

A Malignant Case of Constrictive Pericarditis



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INTRODUCTION

Primary pericardial tumors are rare conditions that can mimic classic constrictive pericarditis. Multimodality imaging can provide enhanced tissue characterization of the pericardium, which improves noninvasive diagnostic performance, particularly in rare cases, such as primary pericardial tumors, presenting with constrictive pathophysiology.

CASE PRESENTATION

A 74-year-old man presented to the emergency department with a 3-month history of progressive dyspnea, fatigue, and pedal edema. His past medical history is pertinent for smoking, Hodgkin's lymphoma treated with radiation therapy, and prostate cancer treated with radical prostatectomy. Cardiopulmonary examination revealed jugular venous distension with positive Kussmaul's sign, decreased breath sounds at the right lung base, an irregular cardiac rhythm, an early peaking 2/6 systolic ejection murmur over the right upper sternal border, and an absent pericardial knock. There was peripheral pitting edema, along with hepatomegaly.

An initial electrocardiogram demonstrated atrial fibrillation with a ventricular response rate of 120 beats per minute. Chest x-ray revealed a right-sided pleural effusion and cardiomegaly without pericardial calcification. Chest computed tomography (CT) with contrast demonstrated a circumferential organized pericardial collection (Figure 1). On CT, the mean density of the right-sided simple pleural effusion was ~ 7 Hounsfield units. In contrast, the density of the pericardial collection was higher, with a mean value of ~ 30 – 45 Hounsfield units, suggesting the presence of a highly proteinaceous fluid collection. A limited transthoracic echocardiogram confirmed the circumferential pericardial effusion, as shown in Figure 2A-D and Video 1. Respiratory variation in transmitral and transtricuspid flow is demonstrated in Figure 3, while annulus reversus on tissue Doppler imaging is demonstrated in Figure 4. Subxiphoid pericardiocentesis (Video 2) yielded 1 L of bloody fluid, with scant reactive mesothelial cells but no malignant cells.

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Subsequently, the patient was discharged on beta-blockers, diuretics, nonsteroidal anti-inflammatory analgesia, and colchicine. Over the next few months, he was readmitted to the hospital on several occasions with debilitating shortness of breath and signs of volume overload. A subsequent transthoracic echocardiogram, 6 months after the initial echocardiogram, revealed a circumferential pericardial effusion, diastolic septal bounce, and mild aortic stenosis with calcification of the aortomitral continuity. Myocardial strain imaging revealed reduced peak systolic strain in the anterolateral left ventricular (LV) segments (-13% to -17%), with preserved peak systolic strain in anteroseptum (-24% ; Figure 5). Transesophageal echocardiogram performed prior to cardioversion during an episode of atrial fibrillation was again notable for a circumferential pericardial collection (Video 3A-C).

Cardiac magnetic resonance imaging (CMR) was performed to further investigate the pericardial collection. It demonstrated decreased signal intensity in the pericardial collection, compared with pleural effusions on steady-state free-precession (SSFP) sequences before gadolinium administration (Figure 6A, Videos 4A and 5A). After gadolinium administration, the signal intensity within the pericardial collection increased (Figure 6B, Videos 4B and 5B), suggesting hypervascularity. Further tissue characterization achieved with fat saturation T2-weighted imaging revealed a hyperintense signal throughout the pericardial collection, supporting the diagnosis of a pericardial tumor (Figure 7). Furthermore, delayed gadolinium enhancement

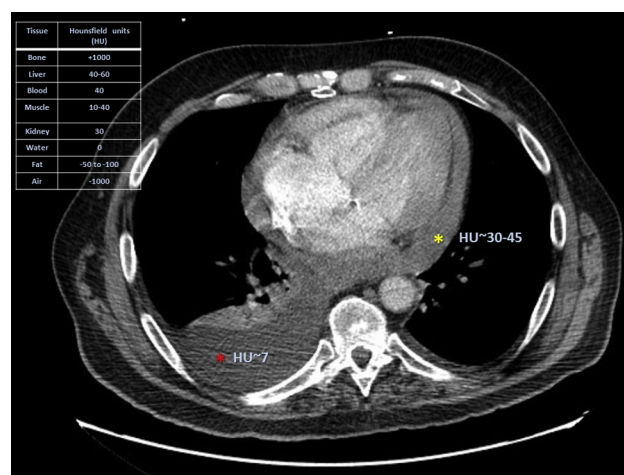


Figure 1 Chest CT with contrast showing a circumferential organized pericardial collection. The density of the right-sided simple pleural effusion had a mean value of ~ 7 Hounsfield units (*red asterisk*). In contrast, the density of the pericardial collection was higher, with a mean value of ~ 30 – 45 Hounsfield units (*yellow asterisk*), suggesting the presence of highly proteinaceous fluid collection.

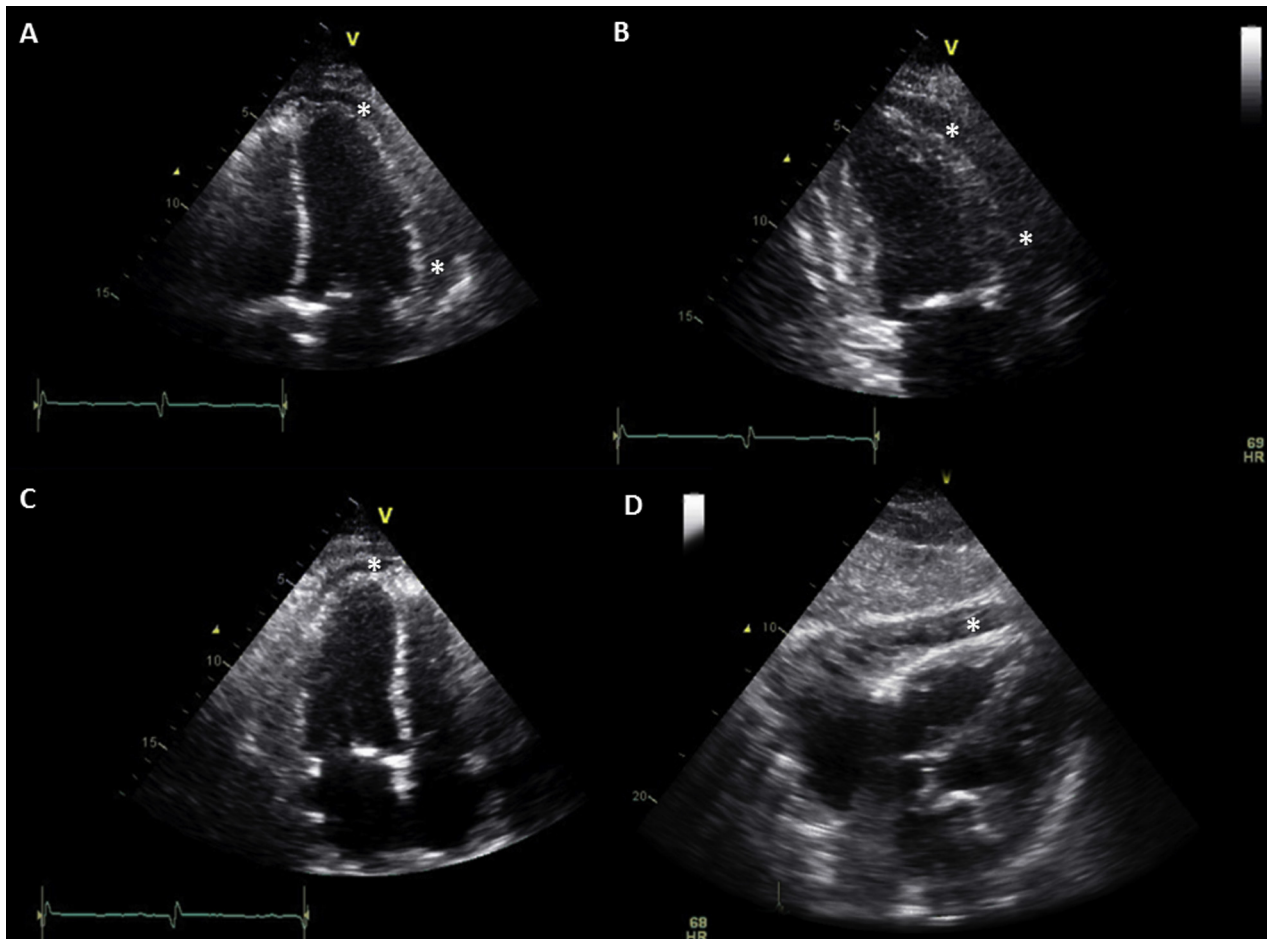


Figure 2 Transthoracic echocardiogram showing a circumferential organized pericardial effusion (asterisk) in the four-chamber (A), two-chamber (B), three-chamber (C), and subcostal views (D).

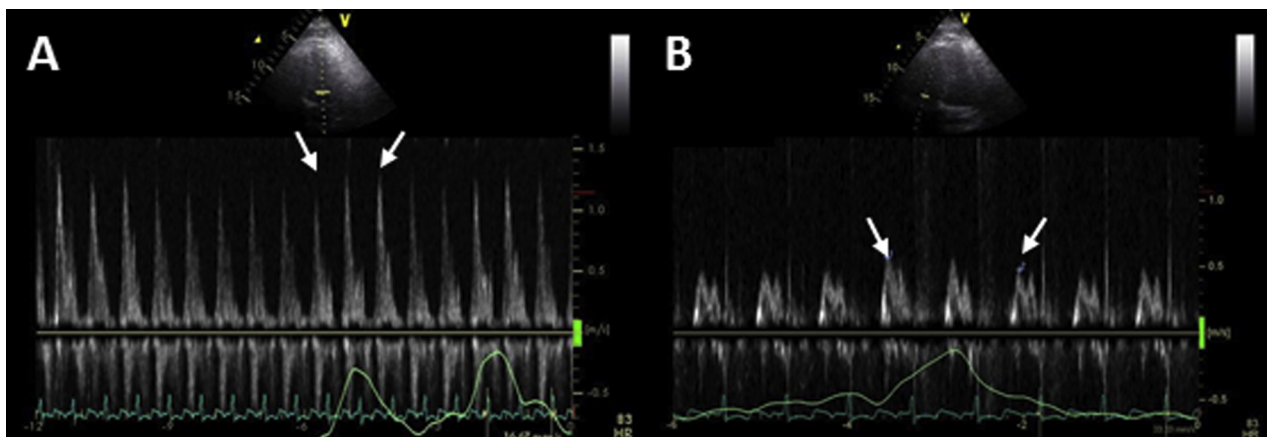


Figure 3 (A) Transmitral pulsed wave Doppler at the level of the mitral leaflet tips with respirometry tracing seen in the second half of the clip, demonstrating significant respiratory variation of more than 25% when comparing the first beat of inspiration with the first beat of expiration (arrows). (B) Transtricuspid pulse wave Doppler at low speed with sample volume at the level of the tricuspid leaflet tips with suboptimal respirometry tracing seen, attempting to demonstrate a significant respiratory variation of more than 40% when comparing the first beat of inspiration with the first beat of expiration (arrows).

images revealed increased though heterogeneous signal intensity, further supporting the diagnosis of a soft-tissue tumor (Figure 8). Additionally, there were classic features of constrictive patho-

physiology as demonstrated by the free-breathing SSFP sequence (Video 6), where the circumferential pericardial tissue results in significant respirophasic septal shift.

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