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Case report

Life-threatening parkinsonism-hyperpyrexia syndrome following bilateral deep brain stimulation of the subthalamic nucleus

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ABSTRACT

Parkinsonism-hyperpyrexia syndrome (PHS), or neuroleptic malignant syndrome (NMS), is a neurophysiologic reaction to the acute withdrawal/decrease of central dopamine levels. It is a severe complication characterized by rigidity, change in consciousness level, fever, hypertension, and autonomic instability, that can be fatal. To the best of our knowledge, PHS following deep brain stimulation (DBS) of subthalamic nucleus (STN) surgery due to anti-Parkinson drug discontinuation has been previously reported only six times. Half of these cases resulted in fatalities. Herein, we report on an early diagnosed case of PHS following bilateral STN-DBS which was successfully treated with the administration of dopamine agonists, fluid replacement, and activation of DBS.

1. Introduction

Deep brain stimulation (DBS) of the subthalamic nucleus (STN) is an established option for the treatment of advanced Parkinson's disease (PD). Patients with tremor-affected quality of life, medically uncontrollable motor fluctuations, or dyskinesia are candidates for DBS [1]. These patients usually have a long history of multiple, high-dose, anti-PD drug usage [2]. Prior to DBS surgery, the anti-PD drugs are temporarily discontinued to better observe patients' responses during surgery in most centers [2]. There is also a common tendency to reduce the use of anti-PD drugs postoperatively. The sudden withdrawal of anti-PD drugs may trigger a severe, fatal complication known as parkinsonism-hyperpyrexia syndrome (PHS), which is characterized by rigidity, change in consciousness level, fever, hypertension, and autonomic instabilities [2,3]. To the best of our knowledge, although anti-PD drug withdrawal is routinely used in clinical practice,

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to date, only six cases of PHS have been reported following STN-DBS surgery [2–7]. Fifty percent of these cases resulted in fatalities despite aggressive medical treatment efforts [3–5]. Herein, we report on a case of PHS which was recognized early on, following bilateral STN-DBS, and successfully treated with the administration of dopamine agonists, fluid replacement, and DBS activation.

2. Case report

A 61-year-old male with a 14-year history of PD was admitted for DBS surgery. His symptoms had progressed over the years with a predominant tremor. The Unified Parkinson Disease Rating Scale Part III (UPDRS-III) showed an improvement of 24% on the levodopa test. Although motor response to levodopa test is accepted as the primary selection criteria for DBS in Parkinson disease, in our patient who do not meet the 33% threshold, DBS was performed for medication-refractory tremor. The patient had been taking levodopa-benserazide, 600–150 mg/day, trihexyphenidyl 6 mg/day, and amantadine 300 mg/day preoperatively. 12 h before the operation, medications that the patient had been taking were withdrawn in order to observe the clinical response to the microelectrode stimulation. On the day of surgery, following magnetic resonance imaging (MRI) of the brain, a stereotactic frame (Integra CRW, New Jersey, USA) was placed onto patients head and patient underwent computed tomography (CT) scan. Then, the MRI and CT images were auto-fused using an image fusion program (Atlas Integra Software, New Jersey, USA) in order to target anterolateral side of right and left STN. Bilateral microeletrode recordings and macro stimulation tests were performed (Alfa Omega NeuroNav Micro Recording System, Nazareth, Israel). DBS electrodes were placed then bilaterally to anterolateral sides of both STNs (St. Jude Medical 6149 40 cm lead, Minnesota, USA). The cranial skin incisions were closed and the patient was brought to the radiology unit. A control cranial MRI revealed that the left sided STN electrode position found to be inappropriate (Fig. 1a and b). The left sided electrode was corrected under microelectrode recording and macro stimulation tests. Then permanent electrode positioning was confirmed with cranial MRI (Fig. 2a–c). The stereotactic frame was removed, the patient was intubated. Under general anesthesia, the impulse generator (St. Jude Medical Libra XP 6644, Minnesota, USA) was implanted and connected to the leads. The total length of this procedure was 12 h. Our protocol dictates that every patient who undergoes DBS surgery is routinely followed up overnight in the intensive care unit (ICU). Starting from the sixth postoperative hour in the ICU, the patient suffered from a high fever (up to 39 °C), confusion, generalized tremor, elevated blood pressure, and extreme axial rigidity. Autonomic dysfunctional attacks presented with tachycardia, blood pressure fluctuations, and diaphoresis. Deterioration of consciousness was also encountered. Intravenous (IV) hydration and empiric antibiotic treatment (vancomycin 4×500 mg and meronem 2×2000 mg) were administered immediately to counteract a possible central nervous system infection, although it is unusual in such an early postoperative period. A postoperative cranial CT scan showed no abnormalities except mild pneumocephalus and

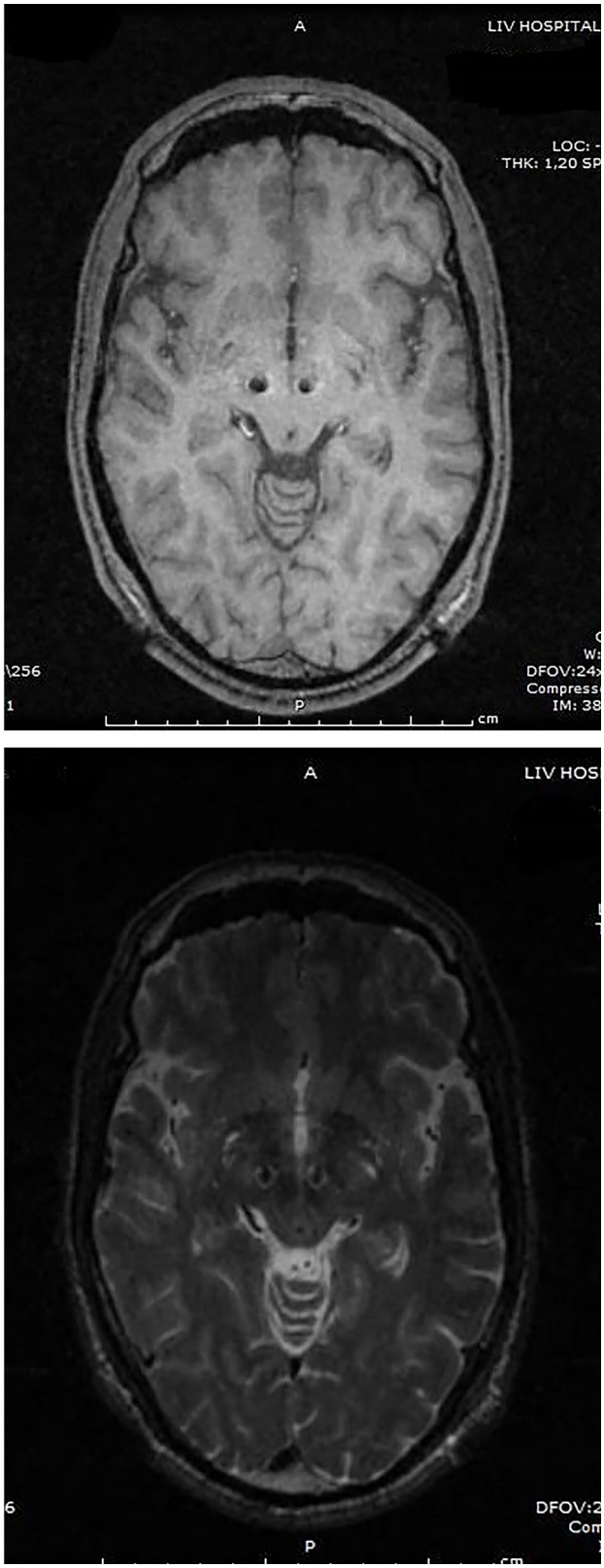

Fig. 1 – (a) and (b) Control MRI scan showed that the position of the depth electrode is appropriate on the right side, however too much medially located in the left side as shown by the axial T1 (a) and T2-weighted (b) images.

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