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INTERMEDIATE

CASE REPORT: CLINICAL CASE

Lipomatous Hypertrophy of the Interatrial Septum Manifesting as Third Degree Atrioventricular Block

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ABSTRACT

A 55-year-old patient was found to have complete heart block during preoperative assessment. Cardiac magnetic resonance imaging revealed an interatrial mass suggestive of primary cardiac tumor. Extensive evaluation including intracardiac biopsy and finally open resection revealed lipomatous hypertrophy masquerading as tumor. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:2235–9) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 55-year-old patient with a history of hypertension and osteoarthritis presented for pre-operative evaluation for total knee replacement. Assessment included an electrocardiogram, which revealed sinus rhythm with complete heart block (CHB) and a junctional escape at 50 beats/min (**Figure 1**). Initial examination was notable for a blood pressure of 180/86 mm Hg and occasional cannon atrial waves. A transthoracic echocardiogram showed a left ventricular ejection fraction of >65%, hypertrophied interatrial septum (IAS), and a 2-cm circumferential pericardial effusion with concerns for infiltrative heart disease (**Figure 2A**, Video 1). Cardiac magnetic resonance imaging (MRI) revealed a large, well-circumscribed, sessile IAS mass extending into the cranial portion of the left atrium, aortic root, main pulmonary artery, and mediastinum (**Figure 2B**) with delayed perfusion (Video 2).

LEARNING OBJECTIVES

- To review the differential diagnosis of intracardiac masses.
- To outline the diagnostic options available for assessment of intracardiac masses.

PAST MEDICAL HISTORY

This patient had a history of hypertension, osteoarthritis, depression, and obesity (body mass index, 30.9 kg/m^2). Surgical history included Cesarean section delivery and laparoscopic Roux-en-Y gastric bypass surgery.

DIFFERENTIAL DIAGNOSIS

Initial differential diagnosis included primary cardiac sarcoma, cardiac or invasive lymphoma, atrial myxoma, angiosarcoma, metastatic disease, invasive

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ABBREVIATIONS AND ACRONYMS

CHB = complete heart block

IAS = interatrial septum

LA = left atrium

LASH = lipomatous hypertrophy of the interatrial septum

MRI = magnetic resonance imaging

primary pericardial mesothelioma, or inflammatory pseudotumor.

INVESTIGATIONS

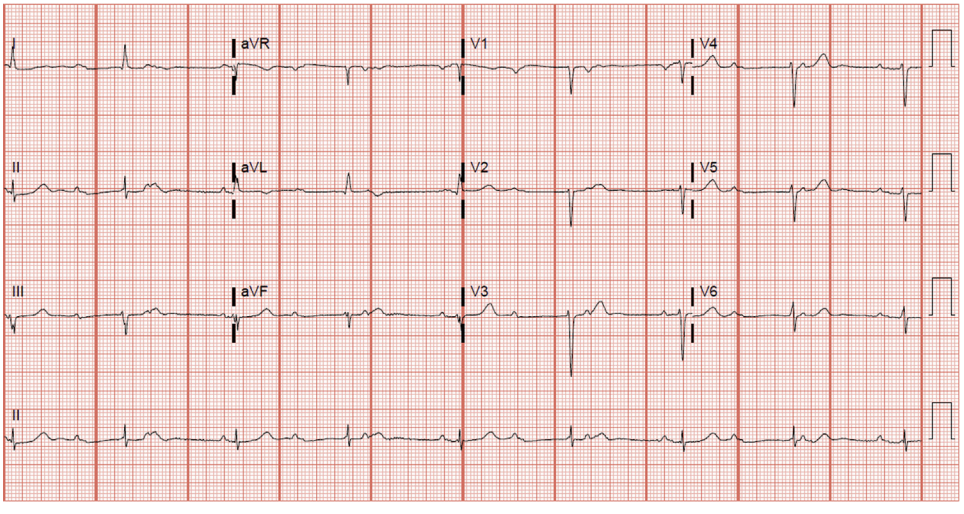
Computed tomography angiography of the heart confirmed an infiltrative cardiac mass involving the IAS with complex pericardial effusion and apparent pericardial masses (Figure 2C). Pericardial window and fluid analysis was performed showing mixed inflammatory cells (86% lymphocytes; 16% B cells; Kappa:Lambda 1.9:1; T cells 70%; and CD4:CD8: 9.1:1), mesothelial cells, and mesothelial-lined fibrous tissue consistent with pericardium. Fluorodeoxyglucose positron-emission tomography showed mildly increased metabolic activity along the IAS, aortic root, and ascending aorta corresponding to infiltrative soft tissue (Figure 3). There were no other abnormal areas of fluorodeoxyglucose uptake.

Intracardiac echocardiography (ICE)-guided atrial septal biopsy was performed. The right femoral vein was accessed with an 8.5-F short sheath and 8.5-F medium curl Agilis (St. Jude Medical, Minnetonka, Minnesota). An 8-F AcuNav ultrasound catheter (Siemens Medical Solutions, USA, Inc., Mountain View, California) was advanced into the right atrium through the short sheath. A 2.4 mm × 50 cm nonfenestrated endomyocardial biopsy forceps (Argon Medical Device, Inc., Athens, Texas) was inserted through the deflectable Agilis sheath and guided towards the IAS using ICE guidance (Figure 4). Several biopsy specimens were obtained and reviewed by both our internal pathology department and the Mayo Clinic (Rochester, Minnesota). This showed strips of endocardium with adherent fibrin-rich thrombus and some underlying endocardial inflammation (Figure 5). Fibrin-rich thrombus may be seen on or even adjacent to mass lesions; therefore, it was not possible to conclude that the sampling was representative of the entire mass. Electrophysiology study was not pursued given the clear evidence of CHB.

Ultimately, the patient underwent excision of the intracardiac mass and excisional biopsy of multiple pericardial and periaortic sites via median sternotomy under cardiopulmonary bypass. Initial specimen biopsy showed reactive inflammatory tissue. Tissue obtained from around the aorta came back as inflammatory, potentially granulomatous type without any evidence of malignancy. The IAS mass was a friable, thickened inflammatory looking mass and not a discrete tumor; however, 18 biopsy specimens had been taken at that point.

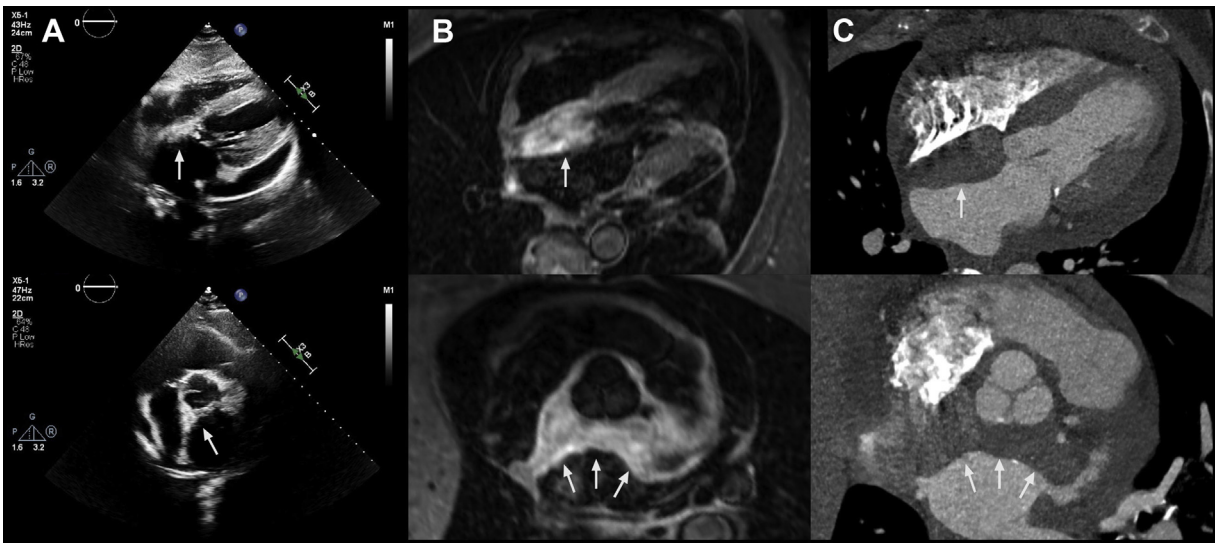
Pathology specimens from the pericardium showed histiocytes, giant cells, markers of chronic and acute inflammation including eosinophils, focal necrosis, and fibrin. There was no amyloid deposition (negative Congo Red stain), and no mycobacteria or fungal organisms. The mass consisted of reactive bland fibrous tissue, chronic inflammation, fibrin, myocytes, and adipocytes suspicious for lipomatous hypertrophy of the interatrial septum (LASH). Final pathology review

FIGURE 1 Electrocardiogram



Electrocardiogram showing complete heart block.

FIGURE 2 Cardiac Mass Multimodality Imaging



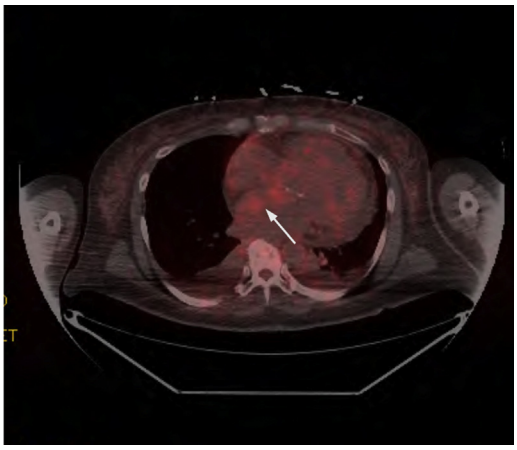
Interatrial septum mass with local extension **(white arrows). (A)** Transthoracic echocardiogram. **(B)** Cardiac magnetic resonance image. **(C)** Cardiac computed tomography angiography.

was consistent with inflamed pericardium and LASH with a reactive benign component.

MANAGEMENT

After resecting the interatrial septal mass, the atrial septal defect was repaired. A dual chamber pacemaker was placed due to persistent CHB and risk of invasion into the bundle of His. Based on final pathology, no further oncologic therapies were warranted.

FIGURE 3 Fluorodeoxyglucose-Positron-Emission Tomography Imaging



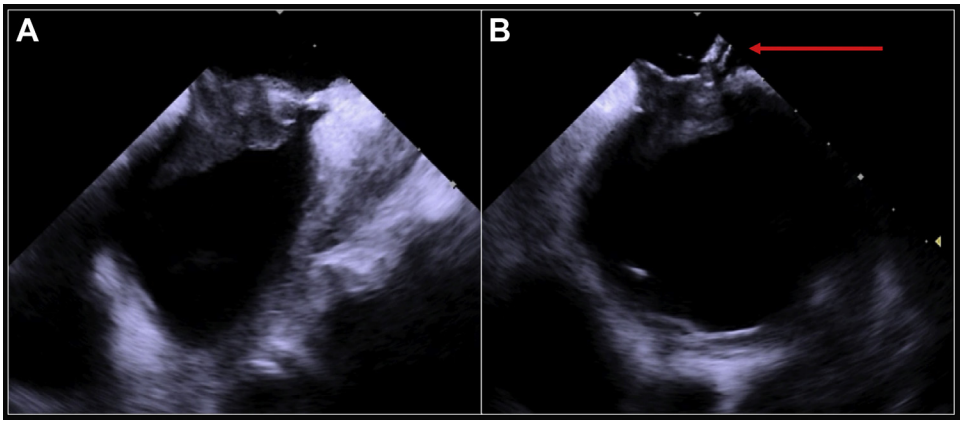
Increased metabolic activity along the interatrial septum **(white arrow).**

DISCUSSION

LASH is typically a benign condition that can be complicated by atrial arrhythmias. The mass is noncapsulated unlike cardiac lipomas, which are encapsulated and contain few myocytes (1). It can be incidentally noted on transthoracic echocardiogram while potentially confirmed by computed tomography (CT) or MRI. The utility of multimodality imaging to analyze the IAS has been previously described (2). In this case, both CT and MRI were highly concerning for a malignant process prompting the need for a tissue specimen. Specifically, the mass was isointense compared to the myocardium under T1 imaging and did not saturate during T1 imaging with fat saturation. When examined with T2 short tau-inversion recovery, the IAS mass was hyperintense to the myocardium (**Figure 2B**). Based on the enhancement pattern, sarcoma was thought to be the most likely diagnosis. Furthermore, imaging confirmed extension of the underlying process into the ventricular outflow tract and aortic root, and CHB suggested invasion into the specialized conduction fibers of the atrioventricular (AV) node.

Prospective investigation has shown that LASH has an estimated incidence of 2.2% (3). It has been

FIGURE 4 Intracardiac Echocardiography–Guided Endomyocardial Biopsy

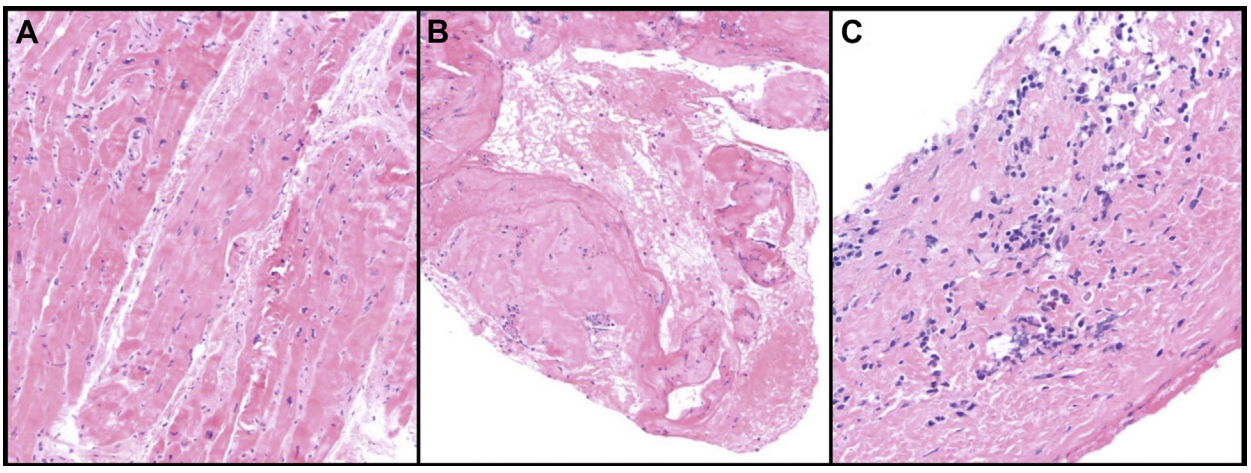


(A) Intracardiac echocardiogram from the right atrium viewing the mass-like interatrial septum. **(B)** The endomyocardial biopsy forceps are guided towards the interatrial septum with the help of intracardiac echocardiogram and a deflectable sheath **(red arrow).**

suggested that LASH can cause AV block; however, to our knowledge, ours is the first published case in which the inflammatory milieu included extensive anterior invasion into the AV node resulting in complete heart block (1). CT studies typically show a homogenous mass of fat attenuation without any significant contrast enhancement as well as sparing of the fossa ovalis and constriction of the central septum resulting in a characteristic "dumbbell" shape of the mass (3,4). In contrast, our patient had an enhancing lesion that appeared infiltrative and did not spare the fossa ovalis. Finally, it has been previously suggested that LASH can be associated with recurrent pericardial effusion, although data is limited (5).

Intracardiac masses present diagnostic challenges because of the potential difficulty in safely obtaining a tissue diagnosis. Based on autopsy data, cardiac tumors are rare, with a 0.056% incidence of primary tumors and 1.23% of secondary cardiac tumors (6). Accurate tissue diagnosis is paramount to prognosis and formulating a treatment strategy because of the potential lethality of cardiac tumors. ICE-guided endomyocardial biopsy has been previously described (7,8). A negative ICE-guided biopsy still

FIGURE 5 Endomyocardial Biopsy Pathology



Intracardiac echocardiogram-guided intra-atrial biopsy specimens. **(A)** Myocytes with hypertrophy and loose fibrosis. **(B)** Fibrin. **(C)** Small lymphocytes in loose fibrous background.

merits open biopsy; however, it is a useful diagnostic tool that can spare excisional biopsy and complex reconstruction.

FOLLOW-UP

Once final tissue diagnosis was confirmed, the patient received a dual-chamber pacemaker with routine device wound check and follow-up appointments. There is no definitive recommendation for follow-up cardiac imaging.

CONCLUSIONS

This case shows the extensive workup that can be required to accurately diagnose cardiac masses. A multidisciplinary approach is helpful and allows a stepwise approach to diagnosis ranging from least to most invasive. Finally, LASH is capable of conduction system invasion leading to CHB, highlighting the serious consequences of an otherwise benign lesion.

AUTHOR DISCLOSURES

All authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS cardiac magnetic resonance, cardiac mass, cardiovascular disease, echocardiography, imaging, interatrial septum, intravascular ultrasound, lipomatous hypertrophy

APPENDIX For supplemental videos, please see the online version of this paper.