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Case Report

Catastrophic Cardiac Amyloidosis

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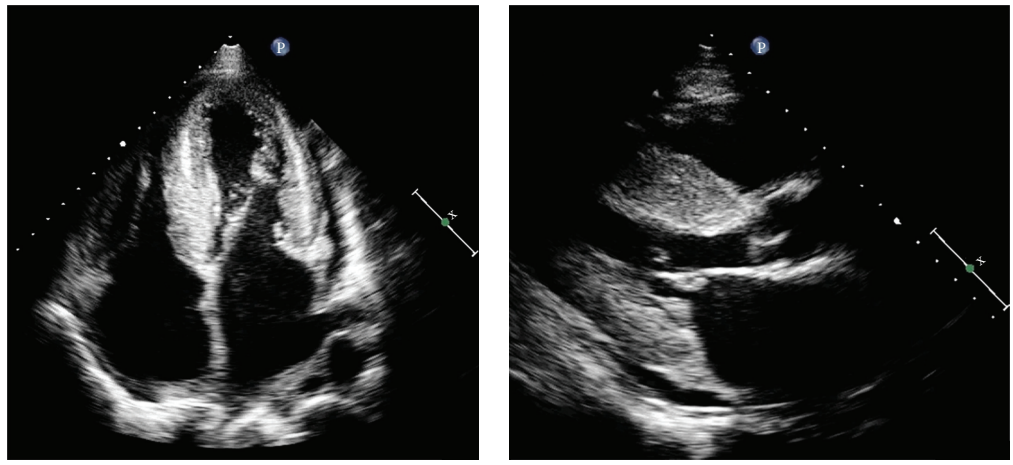
We report a case of a 61-year-old patient presenting with cardiogenic shock. His echocardiogram suggested typical features of cardiac amyloidosis. This case demonstrates that cardiac amyloidosis can present acutely and may be catastrophic.

1. Case Report

A 61-year-old Iranian male, nondiabetic, and nonhypertensive, presented with sudden onset syncope. He had history of exertional dyspnea for two months, being investigated in private clinics. On arrival, he was profusely sweating, afebrile with cold clammy peripheries. Clinically, he was in shock with blood pressure of 55/30 mmHg and pulse 110 beats per minute. His complete blood count was normal, but serum creatinine was 449 mmol/L with urea 25 mmol/L. His Troponin T was 0.9 μg/L and N-terminal pro-B-type natriuretic peptide (BNP) was 978 pg/mL. His ECG showed sinus rhythm with low voltage complexes without ischemic changes. Bedside transthoracic echocardiogram done showed nondilated ventricles with good biventricular function, but there was severe concentric left ventricular wall thickening with "sparkling" appearance, mitral valve leaflet and interatrial septal thickening, biatrial enlargement, and mild circumferential pericardial effusion (Figure 1). All these features were typical of cardiac amyloidosis. Doppler mitral inflow showed restrictive pattern. Tissue Doppler imaging of mitral annulus (MA) showed decreased systolic longitudinal velocity (**S′**) (septal MA 4.1 cm/s, lateral MA 5.3 cm/s) and decreased early diastolic longitudinal velocity (**E′**) (septal MA 4.96 cm/s and lateral 4.29 cm/s) with **E**/**E′** ratio of 13 (Figure 2). He was treated with intravenous fluids, inotropes, but within few hours, he went into pulseless electrical activity and expired.

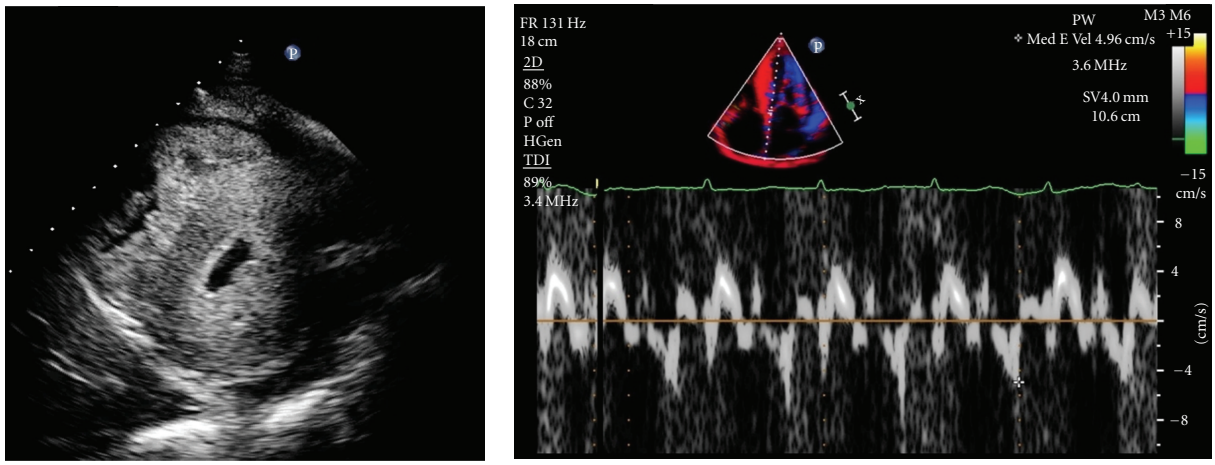
2. Discussion

Cardiac amyloidosis is commonly seen in systemic (primary) AL amyloidosis and senile systemic amyloidosis. In 90% of AL patients, heart is affected pathologically, and in 50% of whom, diastolic dysfunction with physical signs of right heart failure is a presenting feature. Conversely, <5% of patients with AL amyloidosis involving the heart have clinically isolated cardiac disease, but most frequently they have features of multiorgan dysfunction [1]. This was seen in our patient who primarily presented with cardiac amyloidosis but also had renal impairment. In a study, median age at presentation was 59 years (range 29–85), and AL cardiac disease was unusual both in patients under the age of 40 (3.0%) and in non-Caucasians (6.5%) [1]. Case-control studies indicate that echocardiographic evidence of left ventricular wall thickening, biatrial enlargement, and increased echogenicity in conjunction with reduced electrocardiographic voltages is strongly suggestive of cardiac amyloidosis [2]. Myonecrosis and small-vessel ischemia due to amyloid deposit cause an increase in cardiac troponins [3], whereas diastolic dysfunction and increased genetic expression of natriuretic peptide genes in the amyloid infiltrated ventricles cause increase in plasma BNP levels [4]. The diagnosis of AL amyloidosis requires demonstration of amyloid in tissue (either by fine-needle aspiration of abdominal fat or rectum or organ biopsy) and demonstration of a plasma cell dyscrasia (either by bone marrow biopsy showing predominance of Kappa or Lambda light chain producing



(a) (b)

Figure 1: Transthoracic echocardiography in apical 4-chamber view (a) and parasternal view (b), demonstrating a small left ventricle with severe concentric left ventricular wall thickening with "sparkling" appearance, mitral valve leaflet and interatrial septal thickening, biatrial enlargement, and mild circumferential pericardial effusion in patient with cardiac amyloidosis.



(a) (b)

Figure 2: Transthoracic echocardiography in short-axis view (a) demonstrating severe concentric left ventricular wall thickening with "sparkling" appearance in a patient with cardiac amyloidosis. Tissue Doppler imaging (b) illustrating reduced systolic and early diastolic velocities at septal mitral annulus in a patient with cardiac amyloidosis.

plasma cells or by the presence of a monoclonal Ig light chain in the serum or urine electrophoresis and/or elevated serum-free light chain by immunoassay). These tests could not be done in this patient as he had a rapid downhill course. The prognosis is usually poor, with a median survival of 8 months after the onset of heart failure [1], specifically in patients with raised troponins and BNP levels [2]. The exact cause for cardiogenic shock in this patient with preserved biventricular systolic function is not known. Amyloid infiltration of heart affects contractility, electric conduction, and coronary flow [5]. Systolic dysfunction occurs when amyloid fibrils involve more than 25% of the myocardium. Atrial dysfunction is due to inadequate atrial emptying secondary to loss of atrial systolic function and increased after load, even in the presence of sinus electric activity. The extensive deposition in atrial walls results in atrial standstill or atrial electromechanical dissociation [5]. Sudden death can occur due to ventricular tachyarrhythmias, atrioventricular block, and acute electromechanical dissociation. Autonomic nervous system involvement by AL amyloidosis can lead to orthostatic hypotension and syncope. All these factors may be involved in this patient leading to cardiogenic shock. It is reported that, in AL amyloidosis, chemotherapy and stem cell transplantation may arrest and even reverse the disease, with resultant stabilization or improvement of symptoms [2]. Thus, early diagnosis is critical in these patients as newer treatment options can prevent serious complications and death which was evident in our patient.

Disclosures

All authors have read and approved the paper, and no part of this paper is being published or under consideration for publication elsewhere. There is no conflict of interests for any of the authors.

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