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Original Article

Left Atrial Myxoma Presented with an Obstructive Shock, Right Ventricle Dysfunction and Pulmonary Hypertension

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ABSTRACT

Myxoma is a benign primary cardiac tumour, mostly located in the left atrial. A 43 years old woman was referred with a difficulty of breathing for 3 months. The patients also complained about weakness, swollen legs, enlarged abdomen, and blood-tinged cough. The patient appeared weak with blood pressure of 80/50 mmHg and grade III/IV systolic and diastolic murmurs were found. Transthoracic echocardiography and pathology evaluation conclude a cardiac myxoma. The patient was diagnosed with a LA myxoma with an obstructive shock, right ventricular (RV) dysfunction and pulmonary hypertension, thus a surgical approach was done immediately to prevent embolism and sudden death. Cardiac features are most likely a consequence of obstructed LV inflow. Transthoracic echocardiography is a useful modality to determine the size, location, and mobility of the mass. The persistence of RV dysfunction post-surgical may be due to the chronicity of the myxoma.

Key words: Myxoma, Transthoracic echocardiography, Obstructive shock, Right ventricle dysfunction, Pulmonary hypertension.

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INTRODUCTION

Myxoma is a benign primary cardiac tumour. In about 75% of cases, myxoma is located in the left atrial, while occasionally can be found in the right atrial. In general, myxoma is found in solitary, pedunculated, attached to the septal wall near the fossa ovalis.[1] The clinical manifestations vary, ranged from asymptomatic to symptomatic.[2,3] In symptomatic myxoma, clinical triad consists of constitutional symptoms, cardiac symptoms, and embolism may be found.[4]

This following case is going to discuss about left atrial myxoma causing an obstructive shock, high probable pulmonary hypertension, right ventricle dysfunction, and the role of echocardiography in for mass identification and to evaluate the probability of pulmonary hypertension and right ventricle dysfunction.

CASE REPORT

A 43 years old woman was referred to the outpatient installation of tertiary hospital in Indonesia with a chief complaint of difficulty of breathing for 3 months, that have been worsened with orthopnea within the last couple of weeks prior to admission. Patient also felt that her body is getting weak and sore causing activity impairment. There is loss of appetite and loss of body weight about 10 kg within the last 2 months. Additionally, the patient reported swellings on both legs and enlarging abdomen. She also experienced cough with a blood-tinged sputum a day prior to admission. General examination showed a weak general status, fully conscious. Blood pressure was 80/50 mmHg, regular pulse 106 beats per minute, respiratory rate 26x/min with jugular vein distention. On chest auscultation, there were grade III/VI systolic murmur on the left parasternal ICS IV, grade III/IV diastolic murmur on the apex, and fine crackles on both lung bases. Abdomen are distended with a positive shifting dullness. There were also pitting oedema on both legs accompanied with cold extremities. Furthermore, laboratory examinations revealed elevated liver enzymes, potassium and creatinine serum along with oliguria (total urine output of 225 ml in 48 hours).

Transthoracic echocardiography revealed an echogenic mass that moves according to systolic and diastolic phase with a size of 6 cm x 3.8 cm attached to the left interatrial septum with an inflow obstruction of the left ventricle (Figure 1A-C). There was high probability of pulmonary hypertension with tricuspid regurgitant velocity 3.99 m/s, left ventricle eccentricity index 2 cm, pulmonary acceleration time 37 ms (Figure 2A-C) and estimated pulmonary arterial systolic pressure was 78.64 mmHg (estimated right atrial pressure 15 mmHg). There was also right ventricle dysfunction with tricuspid annular plane systolic excursion (TAPSE) 1.6 cm and fractional area change (FAC) 25.4% (Figure 3A-B).

The patient was admitted to the intensive cardiac care unit for shock and congestive heart failure. The patient was decided to have urgent surgical excision of left atrial mass within 48 to 72 hours, after that the excised mass was subjected to histopathological examination (figure 4A). It was confirmed as cardiac myxoma by histopathological examination (figure 4B). Evaluation by transthoracic echocardiography after surgery still found a right ventricular dysfunction with a TAPSE of 1.6 cm.

DISCUSSION

Cardiac tumors are divided into primary and secondary cardiac tumors. Secondary heart tumors have a higher prevalence compared to the primary cardiac tumors. Most primary cardiac tumors are benign in nature, including myxoma with an incidence of 0.5 per million every year.[2,3] Approximately 75% of myxomas are found in the left

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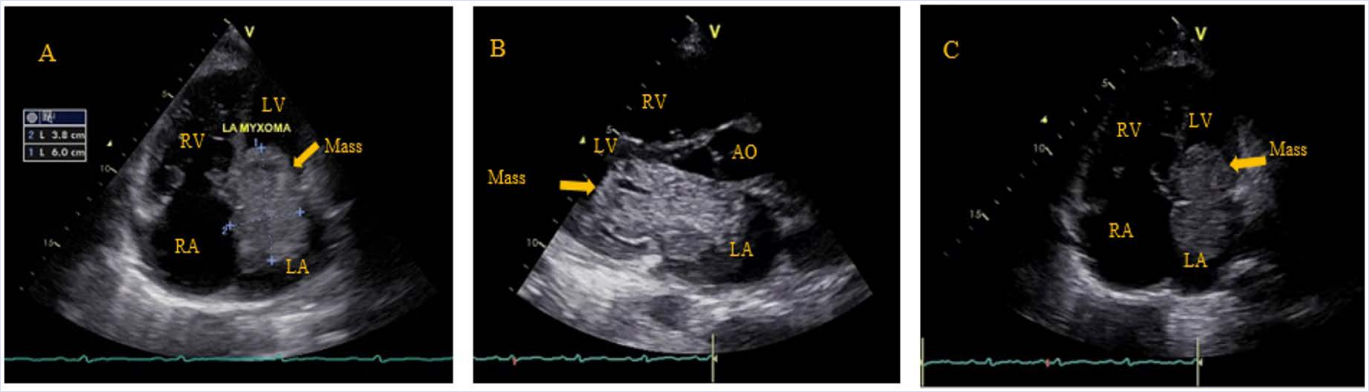

Figure 1A-C: Transthoracic echocardiography (A) apical four chamber view reveals a 3.8 x 6 cm mass which its base is attached to the interatrial septum in the left atrium; (B) Parasternal long axis view showing the mass in the left atrium protruding into the left ventricle through the mitral valve during the diastolic phase; (C) Apical four chamber view shows the mass in the left atrium protruding into the left ventricle through the mitral valve during the diastolic phase.

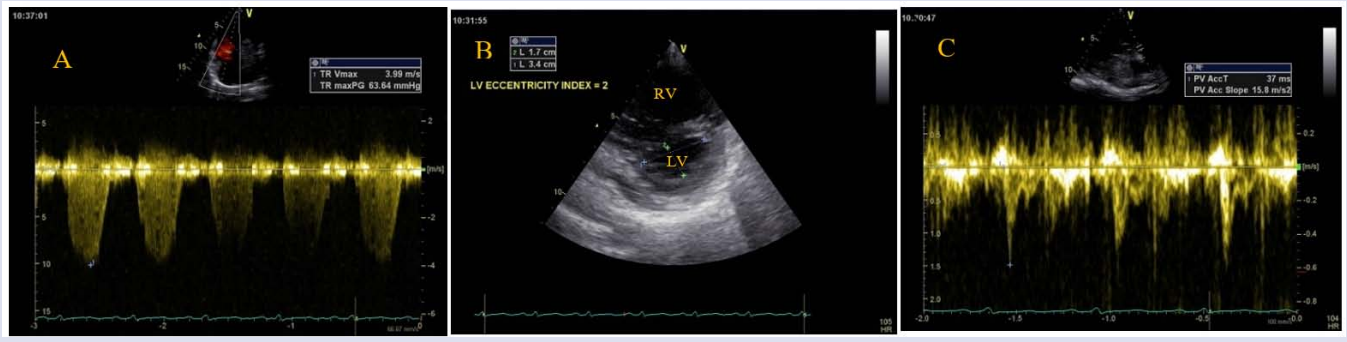

Figure 2A-C: Parameters for assessing the probability of pulmonary hypertension on transthoracic echocardiography (A) Peak tricuspid regurgitation velocity (TR Vmax 3.99 m/s); (B) Left ventricle eccentricity index = 2; (C) Pulmonary acceleration time (PV AccT 37 ms).

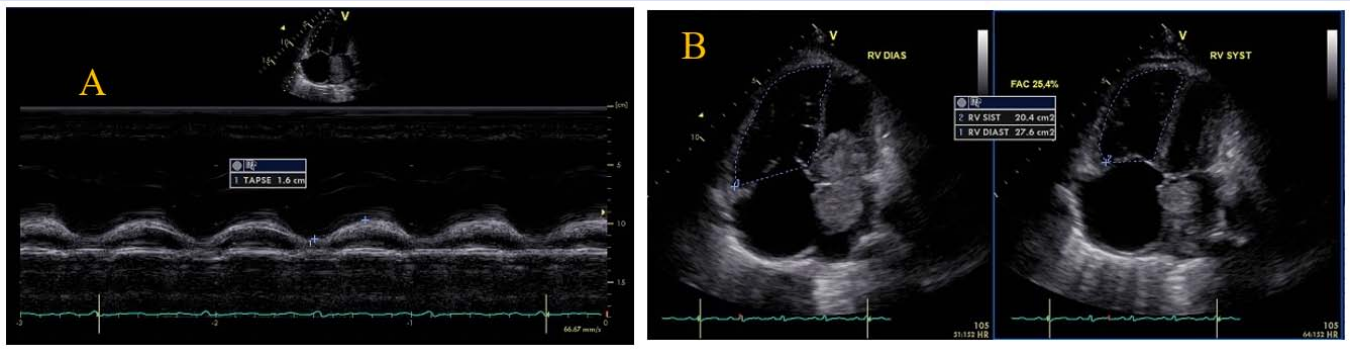

Figure 3A-B: Parameters for assessing the presence of right ventricular systolic dysfunction (A) Tricuspid annular plane systolic excursion (TAPSE) 1.6 cm; (B) Fractional area change (FAC) 25.4%.

atrium with a small proportion can also be found in the right atrium and ventricle. Generally, this condition is more prevalent in women than men. Myxoma may be found on individual at any age, most often occurs between the third and sixth decades.[5,6] In this case, there was a myxoma found in the left atrium attached to the atrial interseptal wall.

Myxoma can exhibit one or more features of the clinical trial that consist of constitutional, cardiac or embolic symptoms. In this case, there are 2 clinical manifestations were found, which are constitutional and cardiac symptoms. The constitutional symptoms appear in the form of arthralgia, fatigue, decreased appetite and weight loss. Cardiac features in this patient are caused by the obstructive shock, heart failure, and highly probable pulmonary hypertension, all secondary to the obstruction of left ventricular inflow. These findings are confirmed with transthoracic echocardiography revealing an echogenic mass with a size 6 cm x 3.8 cm in the left atrium with its base attached to the interatrial septum of the left atrium that protrudes into the left ventricle during diastolic phase causing obstruction of mitral flow thus resembling mitral stenosis. (Figures 1A-C). In one study that reviewed 53 cases of left atrial myxoma, the effects of mitral stenosis occur when the tumor size exceeds 5 cm. The obstructed mitral flow causes obstructive shock because decreased cardiac output thus oxygen transport impairment to vital organs, as in this patient causes kidney failure and oliguria.[4,7-9]

Transthoracic echocardiography showed a high probability of pulmonary hypertension (PVaccT 37 ms and EST PASP 78.64 mmHg) according to

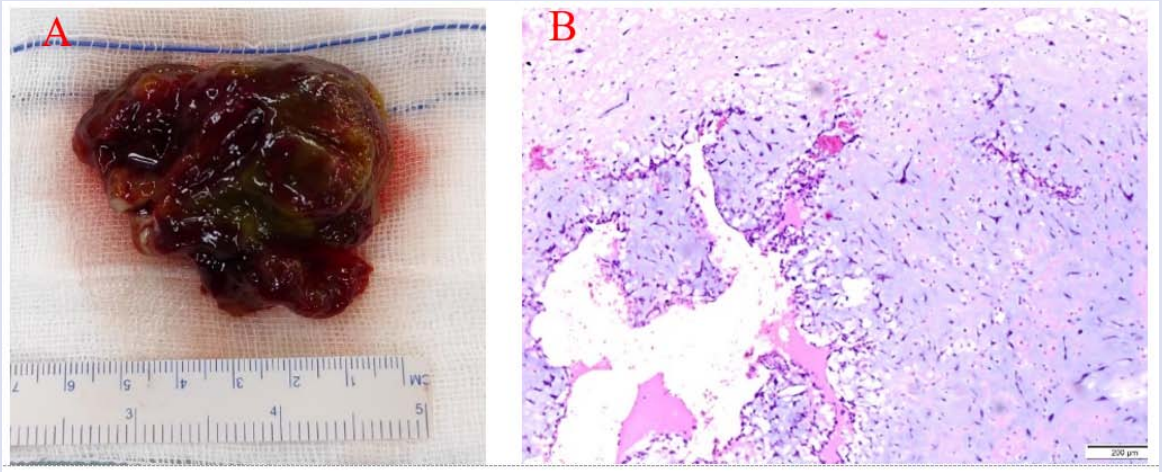

Figure 4A-B: (A) Macroscopic of excised left atrial mass (B) Histopathology of excised left atrial mass concluded a cardiac myxoma.

the tricuspid regurgitation with TR Vmax > 3,4 m/s. Other parameters that support a high probability of pulmonary hypertension are left ventricular eccentricity index > 1.1 and pulmonary acceleration time < 105 ms (Figure 2A-C). The presence of right ventricle dysfunction was determined based on the parameters of the TAPSE < 1.7 cm and FAC < 35% (Figure 3A-B).[10,11] Pulmonary hypertension and right ventricular dysfunction in this patient secondary to the obstructed mitral outflow.[12] The obstruction of mitral flow that resembles mitral stenosis will elevate the pressure in the left atrium causing a backward transmission thus increasing the pulmonary venous pressure. A chronic increased of pulmonary venous pressure triggers a vasoconstriction response in the pulmonary vascular subsequently increase the pressure in the pulmonary arteries. In the end, right ventricle workload is increased, which in turn causing dilation, dysfunction, and can cause right heart failure.[8,13,14]

Several imaging modalities to assist diagnosis of myxoma are transthoracic echocardiography, transesophageal echocardiography, CT scan, CMR, and PET Scan. Transthoracic echocardiography is the most often modality used due to its wide availability in health facilities, with an affordable costs and low radiation exposure. Transthoracic echocardiography is able to determine the size, location, and mobility with the sensitivity of 95%.[1,4,15] In this case, we used transthoracic echocardiography as the first imaging modality to diagnose myxoma and confirmed by the histopathological examination to confirm a cardiac myxoma (Figure 4B). When TTE has confirm the diagnosis of myxoma, surgery should be performed immediately due to the risk of embolism and sudden death. One study stated that surgery should be performed within 48-72 hours.[16]

In post-surgical evaluation with transthoracic echocardiography, the right ventricular dysfunction was still present in our patient. The rationale of this finding may be related to the the chronicity of the myxoma which increases the left atrial pressure secondary to the obstructed mitral outflow causing chronic alteration in the right ventricle or pulmonary vessels.[8]

CONCLUSION

Left atrium myxoma may resembles a mitral stenosis, and showing cardiac manifestations due to obstructed mitral outflow. The chronicity of left atrial myxoma may lead to the development of a highly probable pulmonary hypertension, right ventricular dysfunction and obstructive shock. Therefore, echocardiographic is an important modality needed to assist diagnosis of myxoma, to assess the cardiac and hemodynamic complications of myxoma, also to determine the right time to perform surgical tumour extermination.

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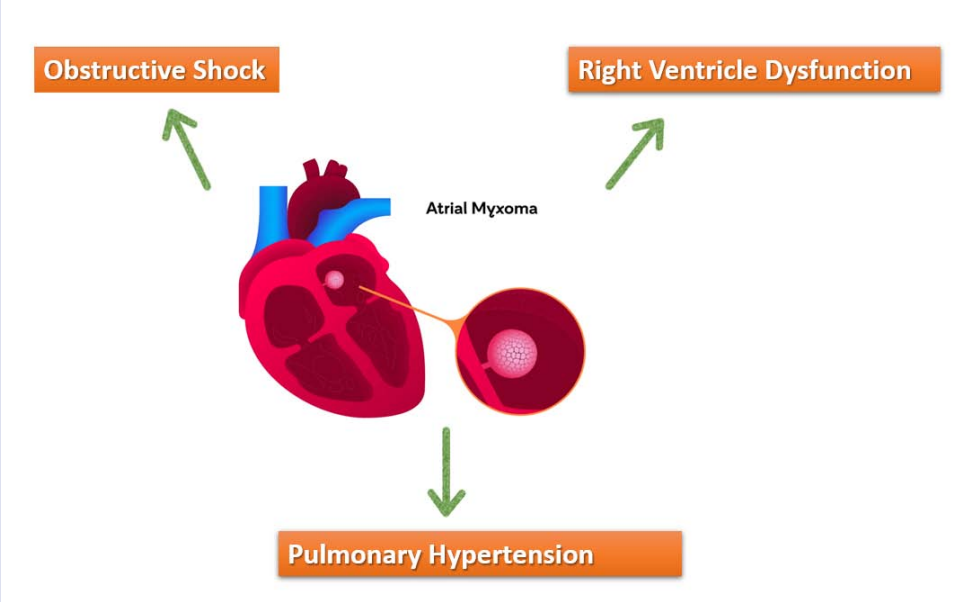
GRAPHICAL ABSTRACT



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