STATUS DYSTONICUS IN TARDIVE DYSTONIA SUCCESSFULLY TREATED BY BILATERAL DEEP BRAIN STIMULATION

(Short report)

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BFMDRS = Burke-Fahn-Marsden Dystonia Rating Scale; DBS = deep brain stimulation; EPS = Extrapyramidal symptoms; GPI = internal part of globus pallidus; SD = status dystonicus

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Case report

We present the case of an 18-year-old boy, who was treated by risperidon (up to 8mg/day) and clonazepam (up to 4mg/day) combination because of schizophrenia since the age of 16. Due to an acute psychotic episode up to 5 mg/day haloperidol was introduced as an add-on therapy to risperidon resulting in acute oculogyric symptoms in July, 2009. After a usage of 16 days, the haloperidol medication was completely stopped, whereas biperiden (2mg tid) was introduced. Subsequently, risperidon was also replaced by olanzapine (10mg/day). However, severe segmental dystonia developed with the combination of retrocollis, toricollis and oculofacial dyskinesia in September, 2009. Because brain MRI was negative and Wilson’s disease, DYT-1 positive dystonia or acoruloplasminemia could not be diagnosed in the background, we proposed a tardive etiology. Meanwhile his tardive dystonia showed slow progression under the combination of olanzapine (10mg/day) and biperiden (10mg/day), therefore, this combination was replaced by clozapine (225mg/day) in October, 2009.

In December 2009, the patient was admitted to our neurological ward in the condition of status dystonicus (SD). The dystonic symptoms differed completely from the previously observed ones. More pronounced and long-lasting fixed dystonic postures were accompanied by continuous, disabling dyskinetic movements of the face and all extremities resulting in extreme pain and several self-injuries (e.g. extremities bumped into furniture). Despite of elevated level of creatine-kinase (CK, 871U/l), no other clinical signs of neuroleptic malignant syndrome could be identified. Therefore, the elevated CK level was thought to be the consequence of extreme muscle activity and injuries. No obvious provoking factor (e.g. infection, metabolic alterations) could be identified in the background of acute worsening.

Because the disabling hyperkinetic symptoms interfered with sleeping and resulted in increased cardiovascular demand (e.g. tachycardia) and extreme pain to the patient, first bolus clonazepam and later continuous intravenous midazolam was applied for achieving an
immediate relief (up to 500mg/day). However, whenever we tried to lower this light sedation, the dystonic symptoms reappeared in a more aggravated manner. Because clonazepam and midazolam were associated with a worsening in the dystonic symptoms after their temporary effects vanished, we switched to continuous propofol sedation (up to 6000mg/day). Because neither light-, nor deep sedation improve the life-threatening symptoms of SD, we decided to implant Medtronic 3389 electrodes into the internal part of globus pallidus (GPI) bilaterally. Few days after initiating GPI deep brain stimulation (DBS), dystonic symptoms began to dramatically improve. Postoperatively the patient received clozapine (225 mg/day) whereas the improvement in dystonic symptoms remained persistent. (Table 1).

The patient and his caregiver gave informed consent to present his case and videos in scientific journals according to the Declaration of Helsinki.

Discussion

Dopamine-receptor-blocking antipsychotic drugs may cause various acute and delayed-onset movement disorders including acute dystonia, parkinsonism, akathisia, tardive dyskinesia and tardive dystonia. Despite tardive movement disorders were described 50 years ago, their medical treatment is still disappointing in some cases. However, a growing number of evidence suggests that tardive dystonia and/or dyskinesia might be an excellent indication for bilateral GPI-DBS with an improvement of 65-100% measured by Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS)[1-3].

Dystonic storm or status dystonicus is an extremely rare form dystonia representing an emergency situation. Severe, disabling and long-lasting muscle contractions characterize SD resulting in increased cardiopulmonary demand, unbearable pain, elevated creatine-kinase levels, and sometimes spontaneous femur fracture[4]. Despite immediate and adequate treatment including sedation, intrathecal baclofen pump or DBS, the mortality of SD is still
approximately 10%[5]. SD is usually triggered by metabolic disturbances, surgery, infections or changes in medication. In our case, however, no provoking factor could be identified.

The present paper might have some interesting observations: To our knowledge, this is the first published case where tardive dystonia resulted in status dystonicus. As mentioned by Mariotti et al[5], benzodiazepine drugs seemed to have a worsening effect on the dystonic symptoms. Whenever the temporary effect of clonazepam or midazolam vanished or were suspended, the dystonic symptoms reappeared in an aggravated manner compared to the severity before these drugs were administrated, which could be interpreted as a possible rebound phenomenon. Our case further supports the impression that drug-refractory tardive symptoms might be good indication for GPI-DBS, even in cases where these symptoms evolve to SD[3].
References


Declaration of interests

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