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ACUTE PERICARDITIS IN AN ADULT WITH CYSTIC FIBROSIS: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Introduction: Cystic fibrosis, an autosomal recessive disorder (1 in 3,000–6,000 births), causes thick mucus and recurrent lung infections. Improved survival has revealed rare complications such as constrictive pericarditis and cardiac tamponade.
Case description: A 24-year-old female with cystic fibrosis, diagnosed at nine months, presented with two weeks of dyspnoea. Examination revealed bilateral rhonchi, facial oedema and non-pitting thigh oedema. Laboratory results showed a white blood cell count of 11.3 $\times 10^9$/l, erythrocyte sedimentation rate of 99 mm/hr and C-reactive protein level of 45.3 mg/dl. Initially admitted for cystic fibrosis exacerbation, she was found to have pericardial effusion. Despite facial and lower extremity swelling, she denied cardiac symptoms. During hospitalisation, she developed haemodynamic instability requiring pericardiocentesis, with echocardiography confirming constrictive pericarditis. Chest computed tomography angiography showed pericardial calcifications; autoimmune workup was negative. She was discharged on colchicine with close follow-up.
Conclusion: Clinicians should have a high index of suspicion for underlying cardiac complications in patients presenting with exacerbations of cystic fibrosis.

KEYWORDS

Cystic fibrosis, pericarditis, pericardiocentesis, pericardial effusion, constrictive pericarditis

LEARNING POINTS

- Pericardial disease – including constrictive pericarditis and tamponade – though rare, can complicate cystic fibrosis in adults and may mimic pulmonary exacerbations. Early cardiac imaging is crucial for timely diagnosis.
- This case highlights the need for a high index of suspicion in cystic fibrosis patients with unexplained dyspnoea, oedema or haemodynamic instability, especially in the setting of pericardial calcifications.
- Medical management with colchicine and diuretics can be effective in selected cystic fibrosis patients with constrictive physiology, potentially avoiding the need for invasive pericardiectomy.

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INTRODUCTION

Cystic fibrosis is an autosomal recessive disorder affecting approximately 1 in 3,000 to 1 in 6,000 live births[1]. It results from mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene on chromosome 7, which encodes the CFTR protein. This protein, expressed in epithelial cells, facilitates chloride and bicarbonate transport across cell membranes[2]. Mutations in the CFTR disrupt ion transport in various organs, including the respiratory tract, skin, pancreas and vas deferens, producing thick, obstructive mucus. In the respiratory system, this mucus impairs clearance, fosters microbial growth – often by resistant organisms such as methicillin-resistant *Staphylococcus aureus* and *Pseudomonas aeruginosa* – and triggers a cycle of infection, inflammation and scarring[3]. Over time, these processes damage bronchoalveolar structures, reducing lung function and increasing oxygen requirements, perpetuating a vicious cycle of pulmonary deterioration[4].

Cystic fibrosis also causes chronic obstructive pancreatitis, pancreatic insufficiency and diabetes mellitus due to islet cell damage. Historically, cystic fibrosis was predominantly a paediatric condition due to limited survival into adulthood. However, advances in treatment – including inhaled mucus-thinning agents, antibiotics, anti-inflammatories, pancreatic enzyme supplements and highly effective CFTR modulators – have significantly improved outcomes. Median life expectancy for individuals with cystic fibrosis born between 2018 and 2022 is projected to exceed 56 years[5].

With this improved survival, the cystic fibrosis population is now primarily composed of adults, who accounted for 59.4% of cases in the United States in 2022. This shift has revealed previously uncommon complications such as pericarditis, tamponade and cor pulmonale, which were rare due to shorter life expectancy. Here, we describe a case of constrictive pericarditis with pericardial thickening and calcification, complicated by moderate pericardial effusion and tamponade symptoms, in a 24-year-old woman with cystic fibrosis.

CASE DESCRIPTION

A 24-year-old female with a history of cystic fibrosis diagnosed at nine months of age, complicated by insulin-dependent diabetes mellitus, pancreatic insufficiency, liver cirrhosis and gastro-oesophageal reflux disease, presented with two weeks of shortness of breath and generalised body swelling. Her symptoms included a productive cough with green sputum and chest tightness, unresponsiveness to inhalers, nebulisers and airway clearance therapies. Two days before admission, she noted progressive swelling in her face and legs. She denied chest pain, orthopnoea, palpitations, nausea, vomiting or dysuria.

Home medications included albuterol, inhaled aztreonam, dornase alfa nebuliser, escitalopram, lipase-protease-amylase, trazodone, and insulin lispro and glargine. On admission, her vitals were blood pressure 100/82 mmHg, heart rate 112 bpm, respiratory rate 20 breaths/min and SpO_2 90% on room air. Physical examination revealed facial swelling, bilateral lung rales and rhonchi, decreased breath sounds, a protuberant abdomen and bilateral non-pitting leg oedema up to the thighs. Initial laboratory tests showed leucocytosis (white blood cells 11,300/µl), elevated alkaline phosphatase, low albumin, erythrocyte sedimentation rate 99 mm/hr and C-reactive protein 45.3 mg/l. Bedside cardiac ultrasound demonstrated moderate pericardial effusion. A chest X-ray (CXR) revealed bilateral infiltrates and computed tomography (CT) angiography confirmed bilateral pleural effusions, moderate pericardial effusion and new pericardial calcifications compared to prior imaging. Abdominal imaging showed mild ascites, nodular liver consistent with cirrhosis and diffuse anasarca.

She was admitted to the intensive care unit (ICU) for acute hypoxic respiratory failure, initiated on oxygen supplementation and empirical antibiotics, and referred to cardiology for pericardial effusion management. Transthoracic echocardiography showed an ejection fraction of 65–70% without tamponade. Autoimmune workup (e.g. anti-CCP, anti-dsDNA) was negative. On day three, she

Characteristic	Pleural fluid	Pericardial fluid
Colour	yellow	red
Appearance	clear	bloody
White blood cells	294	1,050
Red blood cells	47	39,5080
Segmented neutrophils (%)	51	43
Lymphocytes (%)	40	30
Monocytes (%)	6	27
Mesothelial cells (%)	3	-

Table 1. Characteristics of pleural and pericardial fluids obtained from the patient.

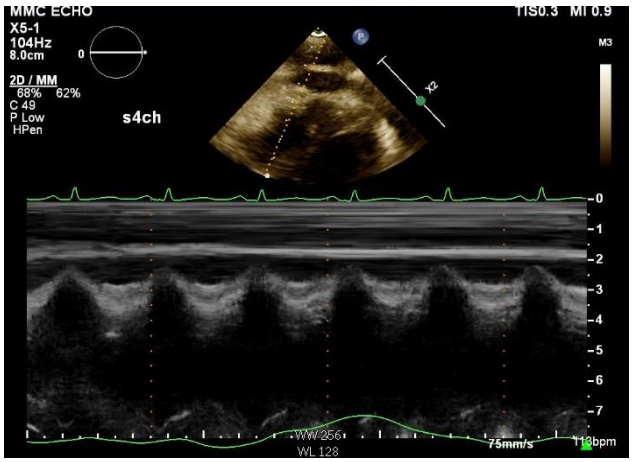


Figure 1. M-Mode right ventricle collapse.

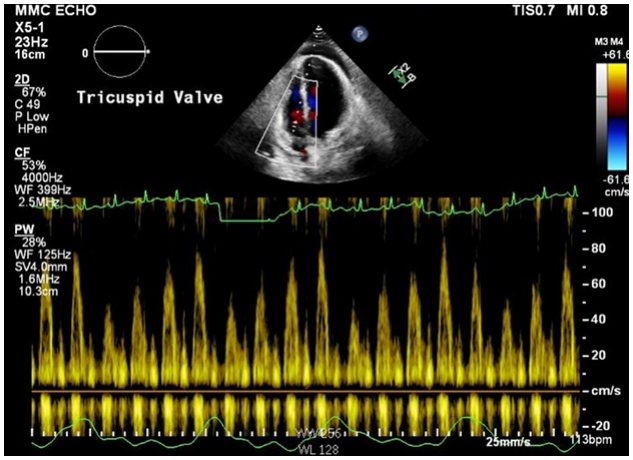


Figure 2. Tricuspid valve respiratory independence.

developed substernal chest pain exacerbated by lying flat. Troponin levels were normal, and an electrocardiogram showed non-specific T-wave changes. Oxygen requirements increased, and a Doppler ultrasound ruled out deep vein thrombosis. Colchicine (0.6 mg daily) was started, and follow-up echocardiography indicated moderate to large effusion with findings suggestive of tamponade.

Pericardiocentesis and right-sided thoracentesis were performed, draining 25 ml of bloody pericardial fluid and 775 ml of clear pleural fluid. Post-pericardiocentesis echocardiography revealed a thickened pericardium indicative of constrictive pericarditis. Fluid analysis is summarised in *Table 1*. The patient was treated with IV furosemide, resulting in significant improvement in respiratory status and anasarca. Right heart catheterisation confirmed constrictive pericarditis with elevated pressures (right atrium pressure 18 mmHg, wedge mean 22 mmHg, pulmonary artery means 33 mmHg, right ventricular end diastolic pressure 24 mmHg, left ventricular end-diastolic pressure 24 mmHg).

Over the following days, her condition steadily improved. She transitioned from high-flow oxygen to a 6 litres nasal cannula and later to room air. Continued diuresis with furosemide alleviated her swelling. She was discharged home with close follow-up after providing informed consent. *Fig. 1 to 5* show the imaging findings.

DISCUSSION

The median survival age for patients with cystic fibrosis has significantly improved, with mortality rates declining in many countries[6]. These advancements are largely attributed to better treatment options and improved management of complications. Constrictive pericarditis, a type of diastolic heart failure caused by a non-compliant pericardium, is a rare condition primarily associated with cardiac surgeries and tuberculosis[7]. Cystic fibrosis, however, is not recognised as a risk factor for constrictive pericarditis, with only a handful

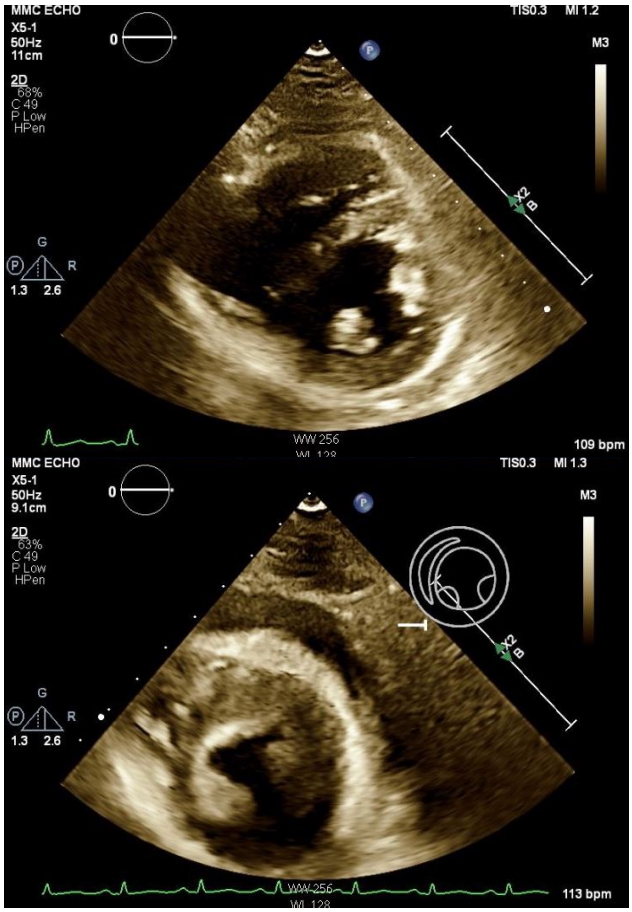


Figure 3. Large pericardial effusion pre-drainage (bottom) and post-drainage ECHO findings (top).

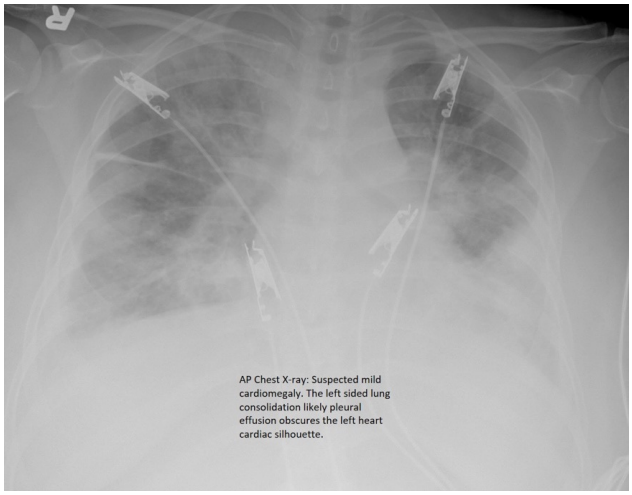


Figure 4. CXR (AP view) showing mild cardiomegaly and a left-sided pleural effusion obscuring the cardiac silhouette.

Reference	Age in years (gender)	Presentation	Diagnostic tool(s)	Management
Brown et al.[8]	31 (M)	The patient reported fever, chills, fatigue, shortness of breath and cough, along with increasing abdominal girth, new bilateral lower extremity swelling and a 20-pound weight gain.	Echocardiogram, chest MRI	Oral colchicine, loop diuretics
McCartney et al.[9]	45 (M)	The patient presented with a cough producing scant mucus, dyspnoea on exertion and new-onset peripheral oedema.	CXR, chest CT, echocardiogram, heart catheterisation	
Betancourt et al.[10]	21 (F)	The patient exhibited progressive malnutrition with a weight of 39 kg, poor appetite, fatigue, malaise, fever, mild dyspnoea and minimal sputum production.	CXR, chest CT, echocardiogram	
O'Grady et al.[11]	34 (M)	The patient reported shortness of breath, minor haemoptysis, loose bowel motions, recurrent chest infections and a one-stone weight loss over the preceding six months.	CXR, echocardiogram	
Zareh et al.[13]	24 (F)	Positional chest pain, dyspnoea on exertion, low-grade fevers and anorexia for 10 days.	CT scan, transthoracic echocardiogram, cardiac catheterisation, cardiac MRI and delayed gadolinium imaging	Ibuprofen, colchicine, pericardiocentesis, partial pericardiectomy
Gregory et al.[12]	24 (F)	Chest pain exacerbated by lying flat, shortness of breath, tachycardia, tachypnoea, low oxygen saturation and facial swelling.	CTA chest, echocardiogram, cardiac catheterisation	NSAIDs, colchicine, steroids, diuretics, pericardiocentesis and antimicrobial therapy based on culture results

Table 2. A summary of all published cases describing constructive pericarditis as a complication of cystic fibrosis.

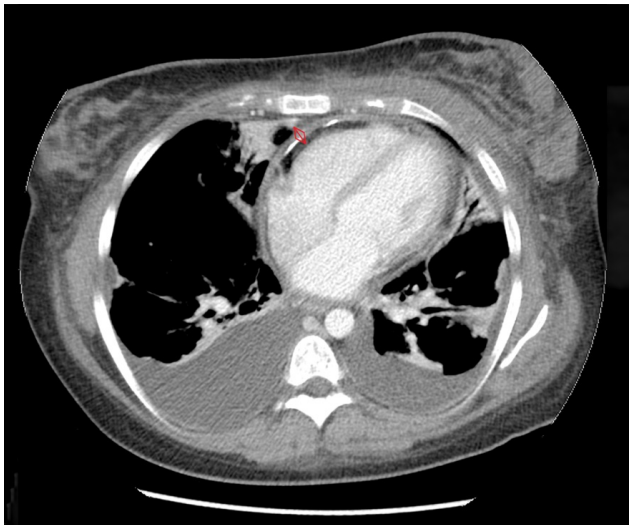


Figure 5. Moderate-sized pericardial effusion accompanied by calcifications in the pericardium; moderate-sized bilateral pleural effusion.

of cases documented in the literature (*Table 2*)[8-13].

The pathophysiology of constrictive pericarditis involves pericardial inflammation leading to fibrosis and calcification, which restricts diastolic filling. In cystic fibrosis patients, chronic pulmonary infections and systemic inflammation may contribute to pericardial involvement, although the exact mechanisms remain unclear. Our case of a 24-year-old female cystic fibrosis patient adds to the growing body of evidence suggesting that constrictive pericarditis, while rare, should be considered a potential complication in cystic fibrosis patients presenting with pulmonary exacerbations or unexplained systemic symptoms.

In our patient, the first indication of constrictive pericarditis was the presence of pericardial effusion and calcification detected on chest CT, performed to assess her pulmonary status. Later during hospitalisation, she developed chest pain exacerbated by lying flat. Pericardiocentesis was employed to differentiate exudative from transudative pericardial effusion, revealing a transudative and sterile fluid. Cardiac catheterisation confirmed the diagnosis and assessed disease severity.

Management strategies for constrictive pericarditis in cystic fibrosis patients remain limited due to the rarity of this condition. Drainage of pericardial effusion through pericardiocentesis or surgery can be considered on a

case-by-case basis. Our patient demonstrated significant improvement with colchicine and diuretics, a therapeutic approach consistent with previously reported cases. However, definitive treatment in refractory cases may require pericardiectomy.

Given the rarity of constrictive pericarditis in cystic fibrosis patients, there are no established screening guidelines. However, clinicians should maintain a high index of suspicion for constrictive pericarditis in cystic fibrosis patients presenting with unexplained dyspnoea, oedema or signs of right-sided heart failure. Early recognition and appropriate management, including consideration of advanced imaging techniques and multidisciplinary care, are crucial for improving outcomes.

Further studies are warranted to explore the prevalence and pathophysiology of constrictive pericarditis in cystic fibrosis patients. Prospective research could help identify risk factors and inform guidelines for screening and management. Additionally, investigating the role of anti-inflammatory therapies in preventing pericardial complications in cystic fibrosis may offer new avenues for treatment.

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