

Deep Brain Stimulator Withdrawal Syndrome

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Objectives

*Illustrate the presentation and pathogenesis of Parkinson Hyperpyrexia Syndrome (PHS).

* Present a very uncommon case of fever related to DBS malfunction in patient with advanced Parkinson's disease

Parkinsonism hyperpyrexia syndrome (PHS) is a neurologic emergency that mimics neuroleptic malignant syndrome. It commonly presents as systemic inflammatory response syndrome (SIRS). The most common trigger for PHS is reduction or withdrawal of anti-Parkinson's medications, especially levodopa. Rare causes of PHS include DBS malfunction due to battery depletion which was reported only in few occasions. This case of PHS was due to DBS battery depletion that presented as sepsis and was successfully treated with changing the DBS battery.

The patient is a 67-year old female that was diagnosed with PD in 1991. Over the years, her treatment included levodopa/carbidopa and pramipexole. In 2007, bilateral STN DBS was implanted. The patient presented to the ER with high-grade fever, altered mental status, poor oral intake. She was febrile 38.5°C, had autonomic instability. Her neurologic exam demonstrated somnolence with lack of response to painful stimuli. Laboratory tests showed, elevated creatine phosphokinase at 1015 U/L. Normal findings on lumbar puncture ruled out CNS infection. On day nine of admission, PHS was suspected due to non-resolving high fever, severe muscular rigidity, altered mental status, autonomic instability and elevated CK levels of 1615 U/L. The patient was treated conservatively; levodopa dose was tripled, with no clinical improvement. Given that the estimated DBS battery life is between three and five years, DBS withdrawal syndrome due to battery depletion was suspected. She underwent successful IPG replacement with rapid clinical improvement within a few hours, all lab values normalized. The patient's rigidity and mental status improved to full recovery until discharge.

Malignant DBS withdrawal syndrome, is a rare disease exclusive to patients with advanced PD as a result of abrupt cessation of DBS activity. Treatment by augmenting the dopaminergic medications should be considered temporary, while immediate DBS

C1

restoration is considered the definitive treatment, preventing an otherwise fatal outcome.