Wild-type TTR neuropathy with cardiomyopathy presenting with burning feet

Figure 1 SPECT-CT in transthyretin (TTR) cardiomyopathy identification

(A) Fusion SPECT-CT shows increase of (99m)Tc-pyrophosphate radiotracer in heart (arrows) with (B) avid post tracer cardiac uptake consistent with TTR cardiac amyloid.

A 79-year-old man presented with a 15-year history of burning feet. His neurologic examination showed absent ankle reflexes and reduced light touch, vibration, and heat pain at the feet. Nerve conductions showed absent sural and reduced peroneal (0.6 mV) and tibial (0.3 mV) motor responses, with normal needle EMG. Diabetes and monoclonal gammopathy were absent. At age 74, he had heart failure and atrial fibrillation. Cardiac SPECT-CT nuclear imaging utilizing (99m)Tc-pyrophosphate radiotracer¹ supported cardiac transthyretin (TTR) amyloid infiltration (figure 1). Epidermal skin biopsy identified amyloid deposition (figure 2). *TTR* gene sequencing was normal and he was diagnosed with wild-type TTR neuropathy and cardiomyopathy.²

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(A) Calf skin biopsy congophilia (B) with apple green birefringent diagnostic of amyloid. (C) PGP9.5 immunostaining with preserved epidermal nerve fiber density (2.8 fibers/mm).

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