

Conversion of 99m technetium-pyrophosphate scintigraphy in a patient with hereditary ATTR amyloidosis: importance of repeat scanning

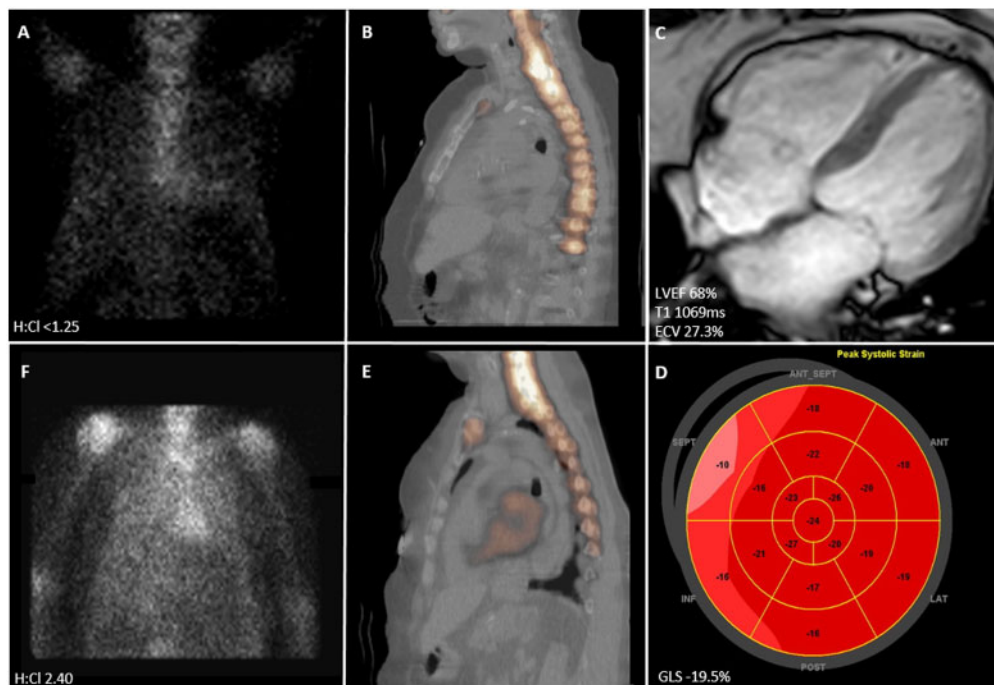
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A 73-year-old female with spinal stenosis, hypertension, and bilateral carpal tunnel syndrome underwent release surgery with tenosynovial biopsy demonstrating transthyretin (TTR) amyloid deposition. Genetic testing for TTR demonstrated Ala81Thr mutation consistent with hereditary amyloid transthyretin (ATTR amyloidosis).

Electrocardiogram was normal ([Supplementary material online, Figure S1A](#)), troponin T was <0.01 ng/mL, and N-terminal pro-hormone of brain natriuretic peptide (NT pro-BNP) was 101 pg/mL. Echocardiography revealed normal global longitudinal strain (GLS) with normal left ventricular (LV) size, no increased wall thickness,



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and left ventricular ejection fraction (LVEF) 62% (Supplementary material online, Figure S2A and B). ^{99m}Tc -pyrophosphate (^{99m}Tc -PYP) showed no uptake on planar or single-photon emission computed tomography (SPECT) imaging and heart-to-contralateral ratio (H:CL) was <1.25 at 3 h (Panels A and B).

Cardiac magnetic resonance at 3 years revealed no gadolinium uptake with normal LV size (end-diastolic volume 84 mL), normal systolic function (LVEF 68%), normal T1 time, and extracellular volume (ECV) (Panel C). At 4 years, the patient remained asymptomatic. ECG was normal (Supplementary material online, Figure S1B), troponin T was <0.01 ng/mL, and NT pro-BNP was 119 pg/mL. Echocardiography showed no changes (Panel D and Supplementary material online, Figure S2C). Repeat ^{99m}Tc -PYP demonstrated grade 3 uptake on SPECT and planar imaging and H:CL of 2.40, consistent with ATTR cardiac amyloidosis (Panels E and F).

To the best of our knowledge, this is the first reported case of conversion of negative ^{99m}Tc -PYP to positive ^{99m}Tc -PYP despite

otherwise normal cardiac evaluation. Current guidelines do not address the need for/frequency of repeat testing in asymptomatic individuals with TTR gene mutations or extra-cardiac TTR deposits. Early identification with repeat testing may facilitate pre-symptomatic initiation of therapy to halt disease progression. This case highlights the importance of follow-up ^{99m}Tc -PYP scanning in these patients.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.