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Successful pallidotomy for post-hyperglycemic hemichorea-ballism

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Chorea and ballism both refer to random abrupt involuntary movements, with choreiform movements typically being more distal and of lower amplitude than ballism. Stroke and heritable diseases (including Huntington's disease) are well-known etiologies, although metabolic, infectious, inflammatory and drug-induced causes have also been decribed [4].

First described in 1960, post-hyperglycemic chorea-ballism (PHGCB) or diabetic striatopathy is now the second most common cause of hemiballism [2,4]. This complication of severe acute hyperglycemia combines (sub)acute-onset hemichorea-ballism and mostly contralateral striatal hyperintensities on T1-weighted magnetic resonance imaging (MRI). PHGCB typically affects patients with unrecognized or undertreated type 2 diabetes, who suffer hyperglycemic/hyperosmotic episodes. There may be a preponderance in elderly Asian women [2,4,5].

Pathophysiologically, a possible mechanism may be that hyperglycemia induces depletion of gamma-amino-butyric acid (GABA), thereby causing disinhibition of the basal ganglia [3]. Although prompt glycemic control quickly resolves most cases, PHGCB may persist, worsen, or recur. In these cases, pharmacotherapeutic options include dopamine-depleting drugs, GABAergic drugs and selective serotonin reuptake inhibitors [2,4,5]. Surgical ablation and deep brain stimulation (DBS) have been rarely reported in medication-refractory cases [1,5].

Herein, we report on a 70-year old right-handed Caucasian woman, who developed subacute left-sided PHGCB 5 years earlier during a non-ketoacidotic hyperglycemic episode (maximal serum glucose 16.2 mmol/L; HbA1c 11.7%), as the presenting symptom of type 2 diabetes. MRI revealed right-sided striatopallidal T1-hyperintensities (Fig. 1A, Video 1).

Supplementary video related to this article can be found at https://doi.org/10.1016/j.parkreldis.2018.11.023.

Her diabetes was medically controlled, and the PHGCB symptoms considerably improved over the following months. However, three years later, poor insulin compliance subsequently triggered a second hyperglycemic episode and PHGCB symptom recurrence, this time predominantly on the right side. Symptoms worsened with distraction, motor activation, and stress. Total Abnormal Involuntary Movement Scale (AIMS) score was 29. Repeat MRI showed left-sided striatopallidal T1-hyperintensities and right-sided striatopallidal atrophy (Fig. 1B, Video 1).

She underwent unsuccessful trials of tetrabenazide (112.5 mg/d; causing parkinsonism), haloperidol (1.5 mg/d; causing akathisia), risperdone (1.75 mg/d; ineffective), amantadine (100 mg/d; ineffective) and botulinum toxin shoulder injections (decreased ballism amplitude without subjective improvement). She continued propranolol and citalopram (both 40 mg/d) with minimal benefit.

Due to the concern about infection given the poorly controlled diabetes, we proposed ablative surgery, targeting the left posteroventral globus pallidus internus (GPi; Supplementary Figure 1-2). Microelectrode recordings from the external pallidum displayed so-called slow frequency discharging cells with pauses (n = 3, mean firing rate 48Hz; range 45–51Hz). In GPi, we encountered relatively dense kinesthetic high-frequency discharging neurons (n = 14; mean firing rate 79Hz; range 69–109Hz; Fig. 1D–E, Video 2). The pallidotomy was uncomplicated and resulted in immediate near-complete arrest of her right-sided PHGCB at rest and marked improvement with stress (AIMS scores 21 and 19 at 4 and 9 months postoperatively, respectively). Interestingly, she experienced mild worsening of her left-sided symptoms after surgery (Video 3).

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The first human stereotactic operation, performed by Spiegel and Wycis in 1947, was a pallidotomy for Huntington's chorea-ballism, and pallidal procedures remain successful for hyperkinetic movement disorders [3]. To date, only four other PHGCB patients have been treated surgically [1,5]. Thalamotomy, pallidotomy and thalamic and pallidal DBS have each improved choreoballistic movements, underlining the role of corticopallidothalamic circuits in chorea-ballism [1,2].

Contrary to one study, where decreased GPi firing rates were described in a single patient with PHGCB [1], and to observations in patients with hemichorea-ballism of different etiologies, our electrophysiological recordings resembled the increased firing of high frequency discharging GPi neurons, as typically found in Parkinson's patients in the 'medication off' state [3]. Only recordings in larger samples of PHGCB patients with a similar disease severity and duration, under comparable pharmacological and anesthetic circumstances, will allow to characterize the typical electrophysiology of PHGCB.

Although our follow-up is limited to 9 months, the long-term benefit

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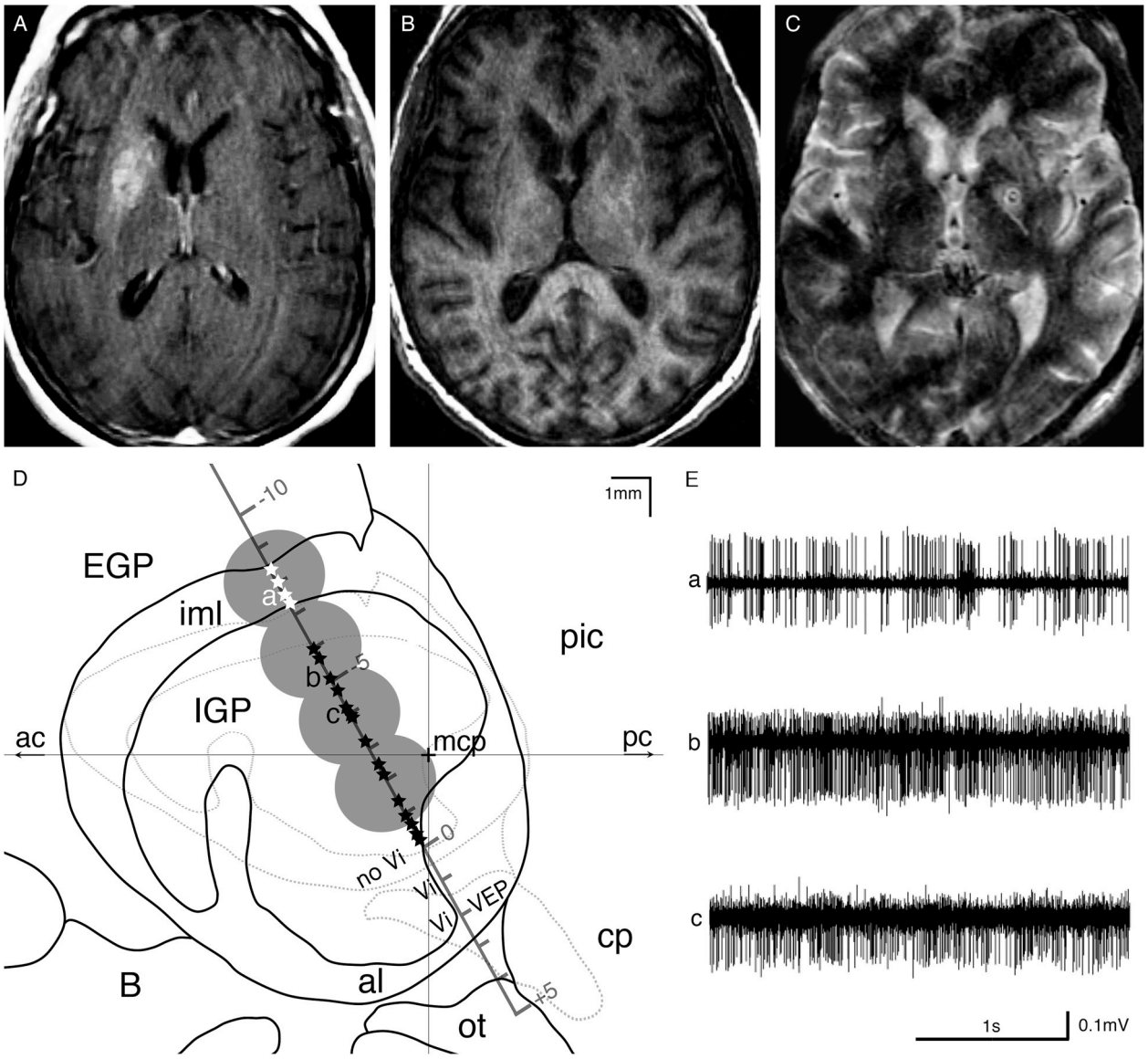

Fig. 1. Radiology, micro-electrode recording and clinical outcome.
(A) Gadolinium-enhanced T1-weighted MRI showing right striatopallidal hyperintensities 2 days after onset of left-predominant PHGCB. (B) Unenhanced T1-weighted MRI 7 days after onset of right-predominant PHGCB, showing left-sided striatopallidal hyperintensities and right-sided striatopallidal atrophy. (C) T2-weighted MRI 1 day postoperatively, showing left pallidal target sign with surrounding edema. (D) The planned trajectory is projected on a simplified parasagittal brain sketch, based on Plate 47/S.l 20.0 of the Schaltenbrand and Wahren stereotactic atlas. White and black stars represent recorded external and internal pallidal neurons, respectively. Visual evoked potentials ("VEP") in response to a flashing operating light, and stimulation-induced phosphenes ("Vi") were recorded. Dotted lines display the atlas drawing corrected based on our electrophysiological recordings. Four merging grey circles indicate the location and size of the radiofrequency lesions. (E) Illustrative recordings 7.5 mm (a: 51Hz, SFD-P), 5.1 mm (b: 109Hz (bigger unit), HFD) and 4.0 mm (c: 78Hz, HFD) dorsal to the target.
Abbreviations: ac: anterior commissure; AIMS: abnormal involuntary movement score; al: ansa lenticularis; B: nucleus basalis; cp: cerebral peduncle; EGP: external globus pallidus; IGP: internal globus pallidus; iml: internal medullary lamina; HFD: high frequency discharging cell; mcp: midcommissural point; ot: optic tract; pc: posterior commissure; pic: posterior limb of internal capsule; PHGCB: post-hyperglycemic chorea-ballism; SFD-P: slow frequency discharging cell with pauses.

of pallidotomy in other hyperkinetic movement disorders gives us cause for optimism in this case, especially given the static nature of PHGCB. This favorable outcome suggests that pallidotomy may be an excellent treatment option in patients like ours with persistent and medication-refractory PHGCB. Given the current global rise in the incidence and prevalence of type 2 diabetes, we expect that neurologists and neurosurgeons will encounter a growing number of PHGCB patients, with an increasing demand for surgical options in refractory cases.

Disclosures

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Declaration of interest

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LM, DJL, RFD and WDH report no conflicts of interest.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.parkreldis.2018.11.023.

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