A case of hedonistic homeostatic dysregulation

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Abstract

Parkinson’s disease (PD) is a common condition especially amongst the elderly population, which is increasing. I present a case of an unusual but also under-recognised syndrome caused by PD therapy, which can have devastating effects on the patient and their family.

Keywords: Parkinson’s disease, hedonistic, homeostatic, dysregulation, dopamine, elderly

Case report

A gentleman, currently 73 years old, was diagnosed 12 years previously with Parkinson’s disease (PD), and he was started on Sinemet with some response. Five years later, Cabergoline a dopamine agonist was added. Two years on, his wife noted he had periods of overactivity, for example, hammering nails into pieces of wood late at night, sexual disinhibition, frequent sudden onsets of sleep and depression. Examination at that time revealed dyskinesias, and so, his PD drug regime was decreased.

One year on, due to increasing fluctuations (on/off periods), he was tried on an Apomorphine infusion (dopamine agonist), but this made him hypersexual and so it was stopped. Cabergoline was also stopped as it was thought to be contributing to his erratic behaviour although this did result in him slowing down.

Three years later, he remained very dyskinetic with bizarre behaviour such as walking around the house naked, talking nonsense for up to an hour at a time and trying to sweep the whole street clear of leaves. He would often get angry and aggressive with his wife and on occasions was hallucinating. At this time, he was unclear how much of his medication he was taking, but it was thought to be equivalent to a total daily dose of 2.53 g of Levo-dopa.

Reviewing him this year, he remains markedly dyskinetic which he fails to recognise as such and therefore takes extra Madopar. He can however have freezing episodes. He continues to have bizarre behaviour such as pushing soil in a wheelbarrow down the street early in the morning, resulting in him falling. He remains on large doses of Levo-dopa and Entacapone but no dopamine agonists. His wife has regular respite care to give her a break.

Discussion

This gentleman displays the classic features of hedonistic homeostatic dysregulation (HHD). This syndrome sometimes called dopamine dysregulation syndrome (DDS) was first described by Giovannoni et al. in 2000 as a neuropsychological disorder associated with substance misuse and addiction to dopaminergic replacement therapy (DRT). It occurs mostly in young-onset male patients with PD. They described a set of core features, which are pathological gambling or shopping, hypersexuality, punding (repetitive and purposeless motor acts), walkabout (walking great distances aimlessly and devoid of specific purpose with awareness of time), alterations in appetite, drug hoarding and social independence or isolation. Lawrence et al. put forward various theories as to how reward systems go awry in compulsive use of DRT.

Diagnostic criteria of HHD Syndrome due to DRT misuse by Giovannoni et al. 2000

- Need for increasing doses of dopamine replacement therapy (DRT) in excess of usual requirements
- Pattern of pathological use
- Impairment in social or occupational functioning
- PD with L-dopa responsiveness
- Development of hypomanic, manic or cyclothymic affective syndrome in relation to DRT
- Development of a withdrawal state
- Duration of disturbance >6/12

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The prevalence is said to be up to 4% of PD patients [2, 5]. In practical terms, it is a difficult disorder to treat [2, 4]. Keeping the lowest medication dose required to overcome motor disabilities is necessary, but quick acting DRT such as dispersible Levo-dopa and intermittent injections of Apomorphine should be avoided. Long-acting DRT including Apomorphine infusions could be tried instead. Other forms of treatment such as psychiatric or psychotherapy especially if depressed or suicidal should be sought, and atypical antipsychotics (e.g. Quetiapine) in manic episodes can be used. Although deep brain stimulation (DBS) may be thought to be useful as it leads to much less DRT requirements, there have been reports of compulsive use of DBS [6].

There may need to be a period of in-patient admission whilst weaning down the medication. On/off diaries can be used for positive reinforcement. A watch needs to be kept for surreptitious DRT ingestion, and on discharge, regulation of drug prescriptions is required as often the patient will try to obtain their drugs from different prescribers for example, their general practitioner and their specialist. Ultimately, prognosis is poor [4].

**Key points**
- HHD is a little known and under-recognised syndrome even by PD specialists and PD nurses.
- It is a very difficult syndrome to treat and is rarely cured.

**Conflicts of interest**
None.

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**References**