

Refractory Ascites as the Only Presenting Symptom of Chronic Calcified Constrictive Pericarditis: A Diagnostic Challenge

Shi-Min Yuan

Department of Cardiothoracic Surgery, The First Hospital of Putian, Teaching Hospital, Fujian Medical University, Putian, Fujian Province, China

ABSTRACT

Extracardiac manifestations of constrictive pericarditis, such as massive ascites and liver cirrhosis, often cover the true situation and lead to a delayed diagnosis. A young female patient was referred to this hospital due to a 4-year history of refractory ascites as the only presenting symptom. A diagnosis of chronic calcified constrictive pericarditis was eventually established based on echocardiography, ultrasonography, and computed tomography. Cardiac catheterization was not performed. Pericardiectomy led to relief of her ascites. Refractory ascites warrants thorough investigation for constrictive pericarditis.

INTRODUCTION

Tuberculosis is a frequent cause of constrictive pericarditis, accounting for 61% of cases [Bashi 1988]. Alternative causative etiologies of the current era include previous heart operation, pericarditis, and radiation therapy [Ling 2000]. Patients with constrictive pericarditis usually manifest circulatory symptoms such as chest pain [Ling 2000; Goyle 2002], dyspnea [Ling 2000], or congestive heart failure [Butany 2004]. Extracardiac manifestations of constrictive pericarditis, including refractory ascites and hepatic cirrhosis, often cover the true situations of the patients and lead to delayed diagnoses [Meszaros 2013]. I describe the diagnostic dilemma of a patient presenting with refractory ascites as an only symptom of constrictive pericarditis.

CASE REPORT

A young 22-year-old female patient was referred to this hospital for treatment of a 4-year refractory ascites. On examination, her vital signs were normal, with no cardiac murmur

or muffled sounds audible. There was no jugular venous distention, paradoxical pulse, or peripheral edema. Her medical history was significant for a history of lung tuberculosis. Abdominal paracentesis and drainage resulted in the shedding of 5 L of light-yellow ascites. Her erythrocyte sedimentation rate was 28 mm/h, but the values for C-reactive protein and D-dimer were within normal ranges. Laboratory work-ups also showed mild hepatic function impairment (total bilirubin, 23.7 $\mu\text{mol/L}$; direct bilirubin, 12.3 $\mu\text{mol/L}$; alanine transaminase, 48 U/L; aspartate transaminase, 49 U/L; and γ -glutamyl transpeptidase 104 U/L) and a high serum-ascites albumin gradient (1.5 g/dL). A low voltage of the QRS complex in all leads was noted in the electrocardiogram. Echocardiography showed normal activities of the ventricular wall and ventricular septum, normal left ventricular ejection fraction (60%), dilation of the left atrium (52 mm) and right atrium (54 mm), mild dilation of the right ventricle (25 mm), a faint bright band along the side of the left heart chambers (Figure 1A), a slightly decreased flow rate in the portal vein (13 cm/s) (Figure 1B), a normal hepatic vein with bidirectional flows (Figure 1, C and D), and a dilated inferior vena cava (Figure 1E). Ultrasonography demonstrated normal liver dimensions, splenomegaly (with a length of 138 mm and a thickness of 51 mm) with a mildly dilated splenic vein (10 mm) (images were not available), and massive ascites (Figure 1, F-J). Computed tomography revealed remarkably thickened fibrous and calcified pericardium over the ventricles (Figure 2A), a dilated inferior vena cava (40 mm), and some pleural effusions (Figure 2B). Abdominal computed tomography showed massive ascites, splenomegaly (with a length of 132 mm and thicknesses of 47 mm and 54 mm), and exomphalos (Figure 2, C and D). Malignant etiologies were ruled out by thorough investigations, and a diagnosis of chronic calcified constrictive pericarditis was established. The patient received a pericardiectomy without the aid of cardiopulmonary bypass.

The anterior and diaphragmatic aspects of the right heart along with the inferior vena cava were covered, and the right atrioventricular groove was tightly embedded by densely calcified pericardium (Figure 3), whereas the left heart was strictly restrained by the fibrous tissues. While the calcification was being peeled off, the right atrium ruptured and severe bleeding occurred. The right atrium was successfully repaired with direct sutures. The patient had a smooth postoperative

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Correspondence: Department of Cardiothoracic Surgery, The First Hospital of Putian, Teaching Hospital, Fujian Medical University, Putian 351100, Fujian Province, China (e-mail: shi_min_yuan@yahoo.com).

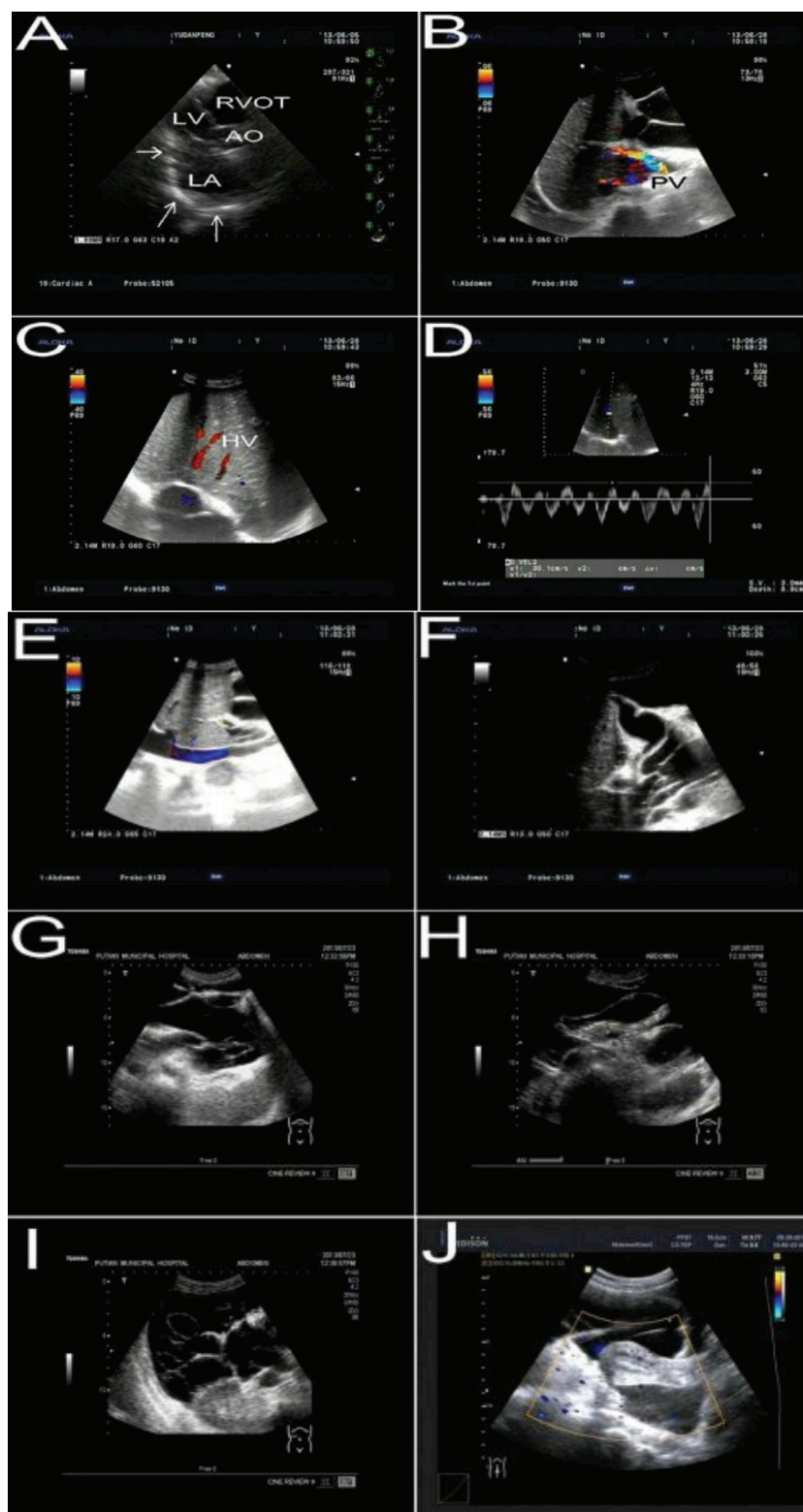


Figure 1. Echocardiography showed dilation of the atria and right ventricle and a faint bright band along the side of the left heart chambers (A), slightly decreased flow rate in the portal vein (13 cm/s) (B), a normal hepatic vein (C), bidirectional flow in the hepatic veins (D), and a dilated inferior vena cava (E). Ultrasonography demonstrated massive ascites near the gall bladder (F), liver (G), pancreas (H), and intestine (I) and in the uterine and the rectouterine pouch (J). AO indicates aorta; HV, hepatic veins; LA, left atrium; LV, left ventricle; PV, portal vein; RVOT, right ventricular outflow tract.

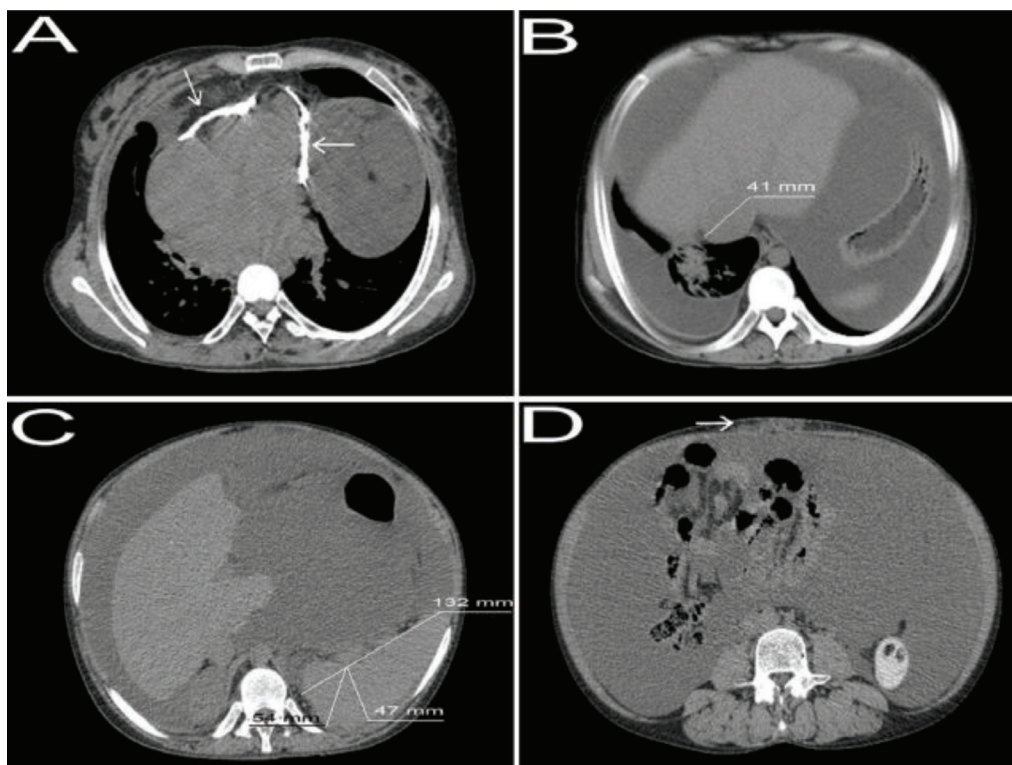


Figure 2. Chest computed tomography showed severely calcified pericardium over the ventricles (arrows) and biatrial dilations (A) and a significantly dilated inferior vena cava and some pleural effusions (B). Abdominal computed tomography showed massive ascites (C and D), splenomegaly (C), and an exomphalos (arrow) (D).

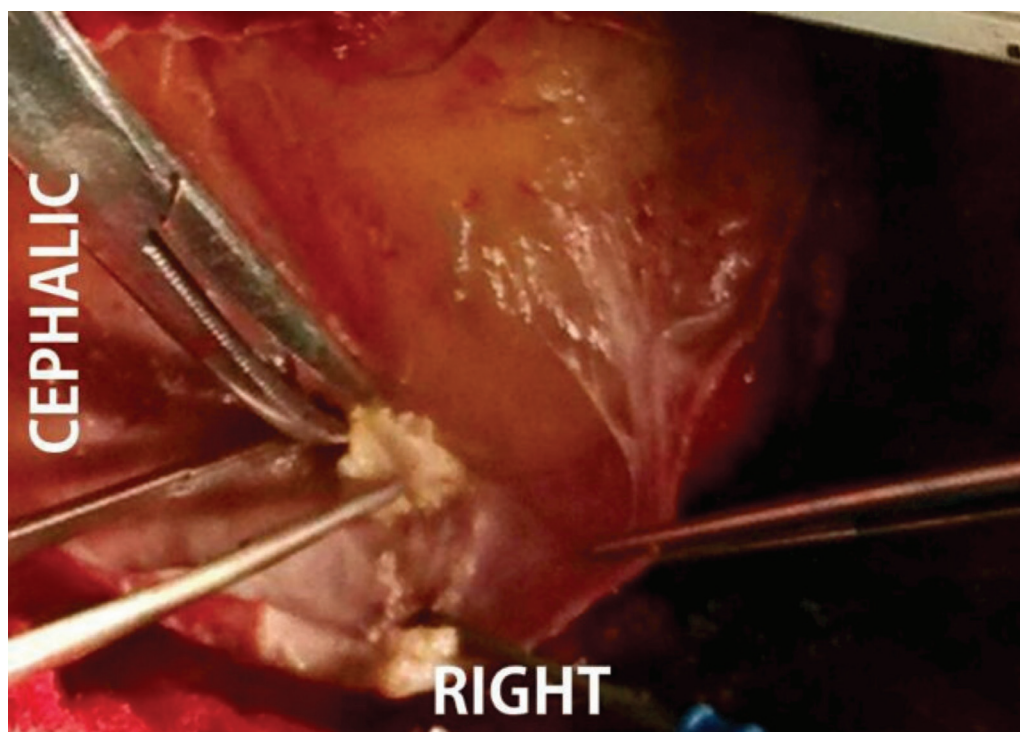


Figure 3. Pericardiectomy was performed to peel severe calcification of the right atrioventricular (AV) groove.

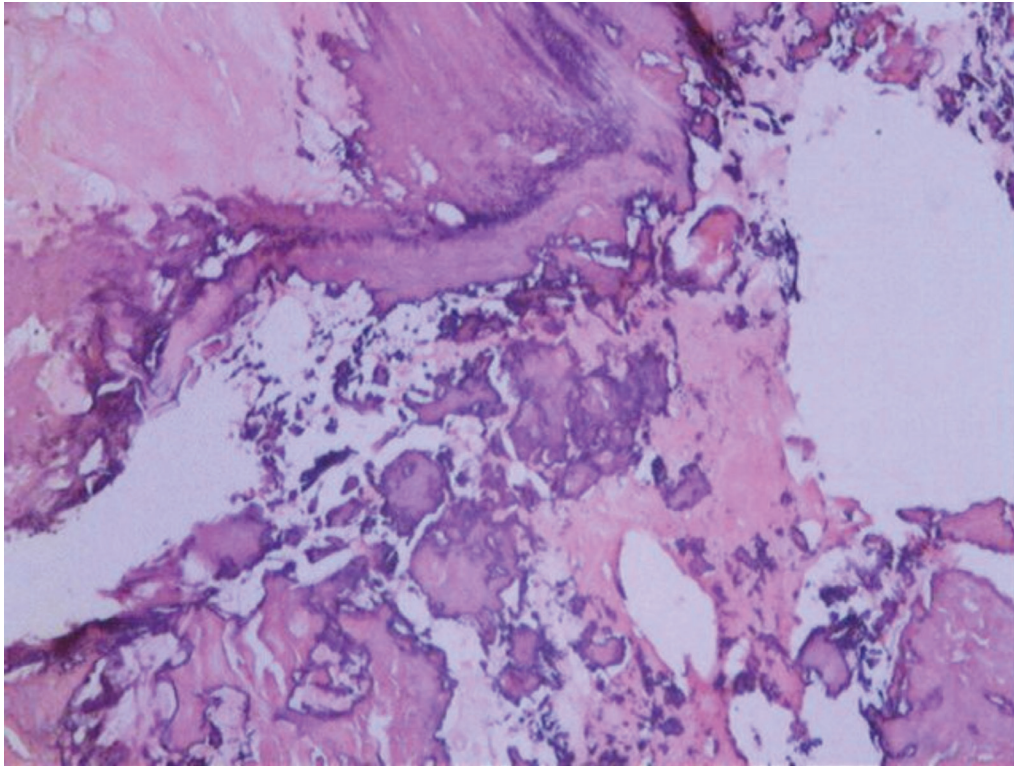


Figure 4. Histology of the resected pericardium showed dense collagen and calcification. Hematoxylin and eosin stain; magnification $\times 400$.

course with reduced ascites. Histology of the removed tissues revealed calcified pericardium (Figure 4). Ongoing regular antituberculosis therapy was advised. An exomphalos repair was expected to be carried out when her hepatic function became normal and her ascites diminished.

DISCUSSION

Constrictive pericarditis often develops in young adults [Bashi 1988]. Calcification of the pericardium may be coarse or fine and is most commonly located on the anterior and diaphragmatic aspects of the heart, covering the atrioventricular groove and right atrium [MacGregor 1987]. Clinical manifestations of constrictive pericarditis vary from patient to patient. Bashi et al. [Bashi 1988] reported that the striking clinical manifestations of constrictive pericarditis were hepatomegaly (100%), elevated jugular vein pressure (100%), muffled heart sounds (98%), ascites (90%), paradoxical pulses (84%), peripheral edema (84%), pleural effusion (54%), and splenomegaly (18%). The present patient was characterized clinically by refractory ascites, splenomegaly, low voltage of the QRS complex, and calcified pericardium, with refractory ascites being the only presenting symptom. Johnson et al. [Johnson 2008] reported that the manifestations of constrictive pericarditis visible on computed tomography were dilation of the inferior vena cava (80%), dilation of the hepatic veins (56%), ascites (56%), pericardial thickening (52%), and

pericardial calcification (40%). Constrictive pericarditis is pathologically manifested by pericardial thickening, pericardial effusion, lymphadenopathy, and calcification, and the calcification tends to be thick, irregular, amorphous, and located in the atrioventricular grooves [O'Leary 2010].

A serum-ascites albumin gradient ≥ 1.1 g/dL and an ascites total protein > 2.5 g/dL are typical of constrictive pericardial disease and showed 97% accuracy for diagnosing portal hypertension [Runyon 1991]. The high serum-ascites albumin gradient of this patient supported the diagnosis of constrictive pericarditis. It was demonstrated that the diagnostic accuracy of cardiac catheterization for constrictive pericarditis was 100% [Bashi 1988]. However, it was not performed as a preoperative diagnostic procedure in the present patient. Echocardiography may demonstrate abrupt changes of posterior wall and ventricular septal motion [Elkayam 1976; Kameda 2007]. Ultrasonography may disclose portal hypertension (portal vein dilation ≥ 13 mm, splenic vein dilation ≥ 11 mm, reduced portal vein flow, splenomegaly ≥ 12 mm, and umbilical vein recanalization) [Aagaard 1982; Berzigotti 2011]. Computed tomography may show significantly increased thickness or calcification of the pericardium [Lima 2011]. Tuberculosis bacilli can be found in the fluid and calcified debris [Bashi 1988]. Histology of the resected calcified pericardium showed a dense collagenous matrix, infiltrations of chronic inflammatory cells, and calcification [Tugcu 2008].

For constrictive pericarditis with extensive calcified pericardium, median sternotomy is the preferred approach for

performing surgery [Bashi 1988], whereas thoracotomy can be associated with high incidence of wound infections and pulmonary complications [Tiruvoipati 2003]. The usual extent of the calcified pericardium resection is up to the phrenic nerve [Lima 2011]. Pericardiectomy may improve the symptoms in a majority of the patients [Peset 2007]. The hospital mortality of constrictive pericarditis patients undergoing pericardiectomy ranged between 5% and 15% [Bashi 1988], and late mortality rates at 5 and 10 years were 1.6% and 9.7%, respectively [Cinar 2006]. The actuarial 5-year survival rate was 75.9% [Bozbuga 2003]. Poor preoperative heart function may indicate significantly decreased long-term survival [Szabó 2013].

Refractory ascites as an only presenting symptom of constrictive pericarditis, as in the present patient, is rare. However, portal hypertension signs were significant, including an elevated serum-ascites albumin gradient, reduced portal flow velocity, a dilated inferior vena cava, and splenomegaly.

In conclusion, extracardiac manifestations of constrictive pericarditis, such as massive ascites and liver cirrhosis, often cover the true situation and lead to a delayed diagnosis. Refractory ascites warrants thorough investigations for constrictive pericarditis. Pericardiectomy is a radical treatment method for constrictive pericarditis.

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