

Cardiac myxoma as a rare acute heart failure etiology in paediatrics: A case report

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Abstract

Cardiac myxoma is extremely rare in children. However, if not treated immediately, it may cause varying symptoms until sudden death. A 9-years old male Javanese child was brought to the emergency department of Prof. Soekandar General Hospital, Mojokerto with progressive dyspnoea since one month which got worse in the left decubitus position. There was no significant past medical history. Physical examination revealed hypotension, mitral stenosis, tricuspid regurgitation, and pulmonary congestion. Transthoracic echocardiography revealed a round pedunculated 3x3.3 cm mass in the Left Atrium that swingingly moved to the Left Ventricle during diastole. This was diagnosed provisionally as Myxoma with a differential of thrombus. After stabilization, he was referred to a tertiary hospital for emergency excision. Histopathology confirmed the myxoma. There were no symptoms and activity limitations during the 6 months follow-up. To the best of our knowledge, this is the first paediatric cardiac myxoma with Acute Heart Failure symptoms reported in Indonesia. Echocardiography is imperative for diagnosing myxoma. Appropriate and timely management results in an excellent outcome.

Keywords: Tricuspid Valve, Diastole, Myxoma, Heart Neoplasms, Mitral Valve Stenosis, Dyspnea, Thrombosis, Hypotension

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Introduction

Cardiac Myxoma (CM) is one of the most common benign primary cardiac tumours in adults, however, it is extremely rare in children. It is mostly found in the left atrium, is mobile and pedunculated.¹ The diagnosis of CM remains challenging due to its variable clinical features

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such as embolism, intracardiac obstruction, or systemic manifestations as heart failure (HF).^{1,2} Due to its rare prevalence in children, diagnosis and treatment are liable to be delayed. This can significantly increase morbidity and mortality.

We report the case of CM in a child who presented to our secondary care hospital with acute heart failure and who was successfully managed.

Case Report

A 9-year-old male Javanese child was brought to the Emergency Department (ED) of Prof. Soekandar General Hospital, Mojokerto in March 2022 due to progressive dyspnoea since 3 days prior to admission. The symptoms got worse in the supine posture and were relieved by sitting up.

Physical activity was limited due to dyspnoea. A week prior, he slept with 2 pillows. His symptom worsened in the left lateral decubitus position and improved in the right lateral decubitus position. A month earlier, he had complained of fatigue in his lower extremities, which had never happened before.

The symptoms did not improve with medications prescribed by primary health care doctors. His mother also complained of low-grade fever for the last 2 months. This month, he lost 3 kg body weight despite an increase in appetite. According to the Centers for Disease Control and Prevention growth curve, his BMI was 13.5kg/m² or <- 3 SD. History of trauma or congenital heart anomaly was denied.

His physical examination showed blood pressure 92/61 mmHg, heart rate 131x per minute, respiratory rate 38x per minute, temperature 37.°C, and oxygen saturation 93% on air. Auscultation revealed a murmur in ICS IV left parasternal line (grade IV/VI) during the systolic phase and in ICS V left midaxillary line (grade IV/V) during the diastolic phase. A Tumour plop was heard in the early diastolic phase. Pulmonary rales were positive, wheezing was negative, and S2 was pronounced. Laboratory examination showed leukocytosis and abnormal ESR. The

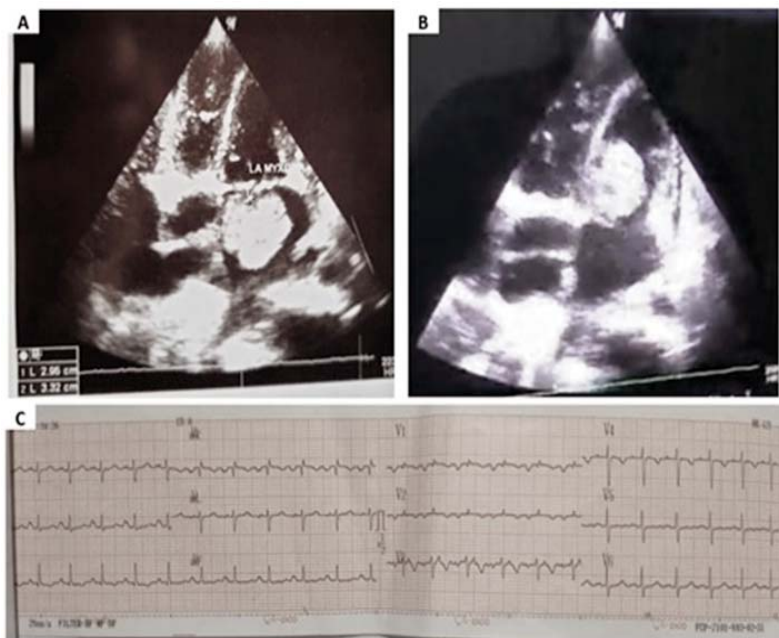


Figure-1: Transthoracic echocardiography examination revealed a mass in left atrial chamber (A) that swung to the left ventricle during diastole (B). Electrocardiography showed LAD, LAE, and non-specific ST change in VI-V4. LAD: left axis deviation, LAE: left atrial enlargement.

Covid-19 nasopharyngeal swab was negative.

ECG revealed sinus tachycardia with left axis deviation (LAD), counterclockwise (CWR), left atrial enlargement (LAE), and non-specific ST change in lead VI-V4 (Fig. 1). Thorax Roentgen showed cardiomegaly with increased pulmonary vascularisation. Transthoracic echocardiography (TTE) revealed a 3x3.3 cm round mass in LA chamber (Fig. 1). The mass was stalked at the basis of the posterior mitral valve with pendulum-like movement that passed the mitral orifice to LV during diastole, strongly suggestive of atrial myxoma. A differential diagnosis of thrombus was made. Relative mitral stenosis was observed during diastolic phase due to tumour obstruction. LA dilatation was observed, while right chamber and annular dilatation was absent. LVEF was 72.7%. In addition, severe PH and severe TR were revealed.

The patient was treated Figure 2 with furosemide, spironolactone, digoxin, and limitation of fluid intake. He was then immediately referred to Dr. Soetomo General Hospital, a tertiary hospital, for emergency open-heart surgery. The mass was solid with a size of 4x3x3 cm in size and a non-villous surface. It contains 0.1-0.9 cm size multiple cysts containing blood and brownish fluid. Histopathological examination confirmed myxoma. The surgery was uneventful. His PH and TR improved. There were no symptoms and activity limitations during 7 months follow-up.

Discussion

CM is the most common primary cardiac tumour in adults, however, its prevalence is extremely low in children. It is 77.7% of all primary cardiac tumours in all age patients,^{3,4} while only 9% of cardiac tumours in the paediatric group.⁵ Rhabdomyoma is the most common primary cardiac tumour in children (63.3%) which has relatively benign symptoms. Compared to rhabdomyoma, cardiac myxoma is relatively more symptomatic and can cause sudden death if not adequately treated in a timely manner.

Around 36-48% of CM are symptomatic, and

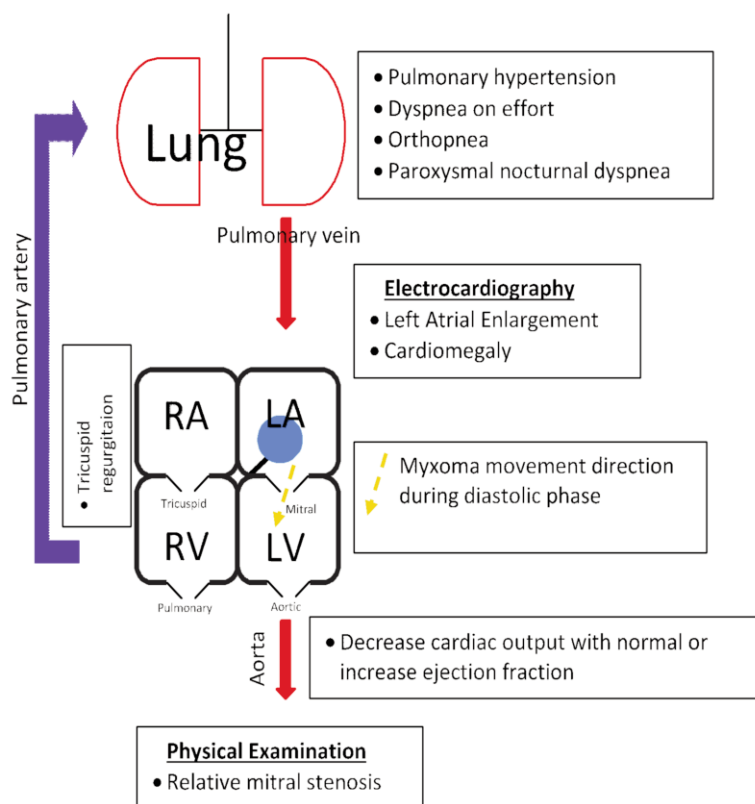


Figure-2: Pathophysiology of heart failure in left atrial myxoma.

its clinical manifestation can be cardiovascular symptoms, embolic symptoms, systemic/constitutional symptoms, or a combination of both.^{6,7} Although cardiovascular manifestation is the most common symptom in adults,^{1,7} it seems that embolic manifestation is more common in the paediatric group.⁸ Our patient had cardiovascular as well as constitutional symptoms.

CM symptoms depend on the size, location, mobility, invasiveness, and friability of the tumour.^{9,10} Large pedunculated LA myxoma located near atrioventricular orifice can move to LV during diastole, impairing LV diastolic filling like as seen in our patient (Fig. 2). Cardiac output is impaired, however, LVEF can be normal or increased as seen in diastolic HF. Backward fluid accumulation stress LA wall, increases pulmonary pressure, and creates pulmonary congestion as seen in congestive HF. Right-sided valve regurgitation may also occur due to backward blood accumulation, as seen in our patient (Fig. 2). Cardiac murmur provides a suspicious abnormality in the heart even in asymptomatic cases.^{7,8} Worsening of dyspnoea in the left lateral decubitus position may indicate a mobile mass in the left heart chamber as it potentially obstructs blood flow.

ECG shows an unspecific abnormality in around half of the cases, commonly sinus tachycardia, LAE, or AF.^{1,11} Chest-X ray is rarely used to diagnose CM, however, it could reveal LAE, cardiomegaly, and pulmonary congestion which can be found in CM.¹

Transthoracic echocardiography (TEE) is an affordable, non-invasive and an accurate modality to diagnose CM.^{7,12} Transthoracic echocardiography can reveal cardiac tumour morphology, size, extension, attachment as well as its haemodynamic manifestation. In confusing cases, trans-oesophageal echocardiography (TEE) provides better visualization of LA or valvular condition than TTE.¹³ However, it may be difficult to differentiate between myxoma and thrombus. The characteristic of mobile pedunculated mass, commonly with a long stalk originating from the endocardial surface of fossa ovalis strongly suggests CM. Left atrial thrombus is usually sessile and located in LA appendage.¹⁴ MRI or enhanced CT scan can be used as another second-line diagnostic modality.

Our patient had an unexplained low-grade fever and weight loss despite increased appetite which may be caused by a proinflammatory cytokine, for instance, IL-6, from a myxoma cell.¹ In the absence of infection and autoimmune signs, the abnormal value of inflammatory parameters such as ESR may indicate pro-inflammatory conditions caused by long-standing myxoma. Higher

energy demand for cardiovascular compensation may also explain his weight loss.

The definitive treatment of CM is surgery, which should be done after the diagnosis. Delayed diagnosis or treatment increases morbidity and mortality. Sudden death can occur due to complete tumour obstruction or a fatal embolic event. Irreversible hemiparesis or other neurology manifestation is common in children.⁸ Irreversible valve abnormality may also occur if the annulus is permanently dilated. Surgical resection is associated with low operative mortality and good long-term outcome. Late myxoma resection had 3x mortality compared to early resection.¹⁵ More than 80% of patients remain without cardiovascular manifestation after resection.¹⁵ Thus, the diagnosis of CM warrants early resection. Recurrent may occur in 5% of surgery.¹²

Conclusion

Cardiac myxoma is an extremely rare etiology of heart failure in a paediatric population. Careful anamnesis, physical examination (e.g cardiac murmur) and followed by echocardiography are important in diagnosing myxoma. Once diagnosis is established, myxoma excision is warranted in timely manner to prevent sudden death.

Consent: The consent from the patient's guardian was obtained for the writing and publication of the manuscript.

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