

IMAGING VIGNETTE

CLINICAL VIGNETTE

Not So Transient Effusive-Constrictive Pericarditis



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ABSTRACT

A 63-year-old male patient presented with chest pain and signs of volume overload. His work-up revealed a diagnosis of transient effusive constrictive pericarditis of idiopathic etiology. Despite treatment with optimal medical therapy, he continued to experience persistent symptoms eventually requiring radical pericardiectomy. (J Am Coll Cardiol Case Rep 2023;28:102088) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

A 63-year-old male patient presented for a 5-month history of chest pain, dyspnea, orthopnea, abdominal fullness, bilateral lower extremity edema, and 7-pound weight gain. Past medical history includes coronary artery disease for which he underwent stent placement in his left anterior descending and left circumflex arteries, paroxysmal atrial fibrillation, hypertension, and hyperlipidemia. Physical examination was pertinent for hypotension with a blood pressure of 92/54 mm Hg, jugular venous distention to the level of the mandibular angle, positive Kussmaul's sign, presence of a pericardial friction rub, and grade 2+ edema in the bilateral lower extremities.

Laboratory studies showed elevated N-terminal pro-B-type natriuretic peptide of 1,282 pg/mL (normal <125 pg/mL) and elevated ultra-sensitive C-reactive protein of 14.8 mg/L (normal <3.1 mg/L). Transthoracic echocardiography showed a large pericardial effusion, septal shift with inspiration, annulus reversus, hepatic vein expiratory diastolic flow reversal, and 27% and 40% respiratory variation across the mitral and tricuspid valves, respectively (**Figure 1A**). Right heart catheterization showed diastolic pressure equalization across all chambers as well as a systolic area index >1.1. Cardiac magnetic resonance showed increased pericardial thickness measuring 5 mm, severe circumferential increased pericardial signal on late gadolinium enhancement, and diffuse increased pericardial signal on T2 short tau inversion recovery imaging (**Figure 1B**). Respirophasic interventricular dependence was present on cine imaging. Based on these findings, a diagnosis of effusive-constrictive pericarditis (ECP) was made.

The patient was initially treated with ibuprofen and colchicine, however, he continued to have persistent signs and symptoms, for which biologics including anakinra to rilonacept, and torsemide were added. Due to inadequate clinical response, prednisone was then added to the regimen of rilonacept, colchicine and ibuprofen. Despite medical therapy, the patient decompensated further and ultimately underwent radical visceral and parietal pericardiectomy with Waffle procedure. Intraoperative findings demonstrated a hemorrhagic pericardial effusion with very thick visceral and parietal pericardium with encasement of the heart within the visceral pericardium (**Figure 1C**). Surgical pathology of the pericardium showed organizing fibrinous

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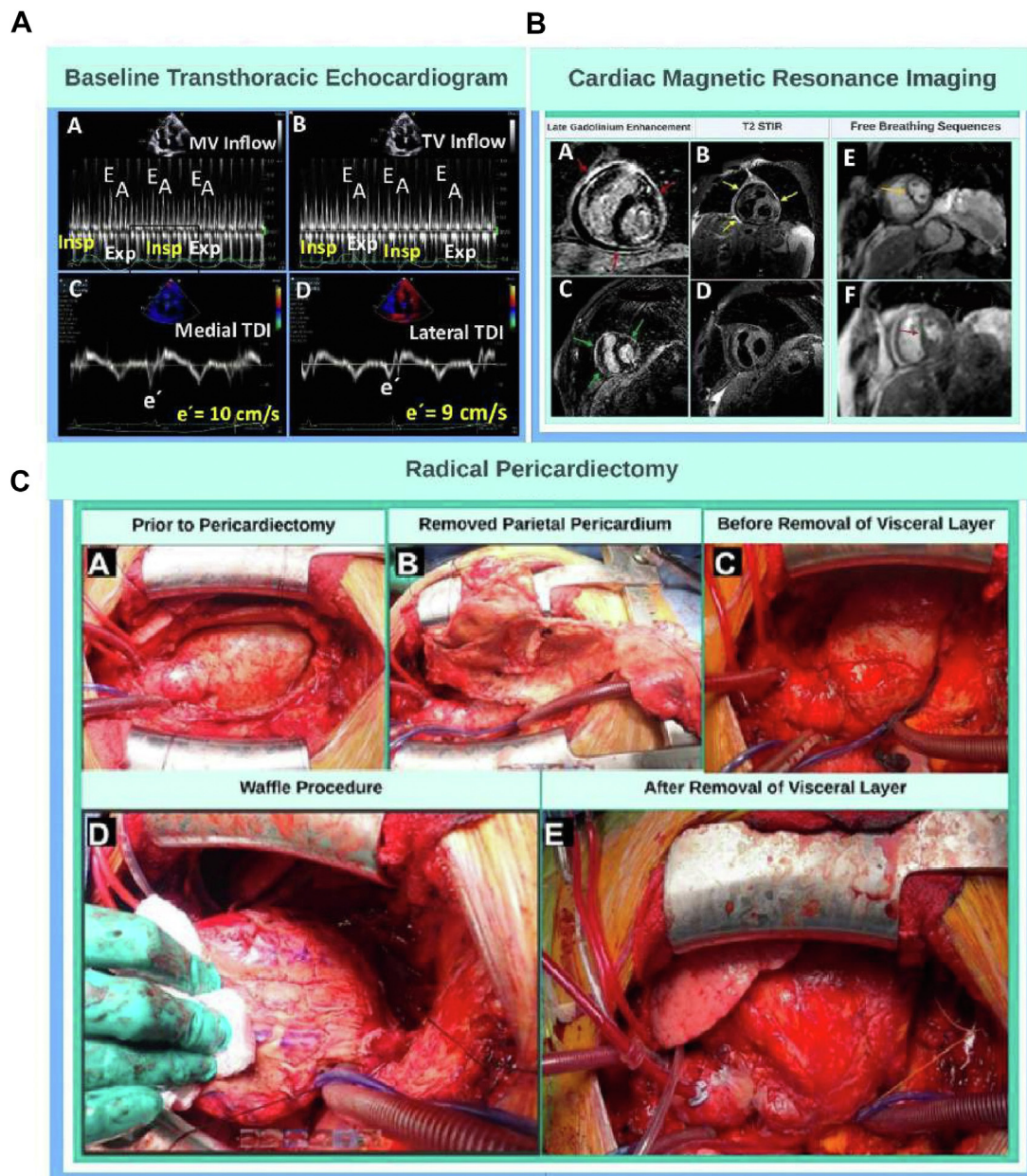
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**ABBREVIATIONS
AND ACRONYMS****ECP** = effusive-constrictive
pericarditis

pericarditis. Bacterial, fungal, and anaerobe cultures were negative, and the patient was diagnosed with idiopathic transient ECP. On follow-up 1 month after pericardiectomy, the patient had marked improvement of his symptoms.

ECP is a rare syndrome first described in 1954 and is characterized by constriction of the heart by the visceral pericardium and fluid accumulation in the pericardial space in the setting of an inelastic

FIGURE 1 Imaging and Gross Surgical Findings Demonstrating Transient Effusive Constrictive Pericarditis

(A) Transthoracic echocardiography demonstrating respiratory variation across (a) the mitral valve (MV) and (b) the tricuspid valve (TV). (C, d) Tissue Doppler imaging revealing annulus reversus with medial e' of 10 cm/s and lateral e' of 9 cm/s. (B) Cardiac magnetic resonance demonstrating (a) moderate circumferential pericardial late gadolinium enhancement and (b) pericardial edema on T2 STIR. There was a mild interval decrease on (c) late gadolinium enhancement and (d) T2 STIR imaging in the subsequent cardiac magnetic resonance. (e, f) Free-breathing sequences demonstrating respiratory variation. (C) Radical pericardiectomy showing (a) thick pericardium tacked on both sides, (b) removed parietal pericardium, (c) before removal of the visceral layer, (d) Waffle procedure, and (e) after removal of the visceral layer. e' = early diastolic mitral annular velocity; STIR = short tau inversion recovery; TDI = tissue Doppler velocity.

pericardium, causing a tamponade-like effect.¹ If there is active inflammation associated with constriction, the condition is deemed transient since anti-inflammatories can help with the symptoms. The clinical presentation is often characterized by jugular venous distention, abdominal distention, lower extremity swelling, a pericardial friction rub, pulsus paradoxus, and Kussmaul's sign.¹ Transthoracic echocardiography can show a pericardial effusion, respirophasic septal shift, inferior vena cava dilation with plethora, exaggerated respiratory variation of mitral and tricuspid valve inflow velocities, and annulus reversus.² Cardiac magnetic resonance can show increased signal intensity on T2 short tau inversion recovery and late gadolinium enhancement imaging indicative of pericardial edema and inflammation.² Cardiac catheterization can show equalization of diastolic pressures, as well as persistent elevation of right atrial pressure following pericardiocentesis.² Anti-inflammatory medications are curative in almost all cases, and pericardiectomy is pursued in cases that persist despite medical treatment.² In this case, the Waffle procedure was employed in which the pericardium is incised longitudinally and transversally where the risk of coronary injury is high.³

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