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Case Report

A Case of Atypical Cor Triatriatum Sinister Persisting into Adulthood

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Abstract

Background: Cor-triatriatum is a rare congenital cardiac anomaly characterized by a fibromuscular membrane that divides the left atrium into two chambers, giving the appearance of three atrial compartments. It typically presents during infancy or childhood due to obstructive symptoms but may remain undiagnosed until adulthood in milder or non-obstructive cases. *Case Presentation:* A 33-year-old Ethiopian male with no known past medical history presented with a one-month history of progressive shortness of breath, orthopnea, paroxysmal nocturnal dyspnea, and bilateral leg swelling. He also reported a productive cough, pleuritic chest pain, and high-grade intermittent fever of similar duration. Physical examination and initial investigations suggested decompensated heart failure and pneumonia. Transthoracic echocardiography revealed findings consistent with cor-triatriatum sinister. Despite initiation of supportive care, intravenous antibiotics, diuretics, and mechanical ventilation, the patient's condition deteriorated. He ultimately developed cardiogenic shock and died during hospitalization. *Conclusion:* Cor-triatriatum is a rare but important cause of heart failure in adult patients. This case demonstrates that congenital heart disease can remain clinically silent for decades and may present with acute decompensation. Awareness and early recognition of this anomaly, especially in resource-limited settings, are essential for timely diagnosis and management. Point-of-care ultrasound can be a valuable tool in such environments for rapid cardiac assessment and should be considered when evaluating patients with unexplained heart failure symptoms.

Keywords

Atypical Cor Triatriatum Sinister, Heart Failure, Young Adult, Case Report

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1. Introduction

Cor triatriatum is characterized by the existence of a fibromuscular membrane within either the left or right atrium, creating the appearance of three atria. It is associated with other cardiac anomalies in 80% of cases [1]. The cause of cor triatriatum is not well understood, and several theories exist on the embryologic origin. The treatment for cor triatriatum may be aggressive or conservative and differs according to the severity of symptoms. Overall, in cases of isolated cor triatriatum, the prognosis is excellent. In most patients, the opening is severely restrictive and about 75% of those born with CT die in infancy. In rare cases, however, the onset of clinical manifestations may be delayed until adulthood if the opening is large. Such adults may be asymptomatic where the diagnosis is incidental, [2] the presenting clinical picture may be dyspnea, hemoptysis, heart failure, [3] pulmonary hypertension and/or atrial fibrillation. Here we report a case of 33 years old male patient who presented with heart failure in whom atypical cor triaatritum has been diagnosed due the vertical orientation of the dividing membrane, Illustrating very unusual and interesting case.

2. Case Presentation

A 33-year-old male patient presented to the emergency department with progressive fatigue, shortness of breath, orthopnea requiring two pillows, and paroxysmal nocturnal dyspnea (PND) that has persisted for one month. His symptoms have worsened over the past two weeks and are accompanied by swelling in both lower extremities. He also reports a productive cough that initially produced whitish, frothy sputum, which has since become blood-stained over the same time frame. Additionally, for the past week, he has experienced low-grade intermittent fever and sharp chest pain on both sides, which worsens with coughing.

For these compliant he sought treatment at a local general hospital, where he was admitted for two weeks and treated with antibiotics (ceftriaxone) and diuretics (furosemide). He was referred to our institution due to inadequate response to treatment. The patient has no prior history of similar complaints, chest heaviness, changes in eye color, hematemesis, diarrhea, constipation, changes in stool color, body weakness, abnormal movements, or urinary or stool incontinence. He also denies any history of dysuria, increased urinary frequency, or changes in urine color. There is no known history of chronic medical conditions in himself or his family. He works as a farmer in a rural area of Ethiopia and is the youngest child in his family. He is currently married but has no children.

Upon physical examination, the patient appeared acutely ill and was in respiratory distress, with a respiratory rate of 28 breaths per minute and an oxygen saturation of 94% while receiving 10 L of supplemental oxygen via a face mask. His blood pressure was 106/74 mmHg, pulse rate of 110 beats per minute, regular and full in volume, and a temperature of 36.9 ℃. Chest examination revealed absent air entry in the lower bilateral posterior two-thirds of the lungs, bronchial breath sounds were noted bilaterally over the upper third lung fields. The cardiovascular examination indicated raised jugular venous pressure at 5 cm above the sternal angle and a grade IV holosystolic murmur best heard at the apex, which radiated to the axilla. Abdominal examination showed a tender, palpable liver extending 5 cm below the right costal margin, with positive shifting dullness. The musculoskeletal examination revealed grade two bilateral pitting edema. No significant findings were noted in other systems.

A bedside ultrasound (point-of-care ultrasound, POCUS) demonstrated bilateral pleural effusions with bilateral B-lines and signs of consolidative changes. The inferior vena cava appeared plethoric (diameter 2.53 cm) with minimal respiratory variation (10%), and there was visible membrane dividing the left atrium on the apical four-chamber view (Figure 1). Ejection fraction was estimated to be 35%. ECG showed a sinus tachycardia with rate of 112 bpm on monitor. High sensitive troponin was normal.

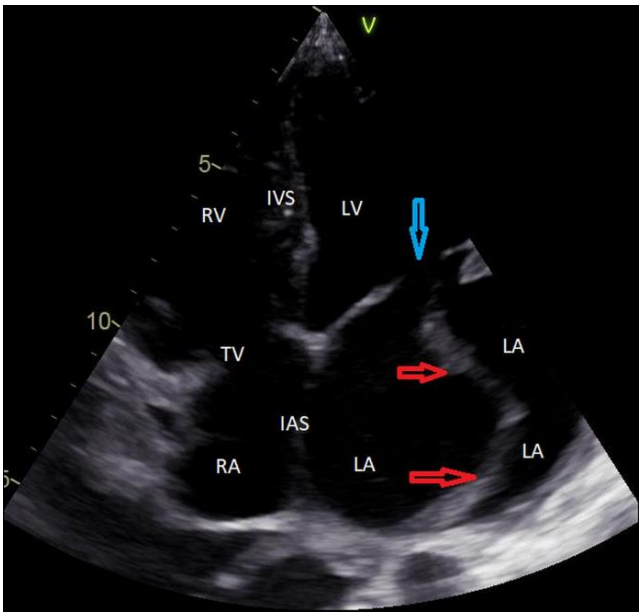

Figure 1. *Transthoracic Echocardiography. Apical four chamber view demonstrating well-formed vertical membranous septum dividing the left atrium in to two (red arrow). Blue arrow, Mitral valve; RA, right atrium; LA, left atrium; LV, left ventricle; RV, right ventricle; IVS, inter-ventricular septum; IAS, inter-atrial septum.*

The patient was diagnosed with NYHA class IV stage C heart failure secondary to congenital heart disease (CHD) due to cor triatriatum, along with hospital-acquired pneumonia (HAP). Diuretics (furosemide) were initiated at a dose of 80 mg Intravenous stat. The patient produced 250 ml of urine and was placed on furosemide 80 mg IV three times daily, along

with spironolactone 25 mg orally once daily. Antibiotic therapy was adjusted to include ceftazidime 1 gm IV TID and vancomycin 1 gm IV BID, considering the diagnosis of hospital-acquired pneumonia after relevant investigations were sent (see Table 1).

The next day, despite the management provided, the patient's respiratory condition deteriorated, with the respiratory rate rising to 40. Blood pressure dropped to 85/60, and the patient became agitated, leading to the decision to intubate. After optimizing hemodynamic status, the patient was intubated and placed on mechanical ventilation. Norepinephrine was initiated at a dose of 0.05 mcg/kg/min, and due to the addition of a diagnosis of cardiogenic shock, the dosage was increased to achieve a mean arterial pressure (MAP) above 65. Unfortunately, the patient passed away despite efforts to optimize vasopressor support and antibiotic therapy.

Table 1. *Investigation summary of the patient upon initial presentation.*

Complete blood count	Patient value
WBC	10.3×10^3
Hemoglobin	12.7 g/dl
hematocrit	37.4%
Red blood cell	5.10×10^6
Platelet count	489×10^3
neutrophil	72.4%
lymphocyte	19.2%
eosinophil	2.8%
Creatinine	0.79 mg/dl
Urea	22.7 mg/dl
SGOT	17.4 IU/L
SGPT	7.5 IU/L
Total bilirubin	0.698 mg/dl
Albumin	2.65 g/dl
Serum electrolyte	
sodium	131
potassium	2.86
chloride	86.2
Pleural fluid analysis	
Cell count	900
Lymphocyte	80%
Neutrophil	20%
Protein	1.1 g/dl
Glucose	123 mg/dl
LDH	120 u/l

3. Discussion

Cor triatriatum is a rare cardiac anomaly that occurs when a fibromuscular membrane divides atrium into 2 distinct chambers [1]. The membrane may be located in the right atrium (cor triatriatum dexter [CTD]) or left atrium (cor triatriatum sinister). CTS results from an improper merging of the common pulmonary vein with the left atrium during heart development, which is more commonly seen than CTD [2]. This cardiac anomaly was first described by Church in 1868 and reported to have an incidence of 0.4% at autopsy of patients with congenital cardiac disease and 0.2% among patients undergoing transesophageal echocardiography [3]. Although traditionally considered a pediatric condition, some cases remain undiagnosed until adulthood like in our patient due to mild or non-obstructive forms [1]. Studies have reported that late presentation in adults is often due to progressive fibrosis, membrane thickening, or superimposed heart disease [4, 5].

The severity of symptoms depends on the degree of obstruction caused by the membrane. Obstructive CTS presents with symptoms similar to mitral stenosis, including progressive dyspnea, orthopnea, PND, and exercise intolerance, [3, 6]. Pulmonary hypertension and right heart failure may develop due to chronic left atrial hypertension [7]. Non-obstructive CTS can remain asymptomatic and be detected incidentally during echocardiography [8]. Cases of atrial fibrillation, systemic embolism, and infective endocarditis due to stasis in the accessory left atrial chamber have also been reported [9, 10]. This aligns with our patient's presentation and highlights the rarity of the case as he remained undiagnosed until adulthood.

The diagnosis is usually established by 2 D TTE (2-dimensional transthoracic echocardiography). Color flow mapping usually demonstrates increases in velocity and turbulent flow, suggesting obstruction that can be assessed by continuous wave Doppler through the membrane [11]. Transesophageal echocardiography is superior to transthoracic imaging to diagnose cor triatriatum, providing better imaging of the left atria, left atria appendage, morphology of the dividing membrane and the degree of obstruction. [12]. Three D echocardiography is a more recent diagnostic tool providing additional information, able to demonstrate the entire membrane, the size, the location and the number of openings in the dividing membrane [12]. It also provides superior anatomical detail, distinguishing CTS from other conditions such as mitral stenosis or supravalvular mitral rings [8]. Cardiac MRI and CT angiography further aid in defining the membrane anatomy and ruling out other congenital anomalies [5, 13]. Histopathological examination

post-surgery reveals a fibromuscular membrane composed of endocardial tissue without valvular structures, confirming the diagnosis [14]. In our patient, further investigations were not possible due to the patient's critical condition, making transport unsafe. However, bedside echocardiography revealed a well-formed, vertically oriented membrane in the left atrium (Figure 1). Additionally, cardiac MRI and histopathology could not be conducted because these resources were unavailable.

The primary treatment for symptomatic CTS is surgical resection of the membrane, which provides definitive relief [1]. Medical management (diuretics, anticoagulation, and heart failure therapy) may be used for symptomatic relief but does not address the underlying defect [7]. In cases of concomitant cardiac surgery (e.g., mitral valve repair or atrial fibrillation ablation), CTS correction is often performed simultaneously [13]. There are also rare reports of successful balloon catheter dilation of the communication between the proximal and distal chambers, but the long-term outcomes are yet to be determined [15]. In our case, the patient presented with congestion and was treated medically with diuretics and antibiotics due to an additional diagnosis of a hospital-acquired infection.

Reported long-term results of surgery are excellent low rates of perioperative mortality and considerable symptomatic relief [5]. Long-term survival following membrane resection is similar to that of the general population, as long as there is no remaining obstruction or pulmonary hypertension [10]. Patients who are diagnosed and treated prior to the development of severe pulmonary hypertension tend to have the most favorable prognosis [10].

This case report has limitations arising from the lack of additional investigations that could have provided a more detailed characterization of the cardiac lesion, such as cardiac MRI and post-mortem histological examination of the tissue. Furthermore, the patient's critical condition and the unavailability of such investigations rendered surgical management unfeasible.

4. Conclusion

Cor triatriatum is a rare cardiac anomaly characterized by a fibromuscular membrane that divides the atrium into two chambers. Although typically diagnosed in children, some cases, like that of our patient, remain undetected until adulthood. The severity of symptoms correlates with the degree of obstruction caused by the membrane. Diagnosis is usually confirmed through 2D transthoracic echocardiography, while histopathological examination post-surgery reveals a fibromuscular membrane made of endocardial tissue without valvular structures. In resource-limited settings like ours, bedside echocardiography and clinical findings play a crucial role. Surgical resection of the membrane is the primary treatment for symptomatic cases and provides definitive relief, while medical management are for symptom control. Long-term outcomes following surgery are generally excellent.

Abbreviations

PND	Paroxysmal Nocturnal Dyspnea
NYHA	New York Heart Association
HAP	Hospital Acquired Pneumonia
CHD	Congenital Heart Disease
ECG	Electrocardiography
MAP	Mean Arterial Pressure
CTD	Cor Triatriatum Dexter
CTS	Cor Triatriatum Sinister
2DTTE	2 Dimensional Transthoracic Echocardiography
MRI	Magnetic Resonance Imaging

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Zelalem Getahun: Conceptualization, Data curation, Supervision, Validation

Alemu Bimrew Mesekere: Data curation, Resources, Writing – review & editing

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Conflicts of Interest

The authors declare no conflicts of interest.

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