Amyloid Cardiomyopathy: All that Sparkles is Amyloid

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A 73 year old female with no past medical history presented to our emergency department (ED) with gradually worsening shortness of breath for a few weeks. She denied any associated complaints of chest pain, palpitations or dizziness. On physical exam, the patient had bilateral rales at the lung bases. The electrocardiogram showed low voltage complexes with right bundle branch block and left anterior fascicular block (Figure 1). The laboratory work came back significant for an elevated BNP (1452 pg/ml) with a normal complete blood count, serum chemistry and negative cardiac biomarkers. The patient was treated with intravenous diuretics in the ED and admitted to a telemetry floor.

The echocardiogram performed on admission revealed concentric bi-ventricular hypertrophy resulting in a restrictive pattern and highly echogenic myocardium with ‘sparkling appearance’, suggestive of Cardiac Amyloidosis (figure 2, Video 1, 2, see supplementary material on website). The LV systolic function was moderately decreased with global hypokinesis and there was bilateral atrial enlargement along with thickening of valve leaflets and inter-atrial septum. A Cardiac MRI also showed delayed nodular enhancement of the myocardium indicative of an infiltrative cardiomyopathy. Patient underwent an abdominal wall fat pad biopsy which came positive for light-chain Amyloidosis.

Amyloid Cardiomyopathy is characterized by ventricular thickening due to amyloid deposition leading to diastolic dysfunction. It most commonly manifests with heart failure, conduction abnormalities or exertional syncope due to inability to augment cardiac output. The combination of increased ventricular mass on echocardiogram and reduced electrocardiographic voltages, along with granular appearance of the myocardium is highly suggestive of Amyloid Cardiomyopathy. The diagnosis can be confirmed by demonstrating amyloid deposits on histologic examination of tissues from abdominal fat pad or kidney in patients with appropriate cardiac findings. Cardiac amyloidosis should be considered in any adult with unexplained heart failure and an echocardiogram showing increased wall thickness with a non-dilated left ventricular cavity, particularly when associated with low voltage on electrocardiogram.

Declarations of Interest
The authors declare no conflicts of interest

Acknowledgements
The authors agree to abide by the requirements of the statement of publishing ethics of the International Cardiovascular Forum Journal.

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Keywords: Cardiac amyloidosis, amyloid cardiomyopathy

Citation: Saxena A, Chadha S, Chen O, Sadiq A, Shani J. Amyloid Cardiomyopathy: All that sparkles is Amyloid. International Cardiovascular Forum Journal. 2016;6:66 DOI: 10.17987/icfj.v6i0.174