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Neurol Neuroimmunol Neuroinflamm
2015;2:e168; doi: 10.1212/NXI.0000000000000168

CEREBELLAR VERMIS HYPERMETABOLISM IN OPSOCLONUS MYOCLONUS WITHOUT ONCONEURAL ANTIBODIES

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A 41-year-old woman presented with dizziness and tremor of the right hand. She had myoclonus of the upper limbs and opsoclonus.[1] Brain MRI including enhanced gadolinium sequences was normal, in particular the cerebellum (figure 1, A and B). CSF analysis disclosed 24 cell/mm^3 with 97% lymphocytes and 0.69 g/L of proteinorachia with oligoclonal bands. A workup for infection and neoplasia was negative except for the presence of atypical neuronal antibodies[2,3] in the CSF detected by immunohistochemistry in a reference center (J.H.) (figure 2, A and B). All other common onconeuronal antibodies associated or not associated with OMS were excluded, particularly anti-Ri, anti-Hu, anti-Ma2, and anti-NMDA receptor.[3] The PET performed 8 days after brain MRI depicted a hypermetabolism[4] in the cerebellar vermis, which was no longer observed at 4-month follow-up (compare figure 1, C and D). The patient recovered almost completely by the time of the second PET. This PET pattern is rarely seen. Whether it is associated with opsoclonus myoclonus syndrome or specific immune reaction requires further studies.

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Author contributions: Dr. Legouy: study concept and design. Dr. Taifas: acquisition of data. Dr. Zeghoudi: analysis and interpretation. Dr. Le Stanc and Prof. Honnorat: critical revision of the manuscript for important intellectual content. Prof. Pico: study supervision.

Study funding: No targeted funding.

Disclosure: The authors report no disclosures. Go to Neurology.org/nn for full disclosure forms. The Article Processing Charge was paid by the authors.

Received December 22, 2014. Accepted in final form September 14, 2015.

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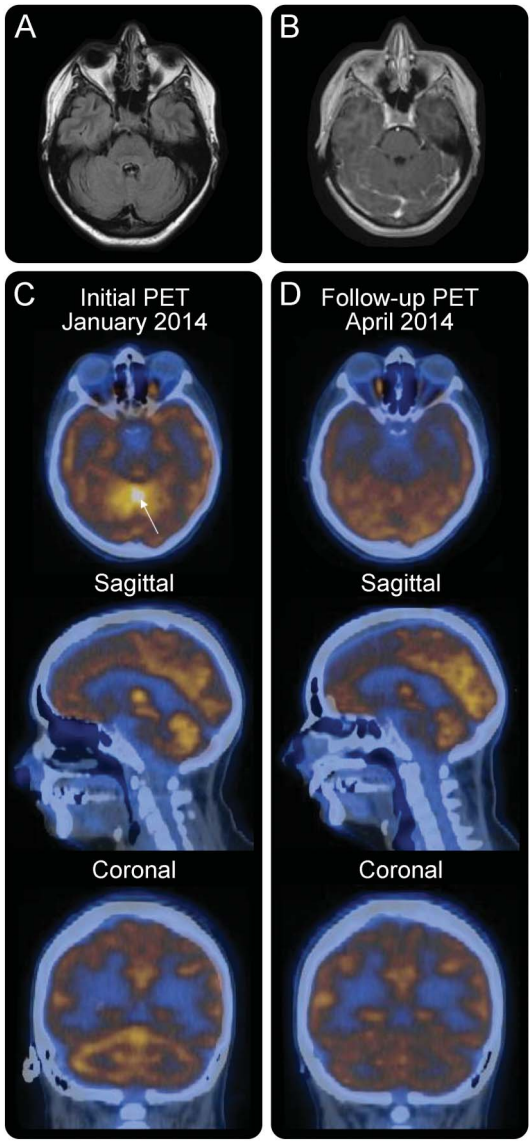
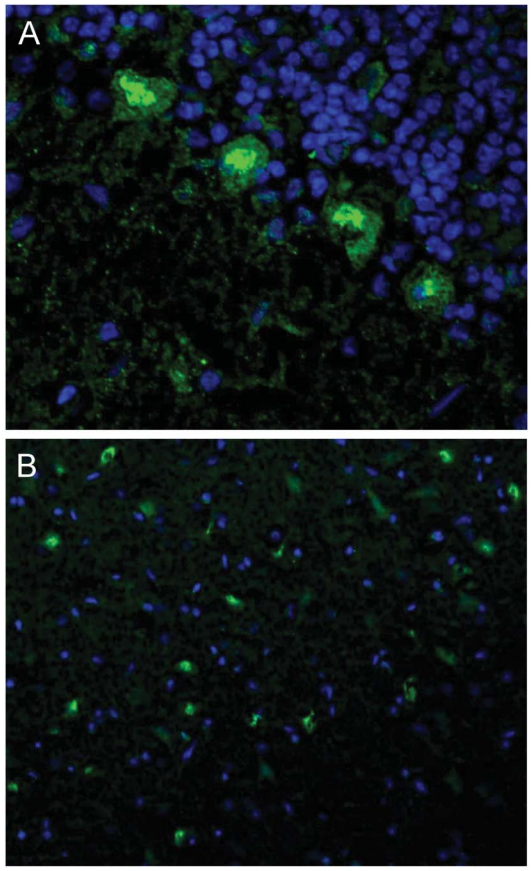
Figure 1 Brain MRI



Brain MRI performed 8 days before PET (A and B). Axial fluid-attenuated inversion recovery (A) and T1 enhanced gadolinium (B) depicted no abnormalities in the cerebellar vermis. Hypermetabolism in the cerebellar vermis (C, white arrow), which disappeared at 4-month follow-up examination (D).

Figure 2 **Detection of atypical anti-neuronal antibodies**



Immunohistofluorescence assay on rat brain sagittal slices incubated with the patient's CSF and revealed by fluorescent anti-human immunoglobulin G. In the striatum, a staining of some neuron nucleus is observed (A, ×200). In the cerebellum, the patient's CSF stained nucleus and weakly the cytoplasm of Purkinje cells (B, ×400).

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