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Case Report

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Cardiac Amyloidosis Revealed by a Hypertensive Crisis: A Case Report and Literature Review

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Abstract

We report the case of a 76-year-old female patient with poorly managed hypertension, admitted for a hypertensive crisis associated with low-voltage on electrocardiogram (ECG). Echocardiography revealed bi-ventricular hypertrophy and bi-atrial dilation, with a bullseye pattern on global longitudinal strain. These findings prompted an amyloidosis workup, which confirmed the diagnosis. This case highlights the importance of considering cardiac amyloidosis in patients with atypical cardiovascular symptoms. A literature review on cardiac amyloidosis is also presented.

Keywords: Cardiac amyloidosis, hypertensive crisis, bi-ventricular hypertrophy, low-voltage ECG, global longitudinal strain, wild-type ATTR, literature review.

Introduction

Cardiac amyloidosis is a rare condition characterized by the deposition of amyloid fibrils in myocardial tissue, leading to structural and functional heart alterations. Although cardiac amyloidosis can present in various forms, including heart failure and arrhythmias, it is often underdiagnosed, particularly in elderly patients. This case describes an atypical presentation with a hypertensive crisis, accompanied by a review of the current literature on this condition.

Case Presentation

We present the case of a 76-year-old woman with poorly managed hypertension, admitted for a severe hypertensive crisis with a blood pressure of 210/110 mmHg. The patient also reported symptoms of heart failure, such as exertional dyspnea and lower limb edema, which had been stabilized with basic medical therapy.

Upon admission, the physical examination revealed signs of moderate volume overload without signs of acute decompensation. An electrocardiogram (ECG) showed diffuse low-voltage without other repolarization abnormalities. The hypertensive crisis was managed with Loxen® (nicardipine), a calcium channel blocker, resulting in effective blood pressure control.

Given the atypical presentation of low-voltage on ECG in a hypertensive patient, a transthoracic echocardiogram was performed to evaluate cardiac function and structure.

Echocardiography revealed marked bi-ventricular hypertrophy with concentric thickening of the ventricular walls, as well as bi-atrial dilation (Figure 1). Moderate valve thickening was also noted (Figure 2). The analysis of global longitudinal strain showed a bullseye pattern (Figure 3), characteristic of amyloid infiltration in the myocardium [1].

In light of the clinical and echocardiographic findings, an amyloidosis workup was initiated. Technetium-labeled diphosphonate scintigraphy (DPD) and a fat pad biopsy confirmed the presence of amyloid deposits. Serum and urine tests for immunoglobulin light chains were negative, ruling out light chain (AL) amyloidosis. Protein typing revealed transthyretin amyloidosis (ATTR).

Discussion

Cardiac amyloidosis is often underdiagnosed and requires a systematic diagnostic approach. The process starts with a high index of clinical suspicion, particularly in patients with unexplained ventricular hypertrophy, low-voltage ECG, or signs of diastolic heart failure. Echocardiography, using strain imaging to detect the bullseye pattern, is a key diagnostic modality [2].

To confirm the diagnosis, technetium-labeled diphosphonate (DPD) scintigraphy is recommended for ATTR amyloidosis, helping to differentiate between transthyretin amyloidosis (ATTR) and light-chain amyloidosis (AL). A biopsy of the salivary gland or heart may be necessary if diagnostic uncertainty persists.

Once the amyloidosis type is confirmed, the therapeutic algorithm is guided by the nature of the amyloid. For ATTR amyloidosis, specific treatments such as tafamidis, patisiran, or inotersen can slow disease progression. For AL amyloidosis, treatment aims to reduce amyloid light chain production through chemotherapy targeting the clonal source, often similar to that used for multiple myeloma [3].

In all cases, multidisciplinary management involving cardiologists, hematologists, and imaging specialists is crucial to optimize outcomes and improve patients' quality of life.

The challenge of amyloidosis lies in its often delayed diagnosis due to the diversity of clinical presentations and its overlap with other cardiac conditions, such as hypertrophy due to chronic hypertension. The integration of new imaging modalities and specific biomarkers has improved detection and differential diagnosis. A better understanding of the clinical manifestations and diagnostic tools for cardiac amyloidosis is essential for optimized management [4].

Conclusion

This case highlights the importance of considering cardiac amyloidosis in elderly patients presenting with ventricular hypertrophy and non-specific ECG abnormalities, such as low voltage, especially in the presence of a hypertensive crisis. Comprehensive echocardiography, along with additional investigations, is essential for accurate diagnosis and appropriate management.

References

1. Maurer MS, Schwartz JH, Gundapaneni B, Elliott P, Merlini G, et al. (2018) Tafamidis Treatment for Patients with Transthyretin Amyloid Cardiomyopathy. New England Journal of Medicine 379: 1007-1016.
   - This phase 3 study demonstrates the effectiveness of tafamidis in slowing the progression of ATTR cardiac amyloidosis, improving overall survival and reducing heart failure hospitalizations.
2. Gillmore JD, Maurer MS, Falk RH, Merlini G, Damy T, et al. (2016) Nonbiopsy Diagnosis of Cardiac Transthyretin Amyloidosis. Circulation 133: 2404-2412.
   - This article describes a non-invasive diagnostic approach for ATTR cardiac amyloidosis using nuclear imaging, avoiding the need for invasive heart biopsies.
3. Falk RH, Alexander KM, Liao R, Dorbala S (2016) AL (Light-Chain) Cardiac Amyloidosis: A Review of Diagnosis and Therapy. JACC: Heart Failure 4: 379-388.
   - This review provides an overview of diagnostic and treatment approaches for light-chain (AL) cardiac amyloidosis, highlighting differences from ATTR.
4. Kittleson MM, Maurer MS, Ambardekar AV, Renee Bullock-Palmer P, Patricia Chang P, et al. (2020) Cardiac Amyloidosis: Evolving Diagnosis and Management: A Scientific Statement from the American Heart Association. Circulation 142.
   - A detailed scientific statement outlining new diagnostic approaches and management strategies for cardiac amyloidosis, especially with advances in available treatments.