

Constrictive Pericarditis and Protein-Losing Enteropathy Is Extremely Severe Hypoalbuminemia Reversible by Pericardiectomy?

Hidenori Moriyama, MD; Takashi Kohno, MD, PhD; Takahiko Nishiyama, MD, PhD; Osamu Hattori, MD; Yuichiro Maekawa, MD, PhD; Kosuke Yoshida, MD; Mitsushige Murata, MD, PhD; Kazuma Okamoto, MD, PhD; Motoaki Sano, MD, PhD; Hideyuki Shimizu, MD, PhD; Keiichi Fukuda, MD, PhD

Protein-losing enteropathy is characterized by excessive loss of serum proteins into the gastrointestinal tract and is a rare complication of constrictive pericarditis.¹ Pericardiectomy, although a radical treatment, is the definitive therapy for constrictive pericarditis. Despite the effectiveness of pericardiectomy in treating constrictive pericarditis, its effectiveness in reversing extremely severe hypoalbuminemia with protein-losing enteropathy has not been established. In addition, treating physicians may question whether such cases are beyond the treatment window for pericardiectomy. Here, we report a case of protein-losing enteropathy caused by constrictive pericarditis, in which pericardiectomy lead to remarkable improvement in extremely severe hypoalbuminemia.

Case Report

A 38-year-old male patient who had previously undergone mitral valve replacement for severe mitral regurgitation and infective endocarditis 12 years prior presented with progressive diarrhea, edema of both lower extremities, and significant weight gain over the past 6 years. In addition to hypoalbuminemia without nephrotic syndrome, liver cirrhosis, or malnutrition, technetium 99 m–labeled human serum albumin-diethylene-triaminepenta-acetic acid abdominal scintigraphy revealed accumulation of radionuclide in the intestines (Figure [A]). He was diagnosed with protein-losing enteropathy and was admitted to our hospital for further investigation. A complete workup of the gastrointestinal tract with upper gastrointestinal endoscopy, small intestine endoscopy, and colonoscopy with multiple biopsies revealed no abnormalities. Lymphangiography also showed no abnormal findings, and contrast-enhanced computed tomography revealed bilateral pleural effusion, ascites and thickened pericardium (Figure [B]). Transthoracic echocardiography showed a thickened pericardium with calcifications beside the inferior and posterior left ventricular wall, marked enlargement of the right and left atria, mild mitral valve regurgitation without prosthetic malfunction, normal ventricular cavity size, and preserved ejection fraction. A septal bounce and septal dip in early diastole (Figure [C]), as well as pronounced respiratory variation

in ventricular filling, were observed in M-mode and Doppler echocardiography, respectively. Hemodynamically, mean right atrial pressure (16 mm Hg) and diastolic right ventricular pressure (17 mm Hg) were markedly increased. Simultaneous pressure recording of the right and left ventricle revealed equalization of end-diastolic pressures with dip and plateau patterns, with respiration-dependent discordance in right and left ventricular systolic pressures (Figure [D]). Based on these findings, a diagnosis of constrictive pericarditis was established, and it was considered to have caused protein-losing enteropathy.

The patient's serum total protein and albumin levels were significantly low on admission (total protein 3.1 g/dL, albumin 0.7 g/dL), and his systemic edema did not improve with treatment with oral diuretics including furosemide and tolvaptan. Pericardiectomy was performed successfully, and the patient had an uneventful postoperative recovery. Diarrhea ceased completely soon after the surgery. In a follow-up examination 3 months later, the patient reported no symptoms; lower extremity edema and pleural effusion had completely resolved. Laboratory data showed that serum total protein and albumin levels had returned to normal (total protein 7.6 g/dL, albumin 4.0 g/dL).

Discussion

A PubMed search of articles published in English until August 2016 using the key words constrictive pericarditis, protein-losing enteropathy, and pericardiectomy retrieved 9 papers: 6 case reports and 3 case series. Among them, serum albumin levels are provided in only 3 articles¹⁻³; we have summarized the recorded changes in serum albumin level after pericardiectomy in the Table. All 3 previously published cases reported improvement in serum albumin levels after surgery. However, hypoalbuminemia was also identified as an independent predictor of mortality after a pericardiectomy,⁴ suggesting that the decision to operate can be a delicate balance of the risk/benefit expected from the pericardiectomy in the presence of hypoalbuminemia. Even though our patient had extremely severe hypoalbuminemia (0.7 g/dL) compared with previously

From the Department of Cardiology (H.M., T.K., T.N., O.H., Y.M., M.M., M.S., K.F.), Department of Cardiovascular Surgery (K.O., H.S.), and Department of Gastroenterology (K.Y.), Keio University School of Medicine, Tokyo, Japan.

Correspondence to Takashi Kohno, MD, PhD, Department of Cardiology, Keio University School of Medicine, Shinanomachi 35, Shinjuku-ku, Tokyo, 160-8582, Japan. E-mail kohno.a2@keio.jp

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Table. Case Reports of Constrictive Pericarditis With Protein-Losing Enteropathy

Author (year)	Case: Age, Sex	Cause	Serum Albumin Level		References
			Before Surgery	After Surgery	
Müller et al (1991)	41, men	TB pericarditis	2.4	3.2	¹
Nikolaidis et al (2005)	76, men	TB pericarditis	2.3	3.8	²
Meijers et al (2006)	74, men	post CABG	1.8	3.1	³
Moriyama et al	38, men	post MVR	0.7	4.0	

Summary of case reports on constrictive pericarditis with protein-losing enteropathy published after 1990. CABG indicates coronary artery bypass graft; MVR, mitral valve replacement; and TB, tuberculous.

reported cases, his serum albumin increased to a level within normal range after the pericardiectomy. Both our results and previous data suggest that patients with constrictive pericarditis may be treated with pericardiectomy successfully even in cases of extremely severe hypoalbuminemia because of protein-losing enteropathy.

Distinguishing hypoalbuminemia that will reverse with pericardiectomy (pericardiectomy-responsive hypoalbuminemia) from hypoalbuminemia that will not (pericardiectomy-independent hypoalbuminemia) can be challenging. Besides the loss of serum proteins into the gastrointestinal tract, other mechanisms resulting in hypoalbuminemia should be considered, including hemodilution, liver dysfunction (liver cirrhosis), inflammation, malnutrition, or cachexia, resulting from heart failure or non-heart failure-related illness. Although pericardiectomy is effective in patients with constrictive pericarditis complicated by severe hypoalbuminemia because of protein-losing enteropathy, the ultimate decision to operate is best made in conjunction with a healthcare team including gastroenterologists, hepatologists, and cardiovascular surgeons.

Disclosures

None.

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KEY WORDS: hypoalbuminemia ■ pericardiectomy ■ pericarditis, constrictive ■ protein-losing enteropathies ■ serum proteins

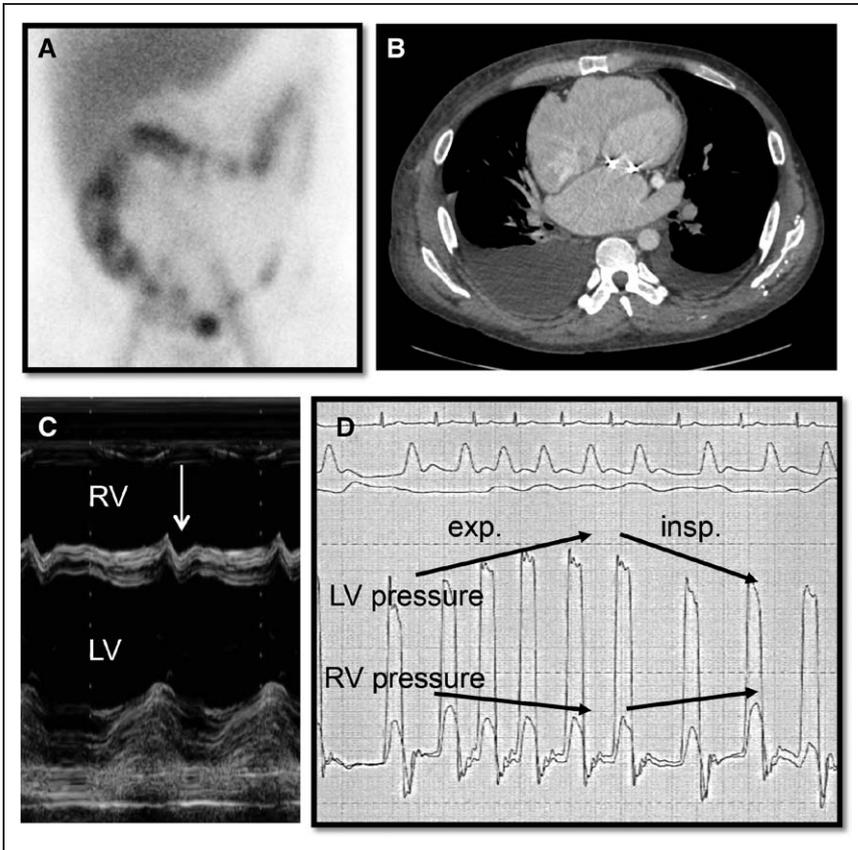


Figure. Imaging studies. **A**, Abdominal scintigraphy 6 h after intravenous administration of technetium-99 m–labeled human serum albumin, in which clear colonic tracer accumulation is present. **B**, Contrast-enhanced computed tomography revealing bilateral pleural effusion and thickened pericardium. **C**, Septal dip (arrow) in early diastole in M-mode echocardiography. **D**, Simultaneous pressure recording of the right and left ventricles (RV and LV, respectively) revealing equalization of the end-diastolic pressures with dip and plateau patterns, with respiration-dependent discordance in biventricular systolic pressures. exp. indicates expiration; and insp., inspiration.

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