International Clinical and Medical Case Reports Journal
Case Report (ISSN: 2832-5788)

Unusual Presentation of Cardiac Amyloidosis in a Vietnam Veteran with Agent Orange Exposure

Meumbur Praise Kpughur-Tule[1], Carly Hubers[2*], Dinakaren Umashankar[1], Aaron Palinkas[1], Ngumimi Peace Kpughur-Tule[1], Mohammad Khan[1], Kendall Conway[2], Alexander M. Satei[3]

[1]Department of Internal Medicine, Wayne State University/Trinity Health Oakland, Pontiac, MI

[2]Wayne State University School of Medicine, Detroit, MI

[3]Department of Radiology, Wayne State University/Trinity Health Oakland, Pontiac, MI

Citation: *Meumbur Praise Kpughur-Tule, Carly Hubers, Dinakaren Umashankar, Aaron Palinkas, Ngumimi Peace Kpughur-Tule, et al. Unusual Presentation of Cardiac Amyloidosis in a Vietnam Veteran with Agent Orange Exposure. Int Clinc Med Case Rep Jour. 2025;4(2):1-3.*

Received Date: 30 January, 2025**;** **Accepted Date:** 02 February, 2025**;** **Published Date:** 04 February, 2025

[*]**Corresponding author:** Carly Hubers, Wayne State University School of Medicine, Detroit, MI

ABSTRACT

Cardiac amyloidosis typically manifests as restrictive cardiomyopathy, but this case presents an unusual overlap of heart failure with reduced ejection fraction, sustained ventricular tachycardia, and severe coronary artery disease in a largely asymptomatic patient. A 77-year-old male with Agent Orange exposure and a history of recurrent premature ventricular contractions experienced syncope, leading to the diagnosis through multimodal imaging and amyloid workup. The coexistence of infiltrative and ischemic pathology complicated management. This case notes the diagnostic challenges in differentiating cardiac amyloidosis from ischemic cardiomyopathy and highlights the potential role of environmental exposures in disease pathogenesis. Recognition of such atypical presentations is crucial for optimizing diagnostic and therapeutic strategies.

INTRODUCTION

Cardiac amyloidosis is a rare cardiomyopathy caused by the extracellular deposition of amyloid fibrils.[1] It typically presents with symptoms resembling right ventricular failure due to restrictive cardiomyopathy, including dyspnea, syncope, elevated jugular venous pressure, hepatic congestion, lower extremity edema, and ascites.[2,3] This case describes an unusual presentation in a largely asymptomatic patient with new-onset syncope, heart failure with reduced ejection fraction (HFrEF), sustained ventricular tachycardia secondary to cardiac amyloidosis and severe coronary artery disease requiring Percutaneous Transluminal Coronary Angioplasty (PTCA).

CASE PRESENTATION

A 77-year-old male presented following an episode of syncope while visiting his late wife's grave. The unwitnessed event lasted 8–10 minutes with urinary incontinence. His medical history included recurrent

premature ventricular contractions (PVCs), hypertension, CKD stage 3A, bilateral carotid stenosis, and prolonged grief disorder. Notably, he had Agent Orange exposure during military service in Vietnam.

On presentation, physical examination was unremarkable, and vital signs were stable. ECG revealed ventricular escape complexes, left ventricular hypertrophy with QRS widening, and first-degree heart block. Laboratory evaluation showed high-sensitivity troponin of 24 ng/L, BNP of 344 pcg/ml, and creatinine of 1.35 mg/dL (eGFR 54 ml/min/1.73 m^2).
A transthoracic echocardiogram revealed an EF of 36%, grade I diastolic dysfunction, and an abnormal strain pattern suggestive of amyloidosis. Cardiac MRI demonstrated myocardial fibrosis with subendocardial to transmural late gadolinium enhancement. Coronary angiography revealed severe multivessel disease. During PTCA, the patient experienced sustained ventricular tachycardia.

Amyloid workup revealed mildly elevated kappa-free light chains (3.90 mg/dL), a normal lambda-free light chain (2.13 mg/dL), and an elevated kappa/lambda ratio of 1.83, with no monoclonal protein detected. The patient underwent successful dual-chamber implantable cardioverter defibrillator (ICD) placement. He was discharged on dual antiplatelet therapy, guideline-directed medical therapy for heart failure, and amiodarone, with plans for cardiac biopsy at follow-up.

DISCUSSION

This case is unique and significant due to the rare and multifaceted presentation of cardiac amyloidosis in a largely asymptomatic patient. While cardiac amyloidosis is typically associated with restrictive cardiomyopathy, this patient presented with HFrEF and sustained ventricular tachycardia, which are unusual in the early clinical course. Such an overlap of arrhythmic and ischemic pathology is rarely documented and underscores the diverse cardiac manifestations of amyloidosis.

The coexistence of severe multivessel coronary artery disease further complicates the clinical picture. Differentiating between ischemic and infiltrative causes of ventricular dysfunction in this patient required advanced imaging modalities, including cardiac MRI and echocardiography with strain analysis, demonstrating the diagnostic complexity of the case.

Additionally, the history of Agent Orange exposure introduces a potentially novel environmental factor in the pathogenesis of cardiac amyloidosis. While dioxin exposure has been linked to various systemic conditions, its role in amyloid fibril deposition remains largely unexplored, presenting an opportunity for further research.[4]
This case highlights the importance of considering cardiac amyloidosis in atypical presentations and calls attention to the need for a multidisciplinary approach in diagnosing and managing such rare and complex cases. Early recognition can enable tailored interventions that significantly impact prognosis.[5]

REFERENCES

1. Jean D Sipe, Merrill D Benson, Joel N Buxbaum, Shu-Ichi Ikeda, Giampaolo Merlini, Maria J M Saraiva, et al. Amyloid fibril proteins and amyloidosis: chemical identification and clinical classification International Society of Amyloidosis 2016 Nomenclature Guidelines. Amyloid 2016; 23:209.
2. Falk RH, Alexander KM, Liao R, Dorbala S. AL (Light-Chain) Cardiac Amyloidosis: A Review of Diagnosis and Therapy. J Am Coll Cardiol. 2016; 68:1323.
3. Gertz MA, Benson MD, Dyck PJ, et al. Diagnosis, Prognosis, and Therapy of Transthyretin Amyloidosis. J Am Coll Cardiol. 2015;66:2451.
4. Committee to Review the Health Effects in Vietnam Veterans of Exposure to Herbicides (Ninth Biennial Update); Board on the Health of Select Populations; Institute of Medicine. Veterans and Agent Orange: Update 2012. Washington (DC): National Academies Press (US); 2014 Mar 6. 12, Cardiovascular and Metabolic Outcomes. Available from: https://www.ncbi.nlm.nih.gov/books/NBK195089/
5. Kelty R Baker. Light Chain Amyloidosid: Epidemiology, Staging and Prognostication. Methodist Debakey Cardiovasc J. 2022;18(2):27-35.