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Patient: Male, 32-year-old
Final Diagnosis: Post pericardiectomy
Symptoms: Acute renal failure • flu like symptoms • hypertension
Clinical Procedure: Pericardial drainage • pericardial window • pericardiectomy • pericardiocentesis

Objective: Rare coexistence of disease or pathology
Background: Effusive-constrictive pericarditis (ECP) is an uncommon clinical syndrome characterized by the coexistence of pericardial effusion and constriction involving the visceral pericardium. This differs from constrictive pericarditis, which presents with thickening of the pericardium without effusions. Specific diagnostic criteria of ECP include the failure of right atrial pressure to decrease by 50% or reach a new level below 10 mmHg after normalization of intrapericardial pressure.

Case Report: We present the case of a 32-year-old obese man with multiple comorbidities who initially presented with flu-like symptoms and pleural effusion with development of constrictive-like symptoms. Despite undergoing numerous pericardiocentesis and appropriate medical management, the patient’s condition failed to improve, leading to the likely diagnosis of effusive-constrictive pericarditis. Cultures of pericardial fluid revealed E. faecium, which required multiple antimicrobial therapy. Despite infection, the exact etiology of ECP remained unknown and likely idiopathic. Common causes of ECP include idiopathic, tuberculosis, cardiac surgery complications, radiation, or neoplasia. Ultimately, the patient underwent a pericardiectomy involving the visceral and parietal pericardium, resulting in hemodynamic stability and resolution of symptoms.

Conclusions: This case highlights the challenges in diagnosing and managing ECP, emphasizing the importance of considering surgical intervention in refractory cases. ECP initially presents as a pericardial effusion, often addressed through pericardiocentesis; however, in a small subset of patients, sustained symptoms and altered hemodynamics persist following pericardiocentesis, necessitating further evaluation and management. The success of pericardiectomy in our patient highlights the potential efficacy of surgical intervention in improving outcomes for patients with ECP.

Keywords: Heart Diseases • Thoracic Surgery

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/943979
Introduction

Effusive-constrictive pericarditis (ECP) is a rare clinical syndrome characterized by the simultaneous presence of pericardial effusion and constriction involving the visceral pericardium. This differs from typical constrictive pericarditis, which features thickening and contraction of the parietal pericardium without effusion. ECP is associated with persistent elevated right atrial pressure after intrapericardial pressure has been reduced by removal of pericardial fluid [1]. The prevalence of this rare hemodynamic syndrome is relatively low; data in the New England Journal of Medicine shows the prevalence of ECP in patients with pericardial disease was 1.3%, with a prevalence of 6.9% in patients with tamponade undergoing catheterization [1]. Overall, there have been few reported cases of ECP, likely due to the rarity of the condition along with the difficulty of establishing the diagnosis [1]. Diagnostic criteria for this condition require precise hemodynamic measurements during pericardiocentesis with simultaneous cardiac catheterization; specifically, intrapericardial and right atria transmural pressures [1]. It is important to recognizing the contribution of the visceral pericardial layer to the pathogenesis of constriction and the implications of surgical removal [1].

Case Report

A 32-year-old man with morbid obesity (BMI 53 kg/m²), intellectual disability, and a history of hypertension presented with flu-like symptoms, anasarca, and acute renal failure. At an outside medical facility, his renal failure was suspected to be due to hemolytic uremic syndrome and was managed with intermittent acute dialysis. A transthoracic echocardiogram revealed a pericardial effusion, leading to subxiphoid tube pericardiostomy. The patient received pulse steroids, due to the possibility of idiopathic origin; however, on initial presentation, the patient presented with flu-like symptoms with 2 positive COVID antibody tests and no history of COVID vaccination, making viral constrictive physiology a possible cause. The cultured bacteria within the pericardium were unlikely to be the primary causative agent, as analysis was not compatible with pyogenic pericarditis, which was indicated in our patient with 900 mL of bloody fluid. Bacterial cultures could have been introduced through interventions such as pericardiocentesis and pericardial window at an outside hospital. Renal stabilization occurred without further dialysis, but recurrent pericardial effusion necessitated repeated interventions due to persistent elevated pressures (Table 1).

Pericardial fluid cultures revealed *E. faecium* during the second pericardiocentesis, followed by *Bacteroides* q week later upon additional pericardiocentesis with drain. The Infectious Diseases Department became involved in the case at this time for antibiotic recommendations, which throughout the hospital course included antimicrobial therapy of Cefepime, Flagyl, Unasym, and Augmentin. The patient exhibited deteriorating renal function, cool extremities, and elevated lactate levels, raising concerns about cardiogenic shock attributed to effusive-constrictive pericarditis. Aggressive diuresis and dobutamine, with persistent filling pressures despite medical management.

The exact etiology of the ECP remained unknown, but was likely of idiopathic origin; however, on initial presentation, the patient presented with flu-like symptoms with 2 positive COVID antibody tests and no history of COVID vaccination, making viral constrictive physiology a possible cause. The cultured bacteria within the pericardium were unlikely to be the primary causative agent, as analysis was not compatible with pyogenic pericarditis, which was indicated in our patient with 900 mL of bloody fluid. Bacterial cultures could have been introduced through interventions such as pericardiocentesis and pericardial window. Other routes of bacterial translocation included a fluid collection on CT abdomen with communication to the inferior mediastinum showing fistulation to the abdominal wall. The CT chest revealed pericardial enhancement concerning for infection and left pleural effusion raising suspicion of the abdominal fluid as the route of bacterial translocation to the pericardial space.

Table 1. Post-second pericardiocentesis RHC findings, initial pericardiocentesis performed at an outside hospital. Elevated filling pressures are shown.

<table>
<thead>
<tr>
<th>Pressure</th>
<th>Value</th>
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<tbody>
<tr>
<td>RA</td>
<td>27/24 mmHg (24)</td>
</tr>
<tr>
<td>PA</td>
<td>30/23 mmHg (27)</td>
</tr>
<tr>
<td>PCWP</td>
<td>18 mmHg</td>
</tr>
<tr>
<td>CO (Fick)</td>
<td>5.48 L/min</td>
</tr>
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RA – right atrium; PW – pulmonary wedge; PA – pulmonary artery; CO – cardiac output.

Table 2. RHC findings following aggressive diuresis and dobutamine, with persistent filling pressures despite medical management.

<table>
<thead>
<tr>
<th>Pressure</th>
<th>Value</th>
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<tbody>
<tr>
<td>RA</td>
<td>16 mmHg</td>
</tr>
<tr>
<td>RV</td>
<td>27/10 (13) mmHg</td>
</tr>
<tr>
<td>PA</td>
<td>29/12 (21) mmHg</td>
</tr>
<tr>
<td>PCWP</td>
<td>18 mmHg</td>
</tr>
<tr>
<td>LVEDP</td>
<td>94/5 (23) mmHg</td>
</tr>
<tr>
<td>CO (Fick)</td>
<td>74/57 L/min</td>
</tr>
</tbody>
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RA – right atrium; RV – right ventricle; PA – pulmonary artery; PCWP – pulmonary capillary wedge pressure; LVEDP – left ventricular end-diastolic pressure; CO – cardiac output.
The patient's suitability for cardiac MRI was hindered by morbid obesity and intellectual disability. Faced with clinical deterioration and a strong suspicion of effusive-constrictive pericarditis, a sternotomy was performed. The parietal pericardium, approximately ½ cm thick, exhibited a leathery texture and was separated from the heart surface by ample space. Excision of the parietal pericardium, extending from phrenic nerve to phrenic nerve, was carried out. The visceral pericardium displayed a white, smooth sheen, incised with a 15-blade knife, revealing bulging epicardial heart fat. An anterior plane was meticulously developed, liberating the anterior part of the left ventricle, followed by the right ventricle, right atrium, and some of the left atrium, thereby freeing the heart from the constrictive layer. This surgical approach of freeing the left heart was taken to reduce volume load risks caused by the right heart and reducing the likelihood of causing pulmonary edema; with the left heart being released first, this maintained proper fluid status within cardiac chambers.

Intraoperatively, the central venous pressure (CVP) dropped from 22 to 15, and the left ventricular cavity size in diastole, measured by transesophageal echocardiography (TEE), increased from 42 cc to 62 cc after pericardiectomy. Systolic function remained preserved. Mediastinal and bilateral pleural drains were strategically placed, and the sternotomy was routinely closed. In the Intensive Care Unit (ICU), the patient's cardiac index reached 3.3 liters/min/m\(^2\) BSA, accompanied by normal pulmonary artery pressures and a CVP of 8. Recovery proved gradual due to persistent renal disease necessitating intermittent dialysis and a compromised functional baseline, leading to tracheostomy for ventilator weaning. Nevertheless, the patient was ultimately discharged home, free from dialysis. Figure 1 provides a complete hospital course timeline.

**Discussion**

The uncommon disease process of effusive-constrictive pericarditis involves constriction of the visceral pericardium and effusion, causing a tamponade-like effect on the heart [2]. Conventional constriction manifests with physical examination findings such as jugular venous distention (JVD), Beck's triad, Kussmaul sign, pericardial knock, and edema [3]. ECG tracings in constrictive pericarditis exhibit reduced voltages. Due to ventricular interdependence within a rigid pericardium, there is an observed increase in right ventricular volume relative to the left ventricle on echocardiography, leading to septal bounce [3]. Echocardiogram findings may reveal a restrictive left ventricular filling pattern. Cardiac catheterization in constrictive pericarditis exhibits increased atrial pressure with prominent x and y descents, equalization of end-diastolic pressures, and a dip-and-plateau or square-root sign of ventricular diastolic pressure [3].

The exact etiology of ECP is often idiopathic, but other possible etiologies include radiation, tuberculous, cardiac surgery complications, neoplasia, and bacterial or fungal infections [1]. The underlying etiologies of ECP are similar to those of constrictive pericardial disease [1]. In developed countries, common causes include idiopathic, viral, post-procedural, radiation, drug-induced, connective tissue disease, post-myocardial infarction, malignancy, trauma, and uremia [4]. In our patient, specifically with the treatment resistant bacteria, bacterial-related inflammation was a possible cause of the initial insult, but the presence of *E. faecium* led to a low suspicion of this, as it is unlikely to cause ECP, and the ECP drainage was more supportive of pyogenic pericarditis, which is unlikely with *E. faecium*. 
Unlike the conventional approach of treating typical pericardial constriction with pericardiectomy once the diagnosis is confirmed, ECP often responds favorably to as-needed (prn) pericardial drainage and anti-inflammatory medications. In a study at the Mayo Clinic examining ECP after pericardiocentesis, only 33 out of 205 patients undergoing pericardiocentesis were diagnosed with ECP, and only 2 patients required pericardiectomy due to persistent constrictive features [2].

Effusive-constrictive pericarditis initially presents as a pericardial effusion, addressed through pericardiocentesis [5]. However, in a subset of patients, sustained symptoms and altered hemodynamics persist after pericardiocentesis, attributed to visceral constrictive pericarditis, leading to the designation of this condition as effusive-constrictive pericarditis.

Accurate diagnosis of effusive-constrictive pericarditis is achieved through simultaneous measurements of intrapericardial and right atrial pressures during pericardiocentesis [6]. It is defined by persistent elevation in right atrial, end-diastolic right ventricular, and left ventricular diastolic pressures, even after intrapericardial pressure reduction through pericardiocentesis [6]. The initial hemodynamic presentation mimics cardiac tamponade, but despite fluid removal, hemodynamic abnormalities manifest as constriction, impacting cardiac function during diastole [6].

While both cardiac tamponade and constrictive pericarditis restrict heart filling, raising systemic and pulmonary venous pressures, the venous pressure waveforms differ between these conditions [7]. In effusive-constrictive pericarditis, the constricting visceral pericardial layer and the overlying pericardial effusion simultaneously contribute to reduced myocardial transmural pressure and filling [6]. This syndrome is considered an end-stage manifestation of persistent pericardial inflammation, characterized by the loss of pericardial elasticity through fibrosis and dystrophic calcification [6].

Initial work-up to evaluate constrictive pericarditis includes echocardiography, as it has excellent sensitivity and specificity for identifying effusions [4]. Constrictive pericarditis on echocardiogram will likely show increased pericardial thickness.

Figure 2. Partial resection of the visceral pericardium; ½ cm leathery texture separated from heart. Arrow identifies the constrictive layer at the start of retraction.

Figure 3. Complete resection of visceral pericardium, with the heart freed from the constrictive layer. The arrow identifies the heart freed from the constrictive layer, with retraction of the constrictive layer by forceps.
with or without calcification [4]. Specific to constrictive pericarditis, a rapid deceleration during diastolic filling, as well as a septal bounce, are specific signs of constriction and ventricular interdependence [4]. In contrast, diagnostic criteria for ECP include the failure of right atrial pressure to decrease by 50% or reach a new level below 10 mmHg after normalization of intrapericardial pressure [6]. Patients with ECP may exhibit symptoms resembling right-sided heart failure and volume overload due to limited intracardiac end-diastolic volume [7].

The initial therapeutic approach emphasizes pericardial fluid removal through pericardiocentesis, but in a subset of cases, fluid re-accumulation may necessitate repeated procedures [6]. Treatment options range from medication management to pericardiectomy, with definitive resolution achieved through the latter, involving the release of the visceral pericardium [7]. Specific treatment approaches depend on the multimodal disease course; with the rarity of ECP, this diagnosis should arise after efforts to drain pericardial effusions have been exhausted. Studies have cited ECP being managed medically due to poor prognosis or high surgical risks [1]. Each patient would benefit from an individualized approach; specifically, our patient was not an ideal surgical candidate due to morbid obesity and severe intellectual disability, but non-surgical options failed, including repeated pericardiocentesis, pericardial windows, diuresis, and dobutamine. Overall, our patient exhibited remarkable hemodynamic improvement following sequential parietal and visceral pericardiectomy.

It is crucial to emphasize that in any pericardiectomy procedure for constriction, the left ventricle should be liberated first, followed by the right ventricle. This sequential approach prevents the liberated right ventricle from potentially overloading the left ventricle, mitigating the risk of intraoperative pulmonary edema. The enhancement in diastolic capacitance was demonstrated through intraoperative transesophageal echocardiography (TEE), revealing a substantial increase in left ventricular diastolic volumes from 42 cc before to 62 cc after pericardiectomy. The heart dynamically emerged from the visceral pericardium; this impactful transformation is visually depicted in Figures 2 and 3.

Conclusions
This case highlights the challenges in diagnosing and managing ECP, with particular focus on surgical intervention in refractory cases. While ECP presents as pericardial effusion, patients with ECP will sustain symptoms and altered hemodynamics after treatment with pericardiocentesis. The success of pericardiectomy can highlight the potential efficacy of surgical intervention in improving outcomes for patients with ECP.

Statement
Patient was seen and managed by University of Texas Medical Branch, Department of Cardiovascular and Thoracic Surgery.

The Institutional Review Board (IRB) waived the need for written consent on 3 April 2023.

Declaration of Figures’ Authenticity
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References: