Teaching NeuroImages:
PET-CT hypermetabolism paralleling muscle hyperactivity in stiff-person syndrome

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A 72-year-old woman with autoimmune hypothyroidism and type 1 diabetes mellitus presented with 1 year of progressively worsening neck, back, and right thigh spasms and leg stiffness. Sudden loud noises would startle her. She ambulated with a walking frame. Serum glutamic acid decarboxylase 65 antibody was markedly elevated (1,118 nmol/L; normal value ≤0.02 nmol/L). EMG demonstrated poor relaxation in thoracic paraspinal muscles and right quadriceps and failure of the acoustic startle response to habituate consistent with stiff-person syndrome.1,2 Whole-body PET-CT imaging, performed to exclude malignancy, demonstrated increased fluorodeoxyglucose metabolism in axial and proximal lower extremity regions that paralleled the clinically most hyperactive muscle groups (figure, A and B). No malignancy was detected.

AUTHOR CONTRIBUTIONS
Dr. Orna O’Toole: first author. Dr. Robert Murphy: imaging expertise. Dr. Jennifer Tracy: advice regarding neurophysiology in stiff-person syndrome. Dr. Andrew McKeon: advice regarding stiff-person syndrome.

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REFERENCES
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