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CASE REPORT

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Atypical parkinsonism and self-mutilation: A new lens on the old concept

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Abstract

We report a case of atypical parkinsonism and self-mutilation.

KEYWORDS

atypical parkinsonism, movement disorders, Parkinson, self-mutilation

1 | INTRODUCTION

Atypical parkinsonism defines a syndrome that consists of parkinsonian features, such as akinesia, rigidity, and tremor, and additional clinical signs which are atypical for Parkinson's disease.[1] These signs including early cognitive decline, severe dysautonomia, early falls,[2] but self-mutilation has not been reported in atypical parkinsonism. Herein, we report a case of atypical parkinsonism and self-mutilation.

2 | CASE REPORT

A 69-year-old woman was referred to our Movement Disorders Clinic because of vertigo and bradykinesia in the last 4 months. She complained of right-hand tremor for 3 months, several episodes of fall, insomnia, memory decline, depression, and urinary retention and frequency the last few months. Also, she reported self-mutilation behavior for 2 years such as scratching her skin around the umbilical area, which was not accompanied by itching, and she felt relaxed after looking at her blood. (Figure 1) She denied any history of hallucination, obsessive-compulsive behaviors and other psychiatric disorders, and REM sleep behavior disorders.

She has a history of diabetes mellitus, hypothyroidism, hypertension, bariatric surgery 7 years ago, and cholecystectomy 2 years ago. She was taking Metformin, Insulin, Metoprolol, Levothyroxine, and Fluoxetine. Her family history was unremarkable.

On clinical examination, she had bradykinesia, bilateral jerky tremor on hands during rest and postural holding, rigidity, wide-based gait, stooped posture, Pisa syndrome, and positive pull test, also, her MOCA was 22. Furthermore, her distal extremities were cold and pale, and her systolic and diastolic blood pressure fell 30 mm/Hg and 20 mm/Hg, respectively, after 3 min standing. She did not have any

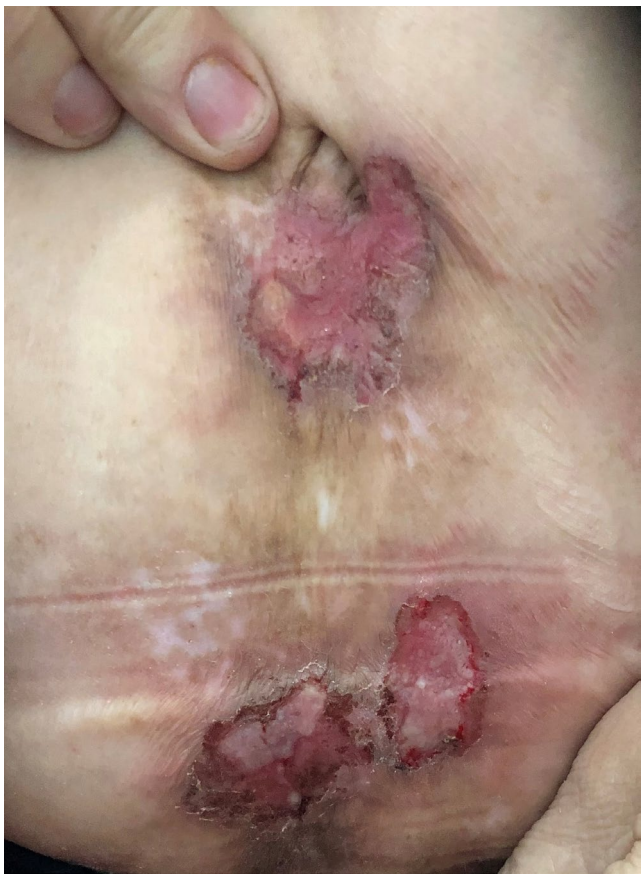

FIGURE 1 Self-inflicted wounds of the abdomen on the periumbilical area

minipolymyoclonus. Brain MRI showed non-specific white matter lesions, but no cerebellar or pontine atrophy at that time. She did not have dystonia, chorea, and pyramidal signs.

We started the following regimen: Levodopa/Benzenoid 100/25 mg gradually increased to four times a day, and Donepezil 2.5 mg a day. On follow-up, the severity of self-mutilation decreased but was not completely resolved, and her parkinsonism improved, also, she had not experienced falling again, but she increased levodopa/Benzenoid herself to 1.5 tablets four times a day, which caused reemerge of unsteadiness and falling and worsening of self-mutilation, and reducing Levodopa/Benzenoid to the previous dosage improved falling but not self-mutilation. On the one-year follow-up, she remained stable on the current regimen in addition to rehabilitation due to wide-based gait and unsteadiness, but still had self-mutilation, which was not bothering her. She developed incontinency, but her cognition remained stable.

3 | DISCUSSION

The well-known differential diagnosis of parkinsonism and self-mutilation is Lesch-Nyhan Syndrome (LNS), which has a central deficiency of dopamine similar to parkinsonism, the age at which dopaminergic neurons are disturbed can explain the different symptoms detected in these two conditions.[3] Upregulation of the striatal D2 receptor occurs in LNS, which is comparable to what can be seen in Parkinson's disease (PD). Therefore, the pathophysiology of hyperkinetic involuntary movements of LNS might be alike to levodopa-induced dyskinesia.[4] Moreover, there are reports that showed bilateral stimulation of the globus pallidus internus (GPi) can improve self-mutilation in LNS, which points to the fact that self-injurious behavior may be associated with basal ganglia dysfunction.[5]

Self-injurious behavior is mostly seen in children and adolescents and infrequently initiates in adulthood. Self-injurious behavior is mainly related to tic severity, obsessive-compulsive disorder, and attention-deficit/hyperactivity disorder,[6] which further supports the role of dopamine in the pathophysiology of self-mutilation behavior.[7] Correspondingly, the key neurotransmitter underlying repetitive behaviors is dopamine, thus abnormalities of dopaminergic neurotransmission may be a risk factor for self-injurious behavior.[8]

Also, self-injurious behavior was recognized in a patient with juvenile parkinsonism who had a F-box Protein 7 (FBXO7) mutation syndrome and received rasagiline therapy. In addition, drug-related self-injurious behavior was also described in a parkinsonian patient after increasing ropinirole dosage.[8]

Two cases with SCA17 have been reported, both of them had chorea, ataxia, and cognitive decline. Although the patients did not have parkinsonism, parkinsonism can a manifestation of SCA17.[7]

Consequently, self-mutilation is theoretically possible in parkinsonism, and to the best of our knowledge, this is the first report of self-mutilation in a patient with atypical parkinsonism, that would lead to a better understanding of the underlying pathophysiological mechanism.

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CONFLICT OF INTEREST

None.

AUTHOR CONTRIBUTION

(1) Research project: A. Conception, B. Organization, C. Execution. **(2)** Manuscript: A. Writing of the first draft, B. Review and Critique. **Mehri Salari:** 1A, 1B, 1C, 2A. **Masoud Etemadifar:** 1A, 1B, 1C, 2B. **Kimia Ghanbari:** 1A, 1B, 1C, 2A.

ETHICAL APPROVAL

We hereby confirm that the present study conforms to the ethical standards and guidelines of the journal.

CONSENT

The patient has given written and informed consent for online publication of her picture.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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