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 4285 μg/mL (upper limit of normal <900 μg/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 amyloidosis. Cardiac magnetic resonance imaging revealed concentric left-ventricular hypertrophy with an ejection fraction of 65%, left-ventricular end-diastolic volume of 146 mL, left-ventricular end-systolic volume of 51 mL, left-ventricular mass of 245 g, and left-atrial volume of 144 mL (Figure 2, Movie I in the online-only Data Supplement). Late gadolinium-enhanced 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 μg/mL) was normal. His ECG showed some recovery of voltages in the limb leads (Figure 5). Repeat cardiac magnetic resonance imaging showed minimal change in left-ventricular volumes, fraction, and mass (left-ventricular end-diastolic volume, 138 mL; left-ventricular end-systolic volume, 43mL; ejection fraction, 69%; left-ventricular mass, 235 g), although left atrial volume was significantly reduced (105 mL; Figure 6, Movie I in the online-only Data Supplement). Late gadolinium-enhanced imaging demonstrated marked regression of the subendocardial hyperenhancement (Figures 7 and 8).

Prognosis for patients with light-chain amyloid and cardiac infiltration has historically been dismal, and extensive cardiac involvement has generally been regarded as a contraindication to stem cell transplantation.1–3 This case suggests that stem cell transplantation can lead to regression of cardiac amyloid and may be considered in selected patients.

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Disclosures
None.

References
Figure 1. ECG showing sinus rhythm with low QRS voltages (<5 mm in the limb leads and <10 mm 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 imaging in the 4-chamber view, showing extensive diffuse subendocardial hyperenhancement involving both ventricles (arrows).

Figure 4. Late gadolinium enhancement 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 (>5 mm 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 imaging in the 4-chamber view, showing significant regression of the subendocardial hyperenhancement.

Figure 8. Post stem cell transplant late gadolinium enhancement imaging in the 3-chamber view, showing significant regression of the subendocardial hyperenhancement.
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