Incidental finding of Transthyretin Cardiac Amyloidosis during Coronary Artery Bypass Grafting

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Introduction

We present a case of Wild Type Transthyretin (ATTRwt) Cardiac Amyloidosis incidentally discovered during two vessel Coronary Artery Bypass Grafting (CABG) for treatment of therapy-resistant diastolic Congestive Heart Failure (CHF).

Transthyretin (ATTR) Cardiac Amyloidosis is a condition characterized by extracellular deposition of misfolded Transthyretin proteins in the myocardium leading to morphological and functional changes to the heart. Deposition of these insoluble proteins lead to thickening and stiffening of cardiac tissue. Resulting cardiac remodeling often leads to diastolic dysfunction and restrictive cardiomyopathy.

ATTR Cardiac Amyloidosis has been historically difficult to diagnose due to diverse clinical manifestations and nonspecific, variable electrocardiogram (ECG) and echocardiogram findings.

Background

Once thought to be a rare condition, recent research suggests that ATTR cardiac amyloidosis may be significantly underdiagnosed and far more prevalent in the general population than previously assumed.

A retrospective analysis of our patient's history reveals the classic symptoms and clinical picture of ATTR Amyloidosis. Yet, the diagnosis was not suspected until the heart was visualized during CABG. This near-miss highlights the pressing need for better screening tools and a routine, standardized screening process for this condition.

Case Presentation

A 78 year old male presented to the Emergency Department with a 30 minute history of chest discomfort, shortness of breath, dizziness, and diaphoresis. His past medical history included heart failure with preserved ejection fraction, hypertension, hypercholesterolemia, Right Bundle Branch Block, Insulin-dependent Type II Diabetes Mellitus, Carpal tunnel syndrome with associated surgical release and elevated BMI.

Consistent cardiology follow-up revealed a six year history of non-specific ECG changes and persistent right bundle branch block despite no prior history of acute cardiac event. Echocardiography performed 11 days prior to ED presentation revealed concentric left ventricular hypertrophy with preserved left ventricular systolic function.

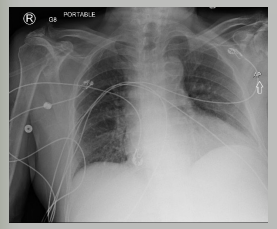

Figure 1: AP Chest X-Ray showing enlarged cardiac silhouette and reduced lung volumes

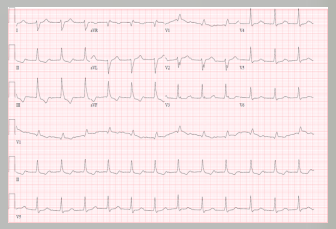

Figure 2: Initial ED ECG demonstrating non-specific changes including 1st degree AV block, Right Axis Deviation and RBBB

The patient's acute chest pain and and shortness of breath improved with administration of lorazepam. At this time he was considered for discharge but due to his comorbidities and consistent concern that something was 'not right', and Percutaneous Coronary Intervention (PCI) was performed.

PCI revealed severe (>90%) obstruction (see figure 1), and the patient was referred for two vessel Coronary Artery Bypass Grafting.

Intraoperatively, the patient's heart was found to be abnormally thickened and fibrosed. Biopsy of cardiac tissue and pathological evaluation using Technetium-99m Pyrophosphate scintigraphy, single-photon emission computed tomography and liquid chromatography tandem mass spectrometry revealed ATTR (transthyretin) cardiac amyloidosis.

Discussion

With treatment available to slow the progression of disease, there is a pressing need for better screening tools for the identification of ATTR amyloidosis. If it were not for the failure of this patient's PCI to resolve his obstruction, CABG would not have been performed and the abnormal gross appearance of the myocardium would not have been identified.

ATTR amyloidosis has many manifestations, which include treatment-resistant heart failure with preserved ejection fraction and musculoskeletal presentations, particularly carpal tunnel syndrome, both of which our patient had. Because the presentation of this disease is typically above the age of 50, it is difficult to distinguish symptoms of heart failure plus age related musculoskeletal complaints from the clinical picture of amyloidosis. Sufficient confirmatory testing exists and include genetic testing and nuclear radiography, neither of which can provide the fast and low-cost features needed of an effective screening tool.

Conclusions

ATTR Amyloidosis is difficult to diagnose, largely because of its non-specific presentation. There is a need for fast and low-cost screening tools to allow for early identification of the disease.

Clinicians should maintain a high index of suspicion for TTR amyloidosis in elderly individuals with treatment-resistant heart failure with preserved ejection fraction, even when presenting in acute settings.

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