

IMAGE IN CARDIOVASCULAR MEDICINE

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Serial electrocardiographic and echocardiographic assessments of cardiac amyloidosis presenting with myocardial ischemia

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A 58-year-old man with a history of type 2 diabetes, hypertension, and dyslipidemia was hospitalized due to effort angina. The B-type natriuretic peptide level at admission was 915 pg/mL. The ST-segment depressions were progressive compared to previous electrocardiograms (Fig. 1A). Serial echocardiograms revealed an evolving thickening of the interventricular septum and progression of diastolic dysfunction (Fig. 1B). Myocardial perfusion scintigraphy showed that the fixed defect in the posterior wall was unchanged, however, the reversible perfusion defects in the inferior and inferoseptal wall of the left ventricle were newly developed (Fig. 1C) compared to the previous scintigraphy. Although coronary angiography showed a chronic total occlusion in the distal posterolateral branch of the left circumflex artery, there was no significant epicardial coronary stenosis that could have caused inferior myocardial ischemia. Bone marrow biopsy demonstrated that the percentage of plasma cells in the bone marrow was 36% (Fig. 1D), and serum electrophoresis and immunofixation detected an IgG-kappa M protein, which was consistent with multiple myeloma. The 99m technetium-pyrophosphate scintigraphy revealed a positive cardiac uptake (Fig. 1E), and the rectal mucosal biopsy showed amyloid deposition (Fig. 1F, G), confirming the diagnosis of cardiac amyloidosis. He died due to the exacerbation of heart failure despite the treatments for heart failure and multiple myeloma. Because there was no progressive stenosis in epicardial coronary arteries, the coronary microvascular dysfunction due to amyloid deposition may have caused myocardial ischemia although it was not assessed by endocardial biopsy, coronary flow reserve or microcirculatory resistance.

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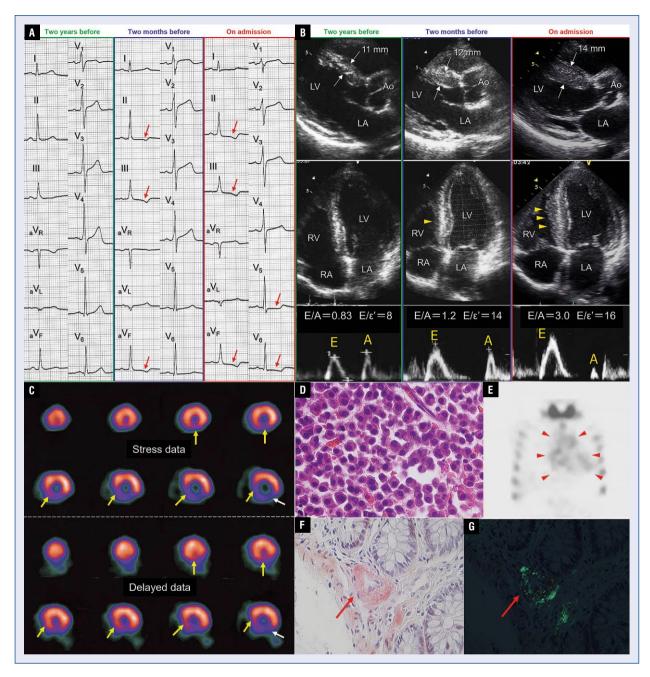


Figure 1. A. Electrocardiograms performed 2 years before (left), 2 months before (center), and on admission (right), showing the progression of ST-segment depressions (red arrows); **B.** Echocardiograms shows thickening of the interventricular septum (white arrows and yellow arrowheads) and diastolic dysfunction; Ao — aorta; LA — left atrium; LV — left ventricle; RA — right atrium; RV — right ventricle; **C.** Stress thallium-201 myocardial perfusion scintigraphy showing reversible perfusion defects (yellow arrows) and fixed defects (white arrows); **D.** Bone marrow biopsy showing an increase in the number of plasma cells (hematoxylin and eosin stain); **E.** 99m technetium-pyrophosphate scintigraphy showing positive uptake in the heart (red arrowheads). Perugini score was estimated to be grade 1; **F, G.** Rectal mucosal biopsy showing amyloid deposition (red arrows, **F**: Congo red stain, **G**: polarization).