CASE REPORT

Parkinson’s Disease Presenting with Oculogyric Crisis in the Off Period

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Abstract

We herein report the case of a 67-year-old Japanese man diagnosed with sporadic Parkinson’s disease (PD) at 52 years of age who presented with oculogyric crisis (OGC) in the off period. Ordinarily, OGC is caused by postencephalitic parkinsonism or the chronic use of antidopaminergic medications. The OGC began at 65 years of age and was associated with the wearing-off of symptoms. The dominant OGC feature was tonic deviations in eye posture induced by looking upward with prominent retrocollis. The administration of control dopaminergic medications led to improvements in the wearing-off phenomenon and OGC. This observation confirms that sporadic PD can induce OGC in the off period.

Key words: Parkinson’s disease, oculogyric crisis, dystonia, wearing-off, off period

(Intern Med 53: 793-795, 2014)
(DOI: 10.2169/internalmedicine.53.1233)

Introduction

Oculogyric crisis (OGC) is characterized by a maximal upward deviation of the eyes in a sustained fashion lasting from a few seconds to hours (1). OGC is considered to be a form of extraocular muscle dystonia (2). OGC is classically caused by postencephalitic parkinsonism or pharmacological effects, including those of antipsychotic agents (2, 3). However, there are only a few reports of OGC in patients with Parkinson’s disease (PD) (4-6). Furthermore, in previous reports with case descriptions, OGC in sporadic PD was observed during the peak-dose period (4, 6). No studies have found an association between the incidence of OGC in sporadic PD and the off-period phenomenon. We herein report the case of a 67-year-old Japanese man diagnosed with sporadic PD who presented with OGC in the off period.

Case Report

A 67-year-old Japanese man was diagnosed with sporadic PD at 52 years of age. His initial symptoms included bradykinesia, muscular rigidity and a resting tremor on the right side of the body. Previously, he had been well, with no family history of neurological disorders in his parents, brothers, sons or other relatives, and had not received any antidopaminergic or antipsychotic medications. Based on the clinical findings, the patient was diagnosed with PD. He was initially started on levodopa, which resulted in a significant improvement of his symptoms.

After 10 years of levodopa therapy, the duration of benefit of the dosing cycle of each medication started to decline (wearing-off phenomenon). Moreover, the patient would complain of OGC as each dose of levodopa wore off (within two hours after taking the medication). The dominant OGC feature was tonic deviation in the posture of the eyes induced by looking upward, with prominent retrocollis (Figure). No other involuntary movements were reported, and no impairment of consciousness was observed.

The patient was referred to our hospital due to a progressive wearing-off phenomenon at 67 years of age. Parkinsonian features, such as bradykinesia, muscular rigidity, resting tremors, mild dysarthria and a disorder of postural reflexes, had developed. He had normal intelligence, with a score of 27/30 on the Mini-Mental State Examination. The investigations included routine laboratory tests of blood and urine, an examination of the cerebrospinal fluid, electroencephalography and magnetic resonance imaging, all of which were normal. [123I]-Metaiodobenzylguanidine imaging revealed a decreased heart-to-mediastinum ratio in the early
Figure. The dominant OGC feature was tonic deviation in the posture of the eyes induced by looking upward, with prominent retrocollis.

and delayed stages (1.47/1.28). The patient was treated with levodopa (600 mg/day), entacapone (400 mg/day) and pramipexole (1.125 mg/day). Adjustments in the schedule of levodopa dosing (current intake, 600 mg/day) and the doses of entacapone (600 mg/day) and pramipexole (2.25 mg/day), as well as the addition of zonisamide (25 mg/day), led to an improvement in the wearing-off phenomenon and OGC.

Discussion

Although OGC caused by sporadic PD has been reported in a few cases, it usually occurs in the peak-dose period of levodopa therapy (4, 6). This case is quite unusual in that the OGC occurred in the off period of levodopa treatment. The pathomechanisms of OGC remain unclear. However, both hypodopaminergic (resulting in relative cholinergic overactivity with supersensitivity of muscarinic receptors) and hyperdopaminergic states have been suggested (7). The basal ganglia may be viewed as the principal subcortical component of a family of circuits that link the thalamus and cerebral cortex, which includes skeletal motor circuits, oculomotor circuits, prefrontal circuits and limbic circuits (8). These pathways appear to play a role in OGC generation (9). OGC has been reported in patients with focal lesions in the basal ganglia and thalamus (10). A dopaminergic state in the basal ganglia can affect these pathways, which may induce OGC as a form of focal dystonia.

In this case, the OGC likely occurred as a symptom of off-period dystonia for two reasons: (i) OGC and cervical dystonia were observed in the off period (OGC and dystonia are closely related to each other) (11) and (ii) reductions in the off period and frequency of OGC were observed following the additional administration of dopaminergic medications.

In addition to that observed in our case, OGC has been noted in both the peak-dose period and off phase (4, 6). This is because (i) in the peak-dose phase, the relative excess of dopamine in the striatum causes an imbalance in the activity between the direct striatal pathway and indirect pathway (12), while (ii) in the off phase, the loss of dopamine content in the striatum potentially leads to a paucity of movement, thereby resulting in a fixed dystonic posture (13). In other previous reports, OGC occurred during the on phase (4, 6); thus, it is possible that, in our case, the OGC may have occurred in a phase of changing the dopamine concentration.

Although further studies are needed to investigate the pathomechanisms of OGC, physicians should be aware of the possible occurrence of this condition in patients with sporadic PD associated with off period dystonia. Our observations also confirm that OGC can be improved by the administration of dopaminergic medications.

The authors state that they have no Conflict of Interest (COI).

Acknowledgement

This work was supported by a grant from the Ministry of Health, Labour and Welfare of Japan and the Ministry of Education, Culture, Sports, Science and Technology of Japan to K. Okamoto.

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