

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

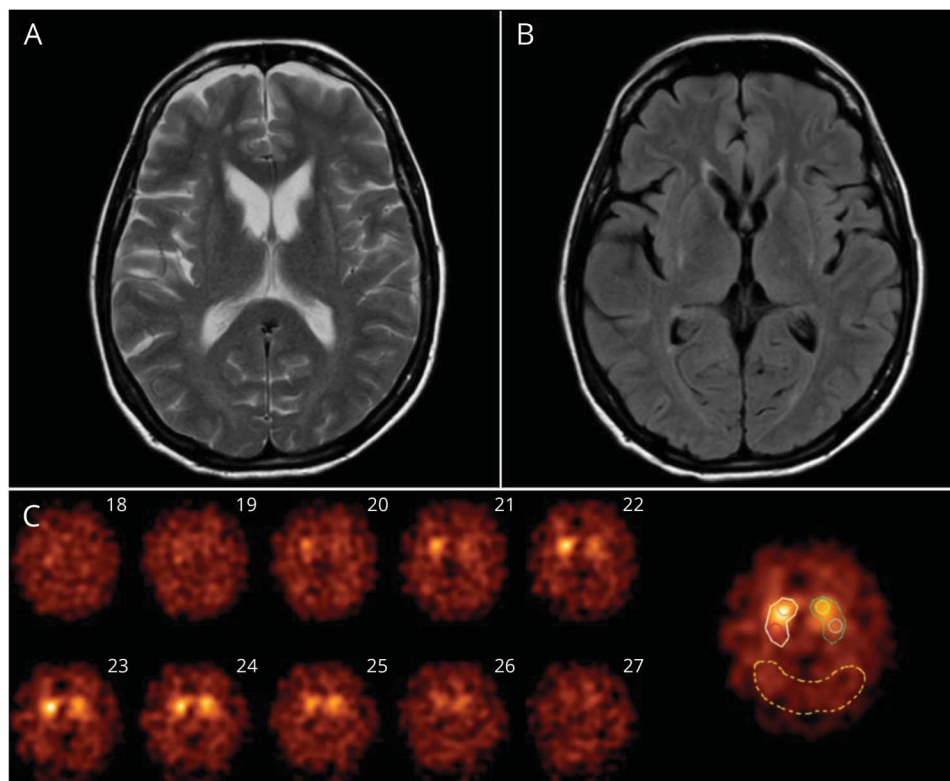
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**Figure** MRI and DaT-SCAN SPECT findings in the adult GM1 gangliosidosis patient



(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 disease<sup>1</sup> (figure, A and B), while DaT-SPECT (<sup>123</sup>I-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.<sup>2</sup> Our report suggests a presynaptic pattern of dopaminergic dysfunction in this disease.

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## 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.

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### Disclosure

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### References

1. Roze E, Paschke E, Lopez N, et al. Dystonia and parkinsonism in GM1 type 3 gangliosidosis. *Mov Disord* 2005;20:1366–1369.
2. Yoshida K, Ikeda S, Kawaguchi K, Yanagisawa N. Adult GM1 gangliosidosis: immunohistochemical and ultrastructural findings in an autopsy case. *Neurology* 1994;44:2376–2382.

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