatomical and radiological features of the lesion, as well as the patient's medical history and age (Table 1).

In aging- related cardiac fat infiltration, the fat deposition is more often encountered in the sub-epicardial regions of the RV free wall (anterolaterally and apically) and it has a prevalence of 85% on histology reports of patients who died of unrelated non-cardiac causes ¬[5,7]. The thickness of the myocardium is often preserved, although it can rarely be thickened in cases of extensive fatty infiltration and involvement of the myocardium and sub-endocardium. Occasionally, fat foci may also be encountered in the Left Ventricle (LV). It is still unclear whether this fatty infiltration is associated with obesity and increased cardiovascular risk, however given the higher prevalence in older patients it is considered part of the normal aging process [7].

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC), is a condition, which histologically is characterized by replacement of the normal myocardium by fibrofatty tissue [2]. This can lead to arrhythmias and sudden cardiac death, with varying involvement of the myocardium depending on the extent of the disease. It is most often encountered in the age group between 15-20 years and involves the RV, accompanied by thinning and dilatation, starting from the epicardium and progressively extending and infiltrating into the endocardial layer. The LV and the intraventricular septum can also be involved in more advanced disease with the same pattern of infiltration [7]. Other important findings may include ventricular aneurysms, as well as segmental hypokinesia [8].

Post-myocardial infarction lipomatous metaplasia, is another common and well recognized condition, encountered often in the middle aged and elderly male population. It usually involves the LV and characteristically is confined within a coronary artery vascular territory. It always originates from the subendocardial myocardium and can extent transmurally, usually involving less than 75% of the myocardium [5]. This progress is thought to be a response to

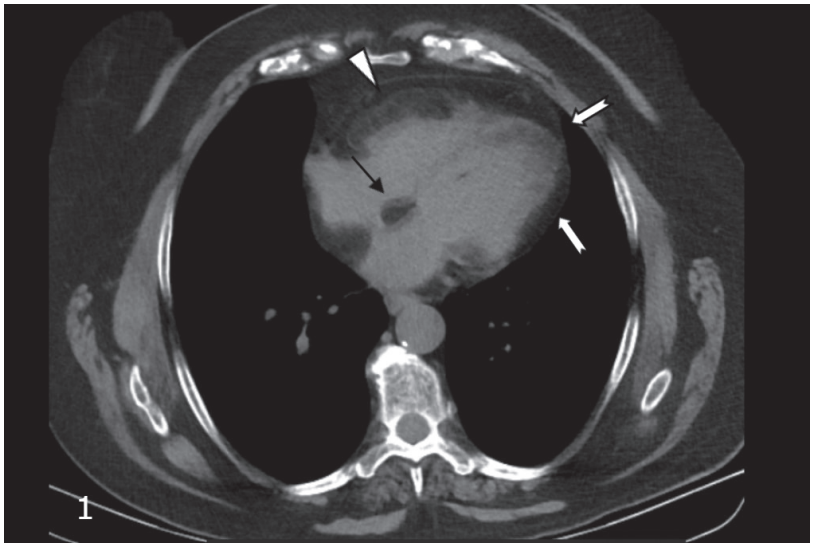
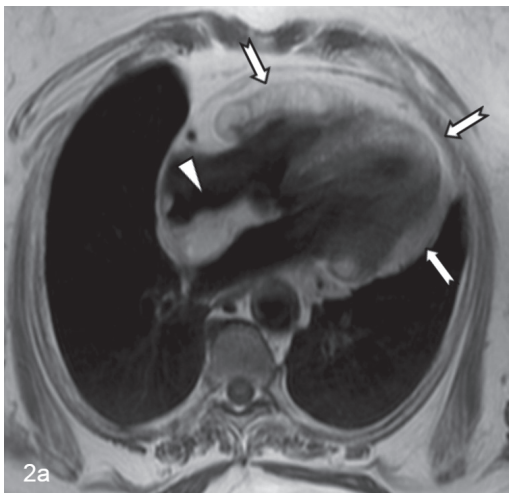


Fig 1. *Axial non contrast CT image demonstrate dumbbell hypertrophy of the interatrial septum (2cm) due to fat deposition (negative attenuation values) (→). Extensive intramyocardial fat deposition is also observed in the subepicardial and midmyocardial layers of RV free wall (▻), as well as in the apex and in the lateral wall of the LV (⟺).*



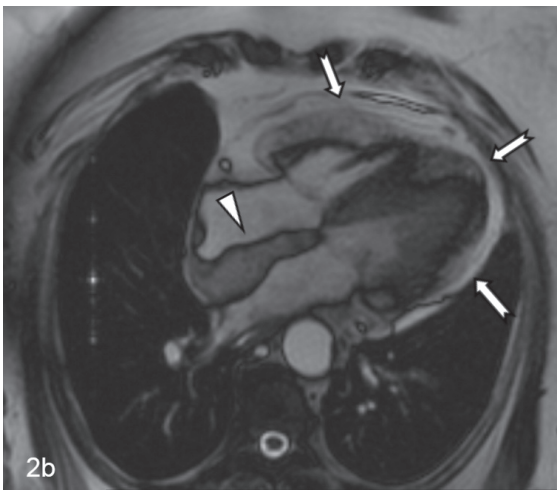


Fig 2. *MRI four chambers view images a.T1W b.T2W TFE and c. STIR. The intramyocardial (⟺) and interatrial (▻) fat accumulation is depicted on T1W and T2 TFE images with identical signal intensity to that of the fat (high signal intensity) and typically with signal drop of on the corresponding STIR image (intramyocardial (⟺), interatrial (▻) confirming the presence of adipose tissue.*

Table 1. Summary of key differential points.

Myocardial fat infiltration	Heart location	Pattern of infiltration	Age group	Additional findings
Age – related	Anterolateral and apical RV free wall, RVOT Rarely fat foci in LV and intraventricular septum in advanced cases	Subepicardial	Elderly	Myocardium usually maintains its thickness
ARVC	RV LV and intraventricular septum in advanced cases	Starting from epicardium and progressing to the endocardium	Young patients 15-20 years old	Thinning of the RV, with dilatation and aneurysms, segmental hypokinesia
Post- myocardial infarction lipomatous metaplasia	Confined within a coronary artery vascular territory, usually involving the LV	Subendocardial myocardium Can extent transmurally, usually involving less than 75% of the myocardium	Middle-aged/ elderly male	At least 6 months post MI
Lipomas	Predilection for right atrium and the LV	Sub-endocardium (50%)	Wide range, most common 40-60 years old	Encapsulated Usually solitary

hypoxia, initially leading to creation of scar tissue, which is subsequently populated by adipocytes. For this process to be noticeable in cross-sectional scans at least 6 months are required. From then onwards CT and MRI images can show progressive increase of fat deposition [5].

Treatment varies widely depending on the underlying condition. When cardiac fat depositions are an incidental finding, attributed to normal aging progress, there is no treatment/follow-up required, but the possibility of a serious underlying condition should always be considered.[4,5]. The patient's age, medical and family history, as well as associated cardiac findings and fat distribution patterns help guide towards the correct diagnosis and management of these patients.

In our case, the extensive fat deposition in sub- epicardial and mid- myocardial layers of both ventricles was depicted with typical radiological characteristics on both cardiac CT and MRI (Fig. 1,2). The diagnosis of age-related cardiac fat was set on account of normal cardiac function tests and an unremarkable medical history. No treatment or further follow up was recommended.

In conclusion, as shown in our case, LHIS can in rare cases be associated with extensive fatty replacement of the ventricular myocardium and pericardium [2]. The combination of CT and MRI can be employed in order to set the diagnosis. The more accurate tissue characterization, which is possible with MRI, can help in revealing additional, potentially concerning findings that may suggest a more sinister diagnosis, such as ARVC. **R**

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Conflict of interest
The authors declared no conflicts of interest.

KEY WORDS

Lipomatous hypertrophy of the interatrial septum (LHIS); Myocardial fat infiltration; Magnetic Resonance imaging; benign cardiac lesion

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