CASE REPORT



Lipomatous Hypertrophy of the Interatrial Septum: Occasional Finding following Control Angio-Tc in a patient with previous Custom-Made Multibranched Thoraco-Abdominal Aortic Endoprosthesis Surgery

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Abstract: Lipomatous hypertrophy of the interatrial septum is a cardiac anomaly characterised by an excessive accumulation of fatty tissue in the interatrial septum that is clinically silent in most cases and therefore often diagnosed incidentally. We report the case of a woman with previous multibranched custom-made thoracoabdominal aortic endoprosthesis surgery with lipomatous hypertrophy of the interatrial septum misdiagnosed during a previous multislice CT-angiography investigation. Previously considered as a rare occurrence, since its first autopsy finding in 1964, the number of cases of lipomatous hypertrophy of the interatrial septum diagnosed has been progressively increasing, also as a collateral finding, thanks to the ever-increasing use of echocardiography in cardiology and the development of ever-better diagnostic investigations (e.g., CT, MRI, PET/CT, etc.) aimed at assessing other problems, as in the case described.

Key words: Lipomatous hipertrophy of the interatrial septum, superior vena cava syndrome, atrial arrhythmias, sudden cardiac death, multislice CT.

Key messages:

- Lipomatous hypertrophy of the interatrial septum is a more common occurrence than previously thought.
- Lipomatous hypertrophy of the interatrial septum, which is usually asymptomatic, can cause obstructive phenomena of the right atrium, arrhythmias and even sudden cardiac death.
- CT studies of thoracic structures (lung parenchyma, thoracic aorta, etc.) may allow for the identification of lipomatous hypertrophy of the interatrial septum as a collateral finding.
- The correct and timely diagnosis of lipomatous hypertrophy of the interatrial septum appears to be increasingly simple thanks to the widespread use of multimodal imaging of the heart and thorax, even if performed with non-specific indications (post-covid chest check, etc.).

Introduction

Lipomatous hypertrophy of the interatrial septum (LHIS), first described by Prior in 1964 (1), is a nosological entity characterised by an infiltration of adipose tissue without capsular delimitation involving the interatrial septum (IS), in particular the limbus of the fossa ovalis (embryological septum secundum) but sparing the fossa ovalis and the membranous atrioventricular septum (2).

LHIS, which is considered a benign nosological entity, is clinically silent in most cases

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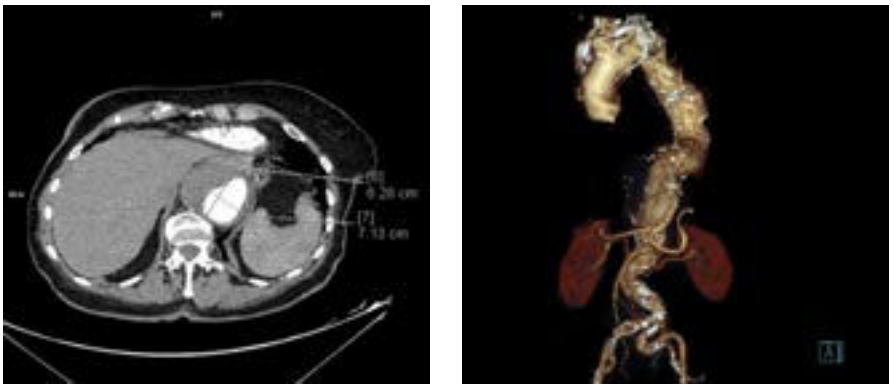


Fig. 1 - Multislice CT angiography scan investigation performed for surgical planning in 2018. Dilatation of the thoraco-abdominal aorta with maximum transverse diameter of the aneurysmal sac near the diaphragmatic hiatus, site of diffuse and irregular parietal thrombotic apposition (A). 3D Volume Rendering reconstruction demonstrating the involvement of the aneurysmal dilatation of the part of the aorta where the celiac tripod, the superior mesenteric artery and the two renal arteries emerge(B).

and often diagnosed incidentally.

At the same time, however, it can be a potentially serious condition causing, in rare cases, right atrium obstructive phenomena, arrhythmias and even sudden cardiac death.

Considered as an uncommon occurrence in the past, the development of ever-improving diagnostics and the increasing use of investigations such as CT scans for studying chest structures (the first non-autopsy, in vivo diagnosis of LHIS using CT scans was made in 1982) (3) is gradually leading to an increase the number of cases identified.

Case Report

We present the case of an 82-year-old woman with lipomatous hypertrophy of the interatrial septum previously misdiagnosed and incidentally found during a diagnostic check-up with thoraco-abdominal aorta multislice CT angiography scan.

In May 2018, following the finding of thoraco-abdominal aortic aneurysm, the Patient was preoperatively screened for adequate surgical planning and underwent thoraco-abdominal aorta multislice CT angiography scan with LightSpeed VCT - GE Medical System CT equipment (slice: 1.25/0. 65 mm) in which aneurysm of the thoraco-abdominal aorta was diagnosed with involvement of the origin of the celiac tripod, the superior mesenteric and the renal arteries bilaterally with maximum transverse dimensions of the aortic aneurysmal sac of 63 x 71 mm near the diaphragmatic hiatus (***Fig. 1***).

Following this diagnosis, in the same year, the Patient underwent a custom-made multibranched thoraco-abdominal aorta endoprosthesis surgery.

In June 2022, following a specialist vascular surgery examination, the Patient was asked to undergo diagnostic monitoring of the previously performed endovascular treatment that had no longer been checked in the post-surgical phase. Therefore, in the same month, the patient underwent a multislice CT angiography scan of the thoraco-abdominal aorta at the UOC of Diagnostic Imaging of the Celio Military Hospital in Rome, using a Somatom Definition Flash – Siemens CT scanner (slice: 1.0/0.6 mm, Kv 120, mAs 182, 80 ml of Iomeron 400 – Bracco with a flow rate of 5 ml/s).

From a vascular point of view, the multislice CT scan documented (***Fig. 2***):

- regular positioning and patency of the thoraco-abdominal endoprosthesis with proximal attachment at the level of the distal part of the aortic arch, immediately downstream of the emergence of the epiaortic vessels, and distal attachment at the level of the aortic carrefour without involvement of the iliac arteries bilaterally;

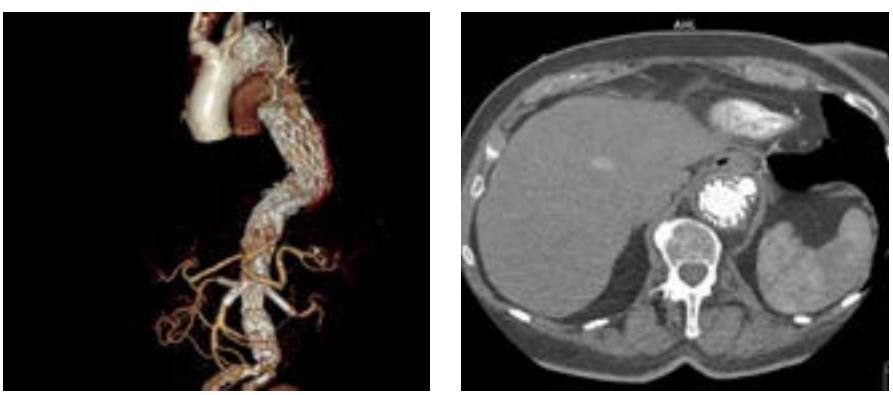


Fig. 2 - Multislice CT angiography scan investigation performed for post-surgical follow-up in 2022. Presence of aortic endoprosthesis with regular positioning of both main aortic branch and visceral prosthetic branches (A). Evident reduction in the size of the aneurysmal sac (B).

- regular positioning and patency of the visceral prosthetic branches of the celiac tripod, the superior mesenteric and the two renal arteries;
- absence of signs of periprosthetic endoleak;
- reduction in the size of the aneurysmal sac.

The subsequent evaluation of the extravascular structures revealed the presence, at the IS, of a coarse hypodense mass (with adipose density) measuring 49 x 30 x 44 mm, without capsule and without evident contrast enhancement (***Fig. 3***), which was responsible for compression phenomena towards the right atrium and the superior vena cava at the entrance to the atrium (***Fig. 4***). In relation to the structural, densitometric pre- and post-contrast characteristics and the localisation, the diagnosis of lipomatous hypertrophy of the interatrial septum was therefore made.

The retrospective re-evaluation of the multislice CT scan carried out pre-surgery in 2018, aimed at detecting the presence or absence of LHIS and the possible evolutionary behaviour over time (e.g., dimensional increase, morphological change, etc.) compared with the recent 2022 investigation, confirmed the existence of lipomatous hypertrophy of the interatrial septum on the previous examination, which was slightly smaller in size and amounted to approximately 38 x 23 x 43 mm (***Fig. 5***).

The LHIS was more difficult to identify in the 2018 investigation due to heart motion artefacts and radiation beam hardening artefacts caused by the presence, during post-contrastographic acquisition, of contrast medium hyperconcetration in the superior cava and within the right atrium (***Fig. 5***). This problem is becoming less and less frequent thanks to the progressive development of CT equipment which, by increasingly reducing acquisition times, determines the presence of minimal or absent artefacts from heart movement and allows acquisition times to be optimised with a consequent reduction in the quantity of contrast medium for computed tomography investigations.

Following this diagnosis, the Patient was referred for a specialist cardiac surgery examination for an appropriate clinical-anamnestic correlation, in order to assess the presence of any symptoms attributable to atrio-caval compression phenomena or arrhythmias that cannot otherwise be explained, and for the planning of the subsequent diagnostic/therapeutic procedure.

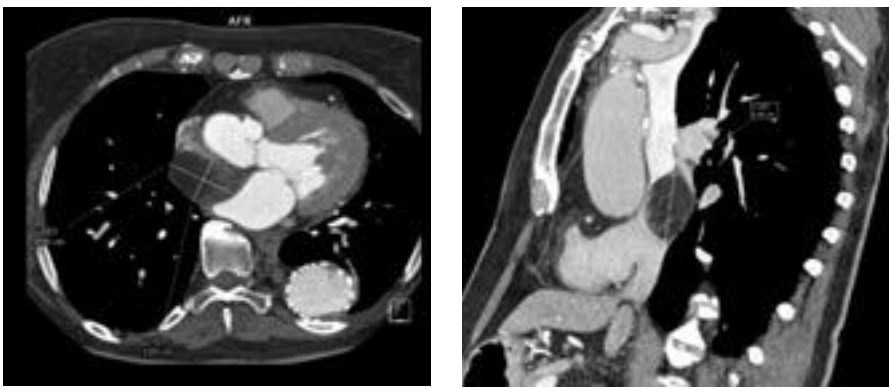


Fig. 3 - Multislice CT angiography scan investigation performed for post-surgical follow-up in 2022. The examination shows the presence of a lipomatous hypertrophy of the interatrial septum with a maximum transverse dimension of 49 x 30 mm (A) and a longitudinal dimension of 44 mm (B).

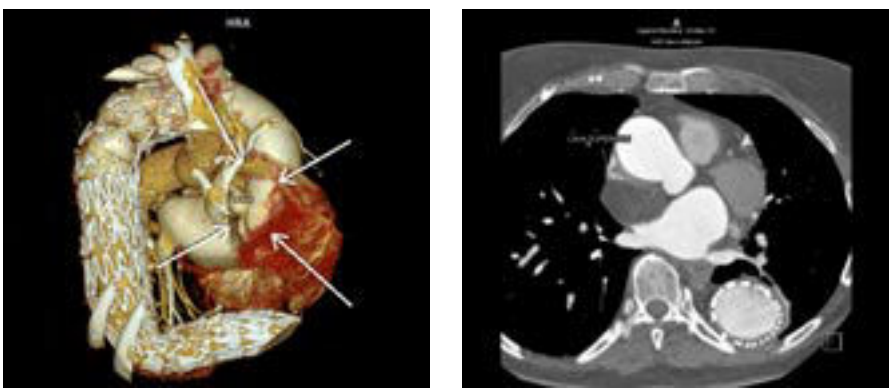


Fig. 4 - Multislice CT angiography scan investigation performed for post-surgical follow-up in 2022. The adipose lesion, which lacks contrast enhancement, causes compression against the superior cava (arrow) at the entrance to the atrium (A). The 3D Volume Rendering reconstruction documents the 'void' (***) caused by the lipomatous hypertrophy of the interatrial septum (margins indicated by the white arrow) responsible for compression on the atrium and superior cava (B).

Discussion

Lipomatous hypertrophy of the interatrial septum (LHIS), first described post-mortem by Prior in 1964, is a cardiac anomaly characterised by an excessive accumulation of adipose tissue in the IS with a thickness greater than 10 mm (4)

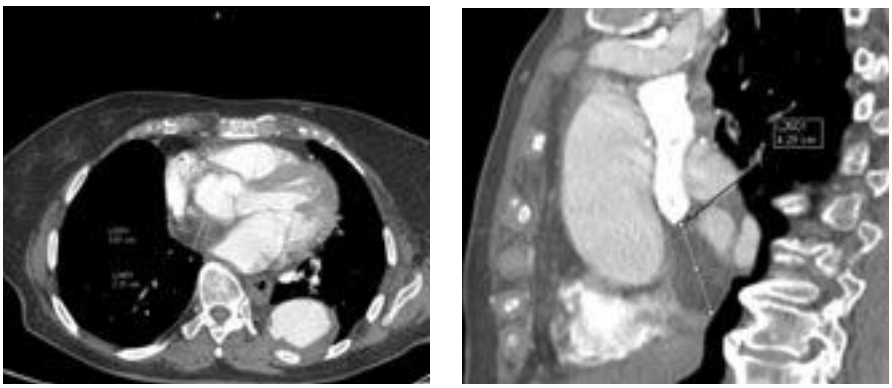


Fig. 5 - Multislice CT angiography scan investigation performed for surgical planning in 2018. The retrospective evaluation of the previous examination documents the pre-existence of lipomatous hypertrophy of the interatrial septum with a maximum size of 38 x 23 x 43 mm (A, B). In this examination, the identification of LHIS is made difficult by the presence of heart motion artefacts and artefacts due to the hardening of the radiation beam by the hyperconcentration of the contrast medium within the superior cava and right atrium (B).

and specifically at the level of the limbus of the fossa ovalis (embryological septum secundum), sometimes associated with a thickening of the crest terminalis but with constant sparing of the fossa ovalis. This particular distribution of adipose tissue often causes the lesion to have an 'hourglass' or 'dumbbell' shape, which, according to some authors, is quite characteristic (5, 6).

The development process of this adipose accumulation, which is often more evident in the right atrium (the limbus is a right atrial structure) – although its underlying mechanism being unknown to date –, appears to be correlated with obesity (7), to affect women more (8, 9) and to be more frequent with advancing age (10). There are studies that also hypothesise an association between LHIS and parenteral nutrition (7).

The actual incidence of LHIS in the population is unknown.

Data from the few studies performed documents an incidence of 1% in autopsy studies (11, 12), 8% in a study using transthoracic echocardiography (13) and 1% to 2.2% in studies using computed tomography (14, 15). This evidence therefore shows that lipomatous hypertrophy is a more common occurrence than initially thought.

Although it is considered to be a benign cardiac abnormality that runs predominantly asymptomatically, it can also be responsible for serious consequences ranging from superior vena cava syndrome (16) to the onset of arrhythmias, even malignant ones, and even sudden cardiac death. Whilst the causes of superior cava syndrome can be easily identified in the compressive phenomena that the adipose mass can exert on the atrio-caval structures, the mechanisms underlying the onset of arrhythmias (the first description of an association between LHIS and arrhythmias by Kluge dates back to 1969) (17), although some hypotheses have been proposed, are still not known (18).

Therefore, given its predominantly asymptomatic course, LHIS is mostly found occasionally or as a hyperechogenic mass in the atrial site during the transthoracic echocardiographic examination, which nowadays is increasingly an integral part of the cardiological examination, or as a collateral finding during CT, MRI or PET/CT examinations (19) also performed with different clinical indications (Covid, thoracic aorta aneurysm, oncological follow-up, etc.).

At the time of identification, especially when LHIS is detected by transthoracic ultrasound examination, it must first be diagnosed differentially together with certain atrial cardiac tumours (lipoma, liposarcoma, myxoma, etc.). Therefore, for a final diagnosis to be reached, further in-depth examinations with second level investigations are necessary to better characterise the lesion, such as cardiac MRI with contrast medium or cardiac CT with contrast medium.

Once the diagnosis has been confirmed, i.e., after the tumour nature of the mass has been excluded, lipomatous hypertrophy of the inter-atrial septum, although mostly asymptomatic throughout life – because of the even serious complications for which it may be responsible (superior cava syndrome, arrhythmias and sudden cardiac death) (15) –, requires optimal diagnostic-therapeutic management that involves adequate clinical-anamnestic correlation and regular follow-up. Surgical resection and reconstruction of the septum, formerly more common, is nowadays reserved only for rare cases of lesions causing circulatory obstruction (16) or leading to malignant arrhythmias (20).

Since the first autopsy description, the number of diagnoses of LHIS has undoubtedly increased, both because of the increasing knowledge in the cardiological field of this entity and the progressive development of diagnostic investigations that allow its identification and correct diagnosis.

In this respect, since 1964, not only have targeted cardiological diagnostics (trans-

thoracic echocardiography, transesophageal echocardiography, cardiac CT and cardiac MRI) increased and improved, but so has the use of investigations (CT, MRI or PET/CT) performed with other indications, which allow, as in the case reported in this article, for the identification of LHIS as a collateral finding.

Despite the progressive increase, the number of diagnoses is still presumably below the actual incidence of LHIS. In imaging, if one excludes a certain limited number of specialists with expertise in cardiology diagnostics, LHIS constitutes a nosological entity whose knowledge is limited and therefore, particularly in the presence of smaller lesions, at risk of being misrecognised.

Conclusions

LHIS is a nosological entity that is considered benign, not without its problems, even serious ones; knowledge of it, combined with modern diagnostics (echocardiography, CT, MRI, PET/CT, etc.), allows for an increasing number of cases to be identified.

A precise and timely diagnosis ensures optimal diagnostic and therapeutic management which, in most cases, involves regular clinical and anamnestic correlation and follow-up, reserving surgical intervention for only a few rare cases.

Disclosures:

The Author declares that he has no relationships relevant to the contents of this paper to disclose.

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