Transthyretin Stabilizer Is Associated With Expanding Apical Sparing Area and Improving Global Cardiac Function in a Patient With Wild-Type Cardiac Amyloidosis

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Transthyretin (TTR) amyloidosis, caused by the deposition of wild-type TTR (ATTRwt) in the heart, is an underdiagnosed cause of heart failure with preserved ejection fraction (HFpEF). The relative apical sparing pattern on 2-D speckle-tracking echocardiography (2D-STE) is frequently seen in cardiac amyloidosis. Tafamidis is a kinetic stabilizer of TTR that inhibits misfolding and amyloidogenesis. We present a case of biopsy-proven WT-TTR, in which the cardiac apical sparing pattern improved with tafamidis treatment.

A 78-year-old man, misdiagnosed with hypertensive cardiomyopathy, was hospitalized with HFpEF. He also had orthostatic hypotension and plantar numbness. Both endomyocardial and intestinal biopsy confirmed the presence of TTR amyloid deposits (Figure A), accompanied by positive late gadolinium enhancement on cardiac magnetic resonance imaging (Figure B). 2D-STE on admission showed a relative apical sparing pattern with global longitudinal strain −5.7%. Heart failure medication was optimized, and treatment with 20mg tafamidis daily was initiated. The area where the myocardial strain was preserved, expanded in the course of treatment (Figure C).

Whether such changes with tafamidis are seen in a larger cohort of ATTRwt patients will be able to be explored in the ATTR-ACT trial,1 which recently announced that it met its primary endpoint of a hierarchical composite of all-cause mortality and recurrent cardiovascular-related hospitalizations.

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Reference