Teaching NeuroImages: Brain MRI and DaT-SPECT imaging in adult GM1 gangliosidosis

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Neurology® 2018;91:e187-e188. doi:10.1212/WNL.0000000000005775

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Figure MRI and DaT-SCAN SPECT findings in the adult GM1 gangliosidosis patient

(A, B) Axial T2-weighted imaging and fluid-attenuated inversion recovery images show increased signal in the putamina. (C) DaT-SPECT demonstrates decreased radiotracer uptake in the bilateral basal ganglia suggestive of nigrostriatal dopaminergic dysfunction. More diffuse involvement of putamen and caudate nucleus can be noted on the left side.

A 58-year-old woman with genetically confirmed adult GM1 gangliosidosis (aGM1-g) presented with generalized dystonia that was later followed by akinetic-rigid parkinsonism. Brain MRI revealed hyperintensities in the bilateral putamen typically observed in this disease¹ (figure, A and B), while DaT-SPECT (¹²³I-Ioflupane) showed decreased radiotracer uptake in both basal ganglia, more evident on the left side (figure, C). Selective involvement of the basal ganglia in aGM1-g is thought to be related to a higher turnover of GM1 ganglioside in this region.² Our report suggests a presynaptic pattern of dopaminergic dysfunction in this disease.

Author contributions

Dr. Antonio Marangi: case report organization, analysis of data, draft of manuscript.

Dr. Matteo Tagliapietra: analysis of data, manuscript review and critique. Dr. Virginia

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Vicenzi: manuscript review and critique. Dr. Isabella Pasquin: analysis of instrumental images, manuscript review and critique. Dr. Alessandro Salviati: case report conception and organization, analysis of data, manuscript review and critique.

Study funding

No targeted funding reported.

Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

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This information is current as of July 9, 2018

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