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ADVANCED

CASE REPORT: CLINICAL CASE

Preoperative Transcatheter Diagnosis of Right Atrial Hemangioma



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ABSTRACT

A 34-year-old man with unremarkable past medical history presents with night sweats and a recent diagnosis of intracardiac mass. The initial diagnostic workup did not provide a definitive diagnosis, so a cardiac biopsy under intracardiac echocardiography guidance was performed, revealing a hemangioma, which was then successfully resected. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2023;15:101857) © 2023 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/>).

Tumors affecting the heart are a rare group of pathologies, secondary tumors originating from other organs being the most common type. Primary cardiac tumors, in particular, account for only 0.3% to 0.7% of all tumors of the heart, and, of these, 25% are malignant.¹

Cardiac hemangioma is a benign tumor originating from endothelial cells. It accounts for only 2% of primary cardiac tumors and is microscopically characterized by vascular channels. Because of the rarity of the pathology and its histology, usually the final diagnosis is performed on the surgical specimen,

mainly because of the high risk related to a biopsy performed on a highly vascularized mass.

We report here the first case of a preoperative diagnosis with cardiac biopsy under intracardiac echocardiography guidance (ICE) of a right atrial hemangioma.

MEDICAL HISTORY

An otherwise healthy 35-year-old man presented to an emergency unit because of a 1-month history of serotine low-grade fever and nocturnal sweats. On admission, arterial pressure was 112/70 mm Hg, cardiac rate was 86 beats/min and oxygen saturation was 98%. Because an oncological pathology was suspected, he previously had a total-body contrast computed tomography (CT) scan that showed a neoformation in the right atrium with a hypodense feature without contrast enhancement (**Figure 1**). On admission, he denied weight loss, itching, dyspnea, chest pain, or palpitations, and no prior family history of cardiovascular or malignant diseases.

LEARNING OBJECTIVES

- To understand the value of a definitive diagnosis of a cardiac mass.
- To highlight the feasibility of a safe vascular tumor biopsy under intracardiac echocardiography guidance.

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**ABBREVIATIONS
AND ACRONYMS****CT** = computed tomography**ICE** = intracardiac
echocardiography guidance**DIFFERENTIAL DIAGNOSIS**

Given the patient's history of fever and nocturnal sweats associated with a mass, the primary diagnosis was an oncological pathology; in particular, a systemic disease was suspected at first. Other differential diagnoses included clot, vegetation, other infective mass, or an imaging pitfall (eg, Chiari network).

INVESTIGATIONS

After hospital admission, the patient underwent transesophageal echocardiography, which allowed detailed evidence of the intracavitary mass in the right atrium (**Figure 2**). The mass had grossly a round feature with a radial dimension of 60 × 67 mm, showing nonhomogeneous echogenicity. It had a broad base of attachment to the antero-superior wall of the right atrium with initial signs of superior vena cava flow obstruction (**Video 1**). No specific anomalies were found on electrocardiogram.

A total-body positron emission tomography-CT scan was performed to exclude a disseminated disease. A focus of hyperactivity was found in the left hemipelvis, which was later revealed by colonoscopy to be a villous adenoma with low-grade dysplasia. At positron emission tomography-CT scan, the cardiac

mass showed a central photopenic area and a peripheral hyperactive rim (**Figure 3**).

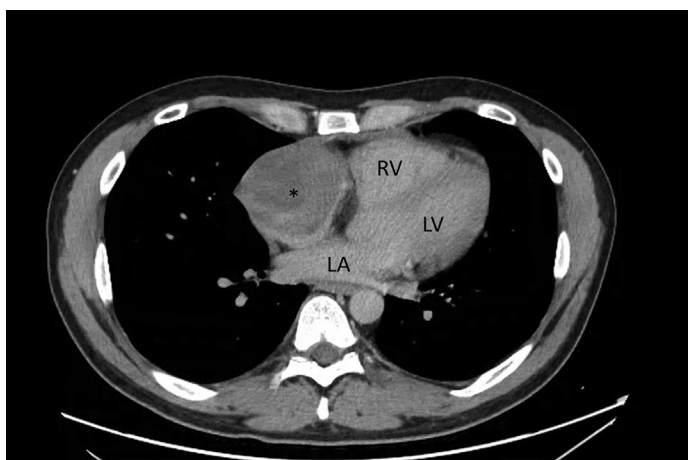
To obtain a specific diagnosis of the cardiac mass, endomyocardial biopsy was performed from a right femoral vein access using a Cordis Biopsy Forceps 7F. To guide the procedure, an ICE guide probe (Abbott View Flex 9F) was advanced in the right atrium via the left femoral vein (**Figure 4**). All the samples were collected under direct ICE vision to confirm the correct position of the bioprobe (**Video 2**). The microscopic examination showed fibrosis and a fibrin-hematic tissue with signs of organization. A fragment of tissue showing dilated vessels with thin walls and CD31-positive endothelial cells also could be seen. No atypical cells were found in the specimen (**Figure 5**). The diagnosis of right atrial cavernous hemangioma was then made.

MANAGEMENT

After a multidisciplinary evaluation, the patient was referred to our cardiac surgery unit to undergo surgical resection of the right atrial neof ormation. After median longitudinal sternotomy, a reactive pericarditis was observed. The right atrial mass, located in the superior-lateral wall of the atrium, was clearly distinguishable from the myocardial tissue because of its increased density (**Figure 6A**). A right atriotomy was then performed and the mass, which measured about 80 × 40 mm, was resected together with a huge, thickened portion of atrial wall and the nearest endocardium, up to the atrio-caval junction (**Figure 6B**). The interatrial septum and the tricuspid valve were not involved and were preserved. The right atrium was reconstructed using a bovine pericardial patch by a continuous running suture with 4/0 polypropylene (**Figure 6C**). The procedure was completed by a wide anterior pericardiectomy.

DISCUSSION

Cardiac hemangioma is a particularly uncommon histotype among primary cardiac tumors. This tumor is usually located in the right atrium and is asymptomatic.² A presumptive diagnosis can be made with imaging exams such as echocardiography, cardiac CT, cardiac magnetic resonance imaging, or coronary angiography in which it can show a characteristic tumor blush sign.³ Nonetheless, the definitive diagnosis is based on the microscopic evaluation, which is generally performed on the explanted specimen. When a vascular tumor is suspected, a preoperative biopsy is generally deferred because of high

FIGURE 1 Total-Body Contrast CT Scan

An intracardiac hypodense mass can be observed in the right atrium. CT = computed tomography; LA = left atrium; LV = left ventricle; RV = right ventricle. *Right atrial mass.

bleeding risk of this procedure. For this reason, the possibility of performing a guided biopsy represents an extremely useful tool to direct the sampling and detect any complications of the procedure.

Here we report a case of successful preoperative histopathological diagnosis of a right atrial hemangioma using cardiac biopsy under ICE guidance. To the best of our knowledge, this is the first reported case of a definitive presurgical histological diagnosis of cardiac hemangioma, by means of a biopsy performed during the cardiac catheterization with ICE guidance. ICE is an emerging imaging modality that allows a high-resolution visualization of cardiac structures, as well as an early recognition of any complications during catheterization (eg, bleeding).⁴ Because of its increased patient tolerance and lack of need for general anesthesia, this technique started to replace transesophageal echocardiography for guiding interventional procedures. The possibility to obtain a

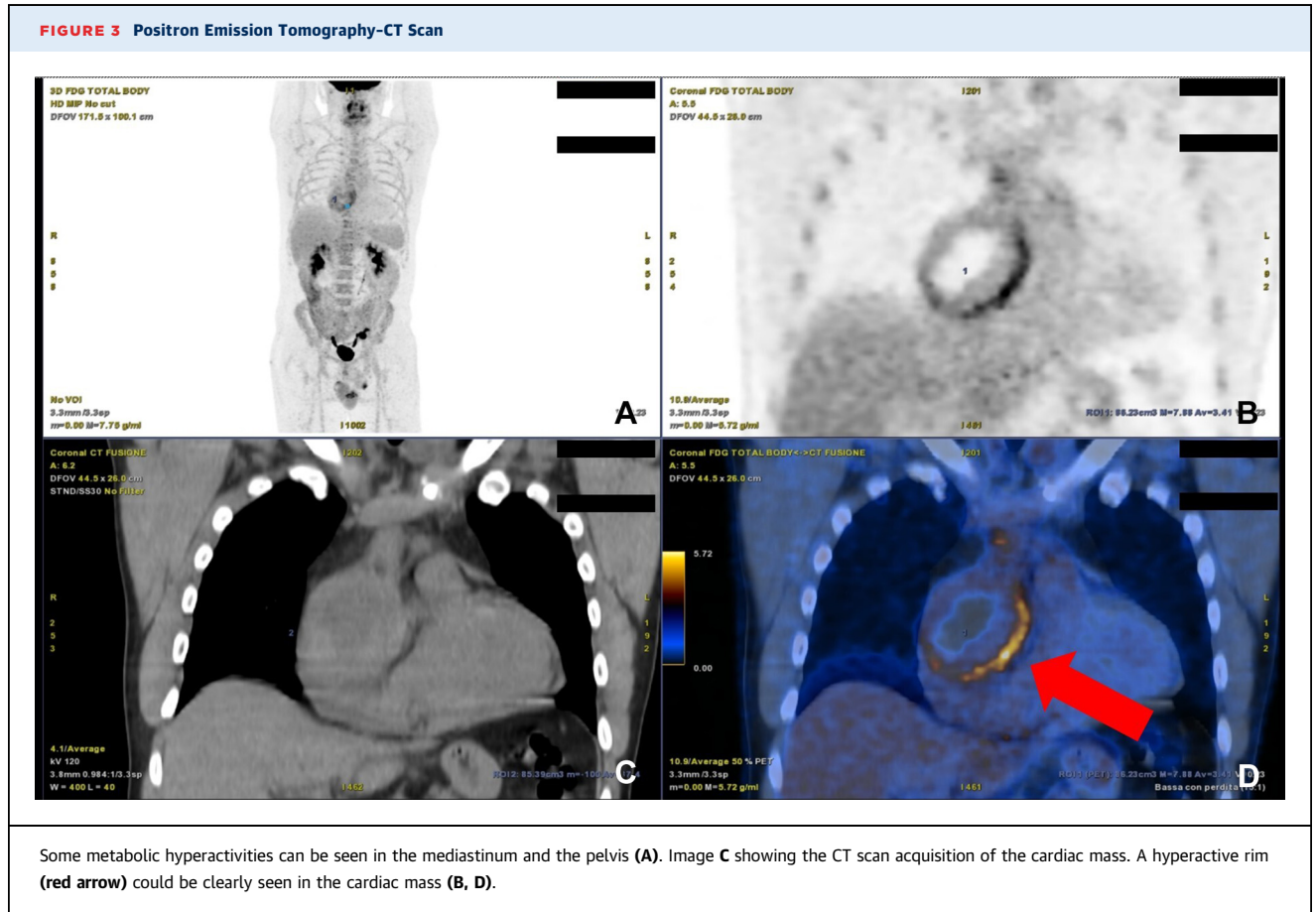
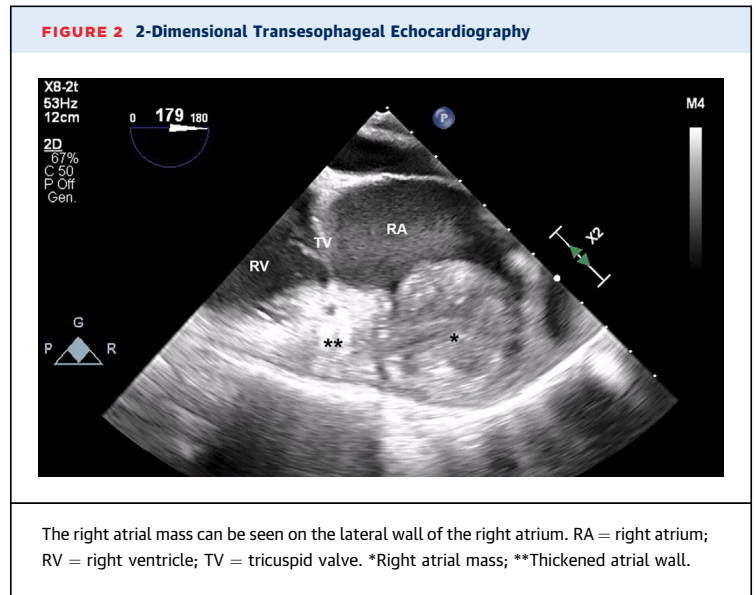
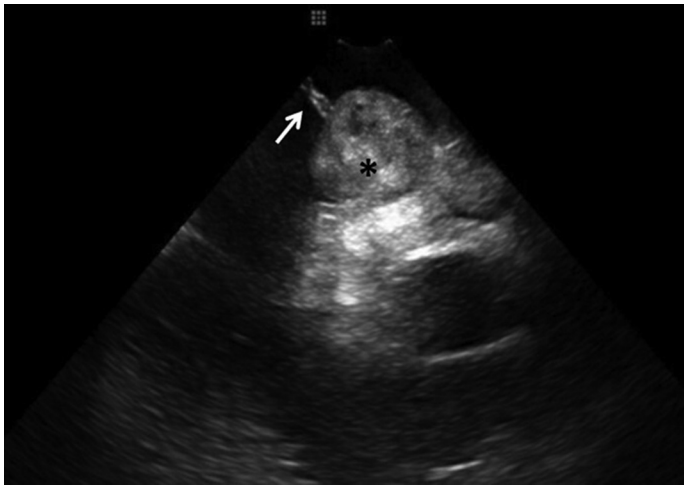


FIGURE 4 Intracardiac Echocardiography Guidance

The biptome (**white arrow**) approaching the right atrial mass (**black asterisk**) can be seen.

definitive diagnosis of a cardiac mass undoubtedly affects the patient's management and should always be pursued to ensure the best therapeutic strategy and minimize any overtreatment. For example, an asymptomatic and hemodynamically insignificant benign mass can be managed only with observation, limiting any invasive treatment.

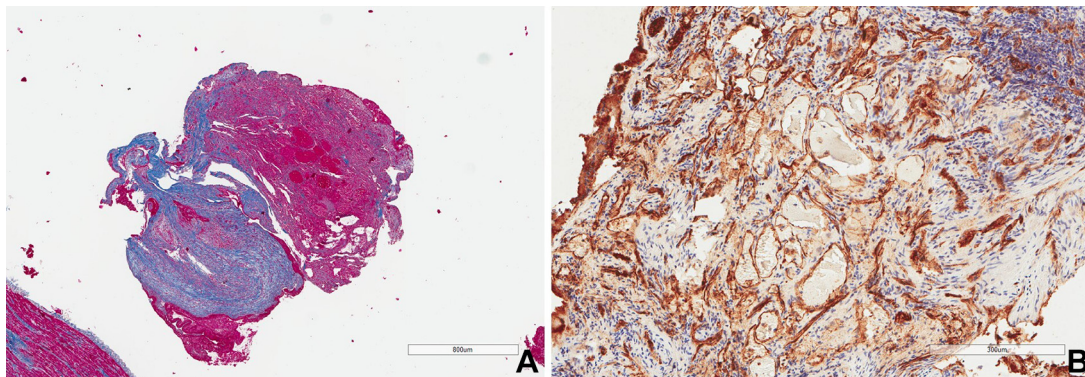
A cardiac mass can be challenging for the surgeon because of the need to balance a complete resection and prevent disease relapse with a conservative surgery.⁵ This is particularly true in young people, who represent a considerable percentage of patients affected by cardiac tumors. A definitive preoperative diagnosis of a benign mass allows the surgeon to perform, when possible, a structure-sparing surgery, as a safe resection margin is not needed, and to correctly orient the therapeutic strategy.

FOLLOW-UP

The postoperative course was uncomplicated, and the patient maintained a sinus rhythm. The postoperative echocardiography showed a nondilated left ventricle with a preserved ejection fraction, no anomalies in the right chambers, and no pericardial effusion. The patient was discharged home in good clinical condition on the seventh postoperative day without the need of further oncologic medications. At 10-month follow-up, no echocardiographic or clinical signs of disease relapse were present.

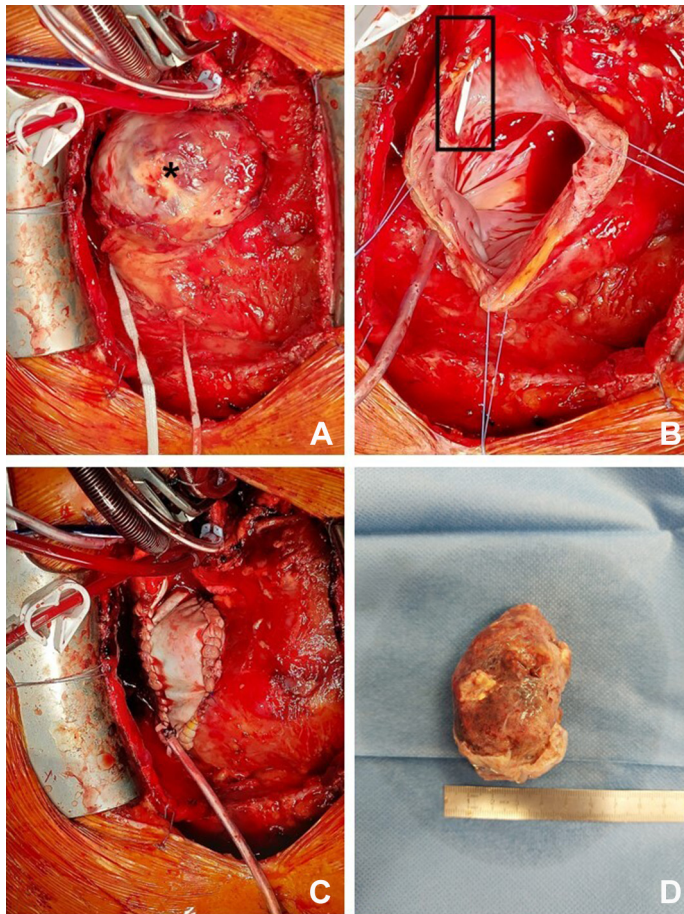
CONCLUSIONS

This clinical case demonstrates that a preoperative diagnosis of a cardiac mass can be safely achieved with biopsy performed during the cardiac catheterization using ICE guidance. A definitive diagnosis

FIGURE 5 Histology Specimen From the Cardiac Biopsy

(A) Azan Mallory stain. (B) Immunohistochemical expression of CD31.

FIGURE 6 Surgical Resection of the Right Atrial Hemangioma



(A) The right atrial hemangioma (indicated by the asterisk) is clearly distinguishable from the nonaffected heart tissue. (B) The right atrium after the resection of the hemangioma; the black box indicates the distal tip of the central line placed in the superior vena cava. (C) Reconstruction of the right atrial wall using a bovine pericardial patch. (D) The right atrial hemangioma resected.

should always be pursued to improve patient management.

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KEY WORDS cancer, echocardiography, ultrasound

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