Parkinsonism Hyperpyrexia Syndrome and Deep Brain Stimulation

Deep Brain Stimulation of the subthalamic nuclei (STN-DBS) represents one of the most effective treatments in patients with Parkinson's disease (PD) with unsatisfactory response to pharmacological therapy. STN-DBS is a neurosurgical approach based on the implantation of electrodes in specific targets of the brain. By means of an electrical pulse generator (a sort of "brain pacemaker") electrical impulses are used to interrupt the aberrant neuron function in PD patients. This therapy proved to have a long-lasting efficacy on motor symptoms in a selected group of parkinsonian patients but some issues related to surgical procedure, to electrical stimulation and/or to hardware dysfunctions need to be taken into account. One of these issues is represented by a very rare syndrome, called Parkinsonism Hyperpyrexia Syndrome (PHS), potentially occurring after the sudden interruption of the stimulation, due to hardware failure or battery depletion. PHS is a life-threatening disorder previously described in association to the withdrawal of dopaminergic therapy, such as levodopa and dopamine-agonists.

In our paper we report the new case of a 63-year-old man with an 18-year history of PD treated by STN-DBS for 5 years, with optimal control of his PD symptoms. Due to battery depletion of the electrical pulse generator and the subsequent sudden interruption of the brain stimulation, the patient rapidly developed a severe rigidity with inability to stand and walk, eating and speaking
difficulties and the onset of tachycardia, hypertension, anxiety and fever. The administration of high doses of oral therapy was not sufficient to compensate the electrical stimulation withdrawal. It was only after the surgical impulse generator replacement, four days after symptom onset, that he began his recovery.

Only other 5 cases of PD patients developing PHS after DBS discontinuation were described in literature. Fatal outcomes, reported in 50% of the cases, were associated with delayed or no restoration of DBS and with older age. Predisposing factors for the development of PHS remain unclear, but we can conclude that the abrupt discontinuation of DBS increases the risk for life-threatening PHS, and rapid restoration of DBS represents the most important therapeutic measure.

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