Role of Cardiovascular Magnetic Resonance Imaging in the Evaluation of Systemic Amyloidosis

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Abstract

Case report: A 65 year old gentleman was admitted with chief complaints of swelling in both lower limbs gradually increasing for 2 months associated with heaviness in legs. He also had shortness of breath, dizziness, fatigue and passage of frothy urine for the same duration. No history of fever, reduced urine output, haematuria, dysuria, nausea, diarrhoea, constipation, loss of weight and appetite, numbness, tingling, hypertension, diabetes, tuberculosis, asthma, previous hospitalizations. Histopathology---Kidney biopsy was done The patient was investigated. Hemogram showed normal leucocyte count raised ESR (38). RBS normal, LFT showed hypoalbuminemia KFT was normal .Coagulation and Thyroid profile was normal. Nephrotic range proteinuria (albuminuria-8.514) .His Chest X ray was normal and ultrasound showed hepatomegaly with benign prostatic hypertrophy (BPH) and left kidney cyst. CT abdomen showed mild hepatomegaly with bilateral pleural effusion and diffuse abdominal wall edema.

In view of the systolic murmur, Echocardiography was done which showed no regional wall motion abnormality of LV. Global LVEF 60%, moderate concentric LVH, increased myocardial echogenicity ?infiltrative cardiomyopathy ?amyloidosis, LA high normal, mild MR and trace TR, Grade 1 diastolic dysfunction (E/E' >15) and minimal pericardial effusion and no intracardiac clot/vegetation.Fig.1and 2..

DISCUSSION

We observed a number of imaging findings, the combination of which appear to be specific to amyloid cardiomyopathy. Typical features of restrictive cardiomyopathy—that is, left ventricular wall thickening, reduced systolic function with decreased ejection fraction, restriction of diastolic filling, and disproportionate atrial enlargement (AE)—were present in all cases. On delayed post contrast images acquired 8–15 min after IV gadolinium administration, a definite widespread homogenous pattern of increased signal on inversion recovery T1- weighted images was observed throughout the myocardium . This pattern differs from common patterns of **enhancement**

associated with other entities such as ischemic infarction, which usually shows intense sub endocardial or transmural enhancement localized to vascular territory; infiltrative diseases such as sarcoidosis or lymphoma, in which enhancement is often focal; and interstitial fibrosis, which may show longitudinal striae of midwall enhancement. The degree of enhancement was considerably less than that seen with replacement fibrosis of myocardial infarction (MI).

CONCLUSION

Cardiac and renal amyloidosis is a rare finding. The presentation was unique and posed a diagnostic challenge. Prognosis of AL chain amyloidosis is poor and mortality is high. In conclusion, we consider the combination of widespread heterogeneous myocardial enhancement with other supporting features of infiltrative myocardial disease to be relatively specific for cardiac amyloidosis. Earlier diagnosis with therapeutic interventions portends a better response to current therapy and prolonged survival. Meaning: The TURP syndrome presents with volumetric overload shock (VOS) best treated with HST, Starling's law is wrong and the correct replacement

is the hydrodynami c of the G tube.

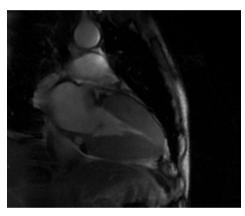


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FIG.1 AND 2. - HYPERTROPHIED LEFT VENTRICLE WITH INCREASED MYOCARDIAL ECHOGENICITY (TYPICAL SPECKLED APPEARANCE OF AMYLOIDOSIS) AND LEFT ATRIA ENLARGEMENT (42MM).



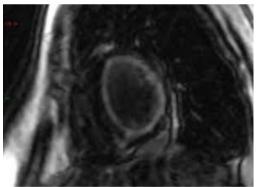


FIGURE 2 AND 3 CARDIAC MRI **SHOWING** CONCENTRIC LEFT VENTRICULAR HYPERTROPHY (LVH) AND DIFFUSE MYOCARDIAL **DELAYED ENHANCEMENT**

Biography:

Reena Anand is working as Head of Cardiac Imaging at Max Super Specialty Hospital, New Delhi, India. She has extensive experience on cardiac MRI from New York University. She has given various national and international talks providing training to emerging cardiac radiologist and cardiac technologists worldwide. She has numerous national and international publications to her credit. She is an active Member of Society of Cardiovascular Magnetic Resonance (SCMR)..



Speaker Publications:

- 1. 1. Banypersad SM, Moon JC, Whelan C, Hawkins PN, Wechalekar AD. Updates in cardiac amyloidosis: a review. J Am Heart Assoc. 2012;1: e000364.
- 2. J.D. Sipe, M.D. Benson, J.N. Buxbaum Amyloid fibril protein nomenclature: 2010 recommendations from the nomenclature committee of the International Society of Amyloidosis Amyloid, 17 (2010), pp. 101-104
- 3. Mohty D, Boulogne C, Magne J, et al. Prognostic value of left atrial function in systemic light-chain amyloidosis: a cardiac magnetic resonance study. Eur Heart J Cardiovasc Imaging. 2016; 17:961-969

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