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## Giant myxoma causing heart failure symptoms

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### Summary

**Background:**

Myxomas arising from the eustachian valve are exceedingly rare.

**Case Report:**

A 72-year-old Jamaican- Chinese woman was evaluated for worsening dyspnea. The 2-dimensional and real time 3-dimensional transesophageal echocardiogram showed a 75 mm length × 44 mm width, multilobulated, mobile mass arising from the eustachian valve occupying the entire right atrial and right ventricular cavities extending into the coronary sinus, right ventricular outflow tract, and proximal inferior vena cava. The patient underwent successful resection of the mass and replacement of the tricuspid valve. Histopathologic examination confirmed the diagnosis of atrial myxoma.

**Conclusions:**

This is the largest myxoma found on a Eustachian valve.

**key words:**

**myxoma • eustachian valve • right atrial myxoma**

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## BACKGROUND

Atrial myxoma is the most common benign cardiac tumor [1]. Primary cardiac tumors are extremely rare with a reported incidence ranging from 0.0017% to 0.28% at the time of autopsy [2,3]. Atrial myxomas comprise approximately 30 to 50 percent of these cases with right atrial myxomas being only a quarter of those cases [4]. Right-sided tumors can present with right-sided heart failure symptoms or even fatal complications such as embolization or obstruction of the outflow tract [1]. Atrial myxomas are the most important cardiac tumors to diagnose, as they have an excellent prognosis following surgical excision [5–7]. We report a giant myxoma (largest reported size) arising from the eustachian valve, a rare location [8]. To the best of our knowledge, there have been only three reported cases [9–11].

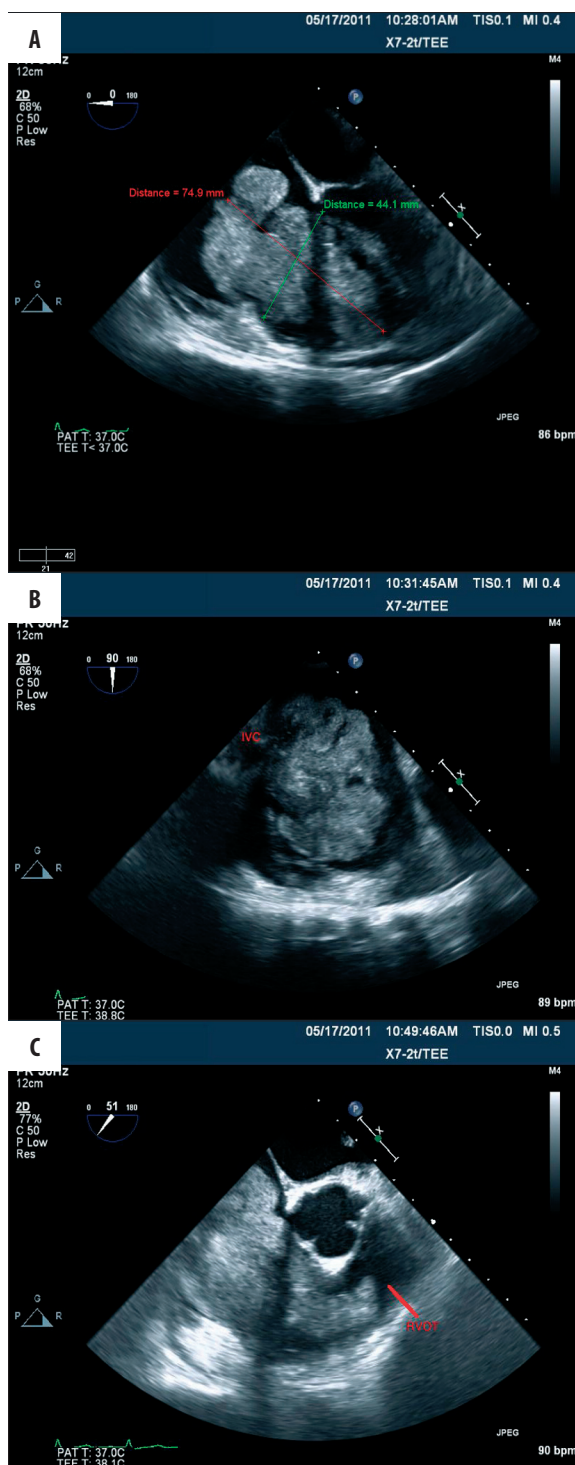
## CASE REPORT

A 72 year-old Jamaican-Chinese woman was transferred to our hospital for further evaluation of a mass seen on the right side of the heart on transthoracic echocardiography. She reported worsening dyspnea for the past one year. She had an unremarkable past medical history and denied any chest pain, syncope or any constitutional symptoms. On admission, her heart rate was regular, and her blood pressure was 130/85 mm Hg. Her physical exam was normal except for distended jugular veins, bilateral lower extremity edema, and a systolic murmur heard at the mid to lower left parasternal area.

Her hemogram revealed an elevated leukocyte count of 13,200/cu mm. The comprehensive metabolic panel was within normal limits. Blood cultures did not reveal any growth after 48 hours of incubation. The 12-lead electrocardiogram demonstrated normal sinus rhythm, q waves in leads III and aVF, left ventricular hypertrophy, and a normal axis. Her chest radiograph was within normal limits.

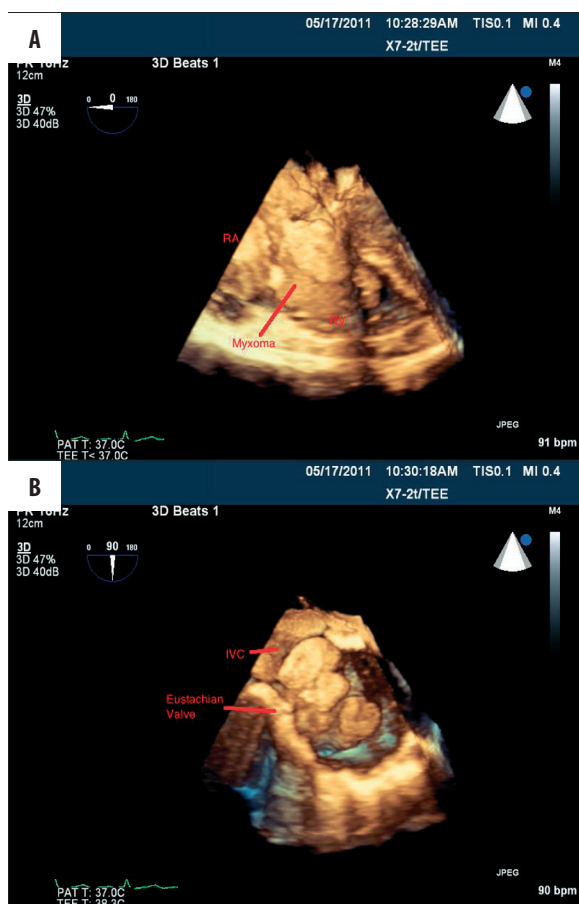
Transthoracic echocardiography (TTE) revealed a large lobular mobile mass originating from the right atrium. To further characterize the mass, we performed transesophageal echocardiography (TEE). Two-dimensional and real time three-dimensional (RT-3D) TEE showed a massive, 75 mm length  $\times$  44 mm width, multilobulated, mobile mass occupying the entire right atrial and right ventricular cavities and extending into the coronary sinus, right ventricular outflow tract, and proximal inferior vena cava (Figures 1 and 2). It appeared to be attached at the site of the eustachian valve. The right ventricular cavity size was dilated with normal wall thickness. Right ventricular systolic function could not be assessed as a result of the occupation of its entire volume with the tumor mass. A moderate to large loculated pericardial effusion was identified anterior to the heart and surrounding the left atrial appendage.

A diagnostic left heart catheterization prior to planned surgery revealed normal coronary arteries. The patient underwent successful operation for the right atrial mass described as a gelatinous friable, multi-lobulated tumor mass attached to the atrial wall at the level of the inferior vena cava and coronary sinus (Figure 3). The gross specimen measured 94 $\times$ 74 mm. The base of the mass was then removed by removing a large wedge of right atrium as well as anterior wall, superior



**Figure 1.** (A) 2-dimensional transesophageal echocardiogram illustrating a giant cardiac myxoma measuring 75 mm length  $\times$  44 mm width. (B) 2-dimensional transesophageal echocardiogram illustrating a giant myxoma protruding into the right ventricular outflow tract. (C) 2-dimensional transesophageal echocardiogram illustrating a giant myxoma with a proximal portion extending into the inferior vena cava.

vena cava and reconstructed with an autologous pericardial patch. Intraoperative transesophageal examination with color flow doppler immediately following tumor resection



**Figure 2.** (A) 3-dimensional echocardiogram illustrating a giant myxoma extending into the inferior vena cava and the attachment site at the level of the eustachian valve. (B) 3-dimensional echocardiogram illustrating a giant myxoma occupying almost the entire cavity of the right atrium (RA) and right ventricle (RV).

revealed severe tricuspid insufficiency. The leaflets appeared to be retracted, restricted and atrophic, probably secondary to long term pressure effects of the mass on the leaflet architecture. A porcine bioprosthetic valve was inserted to replace the tricuspid valve. The tricuspid insufficiency was completely resolved with no perivalvular leak. Histopathologic examination confirmed the diagnosis of myxoma (Figure 4).

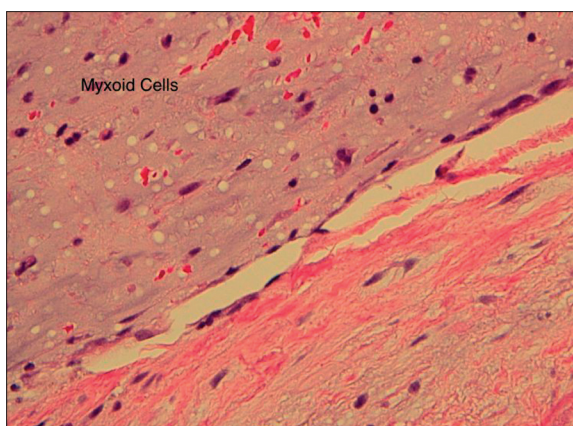
## DISCUSSION

Atrial myxoma is the most common benign cardiac tumor [1]. Primary cardiac tumors are extremely rare with a reported incidence ranging from 0.0017% to 0.28% at the time of autopsy [2,3]. Atrial myxomas comprise approximately 30 to 50 percent of these cases with right atrial myxomas being only a quarter of those cases [4]. Right-sided tumors can present with right-sided heart failure symptoms or even fatal complications such as embolization or obstruction of the outflow tract [1]. Atrial myxomas are the most important cardiac tumors to diagnose, as they have an excellent prognosis following surgical excision [5–7].

The primary modality of investigation of myxoma is the TTE which can provide the size, mobility and possibly the



**Figure 3.** The gross pathological specimen with attachment site via a single stalk(s) and portions that prolapsed into the right ventricular outflow tract (RVOT) as well as the inferior vena cava (IVC).



**Figure 4.** The histopathological specimen showing myxoid tissue.

site of origin [12,13]. Both 2-dimensional and RT-3D TEE aid in surgical planning with superior visualization of tumor attachment sites [14]. Surgical removal significantly decreases the risk of embolic events as well as complete right ventricular outflow obstruction [5,15]. In addition, intraoperative TEE provides significant information regarding tricuspid regurgitation and can assess the need to perform tricuspid annuloplasty.

## CONCLUSIONS

The use of 2-dimensional and 3-dimensional transesophageal echocardiography along with color flow and Doppler imaging aid in the preoperative and intraoperative diagnosis as well as surgical management of atrial myxomas.

## Disclosure

The authors state that they do not have a significant financial interest or other relationship with any product manufacturer or provider of services discussed in this article. The authors do not discuss the use of off-label products, which includes unlabeled, unapproved, or investigative products or devices.

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