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CASE REPORT

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Left ventricular myxoma with carcinoid heart disease: A case report of a 14-year-old child from Jimma Medical Center of Ethiopia

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Key Clinical Message

Understand the importance of considering alternative diagnosis in patients presenting with atypical features, specially when they are not responding to the standard treatment. Understand the importance of considering common presentations of rare cases. Underscoring the critical importance of timely recognition and appropriate management of potentially life-threatening conditions.

KEYWORDS

carcinoid heart disease, cardiac mass, case report, Ethiopia, myxoma, rheumatic heart disease

1 | INTRODUCTION

Carcinoid heart disease is an uncommon cardiac manifestation linked with metastatic neuroendocrine tumors, characterized by fibrous plaque deposition on cardiac valves, which causes valvular dysfunction of variable severity. It is extremely rare in pediatric populations and might appear with unusual symptoms, making diagnosis difficult. Here, we present the case of a 14-year-old child who developed left ventricular myxoma as a result of carcinoid heart disease but was misdiagnosed as having rheumatic heart disease, mainly due to the lack of experts in the most of rural hospitals and unavailability of basic echocardiographic trainings, for 3 years. Due to the disease's rarity and the length of time it took for the confirmatory investigations to be completed, the case was lost, as heart failure and infection therapy were primarily advised.

2 | CASE PRESENTATION

A 14-year-old male adolescent presented with a 2-week history of exacerbated shortness of breath and generalized body swelling, which began in the legs and spread to the abdomen, upper limbs, and face. Over the last year, he has had occasional dry cough, palpitations, loss of appetite, and significant weight loss. Despite past visits to hospital institutions when he was diagnosed with a cardiac condition, the patient did not fully recover as his conditions are not well-addressed. Otherwise, He had no history of psychiatric problems, drug allergies, chicken pox, mumps, smallpox, or any other childhood illnesses. He was raised in Ethiopia's Oromia region, where he was born. His father has nine children, none of them has similar complain.

Upon evaluation, the patient was acutely sick looking on chronic background and in cardiorespiratory distress. His blood pressure was 95/60 mmHg; pulse rate was

112 beats/min and feeble; breathing rate was 30 breaths/min; temperature was 36.7°C and oxygen saturation was 89% with room air. Breath sounds was less audible over the left infrascapular region. The jugular venous pulse was elevated 5 cm above the sternal angle, and there was grade IV holosystolic murmur best heard at apex, hepatomegaly and grade II bilateral pitting edema.

2.1 | Differential diagnosis

Congenital heart disease, rheumatic heart disease, and dilated cardiomyopathies are among the most common differential diagnoses for an adolescent presenting with cough, exertional shortness of breath, body swelling, and an intracardiac mass, which can be a consequence of these conditions. Congenital heart disease refers to a group of birth defects that interfere with the normal function of the heart. Though no evidence has been established in most cases, genetic diseases such as down syndrome, maternal infections such as rubella during pregnancy, smoking, diabetes, and certain medications during pregnancy can all have an impact, which the family denies all.

Further imaging investigations are required to differentiate between the other alternatives because the signs and symptoms are nearly identical, including rapid heartbeat, rapid breathing, swelling of the legs, extreme tiredness and fatigue, as well as cyanosis. No lesions on echocardiography and cardiac computed tomography were suggestive of congenital heart lesions in the patient.

The next most common differential is rheumatic heart disease, particularly in Sub-Saharan Africa, where the case is most prevalent. It is primarily caused by untreated streptococcal throat infections in an overcrowded setting. Patients with rheumatic heart disease have symptoms and signs similar to those of heart failure, which may not be specific, necessitating further evaluation, as it was done for the patient, of whch none of the findings is suggestive of rheumatic heart disease.

Dilated cardiomyopathy is a disorder in which the left ventricle dilates, with or without other chambers, causing chest disconfort, exhaustion, fluttering or palpitations in the chest, a heart murmur, difficulty breathing due to fluid in the lungs, and lower extremity swelling. While certain physical examination results may suggest, they are also not specific, necessitating the use of imaging, though imaging didn't reveal any evidence of dilated cardiomyopathy.

2.2 | Investigations and management

Baseline investigations and diagnostic imaging were used to confirm the diagnosis and rule out the other possible alternative diagnoses. More sophisticated imaging like cardiac magnetic resonance imaging and others were not used as they were not available in the set up. Accordingly, a full blood count revealed 8000 WBC/mm^3, hemoglobin of 13.8 g/dL, and 399,000 platelets/mm^3. CRP was raised twentyfold, as summarized below (Table 1).

PA chest X-ray imaging of the patient revealed Grade I pulmonary edema on top of other complications including cardiomegaly, pleural effusion (Figure 1).

Transthoracic echocardiography revealed thickened tricuspid and pulmonic valves with severe tricuspid regurgitation (V_{max} = 4.6 m/s with pressure gradient of 98 mmHg) and pulmonic valve stenosis (V_{max} = 4.7 m/s with pressure gradient of 88 mmHg), severe pulmonary hypertension, with preserved left ventricular systolic function. There was a 14.4 by 12.3 mm echogenic mass attached to the left ventricular apex, suggestive of left ventricular myxoma (Figure 2).

Electrocardiogram performed on admission demonstrates sinus tachycardia, incomplete right bundle branch block (RBBB), and extreme axis deviation, which is followed by p-pulmonale (Figure 3).

The pre- and post-contrast chest tomography images (Figure 4 and Video S1) below show dilated chambers on the right side of the heart, as well as a hypodense, non-enhancing lesion in the left ventricular apex, suggestive of myomaand bilateral pleural effusion.

3 | RESULTS AND FOLLOW UP

The patient was diagnosed with acute decompensated heart failure due to Cardiac carcinoid disease with left ventricular apical myxoma, which was exacerbated by disease progression and pneumonia. Treatment included frusemide at a dosage of 1 mg/kg, antibiotics, and supportive medical care with close monitoring. As these therapies would be insufficient, it was decided to conduct additional investigations into the patient's condition, including tests of urinary 5-hydroxyindoleacetic acid (5-HIAA) levels and octreotide scintigraphy. Unfortunately, due to the patient's grave condition and significant decompensation, he died before these diagnostic workups and definitive surgical treatment could begin.

4 | DISCUSSION

The case presented underscores the diagnostic difficulty associated with diagnosing unusual cardiac pathologies; the carcinoid heart disease and left ventricular myxoma, particularly in resource-limited settings such as Jimma Medical Center of Ethiopia. In this

TABLE 1 Serological investigation done for the patient during admission.

S	References range	
White cell count (per μL)	80,000	4000-15,000
Neutrophil (%)	84	37–72
Lymphocyte (%)	10	20–50
Hemoglobin (mg/dL)	13.8	11–16.5
Platelet (per μL)	399,000	150,000-450,000
ESR (mm/h)	10	≤20
Creatinine (mg/dL)	0.99	0.5–1.2
Blood urea nitrogen (mg/dL)	12.6	7–20
Sodium (mmol/L)	139	135–145
Potassium (mmol/L)	4.4	3.5–5.5
Chloride (mmol/L)	106	98–107
AST (IU/L)	41.3	0–40
ALT (IU/L)	44.3	0–41
ALP (IU/L)	139	40–130
Random blood glucose (mg/dL)	102	
Prothrombin time (s)	12.9	10–14
APTT (s)	29.1	22–38
INR	1.06	0.7–1.3
TSH (μIU/mL)	0.745	0.27–4.2
VDRL	Non-reactive	
HIV test	Non-reactive	
Serum VDRL test	Reactive	
Hepatitis B surface antigen test	Negative	
Hepatitis C antibody test	Negative	
Urinalysis	Unremarkable	
Pleural fluid analysis	120 cells (90% lymphocytes)	
ASO titer	Nonreactive	
Serum albumin (g/dL)	4.1 g/dL (normal range)	
Pleural fluid cytology	Reactive effusion	
Sputum X-pert for sputum	M. tuberculosis not detected	

Abbreviations: ALP, alkaline phosphatase; APTT, activated partial thromboplastin time; AST/ALT, aspartate transaminase/alanine transaminase; ESR, erythrocyte sedimentation rate; HIV, human immunodeficiency virus; TSH, thyroid stimulating hormone; VDRL, venereal disease research laboratory.

case, the original misdiagnosis of chronic rheumatic heart disease (RHD) highlights the difficulty of distinguishing between distinct cardiac disorders based simply on clinical presentation and inadequate diagnostic techniques.[1]

Carcinoid heart disease is an uncommon consequence of carcinoid syndrome, a neuroendocrine tumor disorder marked by the release of vasoactive chemicals such as serotonin. Carcinoid heart disease develops as a result of endocardial fibrosis and valve dysfunction, which causes cardiac symptoms.[2]

A detailed medical history, physical examination, and assessment of symptoms suggestive of carcinoid syndrome are usually used in the diagnostic diagnosis of carcinoid cardiac disease. Biochemical testing, especially measurement of urinary 5-hydroxyindoleacetic acid (5-HIAA) levels, is critical for detecting increased serotonin metabolites linked to carcinoid activity. Echocardiography is critical for assessing cardiac anatomy and function, with a focus on detecting valve abnormalities and symptoms of right heart involvement. Advanced imaging modalities, such as cardiac MRI and CT scans, provide precise views of heart abnormalities, which aid in treatment planning.[2] However, diagnosing carcinoid heart disease can be difficult due to its ambiguous clinical presentation, which can overlap with other cardiac disorders, as demonstrated in this case.[2]

Treatment for carcinoid heart disease focuses on symptom control and disease management. Somatostatin analogs such as octreotide or lanreotide are frequently used in medical treatment to offset excessive hormone secretion. Valve replacement surgery may be required in cases of severe valvular malfunction as in this case, while heart transplantation is being investigated for patients with end-stage illness. Regular monitoring and surveillance are

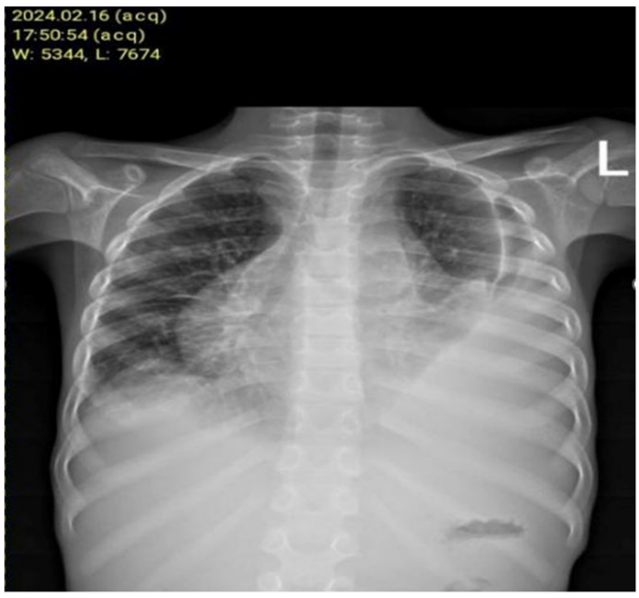

FIGURE 1 PA chest X-ray of the patient showing cardiomegaly, sign of right atrial and right ventricular enlargement, with bilateral pleural effusion with futures of pulmonary edema.

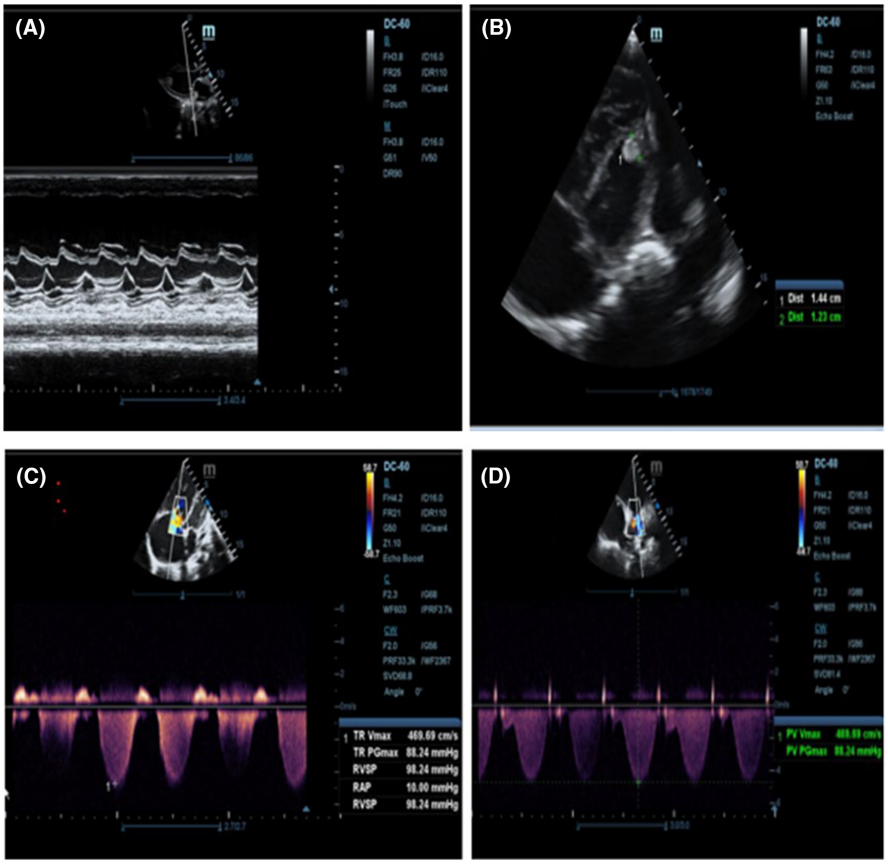

FIGURE 2 Transthoracic echocardiography of the patient showing a 14.4 mm by 12.3 mm echogenic mass attached to the left ventricular apex (A and B), while the CW, PW and color flow Doppler examination reveals severe Tricuspid regurgitation, Severe pulmonic valve stenosis with dilated main pulmonary artery (C and D).

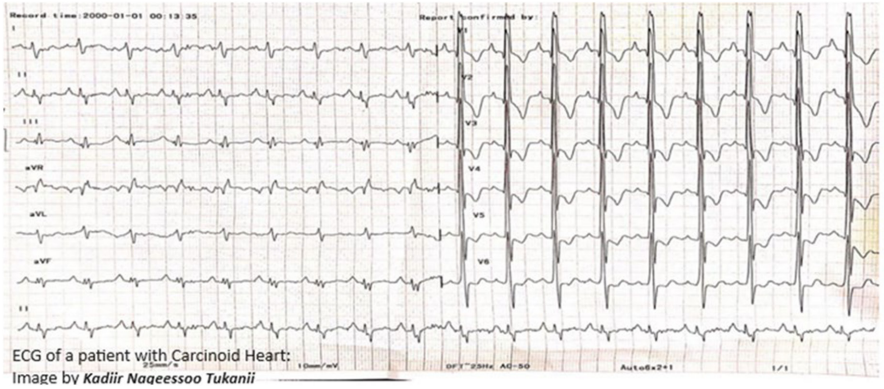

FIGURE 3 A 12-lead electrocardiogram of the patient reveals sinus tachycardia, incomplete right bundle branch block (RBBB), Extreme axis deviation with p-pulmonale.

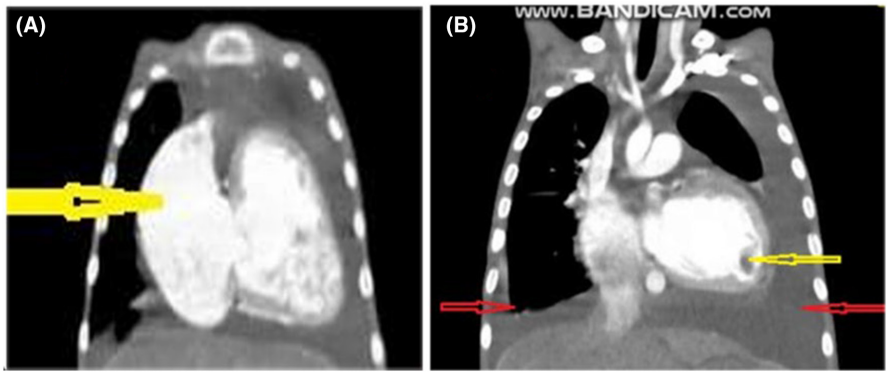

FIGURE 4 Post contrast sagittal CT of chest of the patient demonstrates: Dilated right side cardiac chambers (*yellow arrow*) with hypodense non-enhancing mass in the left ventricular apex (*yellow arrow*), bilateral plural effusion (*red arrow*) (A and B).

critical in detecting illness progression early and providing prompt intervention.[2]

Similarly, left ventricular myxomas are rare primary cardiac tumors that can cause symptoms similar to other cardiac diseases, including RHD. Myxomas can block blood flow, causing symptoms like dyspnea, chest discomfort, and other indicators of heart failure.[3] A left ventricular apical mass is often diagnosed with comprehensive imaging evaluations, such as echocardiography and cardiac MRI, to determine the mass's size, location, and composition. Additional diagnostic tests, such as cardiac catheterization or biopsy, may be used to confirm the diagnosis and rule out other heart diseases. Once detected, care of left ventricular apical tumors frequently necessitates a multidisciplinary approach, with surgical excision being the preferred treatment method to avoid consequences such as embolization, blood flow blockage, or arrhythmias.[4,5]

Our patient was treated only with supportive care as there is no surgical treatment options in the setup and from the fact that the patient's grave condition, exacerbated by severe infections, made it difficult to undertake additional diagnostic investigations and initiate timely referral for surgical intervention. Delayed detection that led to optimal treatment of this very unusual case worth mentioning as it would provide important input for care of similar patients especially in those with resource-limited setup.

4.1 | Limitation

The problems in this instance were numerous. The lack of confirmatory testing for carcinoid heart disease, using urinary 5-hydroxyindoleacetic acid (5-HIAA) levels and octreotide scintigraphy, created a substantial challenge in precisely diagnosing the condition. Similarly, the lack of cardiac catheterization and cardiac surgery, which are required for final management and histopathologic confirmation of the left ventricular mass, hampered the diagnostic and therapeutic process.

5 | CONCLUSION

While this case highlights the diagnostic challenges and limitations associated with diagnosing rare cardiac conditions such as carcinoid heart disease and left ventricular myxoma in resource-constrained settings, it also emphasizes the importance of having high clinical suspicion and early intervention in improving patient outcomes. Addressing these issues requires a holistic approach that includes changes to improve healthcare infrastructure, including establishing basic surgical interventions for similar cases, diagnostic capabilities, and overall healthcare practices.

AUTHOR CONTRIBUTIONS

Elsah Tegene Asefa: Data curation; methodology; validation; writing – review and editing. **Tamirat Godebo Woyimo:** Data curation; investigation; software; validation; writing – original draft. **Kedir Negesso Tukeni:** Conceptualization; data curation; formal analysis; investigation; methodology; writing – original draft; writing – review and editing.

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DATA AVAILABILITY STATEMENT

The data that support the findings of this case study are available within the manuscript.

ETHICS STATEMENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

CONSENT

Written informed consent was obtained from the patient's family to publish this case report in accordance with the journal's patient consent policy.

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

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