IMAGEM EM NEUROLOGIA/IMAGE IN NEUROLOGY

Atypical Parkinsonism Presenting with Isolated Gaze Palsy

Parkinsonismo Atípico com Apresentação Oculomotora Isolada

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We present de case of a 64-year-old male with a 7-year history of slow vertical saccades and eyelid opening apraxia. Two years later, he developed mild relatively symmetrical akinetic-rigid parkinsonism unresponsive to levodopa, without motor fluctuations or dyskinesias. A steady dose of levodopa was nevertheless maintained for the last 5 years without notorious worsening of parkinsonism. In the last two years, mild postural instability became evident. His family also noticed recent behavioural changes without major cognitive impairment. He remained independent for daily activities.

Current exam showed hypomimia, apraxia of eyelid opening and reduced blink rate. Ocular fixation demonstrated frequent square wave jerks, vertical saccades were slow and hypometric and there was partial upgaze restriction superseded with doll's eyes maneuver. Mild rigidity and bradykinesia, without resting tremor was present. His gait had mildly narrow base with shortened stride and bilateral decreased arm swing. Arrising from chair was possible without assistance. The patient took 3 steps back on the pull test but recovered unaided. He scored 24/30 on the Montreal Cognitive Assessment (4-year education). Video-oculography confirmed the bedside findings (**Video 1**).

Brain magnetic resonance imaging (MRI) (1.5 T) revealed the hummingbird sign (i.e., flattening outline of the superior aspect of the midbrain), midbrain atrophy with reduced midbrain to pons area ratio on the midline sagittal plane to approximately 0.14 (normal approximately 0.24) (**Fig. 1A**) and "Mickey mouse" appearance with reduction of the anteroposterior midline midbrain diameter, at the level of the superior colliculi on axial imaging (from interpeduncular fossa to the intercollicular groove: 11.4 mm) (**Fig. 1B**). Dopamine transporter availability evaluation with

Video 1. Eye movement evaluation (part 1): Ocular fixation is unstable, showing frequent square wave jerks. While horizontal saccades are relatively well preserved, vertical saccades are extremely slow and hypometric, more so for downward saccades. Apraxia of eyelid opening is evident. **Evaluation of parkinsonism** (part 2): Mild and relatively symmetrical bradykinesia without resting tremor throughout the evaluation. Evaluation of gait and postural reflexes (part 3): Gait consists of a mildly narrow base with shortened strides and bilateral decreased arm swing. Chair-raise is slightly slower than normal but possible without assistance. The patient recovers unaided from the retropulsion test.

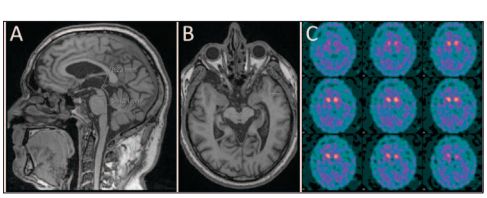

Figure 1. Brain MRI shows hummingbird sign and reduced midbrain to pons area ratio on the midline sagittal plane (Fig. 1A) and "Mickey mouse" appearance with reduction of the anteroposterior midline midbrain diameter at the level of the superior colliculi on axial imaging (Fig. 1B). DaTSCAN shows marked decreased in dopamine transporter availability in the putamen, bilaterally (Fig. 1C).

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1231-2β-carbomethoxy-3β-(4-iodophenyl)-N-(3-fluoropropyl) nortropane (FP-CIT) dopamine transporter SPECT (DaTSCAN) showed marked bilateral putaminal hypometabolism (**Fig. 1C**).

Eyelid apraxia, present from the beginning and aggravated over time, remained the most incapacitating symptom in patient's life. He was already submitted to blepharoplasty 6 years ago without significant improvement and was recently started on botulinum toxin type A, with some benefit.

Progressive supranuclear palsy (PSP) is a clinical syndrome characterized by parkinsonism, supranuclear gaze palsy and postural instability.[1] Its clinical presentation is heterogeneous,[2] probably reflecting the uneven affection of different brain regions.[3] The diagnostic criteria are complex and classify patients according to the predominant clinical features during the first two years of the disease. There are several subtypes, including forms with predominantly oculomotor onset.[4,5] While our patient's symptoms are compatible with the diagnosis of probable PSP, the initial phenotype, examination and clinical course are atypical for the classic presentation of PSP (PSP–Richardson syndrome). Onset with and predominance of oculomotor dysfunction after a 7-year evolution suggests the presence of a PSP subtype with initial predominance of ocular motor dysfunction (PSP-OM). Indeed, PSP-OM can have a more benign course with a significantly lower 5-year mortality rate.[5]

Our patient's MRI revealed several typical signs of PSP. New data regarding specific MRI features in PSP subtypes have been recently published. However, there is no characteristic imaging data concerning PSP-OM subtype.[6] MRI may be useful in the future pointing towards a PSP subtype.

While being a clear representation of PSP heterogeneity, this case shows that the careful determination of the PSP subtype in an individual patient might carry relevant implications for the prognosis and it is essential to offer a tailored prognosis to our patients and families.[2] ■

Contributorship Statement / Declaração de Contribuição

CI: Conception, writing and final approval.

AJ: Conception, writing and final approval.

JL: Critical review with intellectual contribution; and final approval.

FM: Conception, writing critical review and final approval.

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