Pallidal neuronal activity in myoclonus-dystonia syndrome

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Introduction

Pallidal stimulation with high frequencies has proven to be an option in the treatment of therapy-refractory patients with myoclonus-dystonia (MD). Hitherto, there are few reports (see references) on pallidal neuronal activity in this rare movement disorder. The aim of the present study was to further characterize pallidal activity in MD patients. We obtained single-cell activity as well as local field potentials (LFPs) from the external and internal pallidum (GPe and GPi) of two awake MD patients (subjects M+G) and one generally anesthetized MD patient (subject P) undergoing microelectrode-guided stereotaxy for the implantation of deep brain stimulation electrodes in the GPs.

Patients and Methods

1.1) Summary of mean frequencies of all neurons recorded from striatum, GPe and GPi. The individual data points are plotted as a function of distance from target (i.e., GPi).
1.2) Representative magnetic resonance images, taken from subject P. Reconstruction of the trajectory as probe view (‘surgeon’s view’) to show the internuclear boundaries (medullary laminae) as visible on the MRI.
1.3) The coefficient of variation of the interspike intervals (CV) (I(3)) can be taken as a measure of irregularity of neuronal firing. Large values indicate irregular activity, whereas low CV values point at regular spiking activity. The data points of all three patients are plotted as a function of depth.

Figure legends of Results I+II

Figure I(I) demonstrates the presence of rhythmic bursting in a pallidal neuron recorded from the GPi of patient G. The power spectral analysis of the spike train allows the identification of oscillatory activity at around 4-5 Hz, interestingly even in the absence of noticeable myoclonic activity in the limbs (as evidenced by multiple EMG-recordings).

Results I

The firing rate analysis revealed some distinct patterns of activity-levels for these three patients. GPI neurons of patient G fired significantly less than external pallidal neurons (GPI: 8.8±1.6 Hz, mean±SEM, n=15; GPi: 26.1±4.0 Hz, n=13). The firing rates of cells in the GPi of patient M were statistically not different from those obtained in the GPe (GPi: 61.9±8.8 Hz, mean±SEM, n=13; GPe: 31.6±7.3 Hz, n=11). The firing rates of cells in the GPi of patient P were statistically not different from those recorded under general anesthesia.

Results II

Neuronal activity in both the GPe and GPi was modulated by ongoing muscle activity. Tonic neuronal activation preceded episodes of dystonic contractions. Moreover, bursts of oscillatory activity accompanied episodes of rhythmic myoclonic jerks. Cross-correlation analysis of simultaneously recorded neurons revealed transient oscillatory synchrony of pallidal neuronal activity in the 2-3 Hz frequency range during dystonic activity. In contrast, cross-correlograms of unit activity in the absence of muscle activity were flat.