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Temporal encephalocele: a novel indication for magnetic resonance-guided laser interstitial thermal therapy for medically intractable epilepsy

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ABSTRACT – Temporal encephalocele (TE) is a rare but surgically treatable/curable cause of temporal lobe epilepsy (TLE). The surgical intervention varies from local disconnection to extensive anterior temporal lobectomy and amygdalohippocampectomy (ATL/AH). Magnetic resonance-guided laser interstitial thermal therapy (MRgLITT) has evolved as a minimally invasive alternative for intractable epilepsy with good results, however, application of MRgLITT for intractable pediatric epilepsy secondary to TE has not been reported. We present a detailed technical report and clinical experience of MRgLITT in two adolescent children with medically intractable TLE secondary to TE. Pre-surgical evaluation revealed anterior inferior TE with concordant clinico-electrophysiological data. Both the patients underwent MRgLITT after review with the institutional multidisciplinary epilepsy team and discussion with the patient and the family. Both the patients were discharged on post-operative day one and have been seizure-free since the procedure at the last follow-up visit at 18 months and 6 months, respectively. The present series demonstrates first-ever clinical and technical experience of MRgLITT for TE with intractable pediatric epilepsy. The excellent post-operative seizure outcome and favorable post-operative course support MRgLITT as the first line of surgical intervention for TE with intractable TLE and broadens the application of MRgLITT.

Key words: temporal encephalocele, temporal lobe epilepsy, epilepsy surgery, laser ablation, MRgLITT

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Temporal encephalocele (TE) is an abnormal herniation of brain parenchyma through osseus-dural defects of the middle temporal fossa/skull base floor and is quite rare. TE can present with phamacoresistant epilepsy, specifically the anteroinferior TE (Wilkins *et al.*, 1993), which is often under-recognized (Saavalainen *et al.*, 2015).

Identification of TE as a cause of temporal lobe epilepsy (TLE) is important, as TE is a surgically treatable/curable cause of TLE (Saavalainen *et al.*, 2015; Panov *et al.*, 2016; Toledano *et al.*, 2016; Lapalme-Remis *et al.*, 2017). However, the surgical treatment is not well defined and can vary from a focal resection and/or disconnection of the TE to an extensive anterior temporal lobectomy with amygdalohippocampectomy (ATL/AH) (Giulioni *et al.*, 2014; Saavalainen *et al.*, 2015; Panov *et al.*, 2016; Lapalme-Remis *et al.*, 2017; Bannout *et al.*, 2018).

Magnetic resonance-guided laser interstitial thermal therapy (MRgLITT) has emerged as a minimally invasive surgical option for focal epilepsy with promising results (Curry *et al.*, 2012; Willie *et al.*, 2014; Kang *et al.*, 2016). The therapeutic role of MRgLITT in the treatment of pediatric TLE due to TE has not yet been described in the literature. Here, we present an early clinical experience and technical details of MRgLITT for TE.

Case study 1

A 15-year-old female with intractable focal epilepsy of several years duration had seizures which lasted for 20-30 seconds with postictal confusion. Her seizures were characterized by stereotypical behavior arrest or "slowing down" of her actions with staring at a particular spot on the ground. This behavioral manifestation would follow with complex and prominent bimanual motor automatisms, often associated with stiffening of the body and occasionally with drooling. Her neurological examination was unremarkable, though she had diagnosed autism spectrum disorder. Interictal EEG showed slowing with left temporal spikes. Video-EEG confirmed seizure onset from the left temporal lobe. Her CT showed a defect in the floor of the left middle fossa, consistent with a small left anterior temporal lobe encephalocele (*figure 1A, B*). Her MRI similarly revealed a small inferior left anterior temporal lobe encephalocele with tissue protruding outward through the defect in the middle fossa floor (*figure 1C, D*). FDG-PET and resting-state functional MRI showed hypometabolism (*figure 1E, F*) and abnormal connectivity (Boerwinkle *et al.*, 2017) (*figure 1G, H*), respectively, in the left anterior temporal lobe. Our multidisciplinary epilepsy team initially recommended a standard left temporal lobectomy, but the family was reluctant due to potential complications and loss of cognitive function from surgery in the dominant hemisphere and morbidity associated with open surgery. MRgLITT was introduced as an alternative and she underwent MRgLITT for ablation of the anterior TE and its temporal lobe connections. Since surgery, she has been seizure-free (Engel Class 1) for 18 months and is presently being weaned off medications.

Case study 2

A 16-year-old male had intractable complex partial seizures and occasional generalized tonic-clonic seizures, which lasted for one minute, 2-3 times a week. His seizures were characterized by unresponsiveness and upward eye rolling. He also experienced garbled thoughts and seeing flashes of color prior to seizures. His examination was unremarkable, with normal cognition. Though he had multiple *café au lait* spots, genetic analysis for NF-1 was negative. Interictal EEG showed left temporal slowing with T3/F7 spikes. Video-EEG revealed left temporal slowing and epileptiform discharges in the left mid to anterior temporal lobe. MRI revealed multiple left anterior TE (*figure 1I*). PET CT of the head showed bony irregularity in the anterior temporal and adjacent sphenoid wing (*figure 1J*). FDG-PET and rs-fMRI showed hypometabolism and abnormal connectivity, respectively, within the left anterior temporal lobe (*figure 1K, L*). Initially recommended for surgical intervention for a left anterior temporal lobectomy, the option for MRgLITT was discussed, and the family decided for MRgLITT of the TE. Since surgery, he has been seizure-free (Engel Class 1) for six months though continuing lacosamide and zonisamide.

Surgical technique for MRgLITT

One week prior to the surgery, the patients were given dexamethasone, 8 mg daily, in split doses. Following general anesthesia and pre-operative antibiotics, the patients were placed in three-point fixation in a supine position with head turned to the contralateral side and ipsilateral shoulder elevated with a bolster, and the table tilted so that the surgical entry site could be placed at the highest point and as possible, parallel to the floor. The patients were registered using Stealth ™ (Medtronic) and the postero-anterior trajectory was planned for optimal targeting of the anterior middle fossa, specifically, the anterior most part of the intradural component of the encephalocele, close to the dura, carefully incorporating the "neck" of the prolapsed or herniated parenchyma (*figure 2A*). The goal of the ablation was to ablate and/or disconnect the dysplastic tissue from the rest of the temporal lobe. Following normal sterile preparation and draping, the Medtronic Vertek™ frameless navigation guidance was used for marking the entry site. The entry site was incised and the craniostomy performed in line with trajectory with a hand-held power twist drill (3.2 mm).

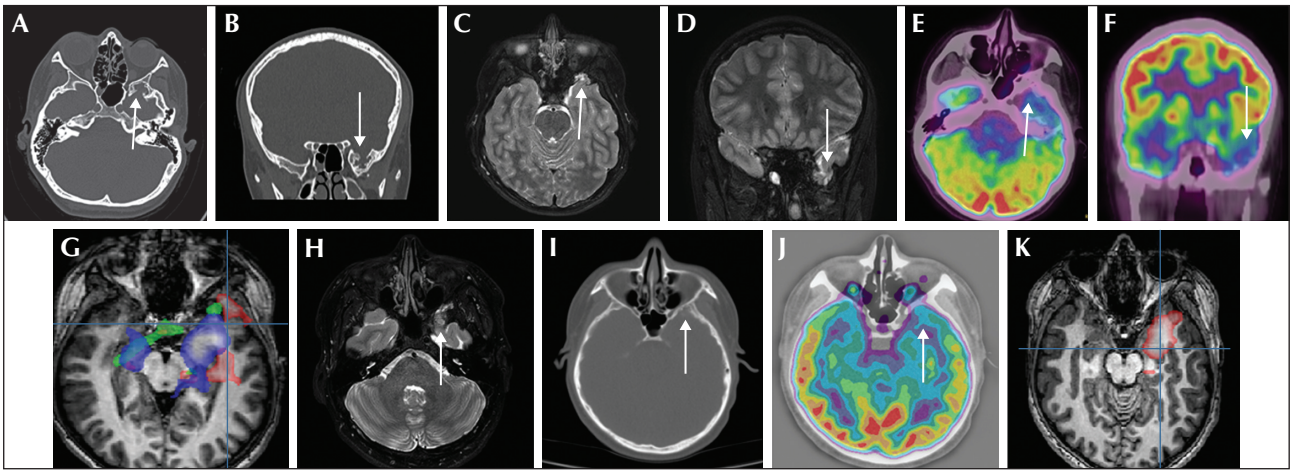

Figure 1. (A, B) Head CT axial (A) and coronal (B) images showing a left anterior inferior skull base defect (white arrow) in Patient 1. (C, D) Brain MRI T2-weighted axial (C) and coronal (D) images showing left anterior inferior temporal encephalocele (white arrow) in Patient 1. (E, F) PET CT axial (E) and coronal (F) images showing anterior temporal hypometabolism (white arrow) in Patient 1. (G) Axial view of resting state functional MRI showing location of abnormal connectivity in Patient 1. (H) Brain MRI T2-weighted axial image showing left anterior inferior temporal encephalocele (white arrow) in Patient 2. (I) PET CT head bone window axial image showing a left anterior inferior skull base defect (white arrow) in Patient 2. (J) PET CT axial view showing anterior temporal hypometabolism (white arrow) in Patient 2. (K) Resting state functional MRI showing location of abnormal connectivity in Patient 2.

The dura was penetrated with a sharp probe using electrocoagulation. The skull bolt was secured along the trajectory and a blunt probe was first passed to target. After removal of the probe, the 10-mm laser sheath was then placed along the tract to the pre-planned target (*figure 2A*), followed by insertion of the laser fiber. The patient was then transferred to the MRI for MRgLITT.

After confirming appropriate placement of the laser system, a test dose of the laser was initiated at 15% of 15 Watts and the heat signature and surrounding tissue heating could be observed using real-time MR thermography. Safety markers were placed at anteromedial, superior and lateral adjacent areas to the TE in order to monitor the temperature map and avoid inadvertent heating of the skull base, cavernous sinus/cavernous sinus dura, mesial temporal lobe, or superior temporal stem. Subsequently, the laser firing was raised to 75% and the extent of the calculated ablation monitored. Pull back of the laser probe in 5-8-mm increments with repeat of the above processes provided further ablation of the surrounding areas of potential abnormal connectivity in the final area of ablation (*figure 2B, C*). Following satisfactory ablation, T1 contrast-enhanced MRI was acquired to visualize the area of ablation (*figure 2D, E, F*). Subsequently, the laser probe and the skull bolt were removed and a single resorbable 4-0 Monocryl™ suture was placed at the insertion site.

Postoperatively, both patients were monitored for one day in the hospital and discharged with a tapering of the dexamethasone regimen over one week. Neither patient suffered a complication or untoward event from the surgical procedure or ablation.

Discussion

TE is a very rare case of intractable TLE. The estimated incidence of TE is 0.3% based on MRI examinations performed for a newly diagnosed epilepsy and 1.9% in patients with medically intractable epilepsy according to one of the largest series from an epilepsy center (Saavalainen *et al.*, 2015). TE in pediatric epilepsy is extremely rare. Based on a meta-analysis of 1,318 pediatric epilepsy patients from 36 different studies, there was not a single case of TE (Englot *et al.*, 2013), however, TE has been described in anecdotal reports or occasionally included in series and/or reviews (Faulkner *et al.*, 2010; Saavalainen *et al.*, 2015; Lapalme-Remis *et al.*, 2017). Though the association of TE with epilepsy is well reported, the detailed chronic and intraoperative electrophysiological description and surgical experience was described only recently by Panov *et al.* (2016). The authors clearly demonstrated epileptiform discharges around the cortical area of the encephalocele in all six surgically treated cases, all of which responded favorably to surgery. The histopathology of the resected tissue and the temporal lobe can show no changes to minimal changes including cortical laminar disorganization, hetrotropia, and gliosis, possibly representing reactive changes (Panov *et al.*, 2016; Bannout *et al.*, 2018).

The surgical approach for TE is not very clear and varies from mere disconnection of the herniated brain tissue to lesionectomy, tailored resection, neocortical resection to standard temporal lobectomy and amygdalohippocampectomy (Giulioni *et al.*, 2014; Saavalainen *et al.*, 2015; Panov *et al.*, 2016;

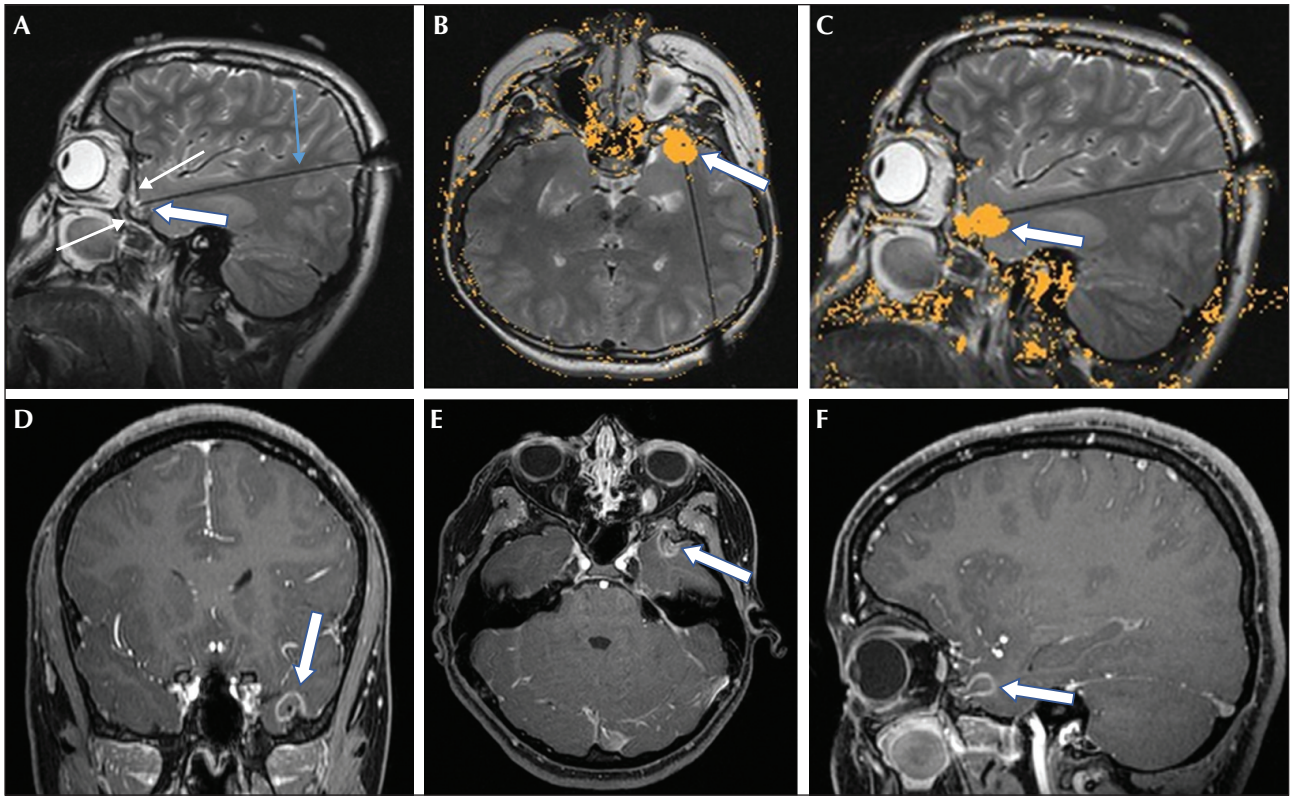

Figure 2. (A) Brain MRI T2-weighted sagittal image showing placement of the laser catheter at the neck of the left temporal encephalocele (blue arrow: laser catheter position; thin white arrow: hypointense line [black line] showing dural margin and dural defect; thick white arrow: relative position of laser catheter with the encephalocele and the dural defect). (B) Procedural MRI thermal map, axial image, showing Arrhenius-based damage estimation in orange (thick white arrow). (C) Procedural MRI thermal map, sagittal image, showing Arrhenius-based damage estimation in orange (thick white arrow). (D) Immediate post-ablation brain MRI, coronal contrast-enhanced T1-weighted image, showing extent of enhancement consistent with successful ablation (thick white arrow). (E) Immediate post-ablation brain MRI, axial contrast-enhanced T1-weighted image, showing extent of enhancement consistent with successful ablation (thick white arrow). (F) Immediate post-ablation brain MRI, sagittal contrast-enhanced T1-weighted image, showing extent of enhancement consistent with successful ablation (thick white arrow).

Lapalme-Remis *et al.*, 2017; Bannout *et al.*, 2018). Post-operatively, most of the patients from the reported case reports or small case series were seizure-free irrespective of the surgical approach (Giulioni *et al.*, 2014; Saavalainen *et al.*, 2015; Panov *et al.*, 2016; Lapalme-Remis *et al.*, 2017; Bannout *et al.*, 2018). In the largest series of TE consisting of 23 patients (Saavalainen *et al.*, 2015), the authors reported surgical outcome for 12 patients who were treated with epilepsy surgery. Nine out of 12 patients were seizure-free (Engel Class I), while two patients had Engel Class II outcome, and only one patient had residual seizures though worthwhile improvement (Engel Class III). The seizure outcomes were comparable in patients who had lobectomy or only local disconnection. No immediate surgical complication was reported in the series. Panov *et al.* reported Engel Class I outcome in five out of six cases, irrespective of the type of surgery (Panov *et al.*, 2016). Only one patient had Engel Class II outcome and that patient had a lobectomy. Interestingly, Bannout *et al.* reported a case of TE with extensive mesial temporal signal changes, and the patient remained seizure-free after disconnection surgery alone and was reported to have preserved hippocampus at the one-year postoperative MRI follow-up visit (Bannout *et al.*, 2018). Even in the surgical series by Panov *et al.*, one patient underwent local disconnection but was also noted to have hippocampal discharges (Panov *et al.*, 2016). This patient remained seizure-free after the disconnection procedure alone. It is worthwhile to note that open surgical repair of the skull base defect and TE has historically been very successful. Various surgical approaches have been described, which not only include intracranial approaches but also otological, endoscopic, and craniofacial approaches, depending upon the location and anatomical details (Wind *et al.*, 2008; Gonen *et al.*, 2016). Extracranial approaches often focus on repair of the osseous and dural defect, addressing CSF leak and its consequences associated with TE. Post-operative

CSF leak remains a challenging issue with the open surgical approaches and even with successful repair, delayed CSF leak can occur (Gonen *et al*., 2016). Moreover, these approaches have been successful but were more invasive and at the same time, associated with the risks related to open surgery, brain manipulation, and even recurrence. The mean recurrence rate has been reported to be around 5%, but some authors reported as high as 28% (Gonen *et al*., 2016).

Extensive resections that include removal of mesial temporal structures and the lateral cortex, particularly in the dominant temporal lobe, are also well known to be associated with recognised consequences of neurocognitive impact, and this, at times, adversely affects QOL post temporal lobectomy. At the same time, with recent advancement in technology and the positive clinical experience, increasingly more and novel indications of medically intractable epilepsy are being treated with MRgLITT. This includes mesial temporal epilepsy, cortical malformations, deep-seated tubers, small tumors, and hypothalamic hamartoma. The impact of this novel technology in these medically intractable cases of epilepsy with sometimes difficult to access lesions/areas has been increasingly accepted and in many instances, has become the preferred surgical therapy/intervention. As an emerging tool and field in epilepsy, MRgLITT has also been used to treat TE,. Bandt and Leuthardt reported a 51-year-old female patient treated with MRgLITT for intractable epilepsy secondary to TE who was seizure-free after surgery at the last follow-up visit at two years (Bandt and Leuthardt, 2016). The same group later mentioned one case of TE treated with MRgLITT from a large series of epilepsy (Kamath *et al*., 2017), however, there were no specific details included for the patient with TE in this series. If seizure outcome is excellent or the same, whether limited to local resection or lobectomy (Faulkner *et al*., 2010), or even with the minimally invasive approach of MRgLITT (Bandt and Leuthardt, 2016), one should consider the more limited approach with the least amount of morbidity. It is also important to note that primary surgical intervention with MRgLITT does not preclude a repeat MRgLITT or an open procedure to try and alleviate seizure burden in case of future recurrence of seizure (Petito *et al*., 2018). With this background, it is reasonable to consider MRgLITT local disconnection for TE as a preferred primary treatment option.

Though MRgLITT is a minimally invasive surgical option and a relatively safe procedure, it is, however, not completely free of complications. Common complications related to MRgLITT include, but are not limited to, hemorrhage, infection, and meningitis/brain infection. An additional specific complication related to TE could be related to the strategic anatomical location within the temporal lobe or CSF space. CSF leak, infection, skull base heating, and visual field defects need to be considered when ablating lesions, especially temporal encephalocele/lesions close to the temporal skull base and within temporal lobe. MRgLITT avoids morbidity associated with open resection, with the potential for a better neuropsychological outcome (Drane *et al*., 2015) though this remains to be validated in larger multicenter studies (Kang *et al*., 2016). MRgLITT also offers an advantage of targeted ablation of the suspected epileptic zone (ZE) for disconnection in TE under real-time monitoring. In this series, the primary goal of the ablation was a disconnection at the stem of the TE, rendering it non-functional by severing its neural connections with the rest of the temporal lobe. The outlet to the TE is thought to be epileptogenic, secondary to the reactive changes seen histopathologically and the often disorganized cortex associated with the encephalocele (Panov *et al*., 2016; Bannout *et al*., 2018). It was hypothesized that it is not important to ablate all of the herniated or prolapsed brain parenchyma, but rather, the focus of ablation being a disconnection of the abnormal tissue from the rest of the brain, rendering it clinically insignificant. Targeting and ablating the residual small herniated tissue may add technical difficulty and may unnecessarily increase the risk of complications, especially CSF leak.

Our present series demonstrates first-ever clinical and technical experience with MRgLITT for intractable pediatric epilepsy secondary to TE with excellent postoperative seizure outcome following disconnection of the TE from the surrounding tissue. While this broadens the application of MRgLITT for intractable epilepsy, a longer-term experience with a larger patient cohort may be required prior to uniform implementation, though this could be difficult in view of the rarity of the lesion.

Conclusion

TE is an under-recognized and rare cause of surgically treatable TLE, through limited resection or tailored resection with good outcome. Local disconnection and limited cortical ablation can be achieved with MRgLITT and this initial series demonstrates that it can be effective and may be considered as a primary surgical option for patients with intractable epilepsy secondary to TE. □

Supplementary data.
Summary didactic slides are available on the www.epilepticdisorders.com website.

Limitations.
The present study is limited by the number of cases with limited time of follow-up, and ablations were not necessarily based on

any intracranial EEG recording. Despite these limitations, seizure freedom following MRgLITT, to date, in these patients has been excellent and encouraging.

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None of the authors have any conflict of interest to declare.

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TEST YOURSELF

(1) Is minimal resection or minimally invasive surgery for temporal encephalocele with intractable epilepsy effective?

(2) What is the rationale for choosing MRgLITT to treat temporal encephalocele with intractable epilepsy?

Note: Reading the manuscript provides an answer to all questions. Correct answers may be accessed on the website, www.epilepticdisorders.com, under the section "The EpiCentre".