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Regression of Cardiac Amyloidosis following Stem Cell Transplantation assessed by Cardiovascular Magnetic Resonance Imaging

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Keywords

cardiovascular magnetic resonance imaging; amyloid; heart failure

A previously fit 52-year-old man presented with severe progressive exertional dyspnea. He was in heart-failure with an elevated jugular-venous-pressure, edema and increased plasma-NTpro-BNP levels of 4,285 $\mu\text{g}/\text{mL}$ (upper limit of normal $<900\mu\text{g}/\text{mL}$). His ECG demonstrated sinus-rhythm with low limb and chest lead voltages (Figure 1). He had significant proteinuria with renal and bone-marrow biopsies confirming Light-chain (AL) amyloidosis. Cardiac-Magnetic-Resonance (CMR) imaging revealed concentric left-ventricular-hypertrophy with an ejection-fraction of 65%, left-ventricular-end-diastolic-volume (LVEDV)=146mL, left-ventricular-endsystolic-volume (LVESV)=51mL, left-ventricular-mass=245g, and left-atrial-volume=144mL (Figure 2, Movie 1). Late-gadolinium-enhanced (LGE) imaging showed extensive diffuse subendocardial hyperenhancement in both ventricles (Figures 3 and 4, **arrows**) consistent with amyloid infiltration. He subsequently underwent successful autologous-stem-cell transplantation.

At follow-up, 2.5 years later, his functional-status had markedly improved and he was exercising regularly. His cardiovascular examination and plasma-NTpro-BNP level (117 $\mu\text{g}/\text{mL}$) was normal. His ECG showed some recovery of voltages in the limb leads (Figure 5). Repeat CMR imaging showed minimal change in left-ventricular volumes, function and mass (LVEDV=138mL, LVESV=43mL, ejection fraction=69%, left-ventricular-mass=235g); although left-atrial volume was significantly reduced (105mL) (Figure 6, Movie 2). LGE imaging demonstrated marked regression of the subendocardial hyperenhancement (Figures 7 and 8).

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Prognosis for patients with AL-amyloid and cardiac-infiltration has historically been dismal and extensive cardiac-involvement has generally been regarded as a contraindication to stem-cell-transplantation¹⁻³. This case suggests that stem-cell-transplantation can lead to regression of cardiac-amyloid and may be considered in selected patients.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

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References

1. Banyersad SM, Moon JC, Whelan C, Hawkins PM, Wechalekar AD. J Am Heart Assoc. 2012; 1:e000364. [PubMed: 23130126]
2. Falk RH, Dubrey SW. Amyloid Heart Disease. Prog Card Dis. 2010; 52:347–61.
3. Falk RH. Diagnosis and Management of the Cardiac Amyloidoses. Circulation. 2005; 112:2047–60. [PubMed: 16186440]

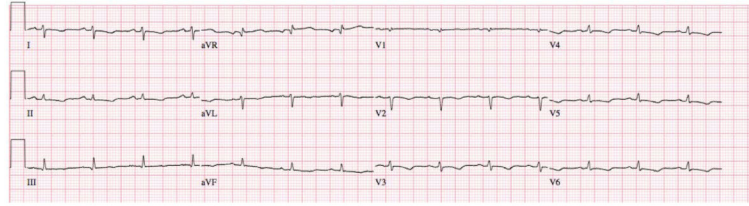


Figure 1.
ECG showing sinus-rhythm with low QRS voltages (<5mm in the limb leads and <10mm in the chest leads).

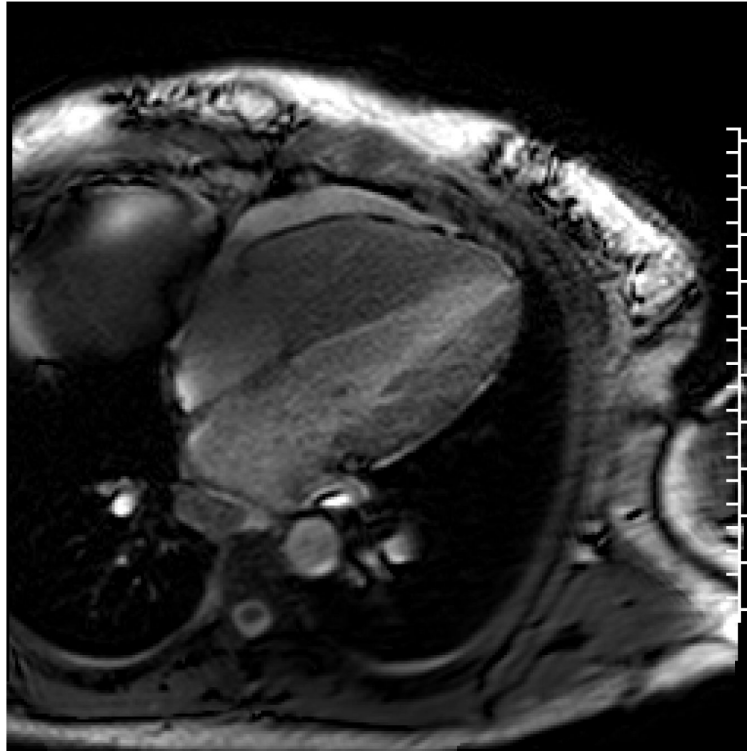


Figure 2. Cine imaging in the 4-chamber view, showing an ejection fraction of 65%, concentric left-ventricular hypertrophy, and an enlarged left atrium.

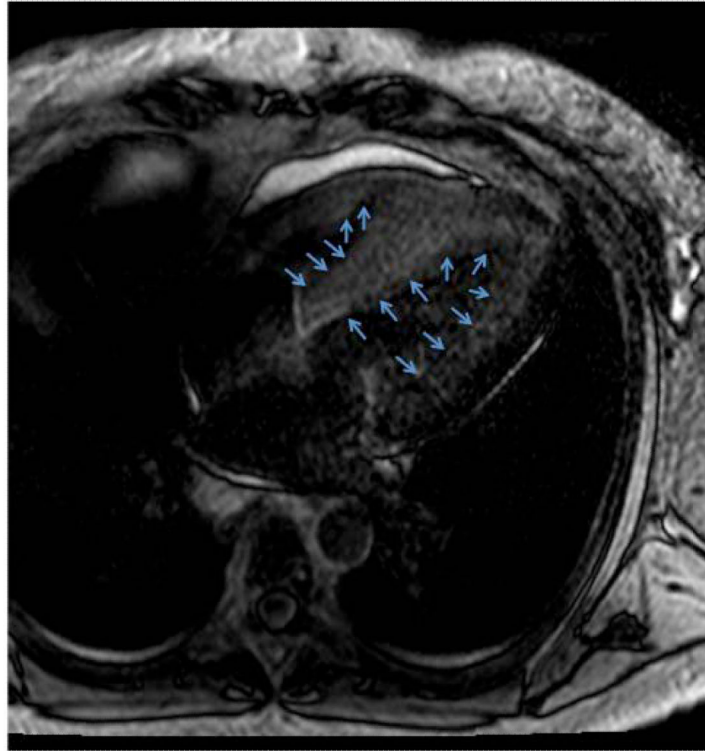


Figure 3. Late Gadolinium Enhancement (LGE) imaging in the 4-chamber view, showing extensive diffuse subendocardial hyperenhancement involving both ventricles (arrows).

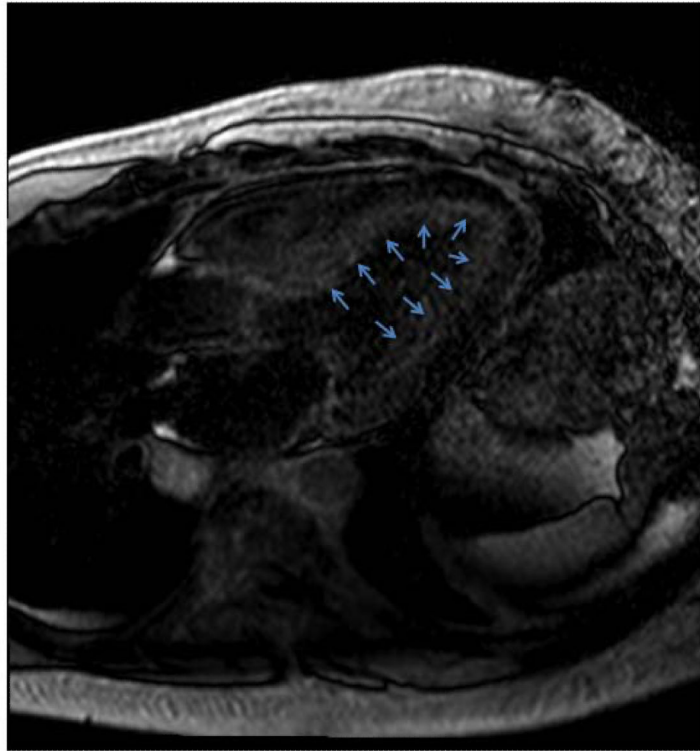


Figure 4. Late Gadolinium Enhancement (LGE) imaging in the 3-chamber view, showing extensive diffuse subendocardial hyperenhancement (arrows).

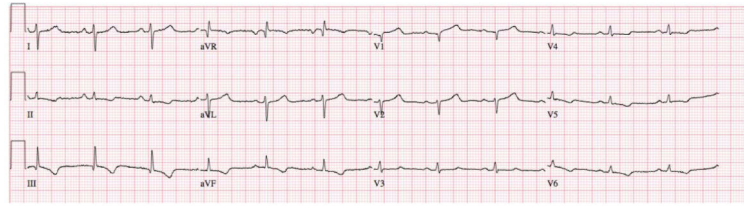


Figure 5.
Post transplant ECG showing sinus-rhythm with some recovery of voltages in the limb leads (>5mm in leads I, III, aVR, aVL).



Figure 6. Post stem cell transplant. Cine imaging in the 4-chamber view, showing an ejection fraction of 69% with concentric left-ventricular hypertrophy. The left atrium has reduced in size.

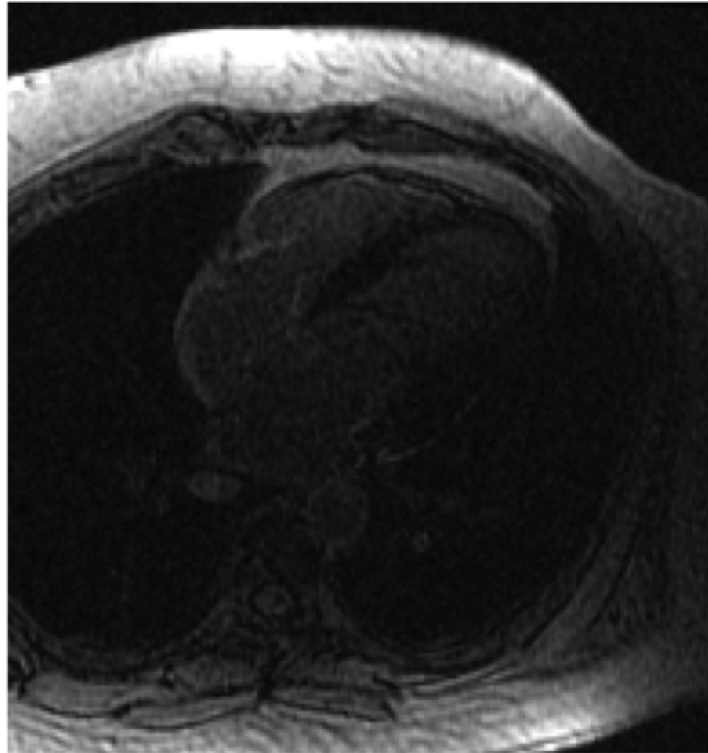


Figure 7. Post stem cell transplant. Late Gadolinium Enhancement (LGE) imaging in the 4-chamber view, showing significant regression of the subendocardial hyperenhancement.

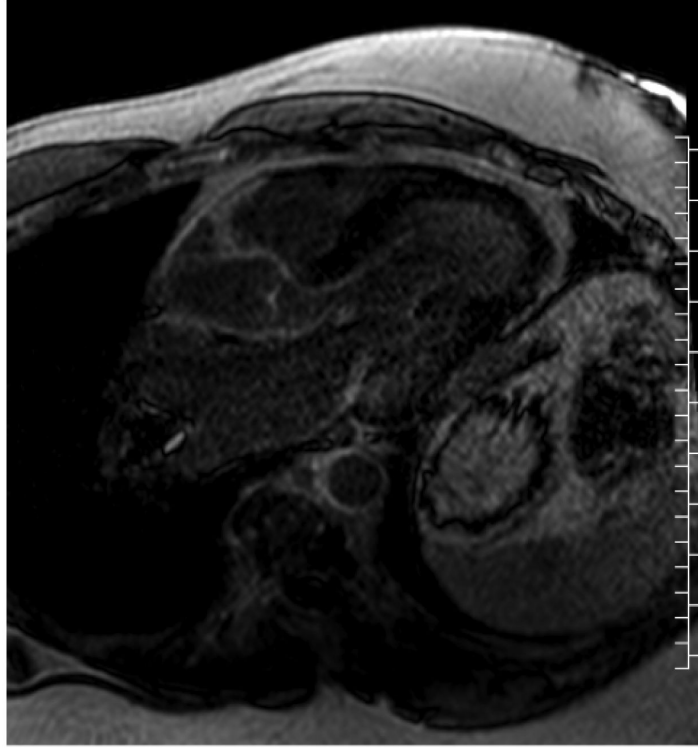


Figure 8. Post stem cell transplant. Late Gadolinium Enhancement (LGE) imaging in the 3-chamber view, showing significant regression of the subendocardial hyperenhancement.