

AN UNUSUAL MANIFESTATION OF CARDIAC AMYLOIDOSIS ON ELECTROCARDIOGRAM

KARDİYAK AMİLOİDOZUN ELEKTROKARDİYOGRAMDA NADİR BİR GÖRÜNÜMÜ

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ABSTRACT

The most common clinical presentation of amyloid light chain amyloidosis (AL-A) includes renal involvement, with a progressive worsening of renal function and congestive heart failure. Unfortunately, the prognosis of cardiac amyloidosis is often very poor. Echocardiography and electrocardiography play cornerstone role in the diagnosis and management of cardiac amyloidosis. We describe a patient with multiple myeloma who was admitted to our clinic with progressive dyspnea due to cardiac amyloidosis. This case reveals an unusual manifestation of cardiac amyloidosis on electrocardiogram.

Key words: Cardiac amyloidosis; ECG; heart failure

ÖZET

Hafif zincirli amiloidozun (AL-A) en sık klinik sunum şekli, progresif kötüleşen renal fonksiyonlar ve kalp yetersizliği ile beraber görülen renal tutulumdur. Maalesef kardiyak amiloidozun prognozu sıklıkla kötüdür. Ekokardiyografi ve elektrokardiyografi, kardiyak amiloidozun tanısı ve tedavisinin yönetiminde köşe taşı rol oynamaktadır. Biz, kardiyak amiloidoz nedeniyle progresif dispne ile kliniğimize başvuran multiple myelom'lu bir hastayı sunduk. Bu vakada kardiyak amiloidozun elektrokardiyogramda nadir bir görünümünü tartıştık.

Anahtar kelimeler: Kardiyak amiloidoz; EKG; kalp yetersizliği

INTRODUCTION

Cardiac involvement is most common and most severe in amyloid light chain amyloidosis (AL-A), but can occur in all types of systemic amyloidosis (3). Cardiac dysfunction is considered to be the most important prognostic factor, patients with congestive heart failure having a median survival close to four months (1). We present a case which reveals an unusual manifestation of cardiac amyloidosis on electrocardiogram.

CASE

A 75-year-old man was referred to our cardiology department with shortness of breath and lower extremity edema. He had no family history of amyloidosis and his relevant medical history was limited to untreated multiple myeloma. His shortness of breath began approximately 3 months earlier with dyspnea on exertion. On physical examination he was ill-appearing with a blood pressure of 135/65 mmHg and heart rate of 83. His jugular venous pressure was markedly elevated. Cardiac exam revealed a regular rate and rhythm. On lung examination, he had bilateral rales at the bases. He had 2+ edema in the lower extremities. Laboratory investigations yielded the following results: urinary protein excretion of 2,8 g/day.

Serum albumin concentration was low, 2,4 mg/dl. Serum creatinine concentration and creatinine clearance were 1,8 mg/dl and 24 ml/min, respectively. Congo red-stained deposits were observed on renal biopsy specimen. A 12-leads electrocardiogram (ECG) revealed, increased limb lead voltage with non-specific lateral ST-T changes (Figure-1). Two-dimensional transthoracic echocardiogram was performed and revealed a small left ventricular cavity with evidence of severe left and right ventricular hypertrophy. The left ventricular ejection fraction was 50%. Both the left and right atria were moderately dilated. Four-chamber views of the ventricles and atria suggested the possibility of a restrictive cardiomyopathy (Figure-2). The combination of these findings with the patients' clinical presentation provided a quite convincing explanation for the etiology of cardiomyopathy, precluding the need for an endomyocardial biopsy.

DISCUSSION

The diagnosis of cardiac amyloidosis requires an evidence of amyloid tissue involvement. Cardiac involvement is a common finding and is the most frequent cause of death in amyloidosis. The prognosis is mainly determined by the presence and extent of heart involvement as well as the

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response to therapy (2). ECG and echocardiography are important tools in the diagnosis. Two dimensional echocardiography features include thickening of the left ventricular walls, increased reflectivity of these walls (the “sparkling” myocardium), thickening of the interatrial septum, biatrial enlargement, thickening and regurgitation of the mitral and tricuspid valves, and the presence of a small pericardial effusion. Treatment of the underlying inflammatory disorder is very important. Randomized studies reveal the superiority of oral prednisolone and melphalan compared with colchicine in systemic AL amyloidosis. There are not enough data on the use of beta-blockers in patients with cardiac amyloidosis. Angiotensin-converting enzyme inhibitors and angiotensin II inhibitors, should be used with caution in cardiac amyloidosis because even small doses may cause profound hypotension. Heart transplantation for cardiac amyloidosis has been performed rarely because of concern about progression of amyloid in other organs and the possibility of amyloid deposition in the donor heart (5).

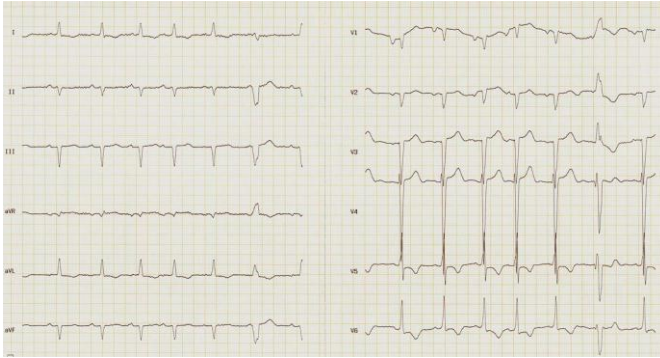


Figure 1. Electrocardiography showing the presence of left ventricular hypertrophy with voltage criteria. This appearance is unusual in patients with cardiac amyloidosis.

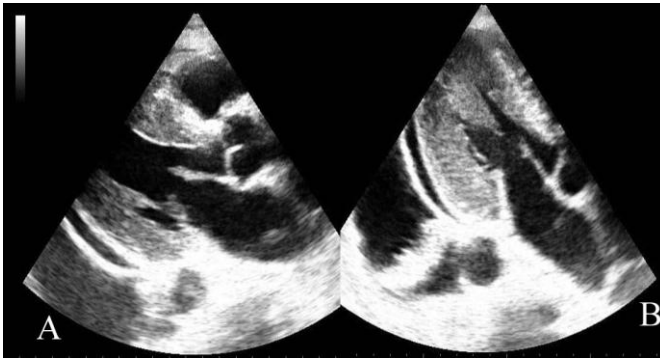


Figure 2. (A) Two-dimensional (2D) echocardiographic parasternal long-axis view from a patient with AL cardiac amyloidosis showing granular “sparkling” ventricular wall appearance, concentric left ventricular wall thickening, and thickened mitral valve leaflets suggesting infiltration. and the presence of a small posterior pericardial effusion. (B) Apical three chamber view.

Characteristic ECG features include pseudoinfarction and low voltage in the limb leads, present in about 47% and 46% respectively in patients with biopsy-proven cardiac amyloidosis. The investigators have reported that if low voltage was present on ECG and interventricular septal thickness was >1.98 cm on echocardiography, the diagnosis of cardiac amyloidosis could be made with a sensitivity of 72% and a specificity of 91% (5). Increased limb lead voltage is extremely uncommon (4). Our case reveals an unusual manifestation of cardiac amyloidosis on electrocardiogram. Probably, this unusual manifestation is a result of an unrelated coexistent condition such as hypertension. Increased limb lead voltage should be considered in ECG features of cardiac amyloidosis. We would like clinicians to be aware of this unusual manifestation of cardiac amyloidosis.

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