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Idiopathic Constrictive Pericardial Disease in a Patient with Cystic Fibrosis

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Abstract Introduction We report on a case of a patient with cystic fibrosis and co-existent constrictive pericardial disease. This unique presentation has been presented only once before in the literature. Our case is the first documented case where a patient with cystic fibrosis has undergone curative pericardial stripping. Case The case is about a patient who was diagnosed with cystic fibrosis at the age of 45. His sweat chloride level was 77.0mmol/L and genotype heterozygous for RII7H and delta F508. His FEV1 was 75% predicted at the time of admission. The patient was hospitalized for a presumed pulmonary exacerbation of his cystic fibrosis. He presented with cough with scant mucous production and no hemoptysis. He also noted dyspnea on exertion and new onset peripheral edema. He was treated for a pulmonary exacerbation with intravenous antibiotics and airway clearance measures and discharged home after two days. He worsened despite optimal therapy for a pulmonary exacerbation. A simultaneous right and left heart catheterization was performed that was suggestive of constrictive pericardial disease. The patient underwent a pericardial stripping that resolved his symptoms. He was discharged home after surgery and has been doing well. Discussion Constrictive pericardial disease is an extremely rare complication that is not frequently seen in cystic fibrosis. Simultaneous left and right heart catheterization can be used to make the diagnosis. Surgery is curative. Although pulmonary exacerbation is the most common cause of shortness of breath in patients with cystic fibrosis other diagnosis should be considered.

Keywords: *cystic fibrosis, constrictive pericarditis, calcifications, pericardial stripping*

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1. Introduction

Constrictive pericardial disease leads to a reduction in pericardial compliance and is most often caused by radiation, infection or trauma [1]. Patients often present with peripheral edema and shortness of breath, and are frequently misdiagnosed with liver disease. The recognition of constrictive pericardial disease is important, as it is potentially curable surgically with pericardial stripping [2]. Simultaneous right and left heart catheterization demonstrating discordance [1,3] of left and right ventricular pressures with inspiration is diagnostic.

Constrictive pericardial disease is extremely rare in patients with cystic fibrosis. There is one previously reported case describing constrictive pericarditis in a patient with cystic fibrosis [4]. Our case is the first reported where a patient with cystic fibrosis underwent pericardial stripping.

2. Case

The patient was diagnosed with cystic fibrosis at the age of 45 after self-referring to an adult cystic fibrosis clinic for repeated prolonged episodes of shortness of breath over the previous four winters. He had been previously diagnosed with walking pneumonia and bronchitis and had been treated with various courses of steroids and antibiotics with minimal relief.

His sweat chloride level was 77.0mmol/L and genotype heterozygous for RII7H and delta F508. His FEV1 was 75% predicted at the time of admission. The patient has congenital absence of vas deferens and type I diabetes without exocrine pancreatic insufficiency. He also has chronic kidney disease.

The patient was hospitalized for treatment of a pulmonary exacerbation. He presented with cough with scant mucous production and no hemoptysis. He also noted dyspnea on exertion and new onset peripheral edema.

He was treated for a pulmonary exacerbation with intravenous antibiotics and airway clearance measures. After two days he was discharged home. Five days later he was readmitted for worsening shortness of breath. A chest CT scan (Figure 1) at that time showed ground glass opacities, pleural effusions and unchanged pericardial calcifications from previous imaging (Figure 2). The patient was again treated for a pulmonary exacerbation.

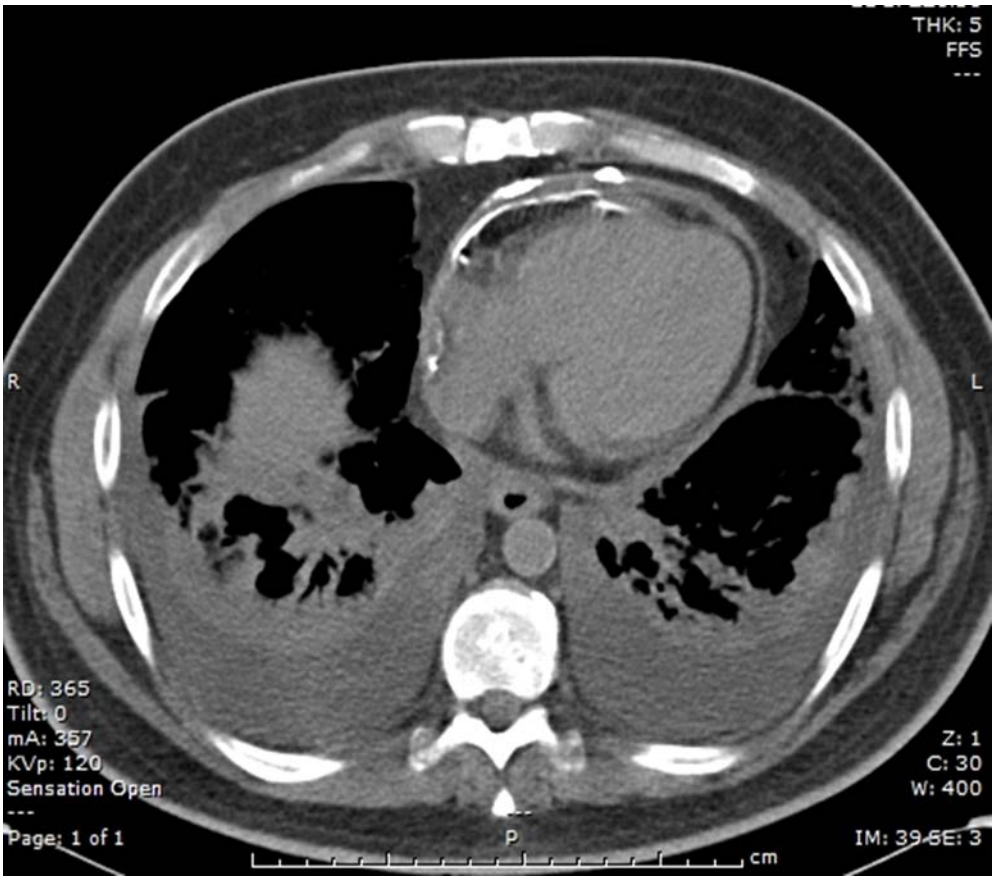

Figure 1. Non-Contrast of the chest on day 3 of patient's second admission. Calcification of the pericardium as well as bilateral pleural effusions are noted. These calcifications had been seen greater than two years previously on CT of the chest and were noted to be unchanged by radiology. The pleural effusions were new

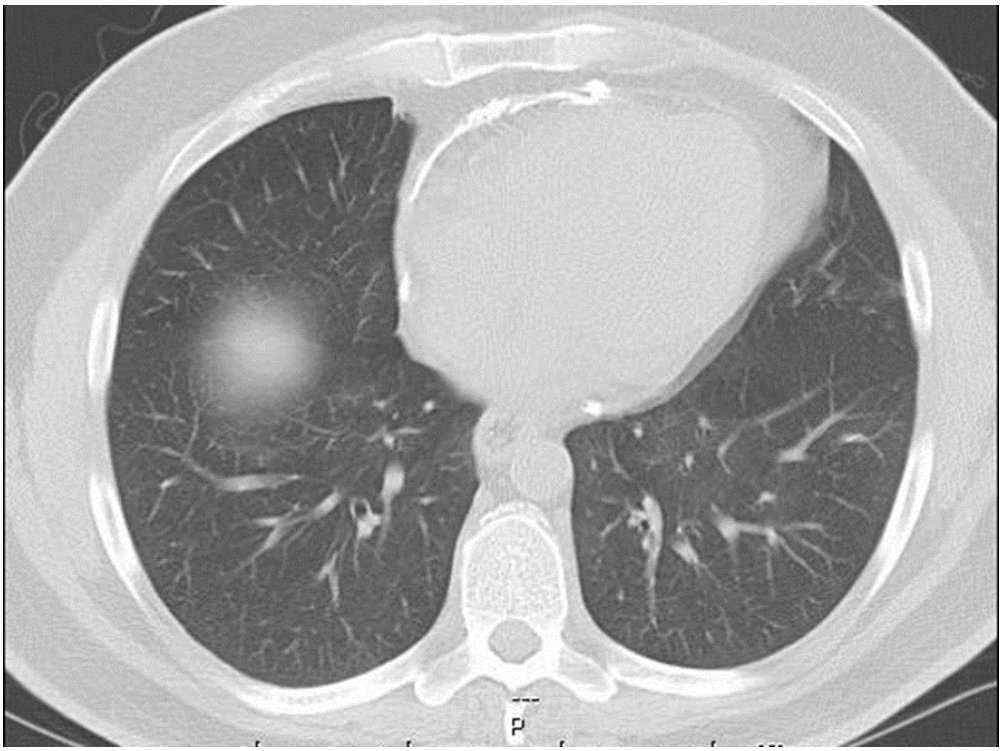

Figure 2. Non contrast CT scan of the chest obtained 10 months prior to admission. Note the presence of pericardial calcifications. The radiologist noted that these had been stable and unchanged from previous imaging two years earlier

The patient underwent bronchoscopy to evaluate for an infectious etiology. Bronchoscopy revealed no mucous plugging and no evidence of infection. The opinion of infectious disease consultant was that the patient's overall clinical picture was likely due to non-infectious etiology.

Volume overload was managed with diuretics and cardiology was consulted. The patient's echocardiogram

showed an ejection fraction of 55% with diastolic function abnormal for his age, without evidence of pulmonary hypertension. The consultant cardiologist suggested that the patient's worsening fluid overload was noncardiogenic in nature and likely due to worsening renal function. The management of volume overload was challenging as his creatinine began to rise in the setting of intravenous diuretic therapy.

The patient slowly deteriorated over three weeks on intravenous antibiotics, airway clearance measures and diuretics. Glycemic control was difficult. His creatinine rose steadily with intravenous diuretics. He was transferred to the intensive care unit for worsening respiratory status but did not require intubation. Intermittent hemodialysis was initiated due to fluid overload in the setting of acute renal failure.

In the fifth week of his hospitalization, cardiology was reconsulted with the specific request of a simultaneous right and left heart catheterization to evaluate for constrictive pericardial disease. The catheterization demonstrated rapid diastolic filling and a relative equalization of the right atrial, right ventricular end diastolic, pulmonary arterial wedge pressure and left ventricular end diastolic pressures, consistent with constrictive physiology (Figure 3a-3d). However, there was a concordance of pressures on simultaneous left and right ventricular tracings, which is more consistent with restrictive physiology (Figure 3).

The presence of pericardial calcifications and findings on catheterization consistent with constrictive pericardial disease, the decision was made to refer the patient to cardiothoracic surgery for pericardial stripping. Investigations for potential causes of constrictive pericardial disease including tuberculosis, fungal and rheumatologic conditions were negative. The patient had no history of trauma, surgery or radiation to the chest.

Following pericardial stripping, surgical pathology demonstrated calcifications but no further diagnostic data was obtained. The patient improved after surgery and was discharged home one week postoperatively. The patient continued to improve as an outpatient and three months after discharge was able to stop dialysis as his kidney function returned. The patient continues to do well two years following presentation.

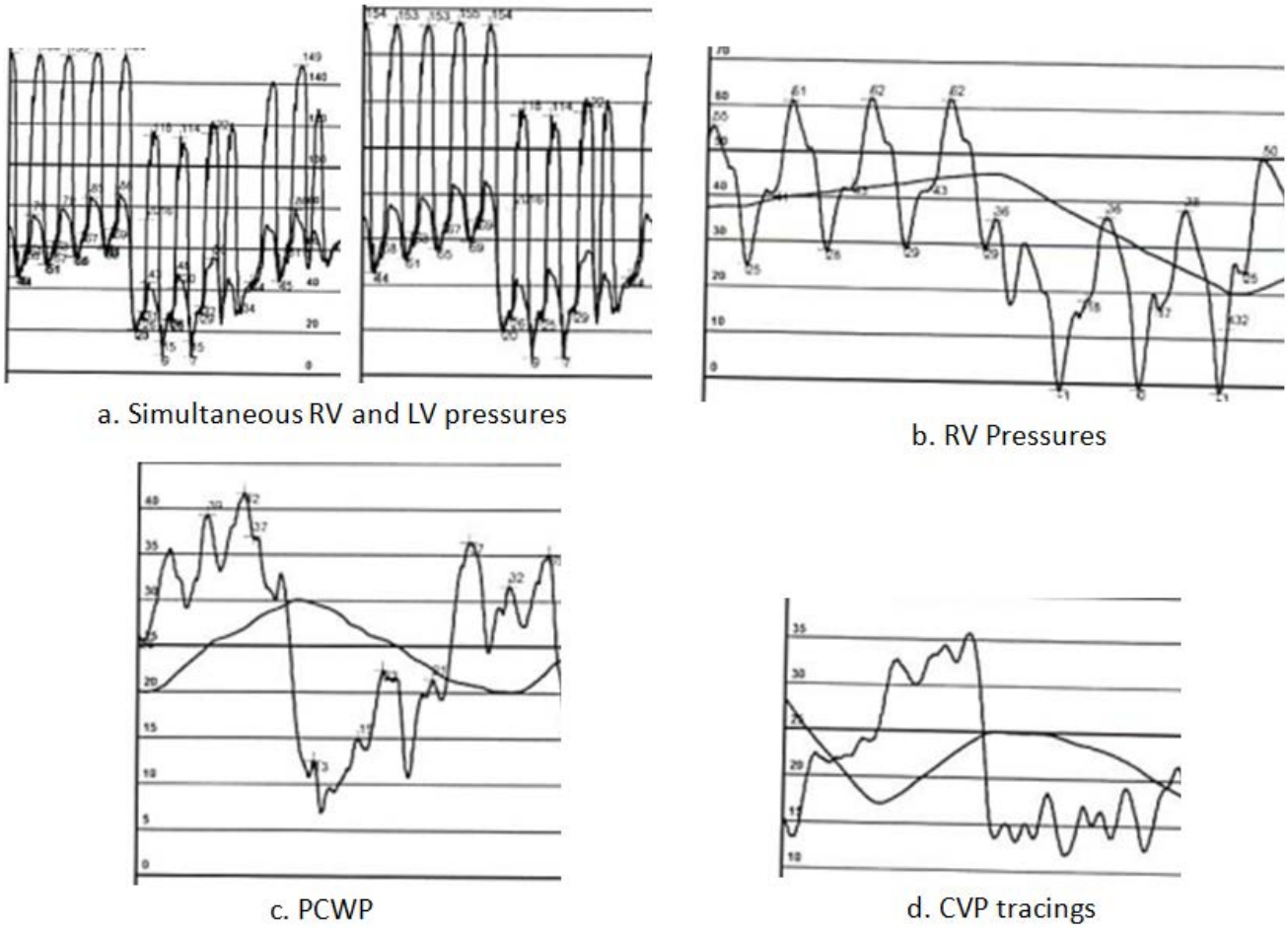

Figure 3.

3. Discussion

Though pulmonary exacerbation is a common cause of dyspnea in patients with cystic fibrosis, it is important to maintain a broad differential and consider other causes particularly when the patient is not improving despite optimal therapy.

Anchoring, a type of cognitive bias [4], led to the delayed diagnosis of constrictive pericarditis in this patient. His shortness of breath made his physicians suspect pulmonary exacerbation. Evidence that did not support that diagnosis, including pericardial calcifications, pleural effusions and peripheral edema, were overlooked initially.

The diagnosis of constrictive pericardial disease was supported with catheterization, although discordance of pressures was not seen. Other features of constrictive pericardial disease were present including a relative equalization of right atrial, right ventricular end diastolic, pulmonary arterial wedge pressure and left ventricular end diastolic pressures. There was also rapid diastolic filling with a leveling off of pressures often referred to as a square root sign.

Constrictive pericardial disease is extremely rare in patients with cystic fibrosis, there is only one such reported case in the literature[5]. Recognizing constrictive pericardial disease in this patient was crucial as he was cured surgically.

Prevalence of constrictive pericardial disease in cystic fibrosis patients is not known. It is our hope that this case sheds light on an extremely rare condition.

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Conflict of Interest

There is no conflict of interest for either authors Dr. Ravi Nayak nor Dr. Conor McCartney

This article does not contain any studies with either human or animal participants.

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