# Multimodality Cardiac Imaging Enhances Diagnosis and Management of Recurrent Atrial Myxomas in Carney Complex

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## INTRODUCTION

Carney complex (CNC) is a syndrome that is characterized by cardiac myxomas, cutaneous and other myxomatous tumors, pigmented lesions of the skin and mucosa, and multiple endocrine and nonendocrine pathologies.[1] It was initially described by J. Aidan Carney in 1985.[2] Patients with CNC may develop it by autosomal dominant inheritance or de novo mutations in the PRKAR1A gene on chromosome 17.[3] Correa *et al*[1] reported the diagnostic criteria required to diagnose this uncommon disease. We present the case of a 53-year-old Caucasian woman with a history of CNC and a fourth occurrence of atrial myxomas who underwent multimodality cardiac imaging to optimize her surgical management.

## CASE PRESENTATION

A 53-year-old Caucasian woman with a medical history of CNC with three prior surgical interventions for cardiac myxomas, removal of multiple fatty breast tumors, facial lentigines, paroxysmal atrial fibrillation status after two ablations, atypical atrial flutter, six transient ischemic attacks, a pulmonary embolism, hyperlipidemia, and neurocardiogenic syncope was evaluated in an outpatient setting for follow-up. She had a cardiac magnetic resonance imaging (MRI) study at an outside facility that reported a right atrial mass measuring 3.1 × 3.1 × 2.9 cm with a stump in the posterior superior right atrial wall closer to the superior vena cava with mild obstruction of blood flow. The patient had a transthoracic echocardiogram within a week that reported a 3.6 × 3.3 cm mass in the right atrial cavity. No other masses were initially reported in the rest of the cardiac chambers. The patient was referred by her primary cardiologist for further evaluation.

Her prior records were obtained and reviewed. In 1998, she first developed left and right atrial myxomas that were surgically removed. In 2012, she had recurrence of a 5 cm left atrial myxoma; there were two broad-base attachment points to the left atrial wall, one posteriorly above the annulus of the mitral valve and one to the right side inferior to the right pulmonary vein, that were removed. Her next recurrence was in 2016; she had a 1.5 cm right atrial myxoma attached via a stalk to the lateral free wall that was removed, and post-surgical echocardiography in 2017 showed no atrial masses.

During our evaluation, a cardiac computed tomography (CT) scan was performed 5 weeks after her echocardiogram for this fourth occurrence of atrial myxomas. It revealed a 4.5 × 4.5 cm right atrial mass with a stalk arising from the posterolateral wall and two additional small left atrial intracavitary masses near the left atrial roof and left atrial appendage ostium, each measuring 1.1-1.2 cm in diameter. She was transitioned from rivaroxaban to warfarin and then heparin before surgery. A fourth cardiac surgery was to be performed at our institution 2 weeks later. On intraoperative transesophageal echocardiography (TEE), her right atrial mass was 5 × 6 × 4 cm, attached to the right atrial wall by a pedunculated stalk (Figure 1), partially occluding both the superior vena cava and inferior vena cava ostium and partially herniating across the tricuspid valve during diastole (Figure 2A, Videos 1 and 2). A sessile mass located on the roof of the left atrium and a mobile mass located at the ostium of the left atrial appendage were also visualized (Figures 3 and 4, Video 3). The right atrial cardiac mass and two left atrial cardiac masses were resected along with the base of the right atrium and roof of the left atrium (Figure 5). Biopsies were obtained from each of the three intracardiac masses, and the pathology was diagnostic for myxomas. To reconstitute the resected left and right atrial tissue, biatrial reconstruction using bovine pericardium was performed inclusive of closure and removal of the left atrial appendage. Despite the extensive replacement of the left and right atrium with pericardial substitute, the patient resumed normal sinus rhythm. However, due to her extensive atrial reconstruction with bovine patches, the patient was placed on indefinite anticoagulation with warfarin.

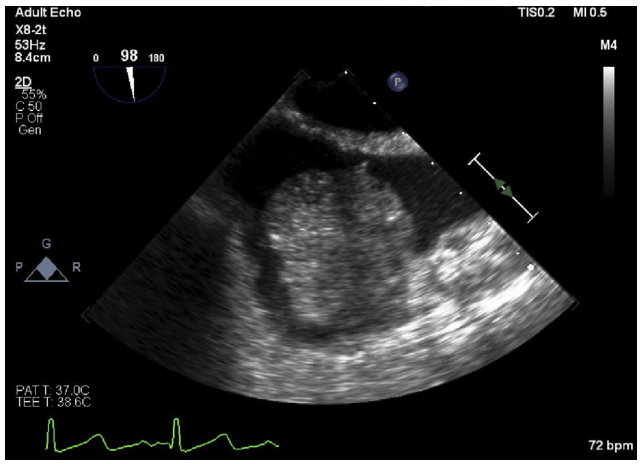


**Figure 1** TEE midesophageal bicaval view: a large pedunculated mass is attached to the posterolateral wall of the right atrium.

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## VIDEO HIGHLIGHTS

**Video 1:** Intraoperative midesophageal four-chamber TEE view in real time with a focus on the right atrium and right ventricle showing a large mass is occupying most of the right atrium with partial herniation across the tricuspid valve during diastole.

**Video 2:** Intraoperative TEE midesophageal bicaval view reveals a large pedunculated mass filling the body of the right atrium.

**Video 3:** Intraoperative three-dimensional TEE "en face" or "surgeon's" view of the left atrium and mitral valve in real time revealing a sessile mass attached at the remnant os of the left atrial appendage and a second small mass attached high on the aortomitral curtain (or dome of left atrium).

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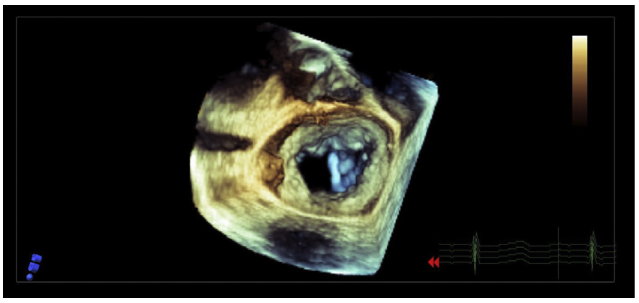

Figure 3 Three-dimensional TEE view of the "en face" mitral valve: the sessile mass is attached at the remnant os of the left atrial appendage. A second small mass is attached high on the aortomitral curtain (or dome of left atrium).

## DISCUSSION

Cardiac myxomas are an important characteristic feature of CNC. Although genetic proof of CNC was not in our patient's records, her history of cardiac myxomas, with confirmatory pathology at our institution, and facial lentigines were sufficient to support a diagnosis of CNC. Cardiac myxomas are found in 20%-40% of patients with CNC.[4] These myxomas can present anywhere in the heart and are seen equally in both men and women.[1] They can recur in the same[5] or different[6] sites and present in one or all four chambers.[7] Prior literature reports that nearly 80% of myxomas are localized to the left atrium, 7%-20% in the right atrium, 2.5%-6% in the right ventricle, 8% in the left ventricle, 2.5% in two or more locations, and <2.5% in both atria of these same patients.[8] Cardiac myxomas seen in sporadic cases may recur in 3% of patients.[9] In contrast, patients with CNC have a reported higher recurrence rate of up to 30%.[10] Cardiac myxoma–related issues are the most common causes of morbidity and mortality in these patients.[11] They are reported to be responsible for mortality in >50% of patients with CNC.[1] Problems related to the myxomas can include mass effect from the tumor, obstruction of a valve or outflow tract, congestive heart failure symptoms, emboli that could result in stroke, cardiomyopathy, arrhythmias, surgical complications, and an increased chance of sudden cardiac death.[11]

The concerns associated with cardiac myxomas warrant careful and appropriate evaluation and management. A thorough clinical assessment for the sequelae of CNC is necessary in all patients with suspected cardiac myxomas. In order to prevent some of the aforementioned myxoma-related problems, surgical resection should be performed soon after the cardiac myxomas are diagnosed.[12] Perioperative risks include mortality in cases of low-output cardiac failure, strokes from embolization, and arrhythmias including atrial fibrillation.[11] In patients requiring recurrent surgery, dense adhesions have been reported to add to surgical risk when compared to only the complications associated with myxomas.[5] Incisions from multiple cardiac surgeries may also disrupt the cardiac tissue and cause fibrosis resulting in atrial dysfunction. In rare situations when the tumor burden deeply invades the myocardium, patients may require consideration for cardiac transplantation.[11] Patients should have an echocardiogram, or alternatively a cardiac CT scan or cardiac MRI, performed 6 months after their surgery for outpatient follow-up.[5]

In our case, we utilized and advise multimodality cardiac imaging for presurgical planning and intraoperative TEE to guide surgical management. The cardiac CT scan in the preoperative period helped identify the left atrial myxomas that were not reported on the echocardiogram and cardiac MRI that were performed outside of our institution prior to our presurgical evaluation of the patient. The intraoperative TEE guided surgical management and ruled out

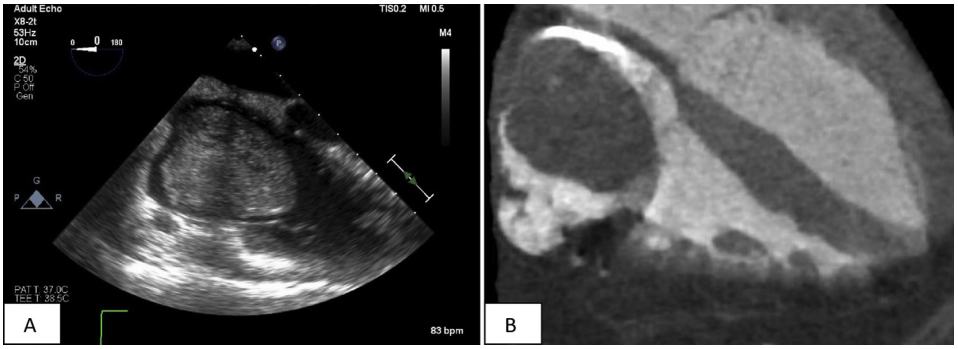


Figure 2 **(A)** Midesophageal four-chamber TEE view with a focus on the right atrium and right ventricle: a large mass is occupying most of the right atrium with partial herniation across the tricuspid valve during diastole. **(B)** Computed tomography heart morphology study showing the right atrial tumor herniating across the tricuspid annulus in diastole.

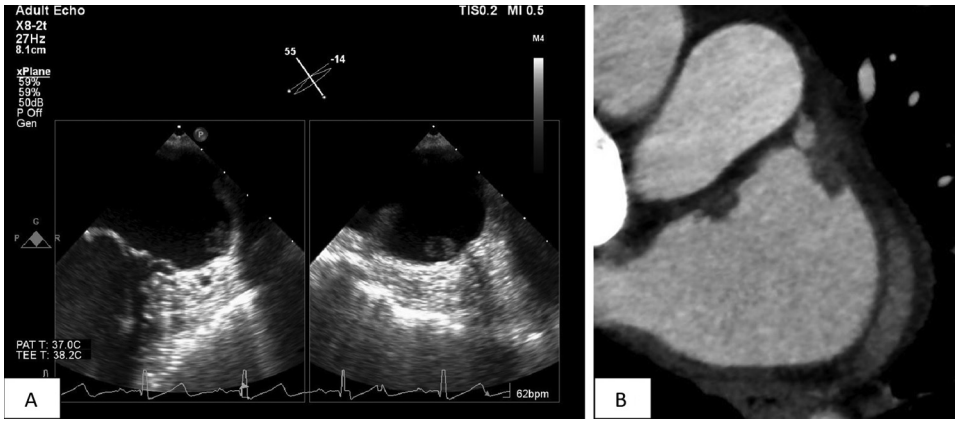


**Figure 4** **(A)** Midesophageal two-chamber X-plane TEE view of left atrial appendage: an irregularly shaped heterogeneous mass attached to the remnant os of the left atrial appendage. **(B)** Computed tomography heart morphology study showing the two small left atrial masses.

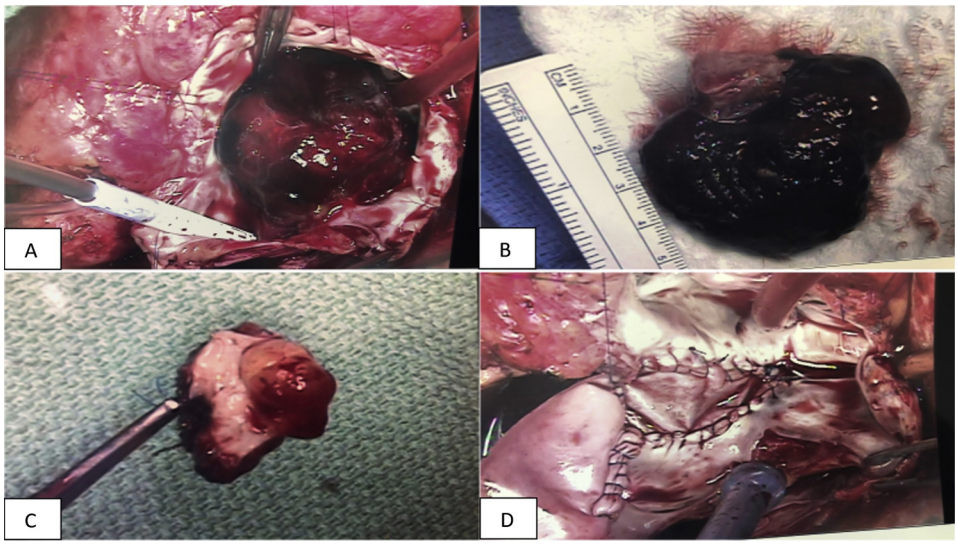


**Figure 5** **(A)** Surgical approach for removal of the right atrial myxoma. **(B)** Right atrial myxoma removed by surgical excision measuring 4.5 × 4.5 cm. **(C)** Tissue removed with one of the myxomas seen in the left atrium. **(D)** Reconstruction of the heart after surgical excision of the left and right atrial myxomas.

perioperative complications. Multimodality cardiac imaging is necessary in CNC patients in order to thoroughly assess for multiple cardiac masses, their location, and their size and for unexpected phenotypical sequelae of CNC.

## CONCLUSION

Patients with CNC may have recurrent cardiac myxomas requiring multiple surgical interventions. In these patients, postsurgical outpatient echocardiography is central to their long-term care. In cases of recurrence, surgery should be performed upon diagnosis of all the myxomas present in the heart. In select patients, adjunctive information provided by preoperative multimodality advanced cardiac imaging and intraoperative echocardiography may better define tumor burden and sequelae not readily identified on transthoracic echocardiography alone.

## SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2021.03.003.

## REFERENCES

1. Correa R, Salpea P, Stratakis CA. Carney complex: an update. Eur J Endocrinol 2015;173:M85-97.
2. Carney AJ, Gordon H, Carpenter PC, Vittal Shenoy B, Go VLW. The complex of myxomas, spotty pigmentation, and endocrine overactivity. Medicine 1985;64:270-83.
3. Takigami M, Kawata M, Kintsu M, Kodaira M, Sogabe K, Kato Y, et al. Familial Carney complex with biatrial cardiac myxoma. J Cardiol Cases 2017;15:155-7.
4. Espiard S, Bertherat J. Carney complex. Front Horm Res 2013;41:50-62.
5. Azzam R, Abdelbar A, Yap KH, Abousteit A. Carney complex: fourth time excision of recurrent atrial myxoma via left thoracotomy. BMJ Case Rep 2014;2014. https://doi.org/10.1136/bcr-2013-201827.
6. Kwon OY, Kim GJ, Jang WS, Lee YO, Cho JY, Lee JT. Fourth recurrence of cardiac myxoma in a patient with the Carney complex. Korean J Thorac Cardiovasc Surg 2016;49:119-21.
7. Saleh Y, Hammad B, Almaghraby A, Abdelkarim O, Seleem M, Abdelnaby M, et al. Carney complex: a rare case of multicentric cardiac myxoma associated with endocrinopathy. Case Rep Cardiol 2018;2018:1-7.
8. Bjessmo S, Ivert T. Cardiac myxoma: 40 years' experience in 63 patients. Ann Thorac Surg 1997;63:697-700.
9. Mahilmaran A, Seshadri M, Nayar PG, Sudarsana G, Abraham KA. Familial cardiac myxoma: Carney's complex. Texas Heart Inst J 2003;30:80-2.
10. Shah IK, Dearani JA, Daly RC, Suri RM, Park SJ, Joyce LD, et al. Cardiac myxomas: a 50-year experience with resection and analysis of risk factors for recurrence. Ann Thorac Surg 2015;100:495-500.
11. Siordia JA. Medical and surgical management of Carney complex. J Card Surg 2015;30:560-7.
12. Wei K, Guo H-W, Fan S-Y, Sun X-G, Hu S-S. Clinical features and surgical results of cardiac myxoma in Carney complex. J Card Surg 2019;34:14-9.