CASE SERIES

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A case series of paediatric atrial myxoma: Clinical challenges and surgical management

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ABSTRACT

Introduction: Cardiac myxomas are rare, benign tumors of the heart, most commonly found in the left atrium, though they can occur in any heart chamber. These tumors can embolize, causing organ infarction, and may present with symptoms such as dyspnea, orthopnea, cough, peripheral edema, palpitations, and fatigue. The clinical presentation varies widely and may mimic other diseases, making early detection and diagnosis critical. The reported incidence of cardiac myxoma at RS Mohammad Hoesin is 5 cases over 2023-2024, consisting of 3 pediatric cases and 2 adult cases. This report emphasizes the significance of thorough history taking, detailed physical examination, echocardiography, and surgical resection in managing atrial myxoma in two paediatric patients.

Case Presentation: We reported three cases: a 16-year-old male, an 11-year-old female, and 12-year-old male, They were presenting with shortness of breath as the chief complaint. The male patient was diagnosed with decompensated heart failure due to a large left atrial myxoma, accompanied by tricuspid and pulmonary regurgitation, and underwent successful surgical removal of the myxoma. The female patient, with a history of contact with tuberculosis, was found to have a mass in the right atrium with a differential diagnosis of myxoma or tuberculoma. She was treated for clinical pulmonary tuberculosis and is currently under medical management. The last patient, male was diagnosed with a large left atrial myxoma, tricuspid regurgitation, and severe mitral regurgitation. The myxoma had removed from the left atrium in this patient.

Conclusion: Paediatric atrial myxoma is a rare condition with a wide range of clinical manifestations, from asymptomatic cases to intracardiac obstruction, embolization, and constitutional symptoms. This often results in misdiagnosis as other more common heart diseases. Transthoracic echocardiography is useful as an initial diagnostic tool, as it effectively determines the tumor's location, size, shape, and connections, aiding in the selection of the optimal management strategy. Surgical excision is the primary treatment, with regular echocardiographic follow-up essential for the early detection of recurrence and long-term patient health.

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INTRODUCTION

Primary cardiac tumors are rare, with an incidence of approximately 200 per 1,000,000 cases, and around 75% of these tumors are benign.[1-3] Among the benign tumors, nearly half are cardiac myxomas, which typically originate from or near the interatrial septum, extending into the left atrium.[4] While cardiac tumors are exceedingly rare in the pediatric population, with prevalence rates ranging between 0.0019% and 0.30% in autopsy studies, myxomas are more common in adults.[5] These myxomas are intracavitary masses characterized by a pedunculated, short, and broad-based attachment to the atrial wall, with a viscous, gelatinous appearance and consistency.[2] The reported incidence of cardiac myxoma at RS Mohammad Hoesin is 5 cases over 2023-2024, consisting of 3 pediatric cases and 2 adult cases.

The clinical manifestations of myxomas vary widely depending on their location, size, and mobility, often presenting as a triad of intracardiac obstruction, embolization, and constitutional symptoms.[1] In one-fifth of the cases, a myxoma can be asymptomatic and discovered as an incidental finding. Most commonly, patients exhibit symptoms of obstructive heart failure, but presentations can also be vague and non-specific, including arrhythmias, systemic embolization, or general constitutional symptoms.[6] In pediatric patients, the tumor may manifest as arrhythmias, congestive heart failure, or hydrops. Postnatally, symptoms may include cyanosis, murmurs, respiratory distress, myocardial dysfunction, valvular insufficiency, and, in severe cases, sudden infant death due to inflow or outflow tract obstruction.[7]

Diagnosing atrial myxoma requires a careful approach, as it can be mistaken for more common heart diseases. The diagnostic process involves a comprehensive medical history, clinical examination, blood tests, and imaging studies, with echocardiography being the preferred method. Additional imaging techniques like chest CT and MRI help in differentiating myxomas from other

cardiac tumors. Precordial examination findings may mimic those of mitral or tricuspid stenosis, but the presence of a tumor plop and changes in physical findings with positional shifts can aid in differentiation.[2]

Radical resection of cardiac myxoma, including surrounding structures invaded by the tumor, is the preferred treatment. Recurrence after surgery is more common in individuals with familial or complex forms of the condition. Genetic analysis offers a valuable tool for identifying hidden myxomas in asymptomatic patients and those with a family history.[1] This case series reviews the clinical manifestations and management of two pediatric atrial myxoma cases, emphasizing the importance of comprehensive history taking, physical examination, echocardiography, and surgical resection. It offers valuable insights into the clinical challenges encountered in treating such cases.

CASE PRESENTATION

Case 1

A 16-year-old male complained of shortness of breath since 7 months prior. Shortness of breath is adversely affected by activity, such as walking to the bathroom. A mucus cough and fluctuated fever accompanied symptoms. Lips and fingertips began to turn cyanotic 4 months ago. Both limbs were swollen, and the abdomen was enlarged 3 months ago. The patient slept with 3 pillows. The patient was referred from Bratana Hospital Jambi with Rheumatic Heart Disease + large left atrium myxoma. Treatment history of benzatine penicillin intramuscular (unknown dose) 11 times.

Upon thorax examination, the chest appears symmetrical. Percussion reveals resonant sounds in both lung fields, with the cardiac apex located at the mid-axillary line on the left side at the 6th intercostal space. Auscultation shows vesicular breath sounds in both lungs without rales or wheezing. Heart sounds (S1 and S2) are regular, with no murmurs or gallops detected. Abnormal physical examination findings included positive shifting dullness and pretibial edema. The lab results indicate anemia, prolonged PT (15.4/18.8), elevated INR (1.39), and a high

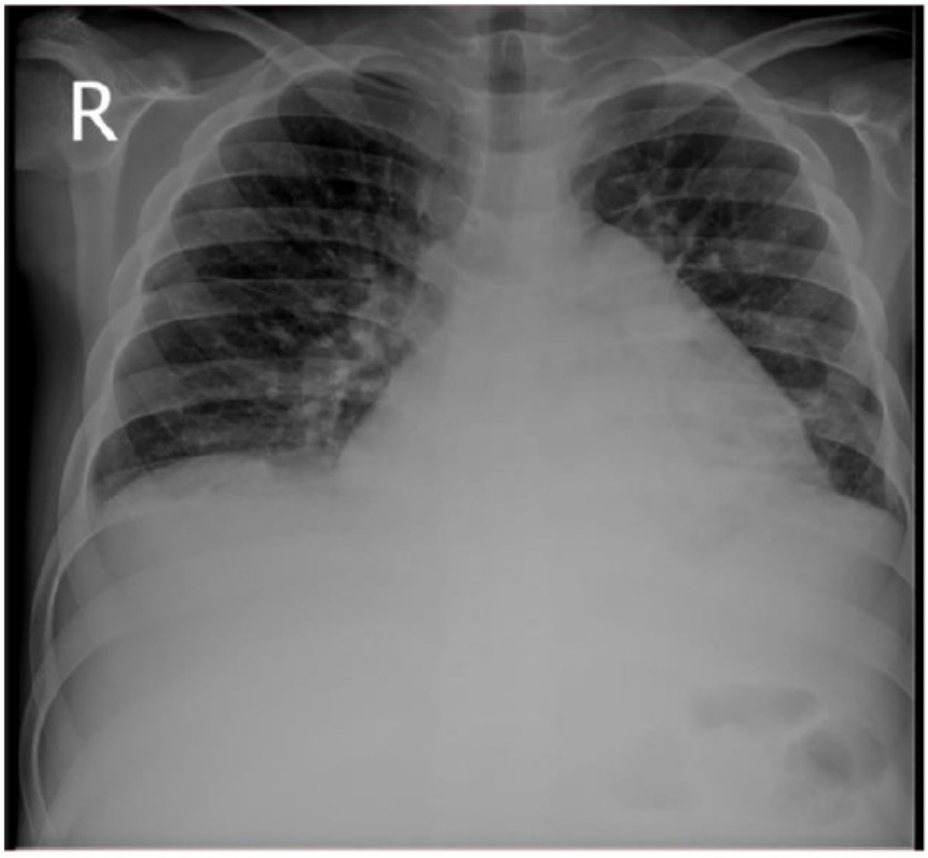

Figure 1. Chest X-ray of patient 1

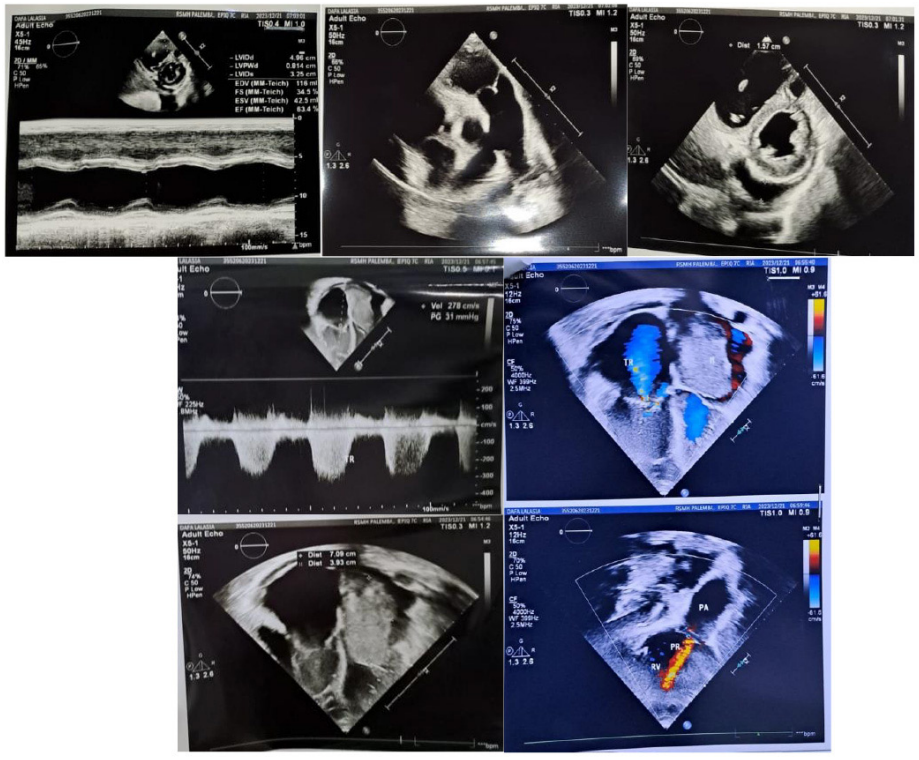

Figure 2. Echocardiography of patient 1

CRP (23.2). Echocardiographic evaluation reveals myxoma in the left atrium + mild tricuspid regurgitation + moderate pulmonary regurgitation + pericardial effusion. Abdominal ultrasound shows hepatomegaly with fatty liver and ascites.

The patient was diagnosed with decomp cordis et. causa left atrial myxoma + mild tricuspid regurgitation + moderate pulmonary regurgitation + pericardial effusion. The treatment plan includes administering intravenous furosemide 40 mg twice daily, captopril 25 mg twice daily orally, sildenafil 15 mg three times daily orally, and calcium supplements (Calos) one tablet twice daily orally. The patient underwent myxoma extirpation. The pathological impression shows cardiac myxoma in the left atrium.

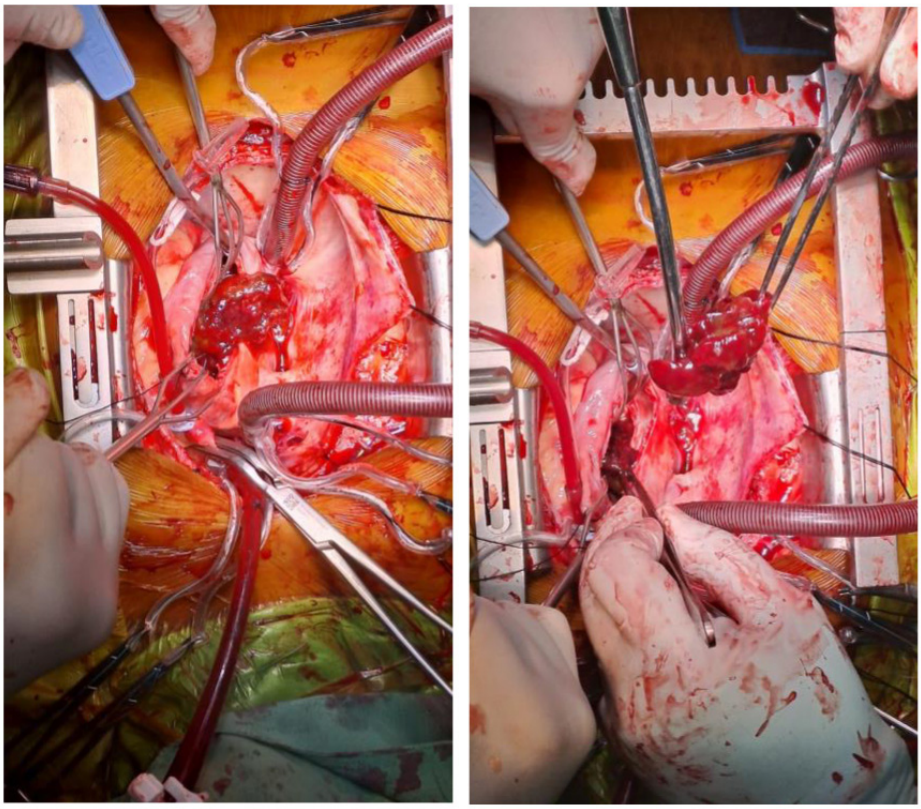

Figure 3. Intraoperative Myxoma Extirpation

Case 2

An 11-year-old girl's mother stated that her child has been experiencing shortness of breath for the past 2 months. Shortness of breath occurred when the child walked long distances and worsened when lying supine. The coughing was intermittent, the fever fluctuated, particularly at night with cold sweats, and there was difficulty gaining weight. The abdomen has been enlarged for the past month. There was a history of contact with tuberculosis patients, and the patient's mother had taken tuberculosis medications. The Mantoux test was positive.

Upon thorax examination, the chest appears symmetrical with a dry wound. Palpation reveals dullness in the right lung field, and the cardiac apex is palpable at the left mid-axillary line. Auscultation shows decreased vesicular breath sounds in the right hemithorax, with fine crackles at the lung bases bilaterally. There is no wheezing, and heart sounds (S1 and S2) are regular with tachycardia, and no murmurs are detected. On abdominal examination, the abdomen is distended. Palpation indicates positive shifting dullness, and auscultation reveals normal bowel sounds. Laboratory results show anemia (Hb 8.9, Ht 29), prolonged PT (15.2/20.6), elevated INR (1.53), and mild metabolic acidosis (base excess -6.9). The thorax x-ray shows increased broncho-vascular markings. The abdominal ultrasound reveals right pleural effusion and ascites.

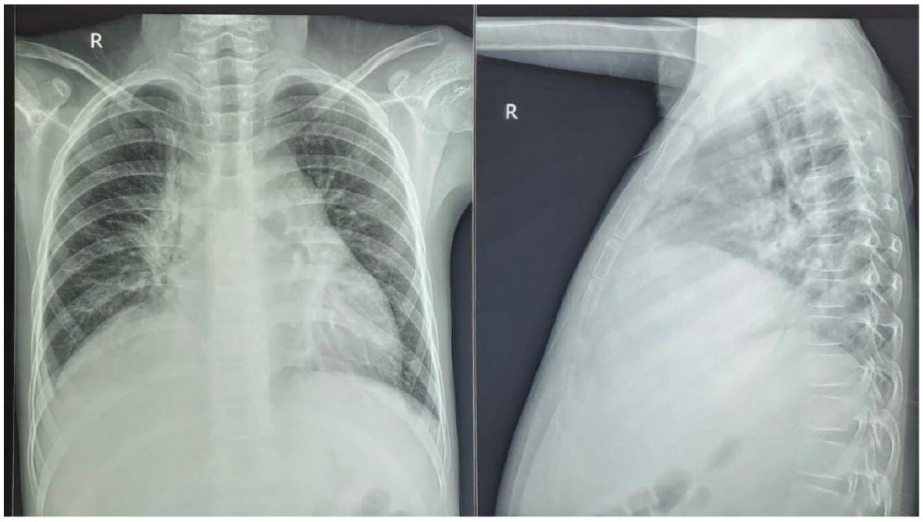

Figure 4. Chest X-ray of Case 2

Echocardiography shows a mass in the left atrium with a differential diagnosis of myxoma or thrombus.

The patient was diagnosed with a mediastinal tumor suspected to be thymoma post-thymectomy + pericarditis post-pericardiectomy + mass in the right atrium suspected to be myxoma or tuberculoma + clinical pulmonary tuberculosis on intensive phase anti-tuberculosis treatment. The treatment plan includes maintaining fluid at 80% of the required amount with D5 1/2 NS at 25 cc/hour. A PRC transfusion of 150 cc has been completed. The patient receives paracetamol 460 mg every 8 hours orally, ampicillin 1 g and cefotaxime 1 g every 8 hours intravenously, furosemide 25 mg twice daily orally, spironolactone 25 mg twice daily orally, and vitamin B6 25

mg once daily orally. Respiratory therapy includes RHZ (rifampicin, isoniazid, and pyrazinamide) once daily, ethambutol 500 mg once daily, prednisolone 3 times a day, and sputum analysis for rapid molecular test. The pathological impression shows fibrotic tissue with collagenization and hyalinization in the pericardial tissue and chronic granulomatous inflammation in the thymus.

Case 3

A 12-year-old male was brought by his mother in a raving condition to the RSMH emergency room suddenly 1 day before entering the hospital. There are no seizures. The patient has been experiencing shortness of breath for 1 month. Shortness of breath occurred when the child walked long distances and worsened when lying. Intermittent coughing, chest pain, dan joint pain. There was a history of lung infection and typhoid fever.

In the thoracic region, the chest appears symmetrical. Palpation revealed dullness. Percussion revealed resonant sounds in both lung fields. Auscultation showed vesicular breath sounds in both lungs without rales or wheezing. Heart sounds (S1 and S2) are regular, with a systolic murmur at the 5th intercostal space at the midclavicular line detected.

Laboratory results show Hb 10.9, Ht 35, prolonged PT (14./18,3), elevated INR (1.32), and prolonged APTT (29,5/33,1). Echocardiography shows a mass in the left atrium with a differential diagnosis of myxoma or Infective Endocarditis.

The patient was diagnosed with decreased consciousness + hemiplegia dextra due to a suspected thrombus due to a mass in the left atrium due to susp myxoma differential diagnosed Infective Endocarditis + severe tricuspid regurgitation + severe mitral regurgitation. The treatment plan includes administering intravenous furosemide 20 mg twice daily, aldactone 12,5 mg twice daily orally, intravenous ampicillin 675 mg four times daily, intravenous gentamycin 80 mg, and intravenous Manitol 135 mg three times daily. The patient underwent myxoma extirpation. The pathological impression shows cardiac myxoma in the left atrium.

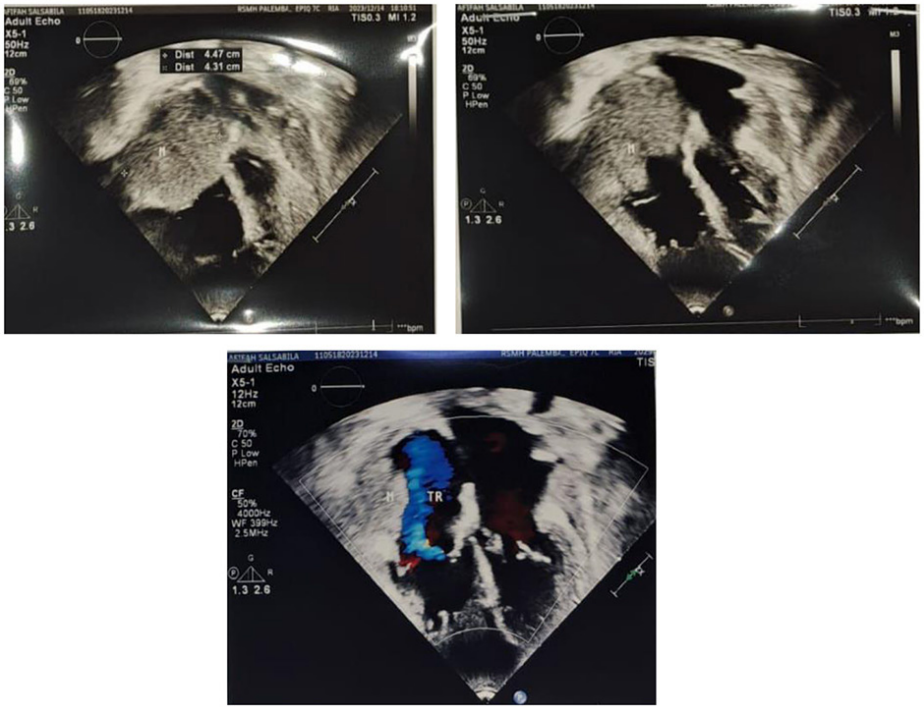

Figure 5. Echocardiography of Case 2

DISCUSSION

Atrial myxoma presents with a wide range of clinical manifestations influenced by the tumor's location, size, and mobility. Patients may experience symptoms such as arrhythmias, intracardiac flow obstruction, embolic events, and constitutional symptoms like fatigue, fever, and weight loss. Left-sided myxomas can obstruct mitral valve flow, mimicking mitral stenosis with symptoms including dyspnea, orthopnea, and syncope. In some cases, the tumor invades the myocardium, causing conduction disturbances that lead to arrhythmias like supraventricular or ventricular tachycardia. Myxomas can also mimic other cardiac conditions, such as mitral regurgitation and pulmonary embolism, and may present without a murmur, complicating diagnosis.[6,7] Additionally, embolic phenomena from the tumor or thrombus can result in cerebrovascular events or pulmonary embolism, depending on the tumor's location. Patients may present with edema and swelling over the body, along with symptoms of jaundice and heart failure. Pediatric cases may present with heart failure, arrhythmias, or hydrops, and the tumor's impact on the cardiac conduction system can lead to life-threatening complications, including ventricular tachycardia and sudden death. The systemic effects of the tumor, including vasculitis, endocarditis, and hematological abnormalities, further complicate the clinical picture.[3,8]

Cardiac myxomas can be asymptomatic in up to 50% of cases, often discovered incidentally during clinical evaluation. Physical examination may reveal signs such as cachexia, fever, cyanosis, clubbing, or rash, with neck vein engorgement and a prominent A wave in the jugular venous pulse. Precordial findings often mimic mitral stenosis, with a loud and widely split first heart sound (S1) due to delayed mitral valve closure caused by tumor prolapse. The pulmonary component of the second heart sound (P2) may vary depending on the presence of pulmonary hypertension. A characteristic "tumor plop," a low-pitched early diastolic sound following the second heart sound (S2), is a key finding, often mistaken for the opening snap of rheumatic mitral stenosis. This tumor plop is caused by the myxoma's impact against the endocardial wall or when its motion is halted. Additional heart sounds, such as a third (S3) or fourth (S4) heart sound, and diastolic murmurs of functional mitral

or tricuspid stenosis may also be present. Occasionally, a systolic murmur indicative of mitral or tricuspid regurgitation can be heard. Notably, the auscultatory findings of cardiac myxomas may change with the patient's position.[1,6]

In the cases presented, both patients exhibited symptoms such as shortness of breath, cyanosis, and edema, which are indicative of underlying cardiac pathology. The 16-year-old male experienced progressively worsening shortness of breath over seven months, accompanied by cyanosis, edema, and an enlarged abdomen—symptoms indicative of heart failure. Despite a history of rheumatic heart disease, the initial physical examination, which revealed resonant lung fields and regular heart sounds, may have initially directed the focus away from the possibility of a cardiac tumor.

Similarly, the 11-year-old female had a history of respiratory symptoms, including intermittent cough, fluctuating fever, and weight loss, coupled with a history of tuberculosis exposure, which may have initially masked the underlying cardiac condition. Physical examination findings, including dullness in the right lung field, decreased vesicular breath sounds, and a distended abdomen, while indicative of possible pleural effusion or heart failure, are also consistent with the presence of a large atrial myxoma. These cases highlight the challenges in diagnosing atrial myxoma in pediatric patients, as the subtle and non-specific symptoms can lead to delays in diagnosis and treatment, emphasizing the importance of thorough evaluation and consideration of cardiac tumors in the differential diagnosis.

Transthoracic echocardiography (TTE) is the primary and most practical tool for diagnosing atrial myxoma. TTE effectively identifies l, making it invaluable for initial diagnosis and preoperative assessment. The classical echocardiographic features of an atrial myxoma include a polypoid or papillary mass attached to the interatrial septum through a stalk, with characteristic mobility, often moving to and from into the cavity and sometimes protruding into the corresponding ventricular cavity across the atrioventricular valve.[1,9]

While transesophageal echocardiography (TEE) provides higher sensitivity and specificity, particularly for detecting small or atypically located tumors, TTE remains the most accessible, non-invasive, and cost-effective option, capable of being performed bedside even in emergencies. Echocardiography is crucial for distinguishing myxomas from other intracardiac masses like thrombi, which typically differ in location, appearance, and mobility. Additionally, Doppler echocardiography assesses the hemodynamic impact of the tumor. Although echocardiography provides comprehensive anatomical and physiological information, histopathological examination remains the gold standard for definitive diagnosis.[10] In this report, cardiac myxoma was initially displayed by TTE.

Surgical management is the primary treatment for pediatric atrial myxoma due to the high risk of cardiovascular complications and embolization. Zhao et al. highlight a surgical approach using median sternotomy with cardiopulmonary bypass (CPB) and myocardial protection via cold crystalloid cardioplegia. To prevent tumor fragmentation and embolization, the aorta and, in some cases, the main pulmonary artery are cross-clamped before cardiac manipulation. Intra-cardiac access is tailored to the tumor's location, with careful removal and thorough flushing of the heart cavity to ensure no residual fragments remain.[11]

Complete tumor excision is crucial, especially in cases with incomplete capsules, to avoid breaking the fragile mass. Pericardial patches are often used to repair defects in the atrial septum or left atrium wall, and valve repairs may be necessary if the tumor affects the mitral or tricuspid valves. The resected tumor is sent for pathological examination to rule out malignancy. Recurrence rates range from 5% to 14%, with incomplete resection being a key risk factor. Close monitoring with regular transthoracic echocardiography (TTE), magnetic resonance imaging (MRI), and clinical evaluations is essential to detect any signs of recurrence early.[11,12]

CONCLUSION

Pediatric atrial myxoma is a rare condition with a wide range of clinical manifestations, from asymptomatic cases to intracardiac obstruction, embolization, and constitutional symptoms. This often results in misdiagnosis as other more common heart diseases. Transthoracic echocardiography is useful as an initial diagnostic tool, as it effectively determines the tumor's location, size, shape, and connections, aiding in the selection of the optimal management strategy. Surgical excision is the primary treatment, with regular echocardiographic follow-up essential for the early detection of recurrence.

DISCLOSURE

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Conflicts of Interest
There are no conflicts of interest from the authors.

Author Contribution
The contribution of each author in writing this article is equal.

Consent for Publication
The patient gave written informed consent for the publication of this report and any related photos.

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