Conclusions

Our findings revealed the presence of a disease-related, sex-specific functional basal ganglia architecture in a cohort of early PD patients. These findings may be related to the presence of different gender-specific nigrostriatal dopaminergic pathways and might be potentially used to predict disease progression over time.

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Alpha-synuclein pathology and enteric glia in advanced Parkinson's disease: A study from gastrointestinal biopsies

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Background and aims

In Parkinson's Disease (PD), recent evidence points towards the involvement of gut-brain axis as one of the primary physio-pathological mechanisms underlying α-Syn aggregation and propagation to CNS. Furthermore, gastrointestinal dysfunctions represent one of the main non-motor symptoms in PD, often preceding the development of proper motor symptoms. Our aim was to investigate the enteric nervous system (ENS) in PD by characterizing α-Syn alterations and glial responses in stomach-duodenum biopsies of PD patients.

Methods

16 patients with advanced PD which underwent Duodopa Percutaneous Endoscopic Gastrostomy and Jejunal Tube (PEG-J) placement were included in the study. A mean of 4 (2 mm3) wall biopsies were sampled from each patient. Immunohistochemistry was performed with anti-aggregated-α-Syn (5G4) and GFAP antibodies. The presence of phospho-α-Syn in conjunction with βIII-tubulin was investigated by immunofluorescence. Morphometrical-semi-quantitative analysis was performed to characterize 5G4+ and GFAP+ density and size. Duodenal control biopsies were included from 8 age-and-sex-matched patients undergoing routine diagnostic endoscopy.

Results

Elevated immunoreactivity for both phosphorylated and aggregated α-Syn was identified in all biopsies of PD patients compared to controls. Phospho-α-Syn partially colocalized with βIII-tubulin, but was found also outside enteric neurons. Evaluation of enteric glia cells revealed an increased size and density when compared with controls ($^{***}P = 0.0002$) suggesting reactive gliosis.

Conclusions

The ENS could be one of the earliest implicated structures in the patho-physiology of PD. The analysis of enteric glia could represent a precocious biological marker of the disease, as its responses to pathological α-Syn could unveil a link between gastrointestinal neural and immune systems in PD inflammation.

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Long-term clinical, neurophysiological and neuroimaging follow-up of a patient with hemiparkinson-hemiatrophy syndrome

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Background and aims

Hemiparkinsonism-hemiatrophy syndrome (HPHA) is a rare form of secondary parkinsonism characterized by unilateral body atrophy associated to early onset ipsilateral parkinsonism, with slow progression, inconstant contralateral involvement and variable response to levodopa. For the first time, we describe a long-term clinical and instrumental follow-up of a patient with HPHA.

Methods

Case report: a 16-year-old male developed a gradually worsening mild slowness and stiffness of left limbs. At age 19, he showed slight rigidity, bradykinesia and distal dystonic posturing on his left side (UPDRS-III motor scores: 30) and mild ipsilateral somatic atrophy. Longitudinal assessment: at baseline, neurological examination after L-Dopa administration, brain MRI, [123I]-FP-CIT DAT-SCAN, genetic analysis for PINK1 and PRKN gene, transcranial magnetic stimulation (TMS) before and after DOPA; during the 18-year follow-up: repeated clinical assessments and instrumental evaluations (i.e. neuroimaging after 15 years, yearly TMS during the first four years).

Results

At baseline: 1. unremarkable brain MRI; 2. negative genetic analysis; 3. bilateral asymmetric (right<left) decrease of striatal transporter binding on DAT-SCAN; 4. significant clinical improvement (UPDRS-III: 11) after 200-mg oral levodopa; 5. reduced TMS short intracortical inhibition in the right hemisphere, unmodified after levodopa. Diagnosis of HPHA syndrome was confirmed and rotigotine, 4 mg/die, was started with clinical improvement. Neurological exam showed a slight worsening of limb dystonia in the 18 years of follow-up (UPDRS-III: 32). No changes were found on TMS, brain MRI and DAT-SCAN.

Conclusions

The present long-term multimodal clinical and instrumental assessment confirmed the slow clinical progression of HPHA, whereas structural/metabolic brain damages and neurophysiological findings remained unchanged.

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Test-retest reliability of static postural balance variables in natural and feet-together stance conditions

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Background and aims

The aim of this study was to investigate the test-retest reliability of postural balance variables during the natural and feet-together stance conditions. It is important to confirm the reliability of postural balance variables because the results of balance tests would be different with different balance test protocols, i.e., stance conditions such as natural and feet-together stances.