

A Rare Case of an Intracardiac Hemangioma Causing Significant Right Ventricular Outflow Tract Obstruction

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Abstract

Introduction: Cardiac hemangiomas are rare benign primary tumors. We present the successful management of a patient with a hemangioma causing significant right ventricular outflow tract (RVOT) obstruction.

Clinical Presentation: A 54-year-old female with no comorbidities presented with progressive right-sided heart failure symptoms. Examination revealed a prominent right ventricular heave, irregular cardiac rhythm, murmurs consistent with pulmonic stenosis and tricuspid regurgitation, ascites, and bipedal edema. Given the echo features of the mass and the patient's clinical course, we favored a benign cardiac tumor over malignancy. Differentials included myxoma, fibroma, and papillary fibroelastoma. Medical management included enoxaparin and carvedilol as anticoagulation and rate control for the atrial flutter, respectively. Surgical treatment consisted of tumor excision and tricuspid valve annuloplasty.

Results: Intra-operatively, the stalk was indeed attached to the RV free wall. Histopathology was consistent with primary

cardiac hemangioma. The patient's post-operative course was complicated by pneumonia, acute kidney injury, and tracheostomy, but she was eventually discharged improved after a month in the ICU.

Significance: This report highlights a rare primary cardiac tumor in an unusual location. While there have been several reports in the world literature on cardiac hemangiomas, less than ten cases have been shown to have significant RVOT obstruction as in our patient.

Conclusion: Cardiac hemangiomas should be part of the differential diagnosis for an intracardiac mass in the right ventricle. Meticulous echocardiography can be a non-invasive and inexpensive aid to diagnosis and pre-operative planning.

Key words: cardiac hemangiomas, right ventricular outflow tract obstruction, RVOT

Introduction

The differential diagnosis of intracardiac masses covers a wide spectrum, including primary cardiac tumors, metastasis from distant malignancies, thrombi, and vegetations. The exact location within the heart as well as the patient's age at presentation can give some clues as to the nature of the mass. We present a patient with a right ventricular outflow tract (RVOT) mass causing predominantly right-sided heart failure symptoms, and the detailed approach that we undertook to arrive at the diagnosis before proceeding to surgical intervention.

Our objectives for this case report are:

- 1) To present a case of an intra-cardiac mass obstructing the right ventricular outflow tract (RVOT) and leading to right-sided heart failure symptoms
- 2) To discuss the differential diagnosis of a mass in the RVOT
- 3) To demonstrate how various modalities in echocardiography were utilized for the diagnosis and surgical planning of a patient with an obstructive RVOT mass
 - a. 2D echocardiography
 - b. Doppler echocardiography
 - c. Real-time 3D echocardiography (4D echocardiography)

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Clinical Presentation

A 54-year-old female, with no known co-morbid diseases and with good functional capacity prior to illness, had a transthoracic echocardiogram (TTE) done as part of employment requirements. Although asymptomatic during this period, she was told that she had an intracardiac ovoid cystic mass measuring 2.9 x 2.6 cm on the pulmonic valve, for which the primary consideration was a papillary fibroelastoma. The mass was exhibiting pulmonic stenosis physiology, with a peak gradient of 124.8 mmHg across the mass, a dilated right atrium (RA), and concentric right ventricular (RV) hypertrophy with depressed systolic function (tricuspid annular plane systolic excursion or TAPSE = 11.8 mm). Left ventricular ejection fraction remained preserved at 84% by Teicholz method.

There was no known personal or family history of malignancy or congenital heart disease. The patient denied any vices, including intravenous drug use. At this time, given financial constraints and her lack of symptoms, the patient opted to defer the advised surgical intervention for the intracardiac mass.

Six months later, the patient started developing shortness of breath on exertion, easy fatigability, gradually increasing abdominal girth, and occasional bipedal edema. Persistence of her symptoms prompted consult, eventually bringing her to the Philippine General Hospital for further work-up and definitive management.

On physical exam, the patient had stable vital signs. Neck veins were not distended. Breath sounds were decreased in bilateral lung bases. She had a prominent left parasternal heave, and her apex beat was displaced to the sixth intercostal space (ICS) at the left anterior axillary line. Heart sounds were distinct with an irregular rhythm. There was a grade 2/6 mid-to-late systolic murmur at the second ICS of the left parasternal border (LPSB) and a grade 3/6 holosystolic murmur at the fourth to fifth ICS of the LPSB. A fluid wave was elicited on the abdominal exam. She had grade 2 pitting bipedal edema, but no cyanosis or clubbing of the extremities. There were no noted skin lesions or abnormal pigmentation.

A standard 12-lead ECG showed a rhythm of atrial flutter with variable atrioventricular conduction and signs of right ventricular hypertrophy (Figure 1). Although the bilateral pleural effusions on the chest radiograph (Figure 2) make it difficult to delineate the cardiac silhouette, the lack of prominent vascular markings taken with the seemingly rounded apex and prominent right cardiac border point to a predominantly right-sided pathology. Other labs, such as complete blood count, serum chemistry, and coagulation profile, were unremarkable.

With the patient's clinical presentation and initial studies compatible with right-sided heart failure, we performed a detailed repeat TTE study to interrogate the right ventricular outflow tract (RVOT) and the pulmonic valve (PV) for the mass that was detected one year earlier. The mass was easily visible on the parasternal long and short axis (level of aortic valve) views (Figures 3A & 3B), measuring 2.6 x 2.6 cm (compared to previous 2.9 x 2.6 cm), implying that there was no significant growth in the size of the lesion over the course of six months. Modified views taken from the short axis (Figures 3C & 3D) were used to document the prolapsing or plopping motion of the mass through the PV and into the main pulmonary artery (MPA) during systole. Also consistent with the previous TTE, the right-sided chambers were dilated (transverse diameters of 52 mm for the RA and 42 mm for the RV), with evidence of depressed RV function (Figure 4).

It was difficult to delineate the exact point of attachment of the mass based on 2D TTE alone. Cardiac MRI was contemplated for this purpose, but the patient could not tolerate prolonged periods in the supine position. We therefore utilized real-time 3D echocardiographic imaging to better pinpoint the stalk of the mass (Figures 5 and 6). Multi-slice imaging revealed that the mass seemed attached to the wall of the RVOT, instead of the PV as initially hypothesized.

Doppler echocardiography was utilized to measure the increased velocities/pressure gradients and demonstrate turbulent flow across the RVOT mass, thus supporting a pulmonic stenosis physiology (Figure 7). Doppler studies (both color flow and spectral analysis) also showed severe secondary tricuspid regurgitation (Figure 8). A chest CT scan with contrast also demonstrated the presence of the mass as a filling defect in the main pulmonary trunk (Figure 9).

Medical management included enoxaparin and carvedilol as anticoagulation and rate control for the atrial flutter, respectively, as well as low-dose furosemide for mobilization of the edema from the right-sided heart failure. With plans for open-heart surgery, a coronary angiogram was done to rule out significant coronary artery disease that would indicate the need for concomitant bypass grafting. The patient's coronaries were angiographically normal.

Surgical treatment consisted of excision of the RVOT mass and tricuspid valve annuloplasty (Devega technique) (Figure 10). Intra-operatively, the mass was described as grayish with a relatively smooth surface, solid with vascular components, measuring 3 x 4 cm. As deduced from echocardiography, the 1-cm stalk of the mass was attached to the RVOT free wall just proximal to the PV. Microscopic findings showed a mixture of dilated and small caliber vessels lined by attenuated endothelium, separated by a fibrous stroma and occasional fibrous septa. These are



Figure 1. ECG showed atrial flutter with variable AV conduction, right axis deviation and right ventricular hypertrophy

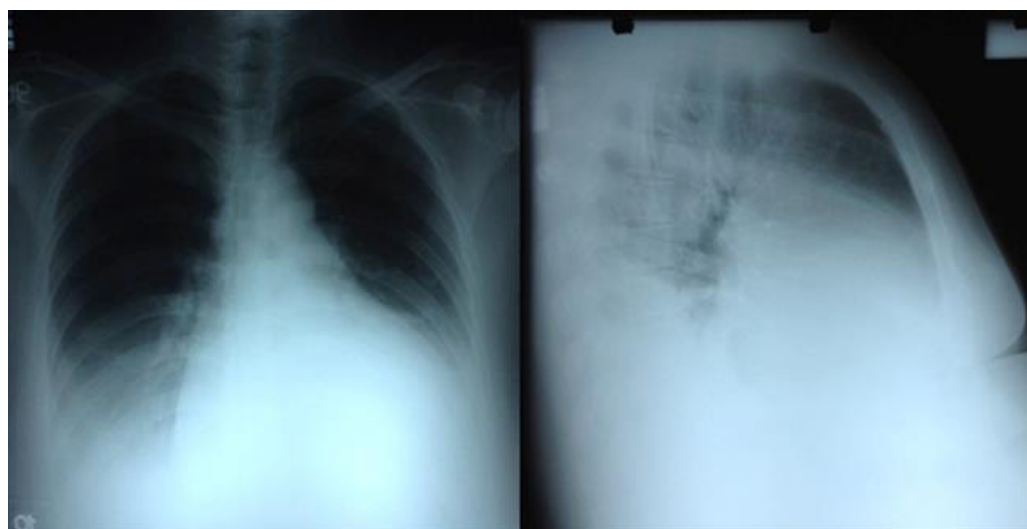


Figure 2: Chest radiography showed a prominent right cardiac border, rounded apex, and a paucity of pulmonary vascular markings, findings suggestive of right-sided pathology.

consistent with a primary cardiac hemangioma. There was also note of small subcentimeter thrombi in the right atrium.

Post-operatively, the patient had a nearly month-long course at the ICU due to sepsis from nosocomial pneumonia. She was successfully managed with inotropic support, broad-spectrum antibiotic therapy (meropenem and colistin), and supportive hemodialysis for acute kidney injury. After a tracheostomy for more effective pulmonary toilette, she was transferred out of the ICU and eventually discharged.

Discussion

RVOT Masses

The dictum with intracardiac masses is that the patient's clinical presentation is generally determined not by the histologic type, but by the specific location of the mass.¹ In our patient, the predominant presentation was right-sided heart failure due to the obstructive nature of the mass in the RVOT, as demonstrated through echocardiographic interrogation. Other manifestations of right ventricular masses include syncope and sudden death.²

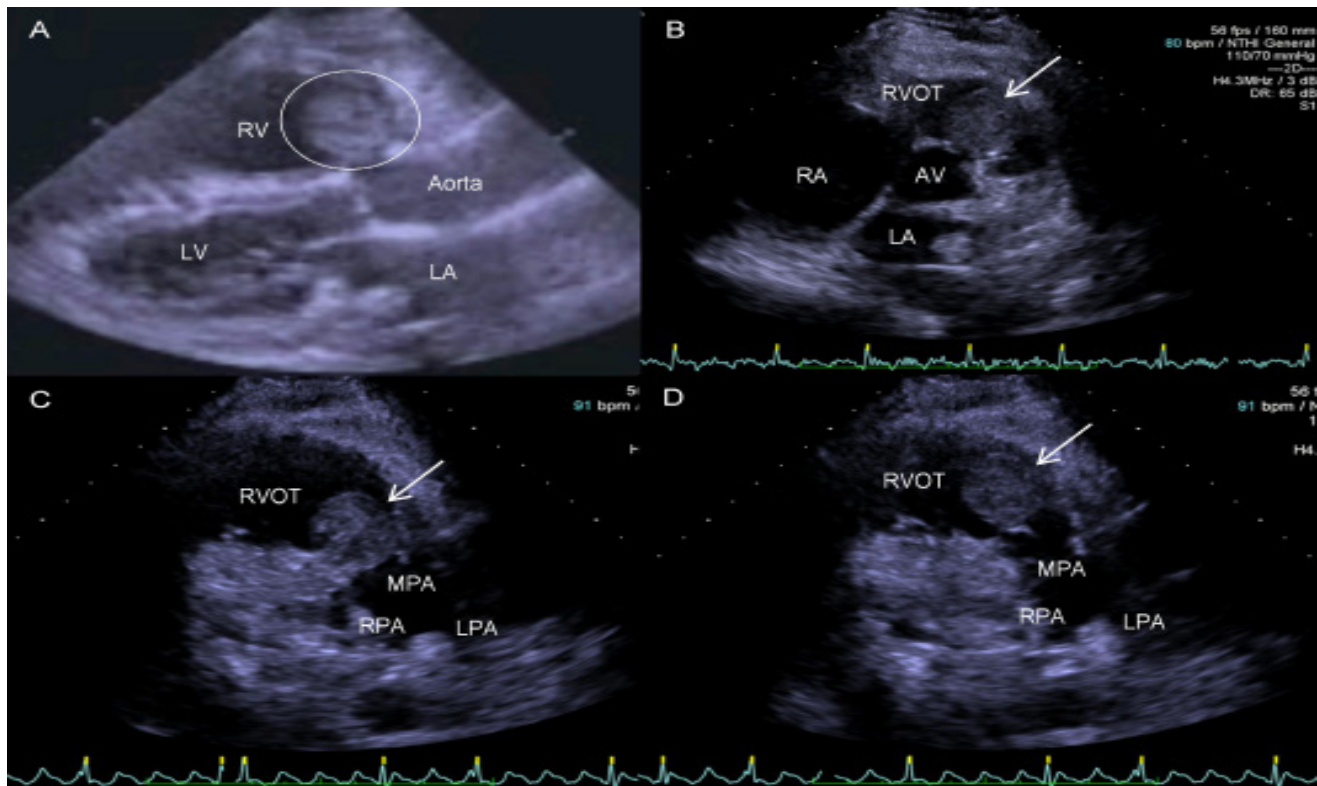


Figure 3. Cardiac Mass in the RVOT

A. Parasternal long axis (PLAX) view showing a dilated right ventricle (RV). The well-circumscribed, non-homogenous, mobile mass (encircled) is intermittently seen during diastole. B. Parasternal short axis (PSAX) view at the level of the aortic valve (AV) demonstrating this well-circumscribed mass (arrow) prolapsing into the outflow tract towards the main pulmonary trunk (MPA) during systole. The attachment of the mass could not be delineated clearly on 2D views. C & D. This was a modified view taken from the short axis to show the RVOT and the MPA. The RVOT, MPA, and right & left pulmonary arteries (RPA & LPA) are visualized during systole (C) and diastole (D). Again, the mass (arrow) was noted to plop towards the MPA during systole.

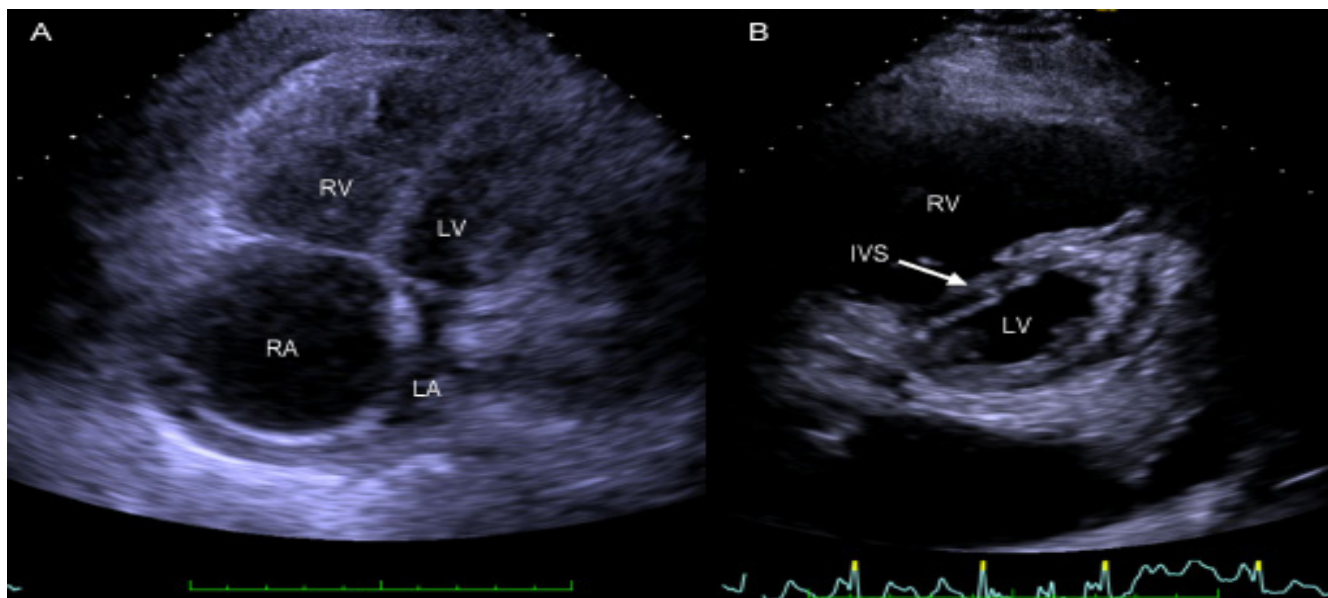


Figure 4. Right ventricular structure and function on 2D echocardiography.

A. Apical four chamber (A4C) view showing a dilated RA and RV. The LA and LV seem to be compressed by the dilated right-sided chambers. The TAPSE was decreased at 11 mm, and the RV fractional area change (RVFAC) was also depressed at 30%. LV systolic function was generally preserved. B. PSAX view at the level of the LV showing flattening of the interventricular septum (IVS) during systole, indicative of RV pressure overload due to obstruction from the RVOT mass.

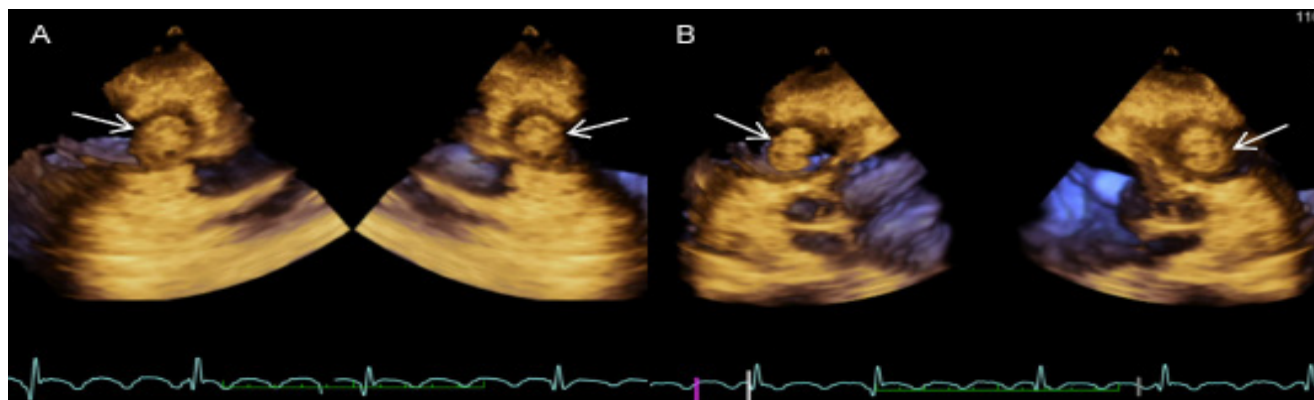


Figure 5. Real-time 3D echocardiography (4D echocardiography), PLAX (A) and PSAX (AV level) (B) views, to better characterize the mass (arrows). Measuring 2.6 x 2.6 cm, it is well-circumscribed, mobile, and intermittently obstructs the RVOT.

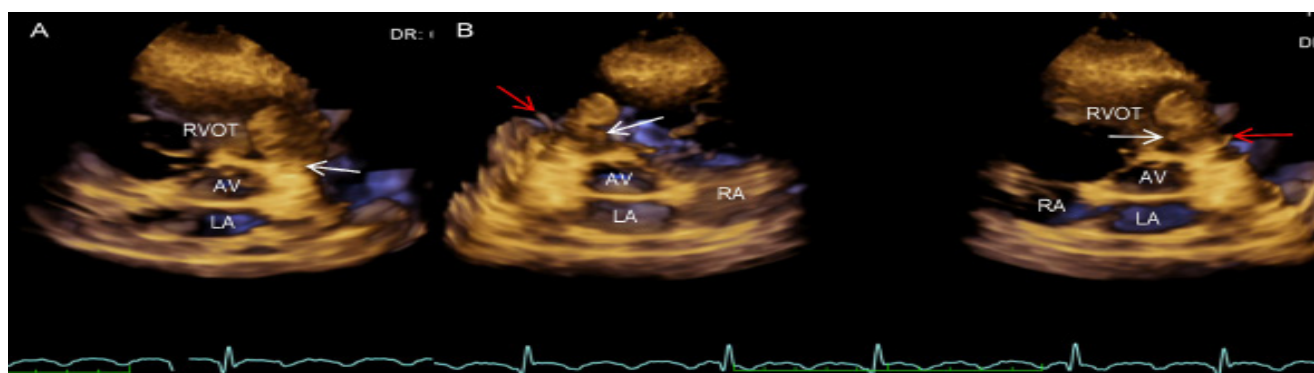


Figure 6. A volumetric approach for data acquisition (multi-slice imaging) was employed to identify the point of attachment of the mass. These are PSAX views (AV level) during systole (A) and diastole (B). The stalk of the mass (white arrows) is more visible during diastole, and seems to be attached to the wall of the RVOT, just adjacent to the annulus of anterior cusp of the pulmonic valve (red arrows). This additional information from 4D echocardiography made papillary fibroelastoma (typically a tumor attached to the valves) a less likely differential.

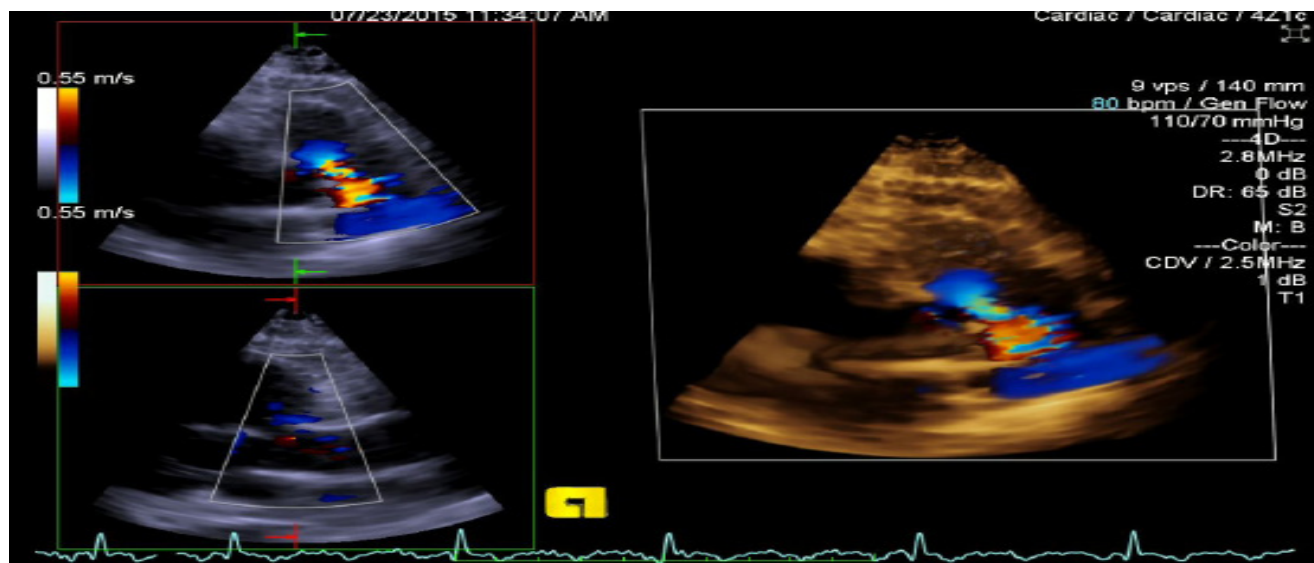


Figure 7. Doppler echocardiography demonstrating RVOT obstruction. First, color flow Doppler was applied at the RVOT demonstrating turbulent mosaic color flow across the mass during systole, indicative of pulmonic stenosis physiology. This was confirmed on spectral tracing of this flow, which revealed a high peak velocity (4.77 m/s) during systole, a peak gradient of 91 mmHg and a mean gradient of 61 mmHg.

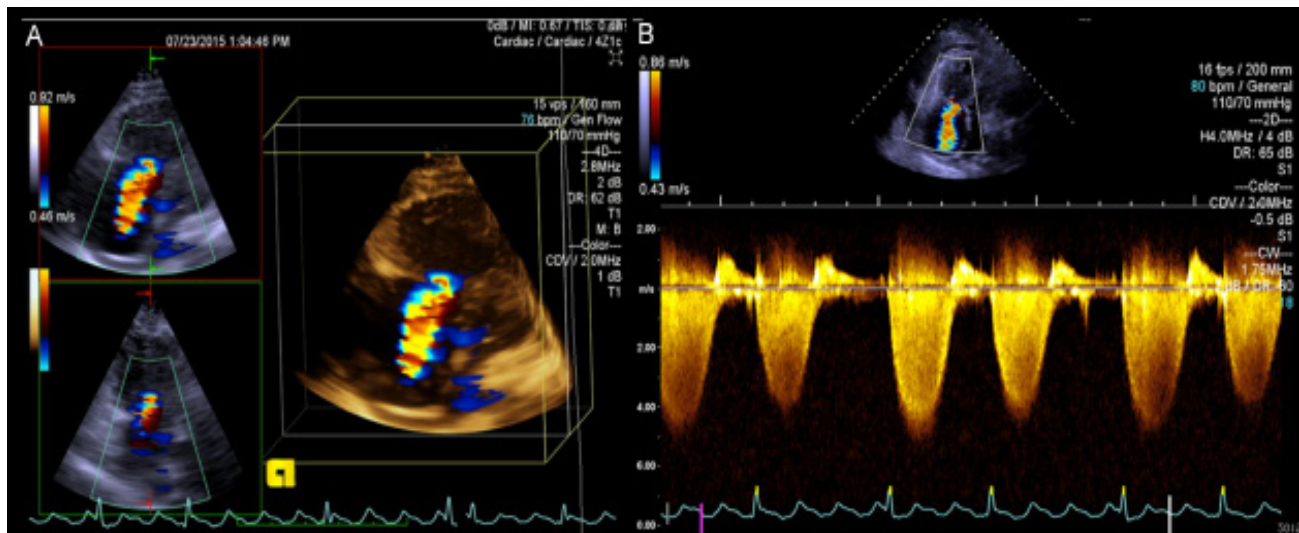


Figure 8. Severe tricuspid regurgitation due to the RVOT obstruction. A. Color flow Doppler showing severe tricuspid regurgitation (TR). This is an indirect sign that there is some degree of RVOT obstruction during systole. B. Spectral tracing demonstrates the severity and holosystolic nature of the TR.

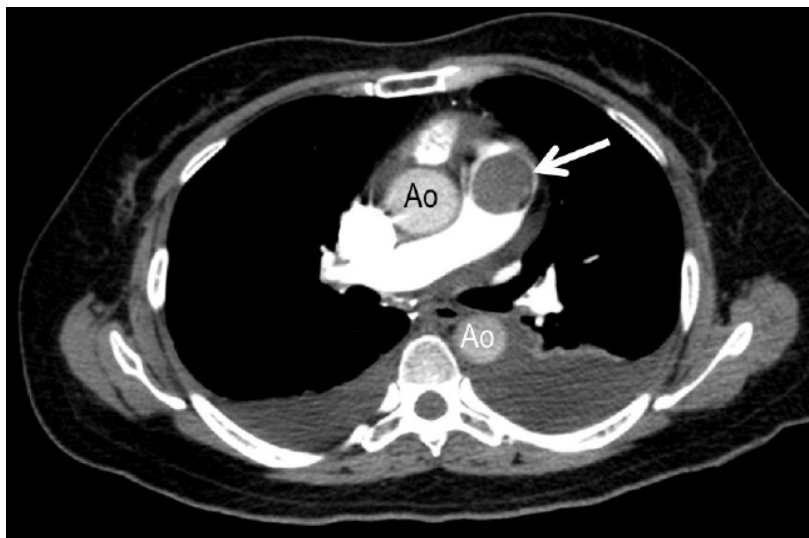


Figure 9. Chest CT scan showed the mass as a filling defect (arrow) in the main pulmonary trunk. Ao = aorta.

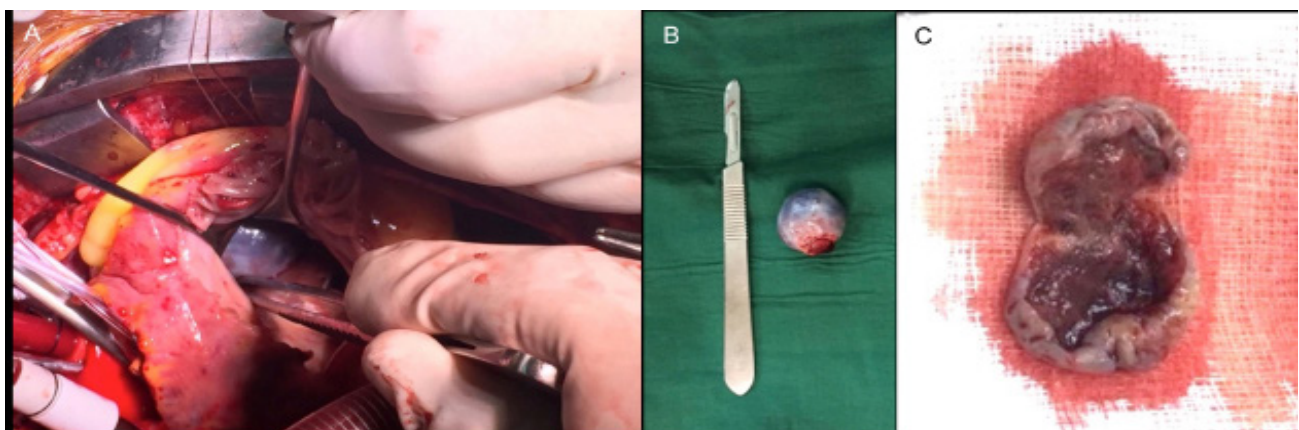


Figure 10. Intraoperative findings. A. Surgical excision of the RVOT mass. B. Grayish, well-circumscribed solid mass (3 x 4 cm) with vascular components. C. Contents of the mass.

Despite the proximity (and initially seeming attachment) to the pulmonic valve, we did not consider a vegetation for the RVOT mass due to the paucity of clinical signs that would satisfy Duke's criteria for infective endocarditis (no history of fever or IV drug use). A thrombus was also considered, given the setting of atrial flutter and depressed RV function, yet this would more likely be located in an area with more stasis, such as the RV apex or a hypokinetic/aneurysmal area of the myocardium, rather than in the RVOT.

We therefore focused our attention on possible neoplasms. Although location of the tumor largely influences the clinical presentation, it is the histologic type that ultimately determines the long-term management and prognosis. By epidemiology, metastatic tumor from a primary malignancy elsewhere is 20-40 times more common than primary cardiac tumors.² A previous case series by Labib et al. of such metastatic tumors causing RVOT obstruction cited a variety of primary malignancies, including melanoma, adenocarcinoma of the breast, colon and pancreas, bronchogenic, renal cell and ovarian carcinomas, and squamous cell cervical carcinoma. On the other hand, primary cardiac malignancy is much less common, with angiosarcomas comprising the vast majority of this group.^{1,3} In their report on the differential diagnosis of RVOT masses, Gopal et al. estimated that an RVOT obstructive tumor is 70 to 140 times more likely to be malignant than benign.² However, echocardiographic features such as the relatively stable size of the mass over a one-year period and its rounded, well-circumscribed contour, combined with a relatively indolent clinical course and lack of an identified primary malignancy on extensive CT imaging, were points against a malignant cardiac tumor.

Benign primary cardiac tumors that were considered for this patient include papillary fibroelastoma (PFE), myxoma, and fibroma. Most PFEs are small (< 1 cm in diameter) and are typically attached to left-sided valves.¹ These characteristics were inconsistent with the mass in our patient, and the likelihood of a PFE was reduced even further on real-time 3D echo when it was demonstrated that the stalk was attached to the RV wall instead of the PV. Aside from aiding in the diagnosis, this crucial echo finding also had important implications in terms of surgical planning and management, as a valvular attachment of the mass would have increased the likelihood of a possible PV replacement if mass excision compromised valve integrity. El Fawal and El Metwally reported a similar case of an obstructive RVOT mass (myxoma by histopathology) that was initially thought to be connected to the PV, but was shown to be actually attached to the RV free wall on cardiac MRI. Three-dimensional echo was able to offer a safer and more feasible alternative to cardiac MRI for delineating the stalk attachment in our patient who could not tolerate lying supine.

Myxomas, the most common cardiac tumor in adults, are usually located in the left atrium (75%). Only 5.0% are located in the ventricles, but there have been several case reports of large myxomas leading to RVOT obstruction.^{2,5,7,8,9,19} Fibromas, while more common in children compared to adults, are typically located in the ventricles and can also lead to RVOT obstruction.^{11,12} The histopathology report, however, yielded a final diagnosis of cardiac hemangioma. While we did not arrive at the exact histologic diagnosis pre-operatively, careful scrutiny of echo features and correlation with clinical characteristics of the patient were the key to at least narrowing the differentials to a benign primary cardiac tumor.

Cardiac Hemangiomas

Cardiac hemangiomas are rare, comprising only approximately 2.8% of the already limited subset of primary cardiac tumors. They are comprised of benign proliferations of endothelial cells, and are histologically similar to hemangiomas found in the skin, subcutaneous tissues, and other parts of the body.¹⁴ Specific morphology can vary, including capillary, cavernous, and arteriovenous forms, with most hemangiomas containing a mix of these subtypes.¹⁵ In a review of 23 cases, Brizard et al.¹⁶ noted that the most frequent clinical presentation was dyspnea on exertion (43%). Signs of right heart failure and a pulmonary stenosis murmur were only encountered in 13% and 21%, respectively.

The epicardium is the most common location for these tumors, but they can also be located in the myocardium and endocardium.¹⁷ In terms of the involved cardiac chamber, Kojima et al. determined that majority (36%) are found in the right ventricle,¹⁸ but only a few cases in the world literature had significant RVOT obstruction as in our patient.^{15,19,20,21,22,23,24,25} In the Philippine setting, there has been only one other published case report of RVOT obstruction.²⁶

While echocardiography is still the diagnostic modality of choice to screen for intracardiac tumors, other tests may provide additional useful information. A cardiac MRI sometimes demonstrates the light bulb sign on T2-weighted images, which is due to a higher signal coming from the sluggish blood flow within the vascular tumor.²⁷ A coronary angiogram, aside from ruling out concomitant coronary disease, can also aid in delineating any vascular feeders of the tumor, giving rise to the classic delayed tumor blush.²⁷ As previously stated, our patient could not tolerate undergoing MRI, and review of the angiogram clips revealed that tumor blush was absent in our patient.

Complete resection is the treatment of choice, although some hemangiomas may be deemed unresectable due to anatomical considerations such as extensive involvement

of the coronaries.¹⁷ There was one reported case of spontaneous resolution of the mass.²⁸

Conclusion

We have presented a rare case of a cardiac hemangioma causing significant RVOT obstruction and right-sided heart failure. We have also demonstrated how the different modalities of echo, coupled with the clinical characteristics of the patient, were utilized to narrow down the differential diagnosis and aid in the surgical planning for our complex case. Knowledge regarding the vital role of echocardiography in the work-up of intracardiac masses is of utmost importance for the astute clinician.

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