

# Idiopathic Effusive Constrictive Pericarditis Accompanying Multiple Complications: A Case Report

**Chaodi Luo**

The First Affiliated Hospital of Xi'an Jiaotong University

**Jing Li**

The First Affiliated Hospital of Xi'an Jiaotong University

**Yang Yan**

The First Affiliated Hospital of Xi'an Jiaotong University

**Dan Han** (✉ [handan1789@163.com](mailto:handan1789@163.com))

The First Affiliated Hospital of Xi'an Jiaotong University

---

## Case report

**Keywords:** Effusive constrictive pericarditis, pericardiectomy, complication

**Posted Date:** November 13th, 2020

**DOI:** <https://doi.org/10.21203/rs.3.rs-105187/v1>

**License:**  This work is licensed under a Creative Commons Attribution 4.0 International License.

[Read Full License](#)

---

# Abstract

## Introduction

Effusive constrictive pericarditis (ECP) is a unique clinical syndrome that is characterized by the coexistence of pericardial effusion and constrictive pericardium. The etiology of ECP usually contains tuberculosis, idiopathic, and neoplastic causes. The early diagnosis, treatment strategy and prognostic predictor of ECP still remain a big problem nowadays due to the sophisticated clinical situations.

## Case Presentation

We here report a rare case of idiopathic ECP with thickened adherent visceral pericardium and normal parietal pericardium, accompanying multiple complications mainly comprising severe tricuspid regurgitation, hypoproteinemia, and proximal deep venous thrombosis. The patient was referred for radical pericardiectomy successfully, but the long-term prognosis may be unfavorable.

## Discussion

This case aims to provide some clinical experience of such situation in which the clinician should weight the benefits and the risks to a particular patient.

## Established Facts And Novel Insights

### Established Facts

1. The non-specific symptoms of Effusive constrictive pericarditis (ECP) are often similar to symptoms such as restrictive cardiomyopathy, tricuspid valve insufficiency, heart failure, and liver cirrhosis. Early diagnosis is still a huge challenge.
2. Patients with idiopathic ECP usually has the best in-hospital and long-term prognosis.

### Novel Insights

1. Proximal deep venous thrombosis (DVT) complicating ECP remains an extremely rare scenario.
2. Prognostic predictor of ECP remain a problem nowadays due to the sophisticated clinical situations.

## Introduction

Effusive constrictive pericarditis (ECP) has been increasingly recognized by clinicians as a unique clinical syndrome that is characterized by the coexistence of pericardial effusion and constrictive pericardium. The etiology of ECP in developing countries usually contains tuberculosis, idiopathic, and neoplastic causes. The clinical presentations are variable depending on the course of the disease, chest pain, fatigue, dyspnea, and leg edema are the most frequent complaints.[1, 2] Since the most distinct pathophysiological feature of ECP is the persistently elevated right atrial pressure after pericardial

effusion drainage, patients might even present with symptoms of constriction or diastolic heart failure for a long time following pericardiocentesis.[3]

Typically, the inner visceral pericardium consists of a monolayer mesothelial cells that cover the surface of the heart supported by fibrous and adipose tissues.[4] In ECP, fibrosis leading to thickened adhesions of both parietal pericardium and visceral pericardium with associated pericardial fluid are commonly observed. However, the visceral pericardium that is responsible for the constrictive component is not typically thickened or calcified of this process.[5] In review of the past case reports, no ECP patient with normal parietal pericardium has been described. Here, we report a rare case of idiopathic ECP with thickened adherent visceral pericardium and normal parietal pericardium, manifesting as long-term right heart failure and accompanying many other complications.

## Case Report

A 23-year-old man was admitted to our hospital with a 7-year history of bilateral leg edema and intermittent shortness of breath. Pericardiocentesis was performed 7 years ago at the local hospital due to massive pericardial effusion. About 7 months ago, the symptoms of bilateral leg edema and abdominal distension reappeared and the echocardiography showed massive pericardial effusion, suggesting effusive pericarditis. Therefore, pericardiocentesis was performed again and oral diuretics were prescribed for him to abate the discomforts. Over the past 1 month, he presented with symptoms of progressive right heart failure, pleural effusion and seroperitoneum, due to fatigue. The echocardiography suggested constrictive pericarditis. Thoracentesis was conducted at the local hospital due to the ineffective drug therapy.

Physical examination on admission showed that heart rate of 91 bpm, abnormal heart sounds, and pulse deficit; blood pressure of 89/60 mmHg; kussmaul respiration; obvious distension of jugular vein, severe bilateral leg edema, abdominal distension and hepatomegaly. The central venous pressure (CVP) was of 330 mmH<sub>2</sub>O. Laboratory findings included severe hypoproteinemia with albumin of 27.5 g/L and globulin of 17 g/L; hypokalemia with serum potassium of 2.38 mmol/L; N-terminal portion of proBNP (NT-proBNP) was of 1267 pg/mL. The chest X-ray showed straightened cardiac silhouette of both sides (shown in Fig. 1). The echocardiography revealed respirophasic septal shift (Supplemental video 1), mild pericardial effusion, severe tricuspid regurgitation (Supplemental video 2), bi-atrial enlargement (the left atrium was 54 mm × 57 mm; the right atrium was 43 mm × 56 mm), stroke volume was of 58 ml; dilated inferior vena cava (27 mm) with disappeared respiratory-induced collapse. Cervical vascular ultrasound showed thrombosis of bilateral internal jugular vein and left brachiocephalic vein (shown in Fig. 2). Computed tomography (CT) indicated pericardial calcification and hepatomegaly (shown in Fig. 3). Consequently, the patient was diagnosed as ECP, New York Heart Association (NYHA) class IV, and therefore referred for radical pericardiectomy. No significant change of the parietal pericardium was observed during the procedure, and the pericardial cavity was smooth with about 50 ml clear pericardial effusion. Additionally, the visceral pericardium was thickened to 1.5 mm and closely adherent to the heart surface, resulting in a constrictive band like an “eggshell” to wrap the heart, which severely influenced the

diastolic and systolic functions. Finally, the thickened visceral layer was successfully removed and the CVP was immediately reduced to 190 mmH<sub>2</sub>O (shown in Fig. 4). The pathologic examination revealed fibrosis suggesting idiopathic pericarditis (shown in Fig. 5). The patient was meanwhile prescribed with hydrochlorothiazide, frusemide, spironolactone, benazepril, digoxin, rivaroxaban, albumin infusion and other symptomatic treatment to alleviate his symptoms of heart failure. The degree of tricuspid regurgitation decreased gradually (shown in Fig. 6) and no symptoms of heart failure were reported during the follow-up period.

## Discussion

This patient is a rare case of idiopathic ECP with thickened adherent visceral pericardium and normal parietal pericardium, presenting with symptoms of right heart failure and accompanying many other complications, which confused the diagnosis, treatment and prognosis.

The early diagnosis of ECP remains to be a huge challenge due to the non-specific symptoms that usually mimic the manifestations of other conditions such as restrictive cardiomyopathy, tricuspid insufficiency, heart failure, and cirrhosis.[6] Therefore, auxiliary tools are necessary to make an accurate diagnosis. Although invasive cardiac catheterization assessing the alterations of right atrial pressure and intrapericardial pressure pre- and post-pericardiocentesis remains the gold standard for the diagnosis of ECP, echocardiography is currently critical and more convenient in evaluation of pericardial disease.[7] The key echo-Doppler hallmark of ECP patient is respirophasic interventricular septal shift due to dissociation of intracardiac and intrathoracic pressures and ventricular interdependence, which can be observed in our patient. Other features of ECP include a higher mean medial mitral e' velocity, a higher prevalence of mitral inflow variation and hepatic vein flow reversal.[5]

Many clinical trials have demonstrated that tricuspid regurgitation tremendously complicates ECP and preoperative higher grade tricuspid regurgitation is frequently associated with increased postoperative mortality.[8] Additionally, tricuspid regurgitation scarcely improves with pericardiectomy alone and valve repair has little benefit on late survival. Therefore, simultaneous tricuspid valve repair should be considered to reduce symptoms only if it does not increase the operative risk.[9, 10] As for this patient, we considered that tricuspid valvuloplasty would increase the immediate risk of the operation, only pericardiectomy was performed. The tricuspid regurgitation gradually decreased to mild-moderate volume and no symptoms of heart failure were reported during the postoperative follow-up.

Albumin infusion (1–2 g/kg) was applied for this patient due to his severe hypoalbuminemia, sodium-water retention, massive pleural effusion, seroperitoneum, and postoperative drainage, until the bad situation had improved. Albumin has many crucial physiological functions including volume expansion, anti-oxidation, anti-inflammation, and endothelial protection, which seems to be an ideal treatment solution for critically ill patients.[11] However, the use of albumin is still controversial. Despite that safety of albumin use has been proven, albumin infusion is expensive and has little impact on the survival rate. [12] Accordingly, more efforts are urgently needed to define exact indications, required dose and response

predictors in regard of albumin use, in order that patients gain the maximum benefit from its administration.

To date, proximal deep venous thrombosis (DVT) complicating ECP remains an extremely rare scenario. Such patients are at high risk of developing further thromboembolic complications including pulmonary embolism (PE), intracardiac thrombi, thrombus incarceration of tricuspid valve, and even heart failure, especially during the procedure. Therefore, ultrasound-guided central venous catheterization to prevent these immediate complications is of great significance.[13, 14] Besides, antithrombotic therapy to prevent further thromboembolic complications is of equal importance. According to the antithrombotic guideline, in patients with proximal DVT or PE, long-term (3 months) anticoagulant therapy such as dabigatran, rivaroxaban is recommended. Aspirin is also suggested to prevent recurrent venous thromboembolism (VTE) after anticoagulant therapy.[15] Central venous catheter placement was performed for our patient in prophylaxis of perioperative thromboembolic complications and gave rivaroxaban (10 mg/d) to aspirin 100 mg/d after being discharged from the hospital for three months to prevent further thromboembolic complications.

The long-term survival post-pericardiectomy depends on many factors especially the underlying etiology and the overall physical condition which mainly incorporates NYHA class III-IV, radiation history, age, serum sodium, serum albumin and serum creatinine levels. Patients with idiopathic ECP usually has the best in-hospital and long-term prognosis, with 7-year Kaplan-Meier survival of 88%. Preoperative NYHA class III-IV has been proved as a robust independent predictor of late death, with long-term survival below 50%.[16–18] Moreover, the Child-Pugh score of 7 or higher, preoperative early diastolic mitral inflow velocity of 71 cm/s or higher and diabetes mellitus have recently been identified as independent prognostic factors of poor prognosis post radical pericardiectomy.[19, 20] Despite that our patient was diagnosed as idiopathic ECP, he was in NYHA class IV, Child-Pugh score was 11 and with multiple complications, therefore, the prognosis may be unfavorable.

## Conclusion

ECP is becoming an increasingly recognized clinical syndrome by clinicians. The early diagnosis, treatment strategy and prognostic predictor of ECP still remain a big problem nowadays due to the sophisticated clinical situations. Our report demonstrates a rare case of ECP with multiple complications in order to provide some experience in such situation in which the clinician should weight the benefits and the risks to a particular patient.

## Abbreviations

CT: Computed tomography

CVP: Central venous pressure

DVT: Deep venous thrombosis

ECP: Effusive constrictive pericarditis

NT-proBNP: N-terminal portion of proBNP

NYHA: New York Heart Association

PE: Pulmonary embolism

VTE: Venous thromboembolism

## **Declarations**

### **Acknowledgements**

Not applicable.

### **Funding**

Not applicable.

### **Availability of data and materials**

The datasets of the current study are available from the corresponding author upon reasonable request.

### **Ethics approval and consent to participate**

The ethics committee of the First Affiliated Hospital of Xi'an Jiaotong University approved the study.

### **Consent for publication**

Consent for publication is obtained from the patient.

### **Competing interests**

The authors declare that they have no competing interests.

### **Author Contributions**

Dr. Chaodi Luo and Dr. Dan Han: collected the primary data and drafted the initial manuscript. Dr. Yang Yan: diagnosed the case and instructed the patient's treatment. Dr. Jing Li: assisted with the treatment of the patient. All authors contributed to discussions and critically appraised the manuscript. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

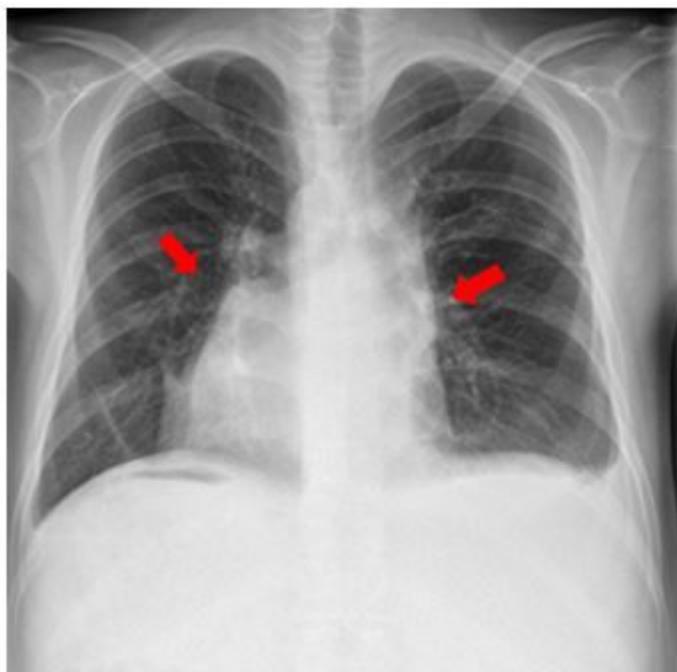
## **References**

1. Miranda WR, Oh JK. Effusive-Constrictive Pericarditis. *Cardiology clinics*. 2017;35(4):551-8.

2. Sagristà-Sauleda J, Angel J, Sánchez A, Permanyer-Miralda G, Soler-Soler J. Effusive-constrictive pericarditis. *The New England journal of medicine*. 2004;350(5):469-75.
3. Shah AD, Molloy DL, Blair JE. Hemodynamic Findings of Effusive-Constrictive Pericarditis. *JACC Cardiovascular interventions*. 2016;9(17):e167-8.
4. Hoit BD. Anatomy and Physiology of the Pericardium. *Cardiology clinics*. 2017;35(4):481-90.
5. Kim KH, Miranda WR, Sinak LJ, Syed FF, Melduni RM, Espinosa RE, et al. Effusive-Constrictive Pericarditis After Pericardiocentesis: Incidence, Associated Findings, and Natural History. *JACC Cardiovascular imaging*. 2018;11(4):534-41.
6. Marta L, Alves M, Peres M, Ferreira R, Ferreira H, Leal M, et al. Effusive-constrictive pericarditis as the manifestation of an unexpected diagnosis. *Revista portuguesa de cardiologia : orgao oficial da Sociedade Portuguesa de Cardiologia = Portuguese journal of cardiology : an official journal of the Portuguese Society of Cardiology*. 2015;34(1):69.e1-6.
7. Miranda WR, Newman DB, Oh JK. Effusive-Constrictive Pericarditis: Doppler Findings. *Current cardiology reports*. 2019;21(11):144.
8. Williams L, Frenneaux M. Is it primarily tricuspid regurgitation, constriction or restriction? *Heart (British Cardiac Society)*. 2009;95(17):1380-1.
9. Góngora E, Dearani JA, Orszulak TA, Schaff HV, Li Z, Sundt TM, 3rd. Tricuspid regurgitation in patients undergoing pericardiectomy for constrictive pericarditis. *The Annals of thoracic surgery*. 2008;85(1):163-70; discussion 70-1.
10. Kandachar PS, Kandachar SS, Maddali MM, Thomas E, Zacharias S. Constrictive Pericarditis and Tricuspid Valve Involvement. *Journal of cardiothoracic and vascular anesthesia*. 2016;30(5):1435-6.
11. Vincent JL, Russell JA, Jacob M, Martin G, Guidet B, Wernerman J, et al. Albumin administration in the acutely ill: what is new and where next? *Crit Care*. 2014;18(4):231.
12. Garcia-Tsao G. Long-term albumin in cirrhosis: is it the ANSWER? *Lancet (London, England)*. 2018;391(10138):2391-2.
13. Ozhan H, Okçun B, Akdemir R. Constrictive pericarditis associated with right atrial and major venous thrombosis. *Acta Cardiol*. 2004;59(4):435-7.
14. Pahade A, Tewari P. Major vessel venous thrombosis in patients of posttubercular chronic constrictive pericarditis undergoing pericardectomy: A rare scenario. *Annals of cardiac anaesthesia*. 2017;20(3):348-50.
15. Kearon C, Akl EA, Ornelas J, Blaivas A, Jimenez D, Bounameaux H, et al. Antithrombotic Therapy for VTE Disease: CHEST Guideline and Expert Panel Report. *Chest*. 2016;149(2):315-52.
16. Szabó G, Schmack B, Bulut C, Soós P, Weymann A, Stadtfeld S, et al. Constrictive pericarditis: risks, aetiologies and outcomes after total pericardiectomy: 24 years of experience. *European journal of cardio-thoracic surgery : official journal of the European Association for Cardio-thoracic Surgery*. 2013;44(6):1023-8; discussion 8.

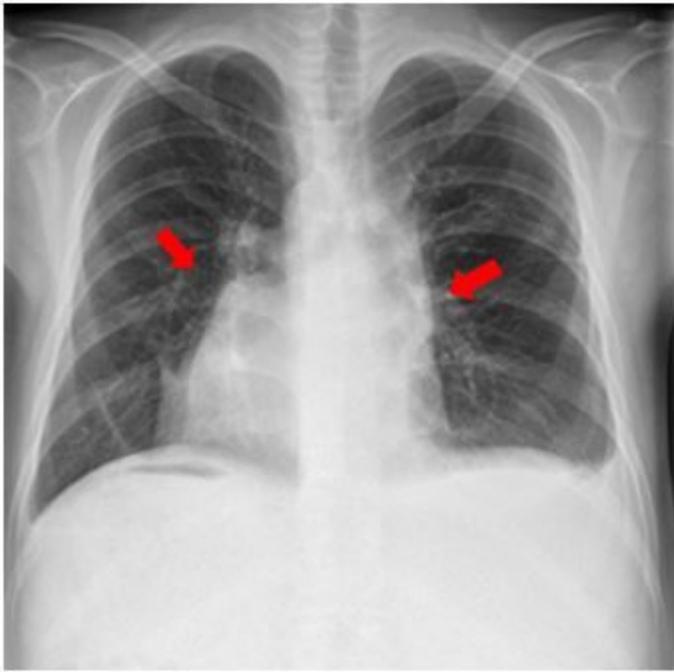
17. Zhu P, Mai M, Wu R, Lu C, Fan R, Zheng S. Pericardiectomy for constrictive pericarditis: single-center experience in China. *Journal of cardiothoracic surgery*. 2015;10:34.
18. Nozohoor S, Johansson M, Koul B, Cunha-Goncalves D. Radical pericardiectomy for chronic constrictive pericarditis. *Journal of cardiac surgery*. 2018;33(6):301-7.
19. Komoda T, Frumkin A, Knosalla C, Hetzer R. Child-Pugh score predicts survival after radical pericardiectomy for constrictive pericarditis. *The Annals of thoracic surgery*. 2013;96(5):1679-85.
20. Kang SH, Song JM, Kim M, Choo SJ, Chung CH, Kang DH, et al. Prognostic predictors in pericardiectomy for chronic constrictive pericarditis. *The Journal of thoracic and cardiovascular surgery*. 2014;147(2):598-605.

## Figures



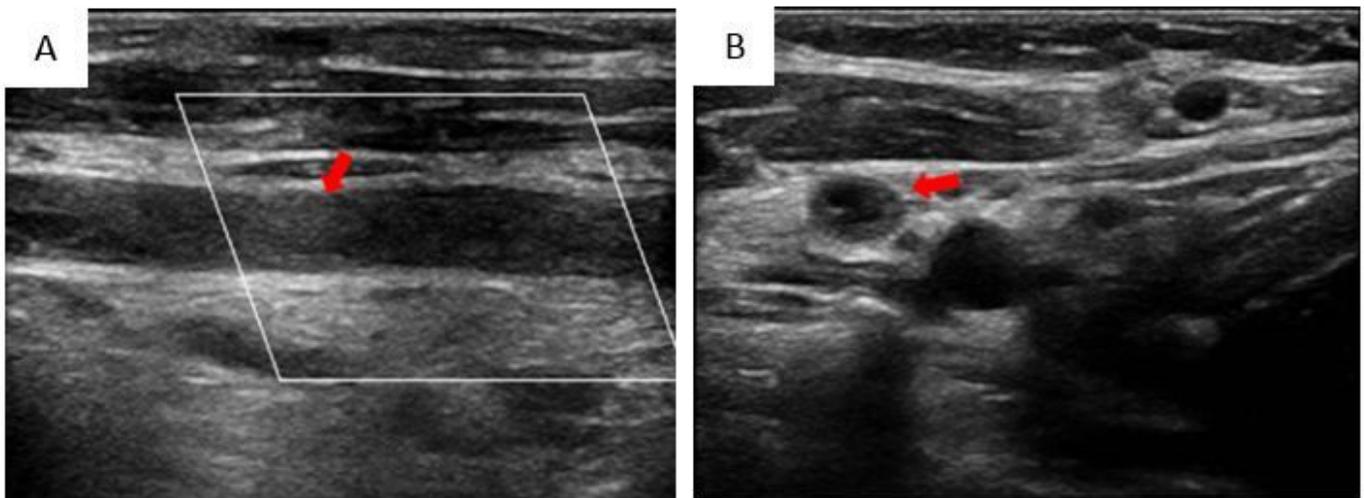
**Figure 1**

Chest X-ray showed straightened cardiac silhouette of both sides (red arrow).



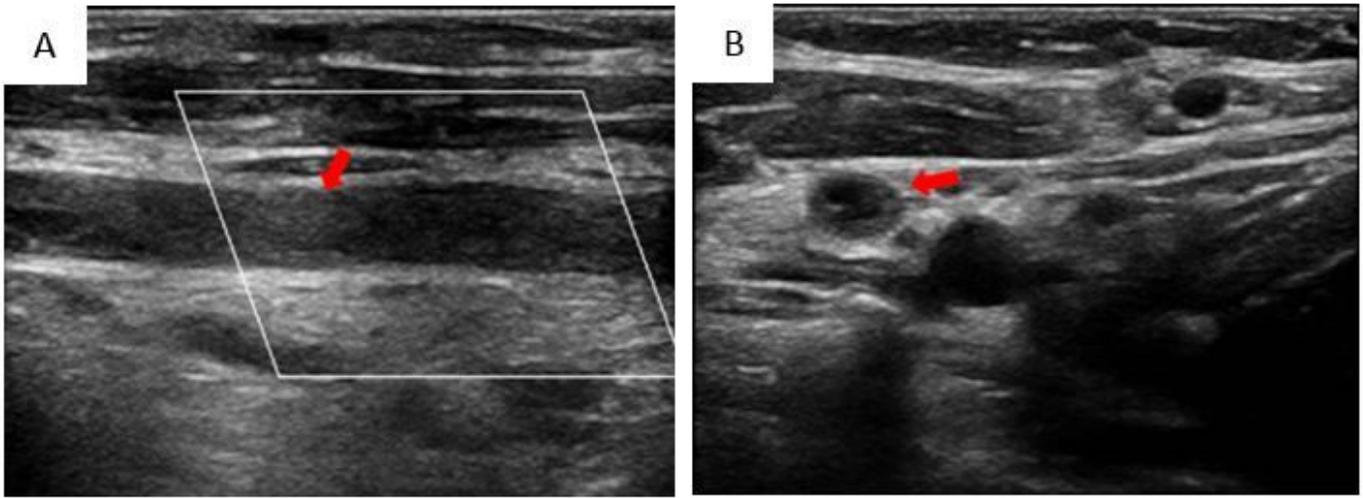
**Figure 1**

Chest X-ray showed straightened cardiac silhouette of both sides (red arrow).



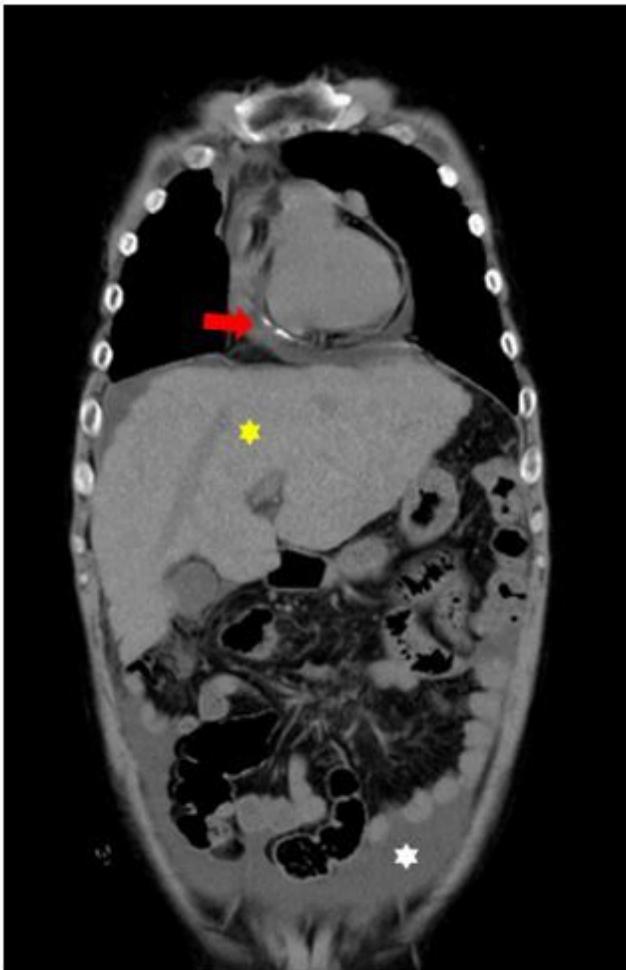
**Figure 2**

Cervical vascular ultrasound showed thrombosis of bilateral internal jugular vein with no blood flow (red arrow).



**Figure 2**

Cervical vascular ultrasound showed thrombosis of bilateral internal jugular vein with no blood flow (red arrow).



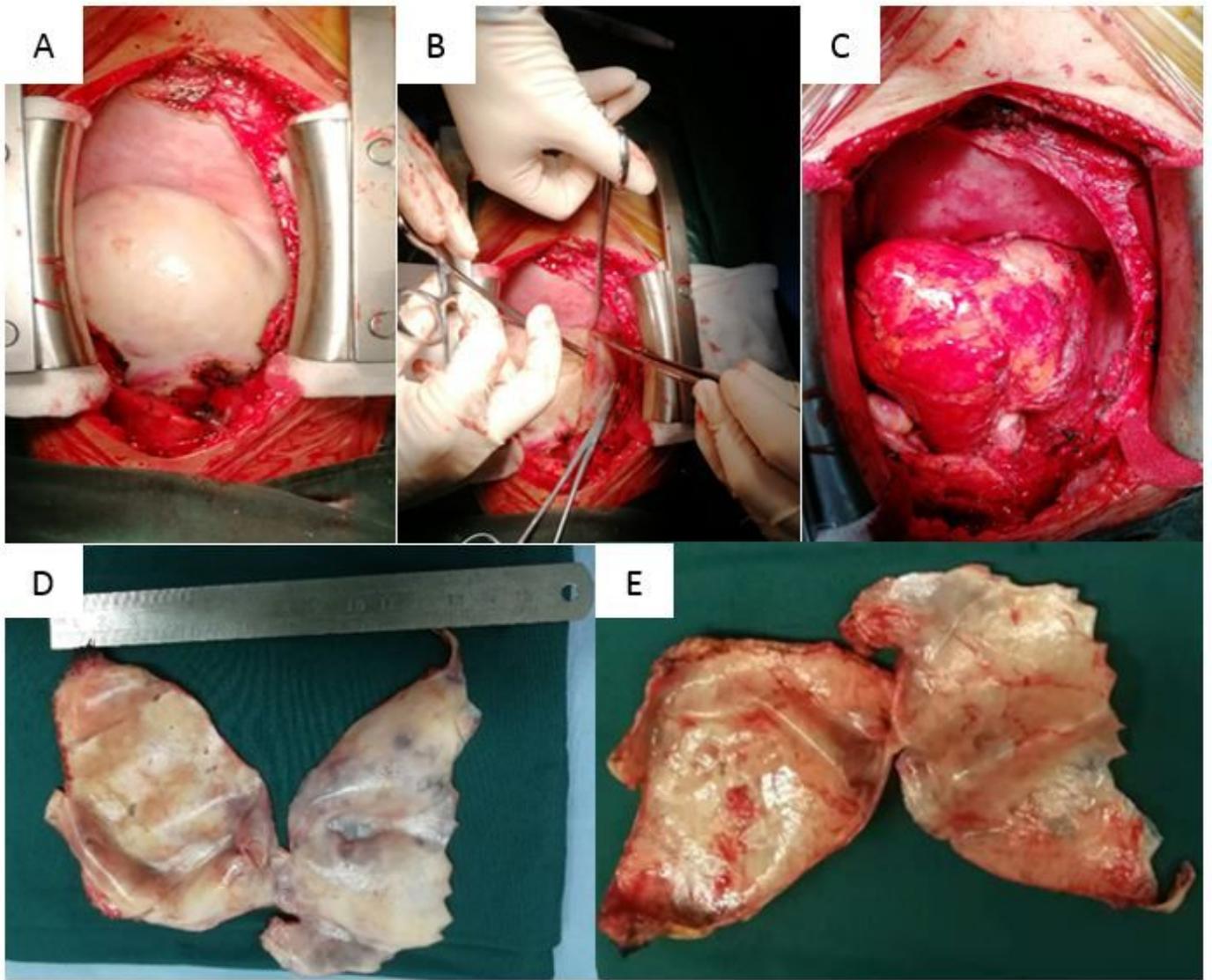
**Figure 3**

Computed tomography (CT) indicated pericardial calcification (red arrow), hepatomegaly (yellow astreoid) and seroperitoneum (white astreoid).



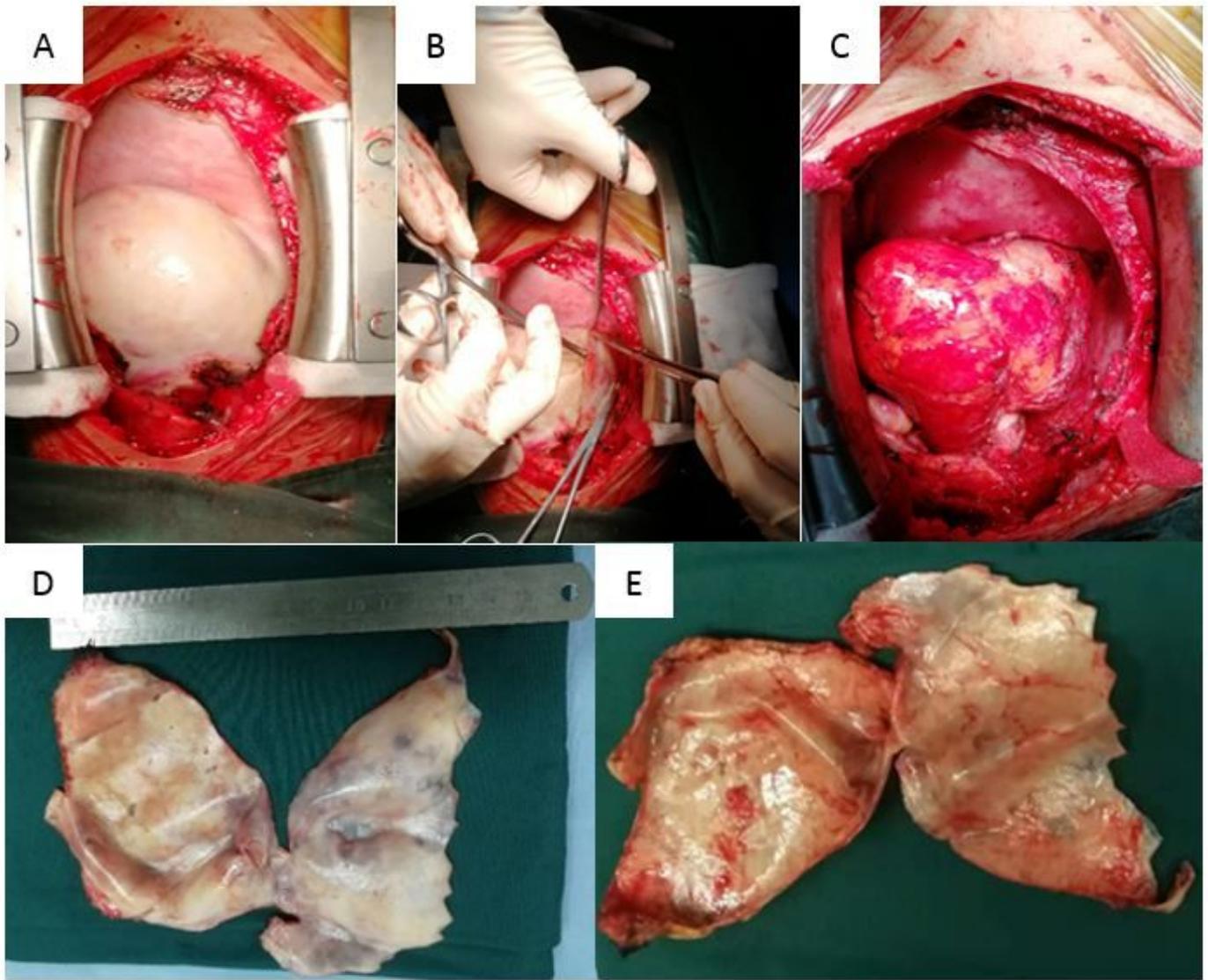
**Figure 3**

Computed tomography (CT) indicated pericardial calcification (red arrow), hepatomegaly (yellow astreoid) and seroperitoneum (white astreoid).



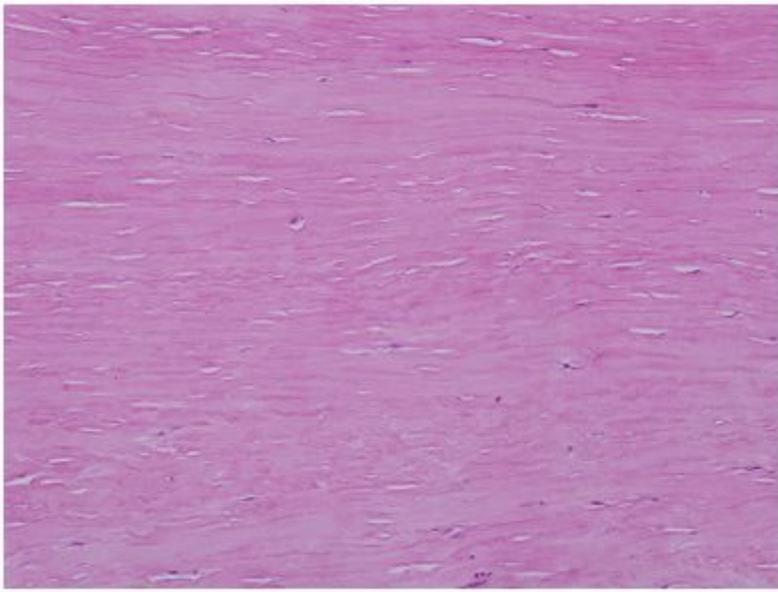
**Figure 4**

a The gross view of heart in situ originally observed at the time of pericardectomy revealed a complete pericardial cavity, a thickened visceral pericardium that closely adherent to the heart surface, resulting in a constrictive band like an “eggshell” to wrap the heart. b The thickened visceral layer was successfully removed from the heart. c The cardiac morphology after peeling off the thickened visceral pericardium. d,e The detached visceral pericardium.



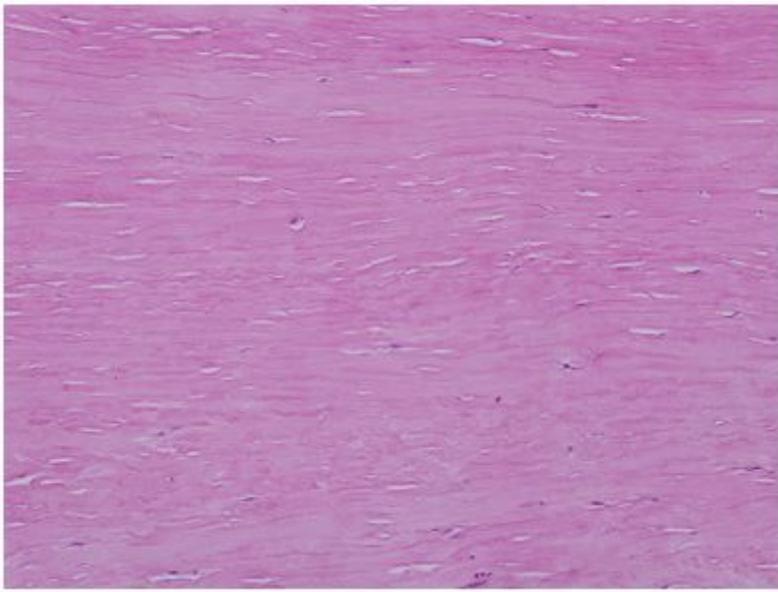
**Figure 4**

a The gross view of heart in situ originally observed at the time of pericardectomy revealed a complete pericardial cavity, a thickened visceral pericardium that closely adherent to the heart surface, resulting in a constrictive band like an “eggshell” to wrap the heart. b The thickened visceral layer was successfully removed from the heart. c The cardiac morphology after peeling off the thickened visceral pericardium. d,e The detached visceral pericardium.



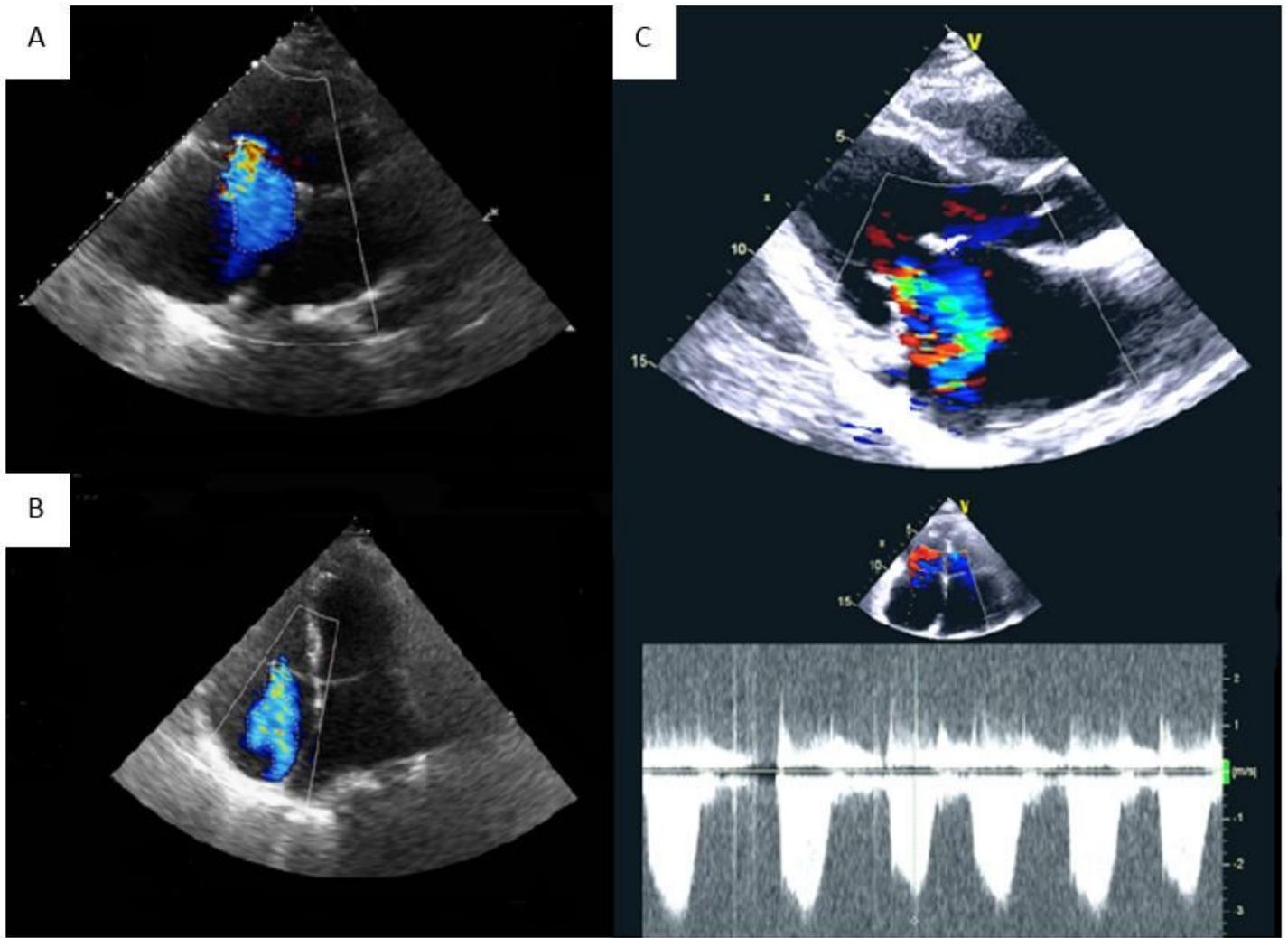
**Figure 5**

The pathologic examination revealed fibrosis suggesting idiopathic pericarditis.



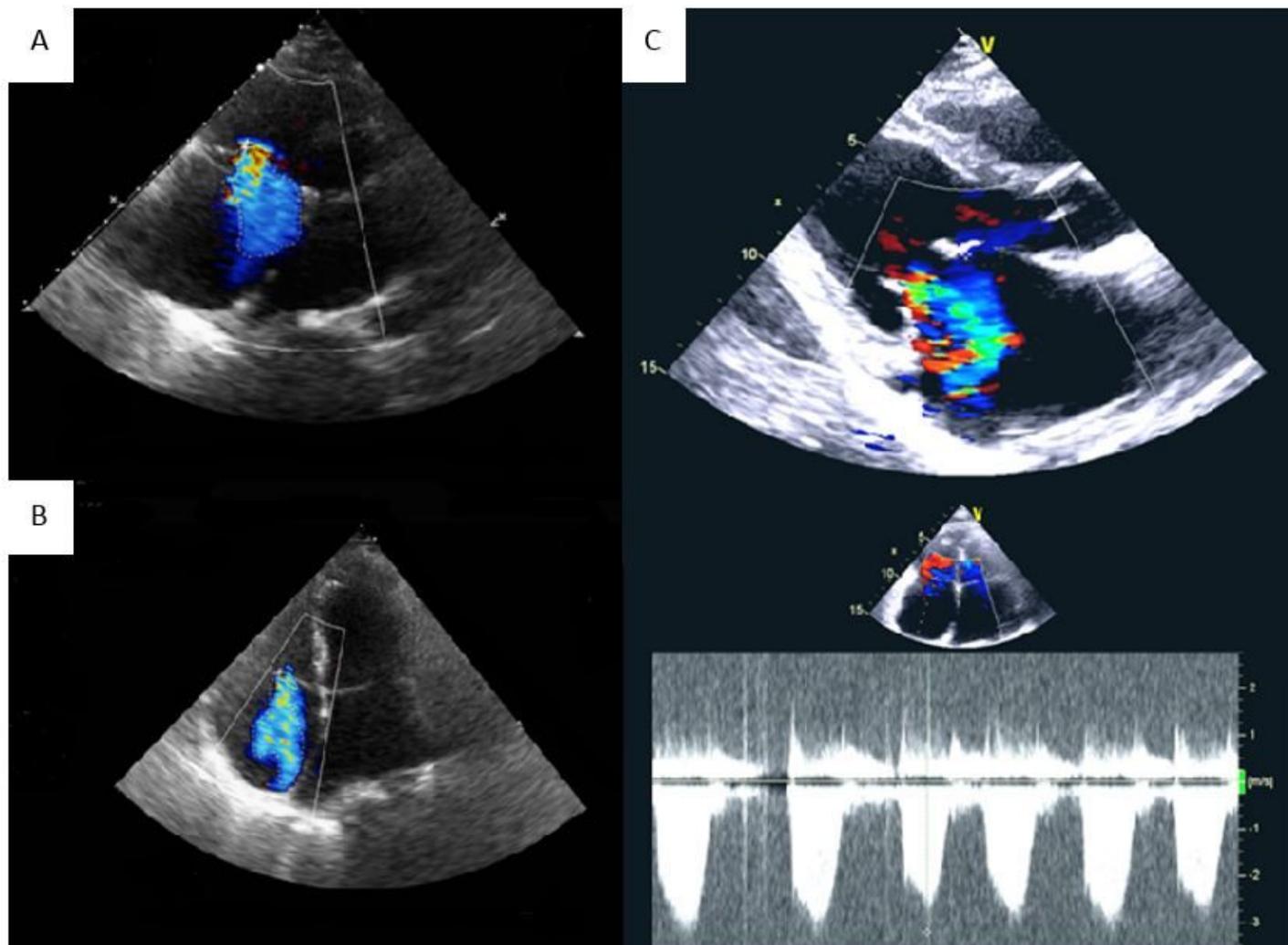
**Figure 5**

The pathologic examination revealed fibrosis suggesting idiopathic pericarditis.



**Figure 6**

The echocardiography showed the degree of tricuspid regurgitation decreased gradually. a Severe tricuspid regurgitation pre-pericardiectomy; b Severe tricuspid regurgitation at 1 week post-pericardiectomy; c Tricuspid regurgitation decreased to mild-moderate volume at 3-month follow-up.



**Figure 6**

The echocardiography showed the degree of tricuspid regurgitation decreased gradually. a Severe tricuspid regurgitation pre-pericardiectomy; b Severe tricuspid regurgitation at 1 week post-pericardiectomy; c Tricuspid regurgitation decreased to mild-moderate volume at 3-month follow-up.

## Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- [CAREchecklist.pdf](#)
- [CAREchecklist.pdf](#)