

Diagnosis and treatment of effusive–constrictive pericarditis: a case report

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Received 6 October 2020; first decision 22 October 2020; accepted 16 April 2021

Background

Effusive–constrictive pericarditis (ECP) is a rare syndrome involving pericardial effusion and concomitant constrictive pericarditis. The hallmark is a persistently elevated right atrial pressure of >10 mmHg or reduction of less than 50% from baseline despite pericardiocentesis. Aetiologies include radiation, infection, malignancy, and autoimmune disease.

Case summary

A 71-year-old man with a history of atrial fibrillation, obesity, hypertension, obstructive sleep apnoea, managed with continuous positive airway pressure presented with acute pericarditis complicated by pericardial effusion leading to cardiac tamponade. He was diagnosed with ECP after pericardiocentesis and was managed surgically with a pericardial window.

Discussion

Early detected cases of ECP can be managed by medical therapy. Therapeutic interventions include pericardiocentesis, balloon pericardiostomy, and pericardiectomy. This report describes a case of new-onset congestive heart failure secondary to ECP.

Keywords

Constrictive • Pericarditis • Cardiac tamponade • Case report

Learning points

- How to make the diagnosis of effusive–constrictive pericarditis (ECP) using history, physical exam, imaging [echocardiogram, magnetic resonance imaging (MRI)], and invasive haemodynamics.
- ECP can be caused by any disease that can lead to pericardial inflammation, including infection, autoimmune disease, or irradiation.
- Cardiac MRI imaging can be utilized to further characterize the pericardium for features of hemodynamic compromise, pericardial inflammation, fibrosis or myocarditis, and to help guiding the treatment approach.

Introduction

Effusive–constrictive pericarditis (ECP) was first described in 1954. The prevalence of ECP varies from 4.5% to 6.9% of patients who present with pericardial effusion.^{1–3} It is a rare syndrome characterized

by the constriction of the heart by the visceral pericardium, with fluid accumulation in the pericardial space causing cardiac tamponade. The syndrome can be caused by any disease with pericardial involvement, including infection, autoimmune disease, or irradiation. Subacute cases present with symptoms of right ventricular failure, such as

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Handling Editor: Konstantinos Stathogiannis

Peer-reviewers: Angel Luis Fernandez Gonzalez; Domenico Filomena

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jugular venous distention (JVD), hepatic congestion, and lower extremity oedema, which is the result of diastolic dysfunction related to pericardial restriction. Acute development of pericardial effusion will present as cardiac tamponade with haemodynamic instability. ECP is often identified when haemodynamic features consistent with constrictive pericarditis persisting after the drainage of pericardial fluid. In particular, a persistently elevated right atrial (RA) pressure of above 10 mmHg or a decline by less than 50% after pericardiocentesis is diagnostic of ECP.³

cava was dilated to 21 mm and decreased less than 50% in size with inspiration (Figure 2). Gentle IV fluid resuscitation was initiated. Right heart catheterization (RHC) showed a decreased cardiac output of 3.7 L/min and a cardiac index of 1.4 L/min/m² with a RA mean pressure of 34 mmHg (Figure 3). A pericardiocentesis in the catheterization laboratory was unsuccessful due to obese body habitus. The patient was subsequently taken to the OR for a subxiphoid pericardiocentesis was performed by cardiothoracic surgery, with drainage of 400 mL of serosanguinous fluid. The pericardial fluid analysis showed no evidence of malignant cells but abundant reactive mesothelial cells. Obtained blood and pericardial fluid cultures remained negative. Cardiac magnetic resonance imaging (cMRI) showed bi-atrial enlargement

Timeline

Day 0	A 71-year-old man with a history of atrial fibrillation, obesity, hypertension, obstructive sleep apnoea managed with continuous positive airway pressure presents with acute cardiac tamponade.
Day 0	Patient is taken to the catheterization laboratory for pericardiocentesis which was unsuccessful due to obese body habitus. Right heart haemodynamic tracings from the attempt are seen in Figure 3A.
Day 1	Patient underwent pericardiocentesis with cardiothoracic surgery.
Day 5	Patients symptoms improved and he was discharged on Colchicine.
Day 12	The patient presented to the emergency department 1 week after discharge with worsening dyspnoea on exertion and was readmitted. A repeat transthoracic echocardiogram revealed a recurrent moderate-sized pericardial effusion.
Day 13	He underwent a repeat pericardiocentesis with a pericardial window procedure, which led to a rapid improvement in the patient's symptoms.
Day 15	Right heart catheterization after pericardial window procedure, demonstrated a persistently elevated mean right atrial (RA) pressure of 13 mmHg, with a rapid y descent.
Day 23	Patient was discharged home in stable condition.
1-month post-discharge	A follow-up echocardiogram showed an estimated RA of 20 mmHg, a left ventricular septal E' velocity of 14 cm/s and a restrictive filling pattern of the left ventricle with an E/A ratio of 1.51 in absence of a pericardial effusion (Figure 5A–D).
Three-month post-discharge	At the 3-month follow-up, the patient remained asymptomatic.

Case presentation

A 71-year-old man with a history of atrial fibrillation; obesity; hypertension; obstructive sleep apnoea, managed with continuous positive airway pressure; presented with 5 days of progressive chest pain, hypoxia, and exertional dyspnoea. Vitals on admission were notable for a SpO₂ of 85% on room air, blood pressure at 104/81 mmHg, and a heart rate of 95 beats per minute. There is a pulsus paradoxus noted with a systolic blood pressure decrease upon inspiration of 20 mmHg. Cardiac auscultation revealed muffled heart sounds with a normal S1 and S2. JVD was noted that increased during inspiration (Kussmaul's sign). Further physical examination is pertinent for lungs that are clear to auscultation and 2+ bilateral lower extremity pitting oedema. Complete blood count was pertinent for a mild anaemia with a haemoglobin of 13.3 g/dL. Inflammatory markers were elevated including a C-reactive protein of 11.8 mg/dL (reference range <0.80 mg/dL) and a sedimentation rate of 37 mm/h (reference range 0–20 mm/h). An electrocardiogram showed sinus tachycardia with low voltage (Figure 1). A transthoracic echocardiogram (TTE) confirmed a large circumferential pericardial effusion, with mild right ventricular collapse during diastole. The inferior vena

and a tubular configuration of the ventricles, suggestive of pericardial constriction. On the myocardial tagging sequences, grid-lines persisted between myocardium and pericardium, indicating pericardial adhesions. Real-time cine cardiac motion imaging revealed a septal bounce on deep inspiration, suggesting increased ventricular coupling, which supported to a diagnosis of constrictive pericarditis (Figure 4A–D). Workup for autoimmune disease was negative. The patient recovered, with no further oxygen requirements, and was discharged on colchicine 0.6 mg twice daily in a stable condition.

The patient presented to the emergency department 1 week after discharge with worsening dyspnoea on exertion and was readmitted. A repeat TTE revealed reaccumulation of a moderate-sized pericardial effusion. Given the respiratory distress, the patient underwent a second pericardiocentesis with a pericardial window procedure, which led to a rapid improvement in the patient's symptoms. A pericardial biopsy obtained during the pericardial window procedure, revealed fibro-adipose tissue with focal chronic inflammation and rare hemosiderin deposits. RHC after pericardiocentesis demonstrated an improved cardiac output of 7.7 L/min, a cardiac index of 2.9 L/min/m², and a persistently elevated mean RA pressure of 13 mmHg, with a rapid y descent.

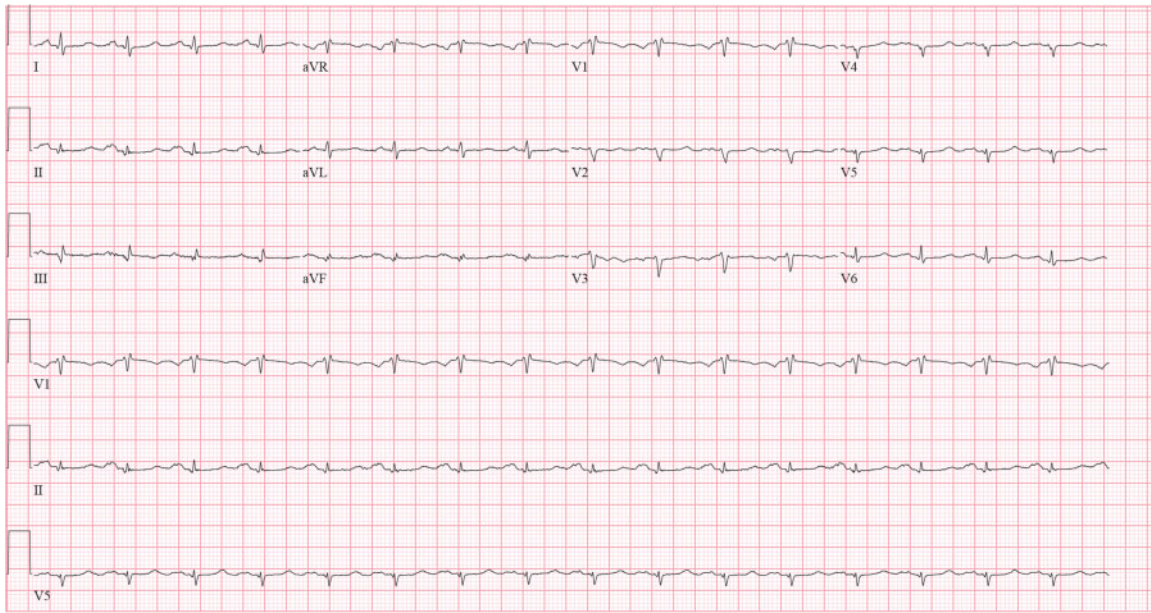


Figure 1 Electrocardiogram on presentation: sinus rhythm, heart rate 98 beats per minute, with low voltage.

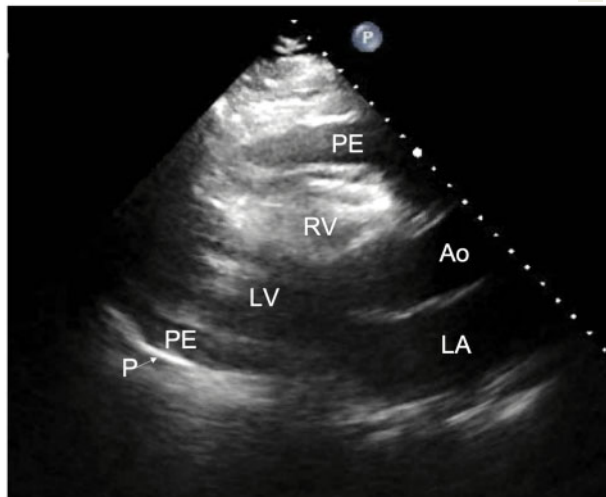


Figure 2 Transthoracic echocardiogram in parasternal long-axis view on presentation showing a large circumferential pericardial effusion with mild right ventricular diastolic collapse suggestive of early cardiac tamponade. LA, left atrium; LV, left ventricle; P, pericardium; PE, pleural effusion; RV, right ventricle.

A follow-up echocardiogram at 1-month post-discharge showed an estimated RA pressure of 20 mmHg, an left ventricular septal E' velocity of 14 cm/s, and a restrictive filling pattern of the left ventricle with an E/A ratio of 1.51 in absence of a pericardial effusion (Figures 3B and 5A–D). At the 3-month follow-up, the patient remained asymptomatic.

Discussion

The underlying aetiologies of ECP are similar to those of effusive and constrictive pericardial disease and include radiation, infection, malignancy, and autoimmune processes. Historically, tuberculosis was one of the most common causes of ECP but the prevalence has decreased through increased screening and treatment. Consequently, radiation therapy for malignancy has now become the most common aetiology of ECP.^{1,3}

The diagnosis of ECP requires a high index of clinical suspicion. A prospective study by Sagristà-Sauleda *et al.*³ evaluated typical clinical findings in patients with ECP. Initial symptoms typically developed within 3 months of presentation. Approximately half of patients had chest pain and fever as the initial prodrome. All patients had signs of right ventricular failure, including JVD and hepatic congestion. Pulsus paradoxus, moreover, was observed in 10 out of 15 patients.³ While all these findings are indicative of CP, they can also occur in cardiac tamponade. TTE findings such as ventricular interdependence and elevated RA pressure can aid when diagnosing constrictive pericardial disease. An increased lateral to medial mitral annulus velocity with decreased movement of the lateral annulus is also diagnostically helpful. It indicates inhibited movement of the lateral ventricular wall due to pericardial constriction. This TTE finding is not seen in restrictive pericardial disease.⁴ cMRI, similarly, can demonstrate the septal bounce, reduced pericardial motion and pericardial thickening. However, T2-weighted cMRI imaging is able to further delineate the pericardial thickening and to distinguish between oedema, inflammation, and fibrosis. These findings can have a direct clinical impact. Conservative medical treatment can be trialed first in active inflammation with oedema prior to surgical treatment.⁵ cMRI can

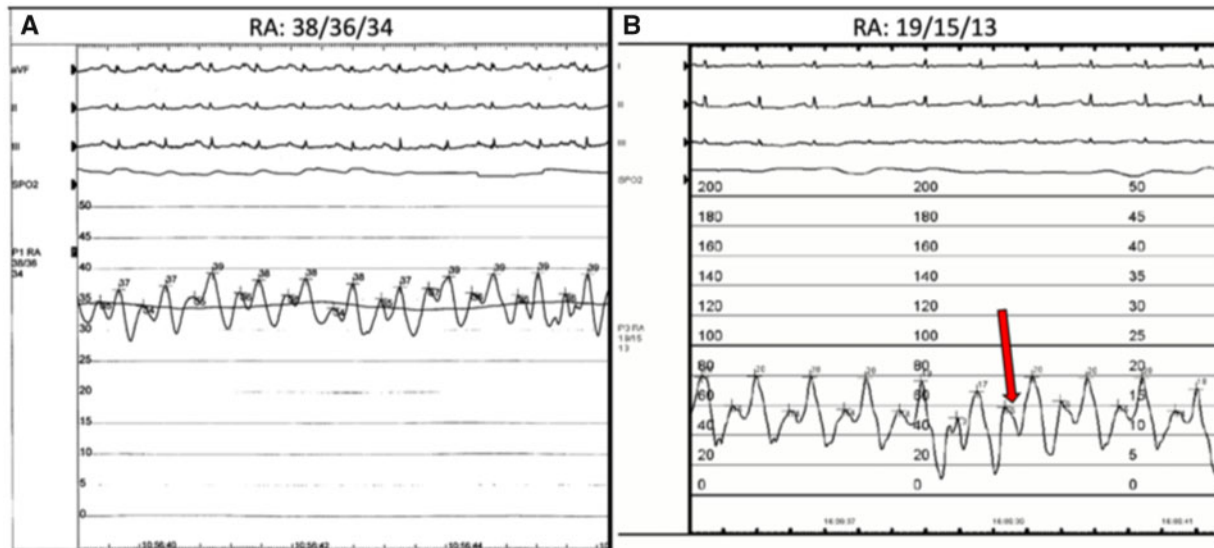


Figure 3 (A) Right atrial pressure on first admission before pericardiocentesis with markedly elevated mean right atrial pressure of 34 mmHg (nl 0–8 mmHg). (B) Right atrial pressure after drainage of pericardial effusion and with pericardial. Persistent elevated mean right atrial pressure of 13 mmHg with a rapid y-descent (red arrow) reflecting constrictive disease. The rapid y descent indicates absence of tamponade physiology.

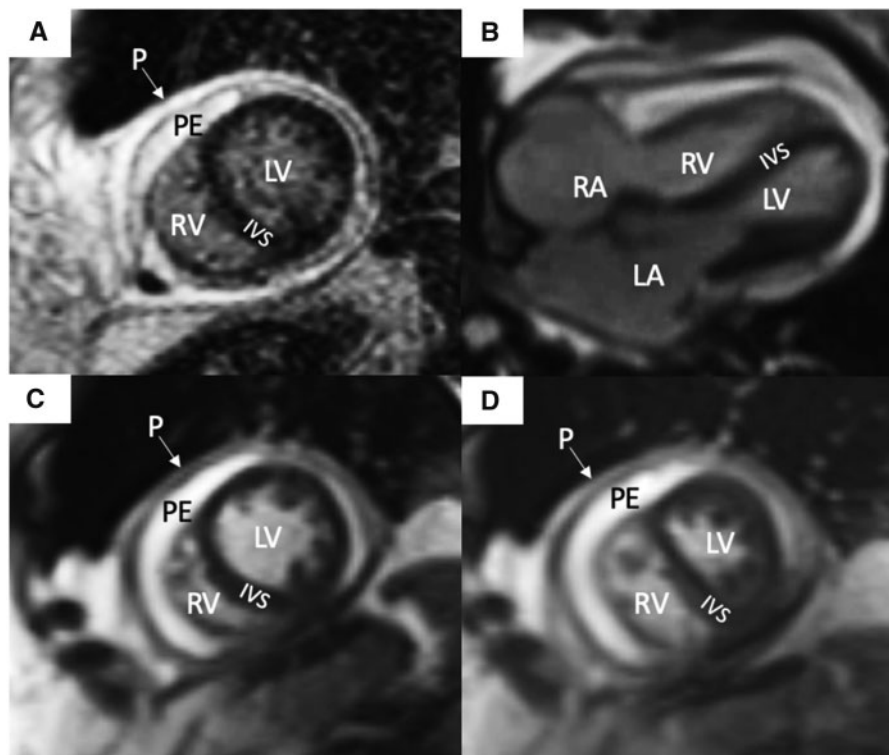


Figure 4 Cardiac magnetic resonance imaging (A) delayed enhancement imaging in short-axis shows circumferential pericardial oedema. (B) Real-time cine imaging in long-axis demonstrates slightly enlarged atria and ventricles have a tubular configuration suggestive of pericardial constriction. (C) Real-time cine imaging in short-axis with (D) diastolic interventricular septal flattening on deep inspiration indicating increased ventricular coupling which diagnosis of pericardial constriction. IVS, interventricular septum; LA, left atrium; LV, left ventricle; P, pericardium; PE, pleural effusion; RA, right atrium; RV, right ventricle.

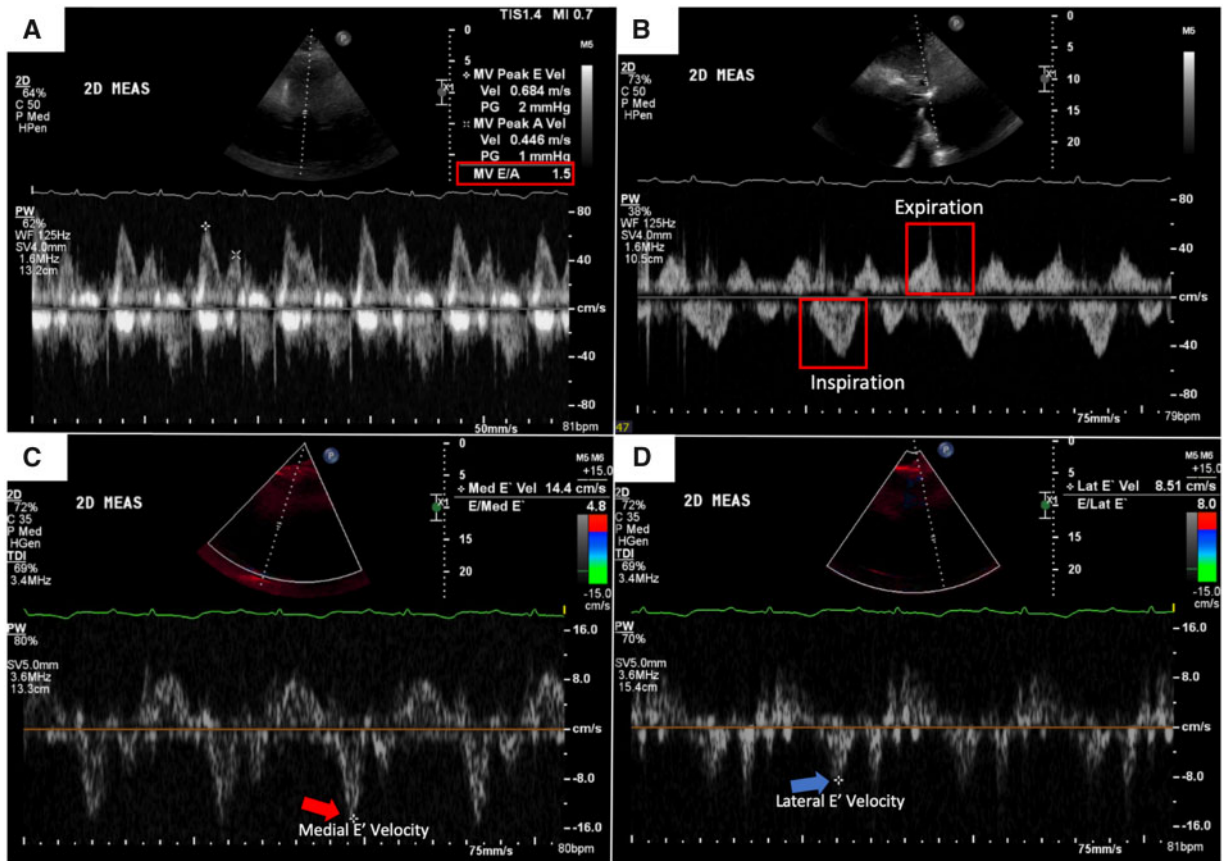
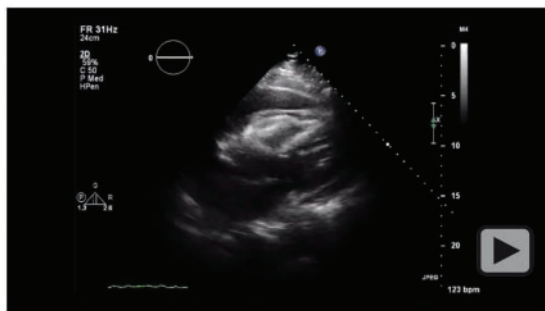
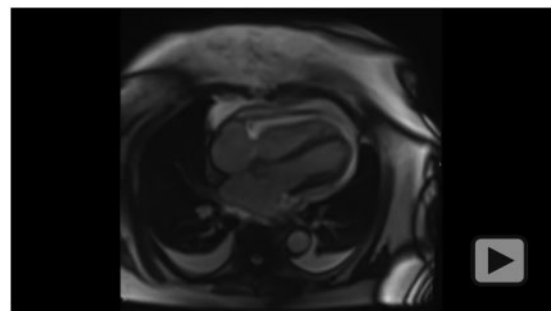


Figure 5 (A) Transthoracic echocardiogram showing mitral valve doppler with an E/A ratio of 1.5 which indicates restrictive mitral inflow velocity. (B) Pulsed wave Doppler of the hepatic vein shows end-diastolic reversal of the blood flow with end-expiratory increment. (C) The inferior vena cava is dilated (red arrow, diameter >21 mm) and does not collapse during inspiration, suggesting an elevated right atrial pressure of 20 mmHg. (D) Tissue Doppler showing the increased medial and reduced lateral mitral annular velocity which is typical for constrictive pericarditis. The constricting pericardium limits movement of the lateral aspect of the heart, while movement is still preserved at the septum. Medial E' velocity: 14.4 cm/s (red arrow) and lateral E' velocity: 8.5 cm/s (blue arrow).



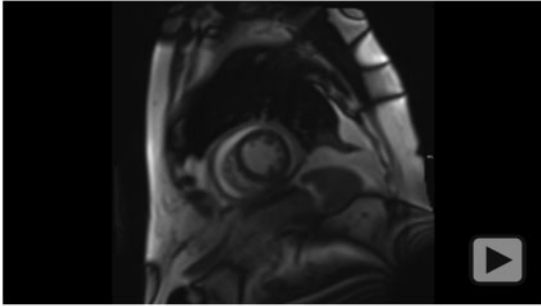
Video 1 Transthoracic Echocardiography: Parasternal longaxis confirmed a large circumferential pericardial effusion, with mild right ventricular collapse during diastole.



Video 2 Cardiac MRI: Real time cine imaging in long axis demonstrates slightly enlarged atria and ventricles with a tubular configuration.

aid in diagnosing concomitant myocarditis by demonstrating typical features including myocardial injury (hyperaemia, necrosis, scarring) in T1-weighted and myocardial oedema in T2-weighted

imaging.⁶ Definitive diagnosis of ECP is made during RHC. On removal of fluid from the pericardial space, a decrease in pericardial pressure is seen. The typical finding in ECP is that the elevated



Video 3 Cardiac MRI: Real time cine imaging in short axis with diastolic interventricular septal flattening on deep inspiration.

RA pressure does not normalize after pericardiocentesis. This, then, is the distinguishing feature of ECP, as compared to pure tamponade physiology.

Treatment for ECP depends on the haemodynamic stability of the patient. The treatment of cardiac tamponade is pericardiocentesis or pericardial window, along with the treatment of the underlying aetiology, if possible.^{6,7} Gentle intravenous fluids are commonly administered as a temporary measure prior to pericardiocentesis. The evidence for this practice is limited, however, administration of 500 mL of intravenous normal saline over 10 min was associated with an increase of 10% in cardiac index in only 47% of patients and a decrease of more than 10% in 31% of the patients.⁵ Evidence regarding the prognosis and optimal treatment is similarly limited. Non-steroidal anti-inflammatory agents or colchicine are considered to be first-line therapies, but up to 16% of patients continue to have symptoms and eventually require extensive pericardiectomy.⁸

Conclusion

ECP remains a rare condition that presents with clinical features of right heart failure and, as a result, can be mistaken for pure cardiac tamponade. Its distinguishing haemodynamic feature is a persistent elevation in RA pressure after pericardiocentesis, which may require pericardiectomy. The presence of the syndrome should be suspected in patients presenting with cardiac tamponade who have typical risk factors, such as rheumatologic diseases, malignancy, and radiation. The most appropriate treatment should be determined and escalated based on the specific clinical presentation.

Lead author biography



My name is Yousif Al-Saiegh, MD. After attending Hannover Medical School in Germany, I made the decision to pursue residency training in the US at Pennsylvania Hospital in Philadelphia. Early on in residency, I developed a strong interest in Cardiology. Currently, I am a PGY-3 Internal Medicine resident starting Cardiology fellowship in July 2021. On a personal level, I enjoy various sports including hiking weight lifting, ping pong, and swimming. I also like exploring new restaurant as well as cooking Mediterranean and Asian food.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

Funding: None declared.

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