2018

Ten patients have undergone surgery for HH under the dual care of Alder Hey tumour and epilepsy neurosurgeons during this period. Eight cases had a midline transcallosal, interforniceal approach with the remaining 2 having a transcallosal, transforaminal approach. All patients had an IOMRI scan, with 40% needing further tumour resection post-IOMRI. Forty percent had a total resection, 3 patients had near-total resection and 3 patients had subtotal resection (~ 30% tumour residual on post-operative MRI). No new neurological complications developed post-operatively. Hypothalamic axis derangements were seen in 3 cases, including 1 diabetes insipidus with hypocortisolaemia, 1 hypodipsia and 1 transient hyperphagia. Eighty percent are seizure free; the remaining two patients have had significant improvements in seizure frequency.

IOMR was used to tailor the ideal tumour resection volume safely based on anatomy of the lesion, which combined with the open transcallosal, interforniceal route performed by surgeons experienced in the approach resulted in excellent, safe and effective seizure control [1)]

2013

14 patients with medically refractory gelastic epilepsy who underwent stereotactic frame-based placement of an MR-compatible laser catheter (1.6 mm diameter) through a 3.2-mm twist drill hole. A U.S. Food and Drug Administration (FDA)-cleared laser surgery system (Visualase, Inc.) was utilized to ablate the HH, using real-time MRI thermometry. Seizure freedom was obtained in 12 (86%) of 14 cases, with mean follow-up of 9 months. There were no permanent surgical complications, neurologic deficits, or neuroendocrine disturbances. One patient had a minor subarachnoid hemorrhage that was asymptomatic. Most patients were discharged home within 1 day. SLA was demonstrated to be a safe and effective minimally invasive tool in the ablation of epileptogenic HH. Because use of SLA for HH is being adopted by other medical centers, further data will be acquired to help treat this difficult disorder [2)].

Fourteen patients underwent transventricular endoscopic resection of HH for treatment-resistant epilepsy. Prior to surgical resection, single neuron recordings from bundled microwires (total of nine contacts) were obtained from HH tissue. Spontaneous activity was recorded for two or three 5-min epochs under steady-state general anesthesia. Off-line analysis included cluster analysis of single unit activity and probability analysis of firing relationships between pairs of neurons. Results: Altogether, 222 neurons were identified (mean 6 neurons per recording epoch). Cluster analysis of single neuron firing utilizing a mixture of Gaussians model identified two distinct populations on the basis of firing rate (median firing frequency 0.6 versus 15.0 spikes per second; $p < 10(-5)$). Cluster analysis identified three populations determined by levels of burst firing (median burst indices of 0.015, 0.18, and 0.39; $p < 10(-15)$). Unbiased analysis of spontaneous single unit behavior showed that 51% of all possible neuron pairs within each recording epoch had a significant level of firing synchrony ($p < 10(-15)$). The subgroup of neurons with higher median firing frequencies was more likely to demonstrate synchronous firing ($p < 10(-7)$).

Hypothalamic hamartoma tissue in vivo contains neurons which fire spontaneously. The activity of single neurons is diverse but distributes into at least two electrophysiological phenoytpes. Functional linkage between single neurons suggests that HH neurons exist within local networks that may contribute to ictogenesis [3)].

1)
van Tonder L, Burn S, Iyer A, Blair J, Didi M, Carter M, Martland T, Mallucci C. Open resection of hypothalamic hamartomas for intractable epilepsy revisited, using intraoperative MRI. Childs Nerv Syst. 2018 May 11. doi: 10.1007/s00381-018-3786-x. [Epub ahead of print] PubMed PMID: 29752488.
2)
Wilfong AA, Curry DJ. Hypothalamic hamartomas: optimal approach to clinical evaluation and diagnosis. Epilepsia. 2013 Dec;54 Suppl 9:109-14. doi: 10.1111/epi.12454. PubMed PMID: 24328883.
3)
Steinmetz PN, Wait SD, Lekovic GP, Rekate HL, Kerrigan JF. Firing behavior and network activity of single neurons in human epileptic hypothalamic hamartoma. Front Neurol. 2013 Dec 27;4:210. doi: 10.3389/fneur.2013.00210. PubMed PMID: 24409165.