



## Case Report

## Combinational elastography prior to pericardiectomy to assess liver condition in patients with constrictive pericarditis



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## ABSTRACT

A 68-year-old man was diagnosed with pericarditis associated with immunoglobulin G4-related disease and was administered prednisolone 2 years prior to presentation. During the process of tapering off from prednisolone 1 year later, edema of the lower legs and pleural effusion worsened. He gradually developed dyspnea on exertion, and laboratory examinations revealed elevated liver enzyme levels. Diuretics were administered; however, the symptoms did not resolve. Transthoracic echocardiography and cardiac catheterization revealed findings consistent with those of constrictive pericarditis. Pericardiectomy was considered and the perioperative risks due to possible recovery from liver dysfunction were discussed. Combinational elastography was subsequently performed. The results indicated the absence of liver fibrosis, suggesting that liver dysfunction was attributable to liver congestion; thus, the liver dysfunction was considered reversible. Subsequently, pericardiectomy was performed. Given that constrictive pericarditis can lead to liver dysfunction due to congestion, the perioperative risk is often controversial when considering surgical interventions.

**Learning objective:** Combinational elastography may be useful in the preoperative evaluation of patients with cardiac diseases complicated by liver dysfunction to distinguish liver fibrosis, understand the pathogenesis of liver dysfunction, and determine subsequent treatment strategies.

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## Introduction

Cardiac diseases that cause right-sided heart failure, such as constrictive pericarditis, are often associated with liver dysfunction owing to congestion. Liver dysfunction is also associated with perioperative risk; therefore, preoperative evaluation of liver status is important. We report a case of constrictive pericarditis caused by immunoglobulin G4 (IgG4)-related pericarditis that developed into liver dysfunction, which was evaluated using combinational elastography prior to pericardiectomy.

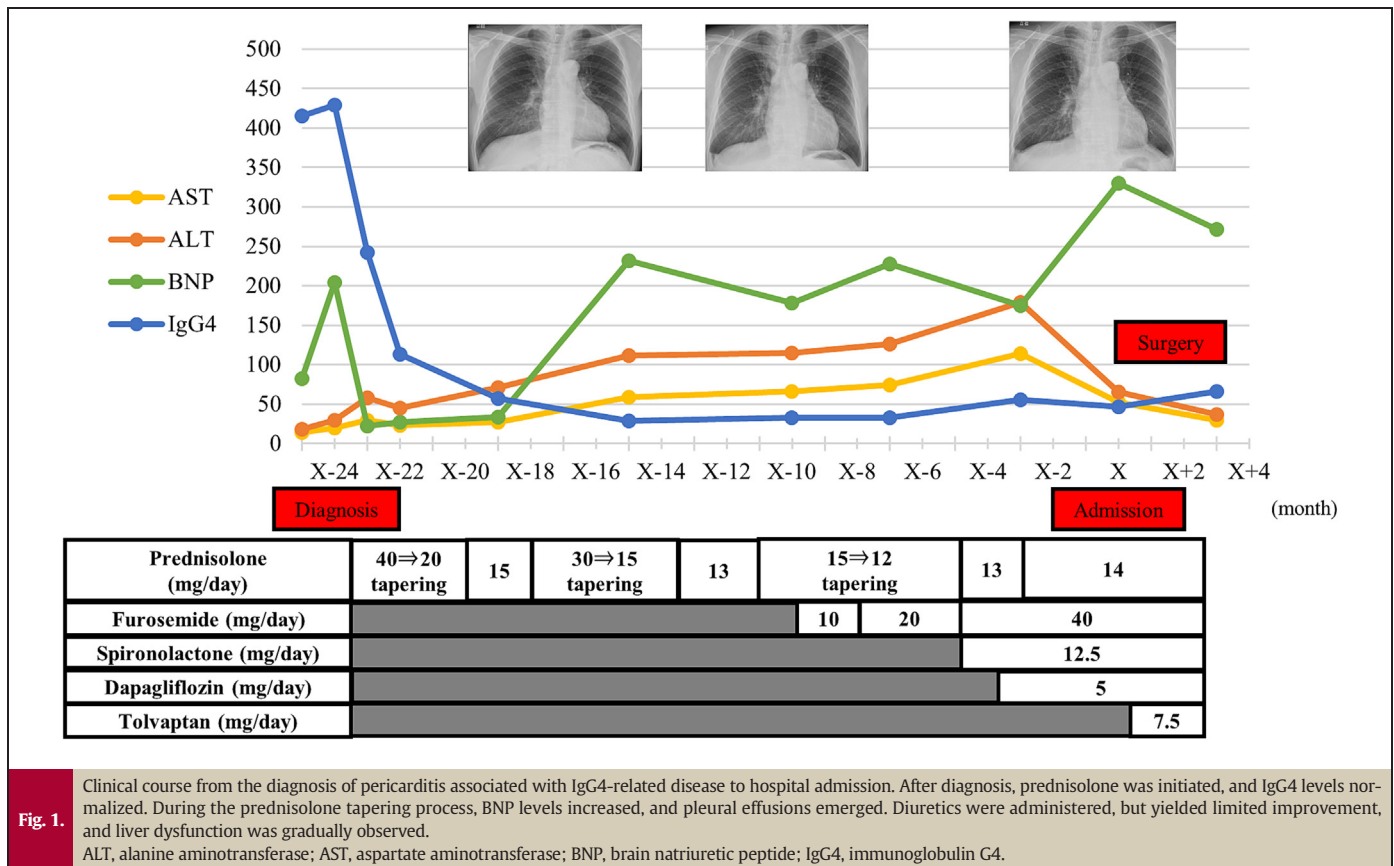
## Case report

A 68-year-old man developed chest pain that increased with inhalation and bending forward 2 years prior to presentation. An elevated inflammatory response, high levels of IgG4 (429 mg/dL), pericardial

effusion, and pericardial thickening were observed. A thoracoscopic pericardial biopsy revealed IgG4-positive cells in the pericardium, consistent with IgG4-related pericarditis. Treatment with prednisolone (40 mg/day) resulted in the prompt normalization of IgG4 levels. The prednisolone dose was gradually reduced during hospital follow-up visits. Approximately 1 year later, the patient developed edema of the lower legs, pleural effusion, and worsening dyspnea on exertion. Despite the addition of diuretics, his symptoms did not improve, and progressive liver dysfunction was observed, leading to hospitalization (Fig. 1).

On admission, his blood pressure was 86/66 mmHg and pulse rate was 78/min. Physical examination revealed jugular venous distention and lower leg edema. Auscultation revealed pericardial knocking. Blood tests showed increased brain natriuretic peptide (330 pg/mL) and slightly elevated C-reactive protein (1.06 mg/dL) levels. IgG4 levels were within the normal range (48.9 mg/dL). Liver enzyme levels were elevated (Online Data). Tests for hepatitis B surface antigen and hepatitis C virus antibodies were negative. Electrocardiography revealed a sinus rhythm and low voltage. Chest radiography revealed pleural effusion and cardiac

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**Fig. 1.** Clinical course from the diagnosis of pericarditis associated with IgG4-related disease to hospital admission. After diagnosis, prednisolone was initiated, and IgG4 levels normalized. During the prednisolone tapering process, BNP levels increased, and pleural effusions emerged. Diuretics were administered, but yielded limited improvement, and liver dysfunction was gradually observed. ALT, alanine aminotransferase; AST, aspartate aminotransferase; BNP, brain natriuretic peptide; IgG4, immunoglobulin G4.

enlargement. Transthoracic echocardiography revealed pericardial adhesions, enlarged inferior vena cava, septal bounce, and marked hepatic vein diastolic flow reversal with expiration. Computed tomography revealed a diffusely thickened pericardium with localized nodular thickening, pleural effusion, and ascites (Fig. 2A). 18F-fluorodeoxyglucose positron emission tomography-computed tomography revealed increased fluorodeoxyglucose uptake in the thickened nodular areas of the pericardium (Fig. 2B). Cardiac catheterization showed a dip-and-plateau pattern in both ventricular pressure waveforms with end-diastolic equalization (Fig. 2C). Right atrial pressure was elevated (Fig. 2D), and the cardiac index decreased to 1.65 L/min/m<sup>2</sup>. The examination results were consistent with constrictive pericarditis.

The symptoms of right heart failure could not be controlled with medication; therefore, pericardiectomy was considered. However, due to liver dysfunction, the risk of perioperative complications was controversial [no encephalopathy, small amount of ascites, bilirubin 2.3 mg/dL, albumin 3.5 mg/dL, prothrombin activity 76 %, Child-Pugh score of 8, and model for end-stage liver disease (MELD) score of 11]. Combinational elastography was conducted to assess the liver condition, revealing a median shear wave velocity (Vs) of 2.79 m/s (interquartile range: 2.73–2.85 m/s) and a median liver fibrosis index (LFI) of 0.433 (interquartile range: 0.119–0.747) (Fig. 3). These data support the suspicion of liver dysfunction caused by liver congestion, ruling out liver fibrosis. Despite the potential perioperative risks associated with liver dysfunction, we believed that treating the underlying cause of constrictive pericarditis could potentially reverse liver congestion. Consequently, the patient underwent pericardiectomy. Due to the strong adhesions, it was difficult to dissect the posterior one-third of the pericardium. Combinational elastography performed after surgery revealed the median Vs of 2.84 m/s, suggesting residual liver congestion. However, the median LFI remained at 0.91, indicating no liver fibrosis progression. Furthermore, the patient experienced improved dyspnea on exertion and a reduction in lower leg edema after surgery. Liver function also improved, with aspartate aminotransferase and alanine aminotransferase

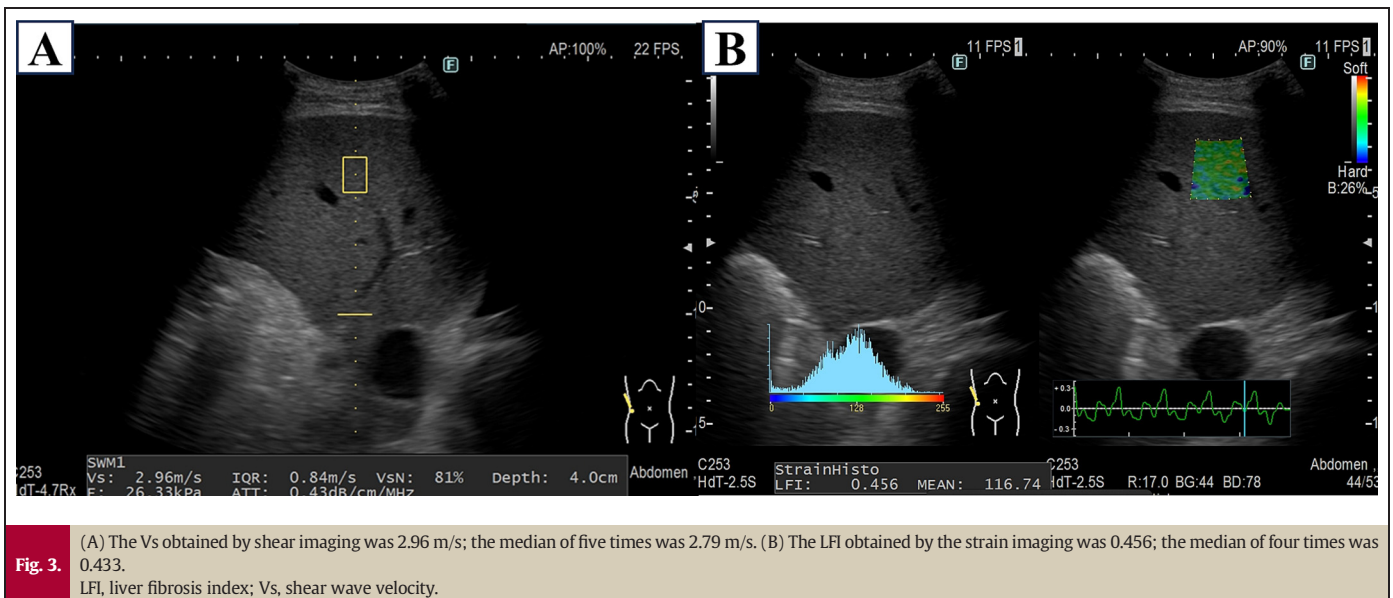
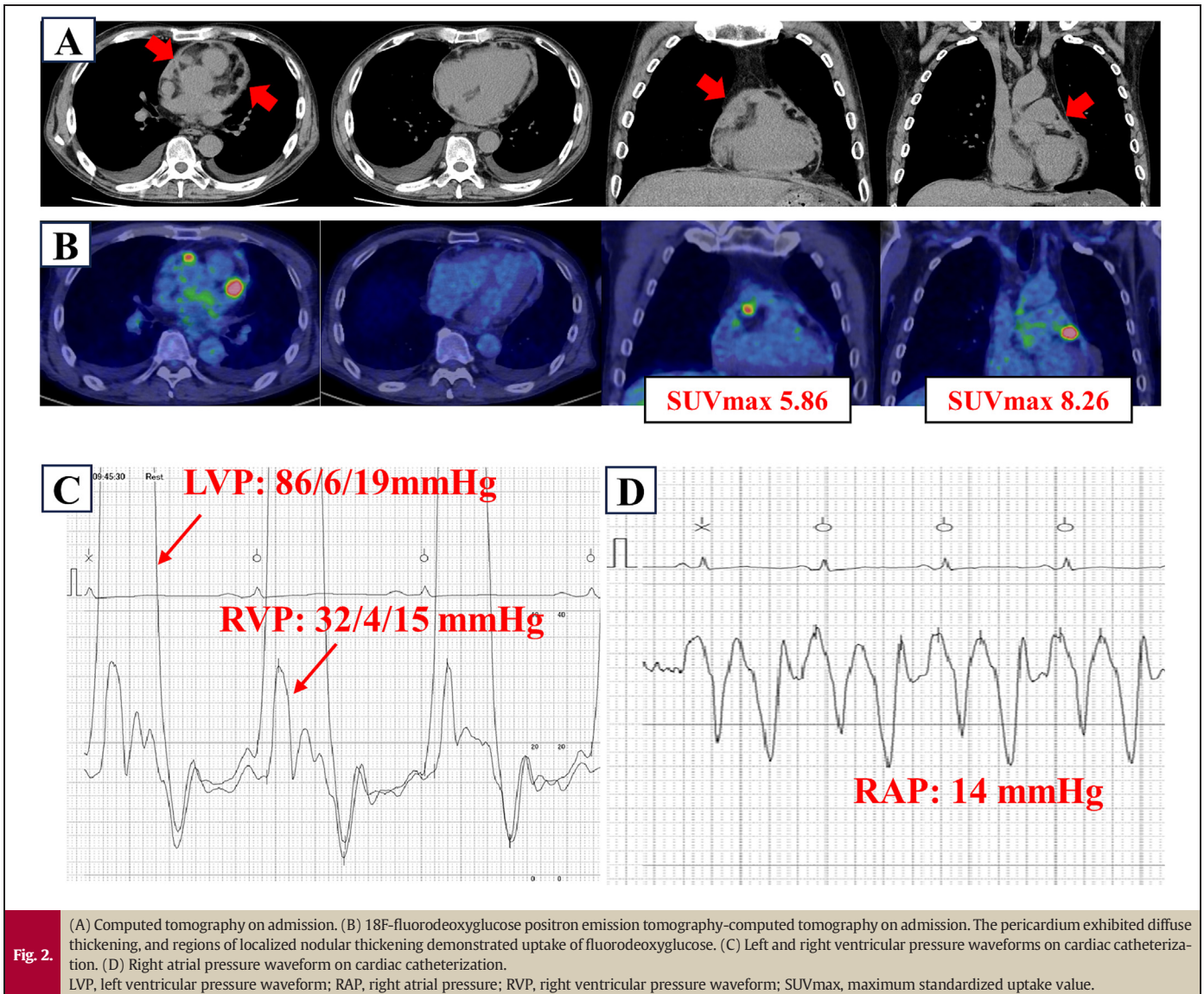
levels of 30 and 37 U/L, respectively (Fig. 1). The patient continues to be followed-up.

**Discussion**

We encountered a case of IgG4-related pericarditis presenting as constrictive pericarditis. Doumen et al. reported 32 patients with IgG4-related pericarditis [1], 24 % initially treated with glucocorticoids alone required additional pericardiectomy or immunosuppressive therapy because of recurrence or an inadequate therapeutic response. Furthermore, five patients initially treated with pericardiectomy alone achieved remission without the need for maintenance therapy; therefore, pericardiectomy could have been considered earlier in our case.

Cardiac diseases that cause right-sided heart failure, such as constrictive pericarditis, are often associated with liver dysfunction owing to congestion. Liver dysfunction is associated with a perioperative cardiac risk. In an analysis of nine studies of patients with cirrhosis undergoing cardiac surgery, the mortality rates for Child-Pugh classes A, B, and C were 5 %, 35 %, and 70 %, respectively [2]. Furthermore, overall survival after pericardiectomy decreased with increasing MELD score; MELD scores ≤7.5, 7.51–15.5, and > 15.5 were associated with overall survival rates of 92.9 %, 69.8 %, and 8.3 %, respectively [3]. In this case, the perioperative risk was discussed because the Child-Pugh and MELD scores were 8 and 11 points, respectively. In patients with cirrhosis, a Child-Pugh score > 7 or MELD score > 13.5 is a contraindication for cardiac surgery [4]. However, if the condition is expected to improve with treatment, such as in the case of liver congestion, surgical intervention may be recommended, despite the perioperative risks. Before surgery, it is important to evaluate whether the liver dysfunction is reversible or irreversible.

Liver biopsy is the gold standard for the diagnosis of liver fibrosis. The stages of liver fibrosis are classified according to the new Inuyama classification: F0, no fibrosis; F1, fibrosis with portal expansion; F2, bridging fibrosis with portal-portal or portal-central linkage; F3,



bridging fibrosis with lobular distortion and disorganization; and F4, cirrhosis. Liver biopsy is an invasive procedure; thus, elastography has emerged as a noninvasive alternative for assessing liver condition [5], which can be categorized into two primary methods: shear wave and strain imaging.

Shear wave imaging enables measurement of the shear wave velocity [Vs (m/s)]. It increased with the progression of liver fibrosis [6]. However, shear wave imaging has limitations as it is influenced not only by liver fibrosis, but also by inflammation, jaundice, and congestion.

Strain imaging quantifies the liver stiffness based on the extent of tissue strain during compression. As liver fibrosis progresses, localized variations in the liver tissue stiffness emerge, leading to speckled image patterns. Liver fibrosis can be objectively assessed using the LFI derived from strain imaging. Yada et al. established a relationship between LFI and fibrosis stage determined through liver biopsy, demonstrating a progressive increase in LFI with higher fibrosis stage [7]. Unlike shear wave imaging, strain imaging remains unaffected by inflammation, jaundice, or congestion, making it a reliable method for accurately assessing liver fibrosis.

Combinational elastography assesses liver pathology by simultaneously performing these two types of measurements [8]. On strain imaging, a low LFI indicates the absence of liver fibrosis. If shear wave imaging results of Vs show elevation in the same patient, it could be indicative of inflammation, jaundice, or congestion rather than liver fibrosis. In addition, when inflammation and jaundice are clinically absent, increased Vs may indicate liver congestion. The liver status can be estimated by subtracting the information obtained from shear wave and strain imaging. Combinational elastography has also been reported to be useful for patients with heart failure [9], which can be used to assess the stages of liver congestion and associated liver fibrosis in patients with heart failure.

In our case, Vs was 2.79 m/s before pericardiectomy, indicating F4 fibrosis [6]. However, the LFI was 0.433, corresponding to F0–F1, suggesting the absence of liver fibrosis [7]. Clinical assessments revealed no contribution from inflammation or jaundice, and elevated Vs was considered to reflect liver congestion. Perioperative risks due to liver dysfunction were discussed before surgery. Based on the findings of combinational elastography, it was determined that the liver condition did not progress to fibrosis and that liver dysfunction resulted from reversible liver congestion. We hypothesized that improvement in right-sided heart failure through pericardiectomy would also lead to a decline in Vs values (i.e. liver congestion). Combinational elastography is a valuable diagnostic tool for assessing the suitability of invasive surgical interventions.

Tricuspid regurgitation is a common cardiac condition that can result in liver congestion due to right-sided heart failure. Chen et al. assessed preoperative liver stiffness through transient elastography in left-sided valve surgery with tricuspid annuloplasty, revealing its independent prognostic value and reversibility if tricuspid regurgitation does not recur within a year [10]. However, this evaluation relied on shear wave imaging alone, and the intertwined pathophysiology of liver fibrosis, congestion, inflammation, and jaundice must be considered. There have

been no reports of preoperative evaluation using combinational elastography for right heart failure, including tricuspid regurgitation and constrictive pericarditis, and further investigation is needed. Furthermore, cardiac catheterization was not available postoperatively in this case. Future studies using combinational elastography and cardiac catheterization may be warranted.

In conclusion, preoperative combinational elastography may enable the simple and noninvasive assessment of liver status in the perioperative management of constrictive pericarditis complicated by liver dysfunction.

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## Consent statement

Written informed consent was obtained from the patient.

## Declaration of competing interest

K.T. is a member of the *Journal of Cardiology Cases* Editorial Team. The authors declare that there is no conflict of interest.

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