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Case Report

Unexpected pain with electrocortical stimulation in a teenager with temporal encephalocele

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ABSTRACT

Temporal lobe encephalocele has emerged as a potentially unrecognized cause of drug-resistant temporal lobe epilepsy (TLE) that can be effectively treated with epilepsy surgery. Here we present a case in which a 17-year-old male with drug-resistant epilepsy and left temporal encephalocele underwent workup for epilepsy surgery, and experienced unexpected pain with electrocortical stimulation. Stimulation of stereo-EEG electrodes in the left temporal pole resulted in severe, unilateral left-sided facial pain due to inadvertent stimulation of the trigeminal nerve. Stereo-EEG showed seizure onset adjacent to encephalocele with no involvement of mesial temporal structures. A temporal pole resection sparing the mesial temporal structures and repair of the sphenoid bone defect was performed. The patient experienced post-operative seizure freedom, with no loss of language function or sensory deficits in the distribution of the trigeminal nerve. This case highlights temporal encephalocele as a potentially overlooked cause of TLE and underscores the anatomical proximity of the trigeminal nerve to the temporal pole, an important consideration for surgical planning.

1. Introduction

Encephalocele is defined as a protrusion of brain tissue and its membranes through defects in the skull [1]. As resolution of imaging has improved, temporal lobe encephalocele has been increasingly identified as a cause of drug-resistant temporal lobe epilepsy (TLE) [2–6]. In such cases, surgical intervention often reduces seizure burden or eliminates seizures entirely [7,8]. Several different surgical approaches may be employed [7,9]; as in all cases of epilepsy surgery, an optimal surgical approach maximizes the likelihood of seizure freedom while minimizing the chance of postoperative neurologic or cognitive deficits. Stereo-EEG (sEEG) may be used to select an optimal surgical approach by localizing the seizure onset zone and identifying adjacent eloquent cortex through electrocortical stimulation [10,11]. Here, we present a case in which a 17-year-old male with drug-resistant TLE underwent sEEG for pre-operative planning. During electrocortical stimulation with sEEG, he experienced unexpected pain that had significant implications for his surgical management.

2. Case report

2.1. Patient presentation

A 17-year-old right-handed male with high-functioning autistic spectrum disorder presented with a two-year history of focal impaired awareness seizures. His seizures lasted 1–2 min. The semiology was characterized by loss of awareness at seizure onset, behavioral arrest, facial grimace to the right, and dystonic stiffening of right upper extremity with hand in a C-shape, or at times stiffening of bilateral upper and lower extremities resulting in falls. Seizures had no associated aura or post-ictal symptoms, and the patient did not remember events due to seizures. His seizures occurred 2–10 times per month, and did not resolve with appropriate doses of clobazam, oxcarbazepine, lacosamide, levetiracetam, carbamazepine, and valproic acid. He was born full term following an uncomplicated pregnancy and had no epilepsy risk factors including trauma, CNS infections, or a family history of epilepsy. His neurological examination was normal. Neuropsychiatric testing was notable for a full-scale IQ of 114, with a strong verbal com-

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prehension index of 132 and lower processing speed index of 86. The patient was initially reluctant to consider epilepsy surgery, but after two additional years of uncontrolled seizures he agreed to a phase one epilepsy surgery evaluation.

2.2. Scalp EEG findings

Video-EEG showed rare spikes in the left anterior temporal region. Over 20 typical seizures were recorded, generally lasting 30–60 s. Most were electrographically nondescript, predominantly characterized by left hemisphere or diffuse delta activity often obscured by EMG artifact, followed by left temporal post-ictal slowing. Some seizures had similar onset but become focal to bilateral tonic-clonic seizures. Some seizures were better localizing, with sharply contoured theta activity in the left temporal head region (Fig. 1A).

2.3. Neuroimaging findings

His initial 3T MRI brain showed mild gliosis and ex vacuo dilation of the left greater than right temporal horn. MRI brain was repeated during his phase one evaluation and showed a small left anterior temporal encephalocele with subtle cortical protrusion into the posterior aspect of the left sphenotemporal buttress (Fig. 2A). No other structural abnormalities were observed. Paired ictal/interictal single-photon emission computed tomography

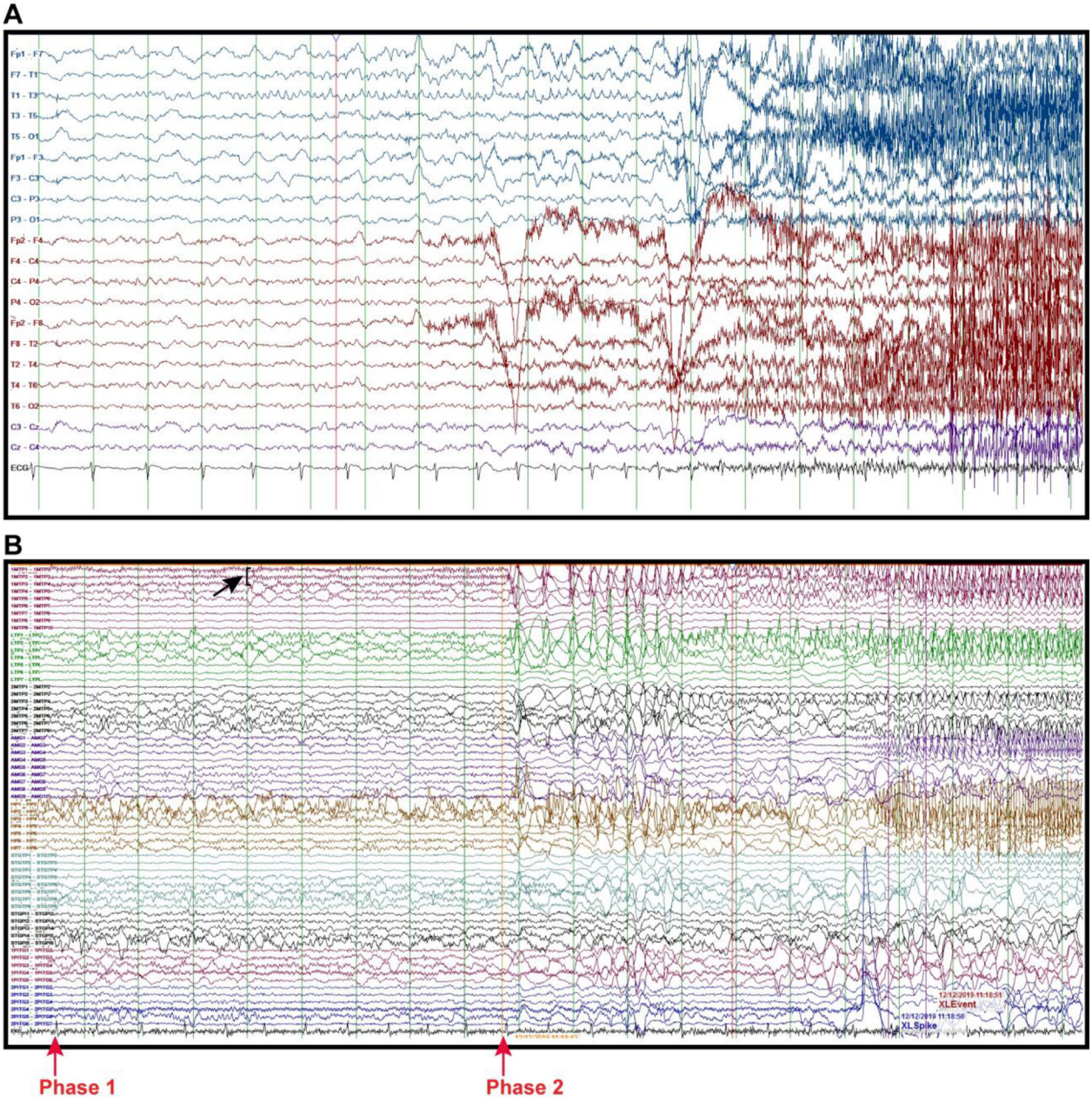

Fig. 1. (A) Scalp EEG recording of the patient's typical electrographic seizure, showing sharply contoured theta activity in the anterior left temporal head region (T1). (B) Representation of the patient's typical electrographic seizure captured by stereo EEG. Phase 1 consists of fast (20 Hz) activity in the deep contacts of 1MTP (top three waveforms, see black arrow). The patient was asymptomatic throughout phase 1 evaluation. Phase 2 monitoring of seizure onset began with the simultaneous onset of 4–8 Hz evolving spikes in the temporal pole, amygdala and hippocampus, clinically accompanied by unresponsiveness and bilateral stiffening of the arms and hands.

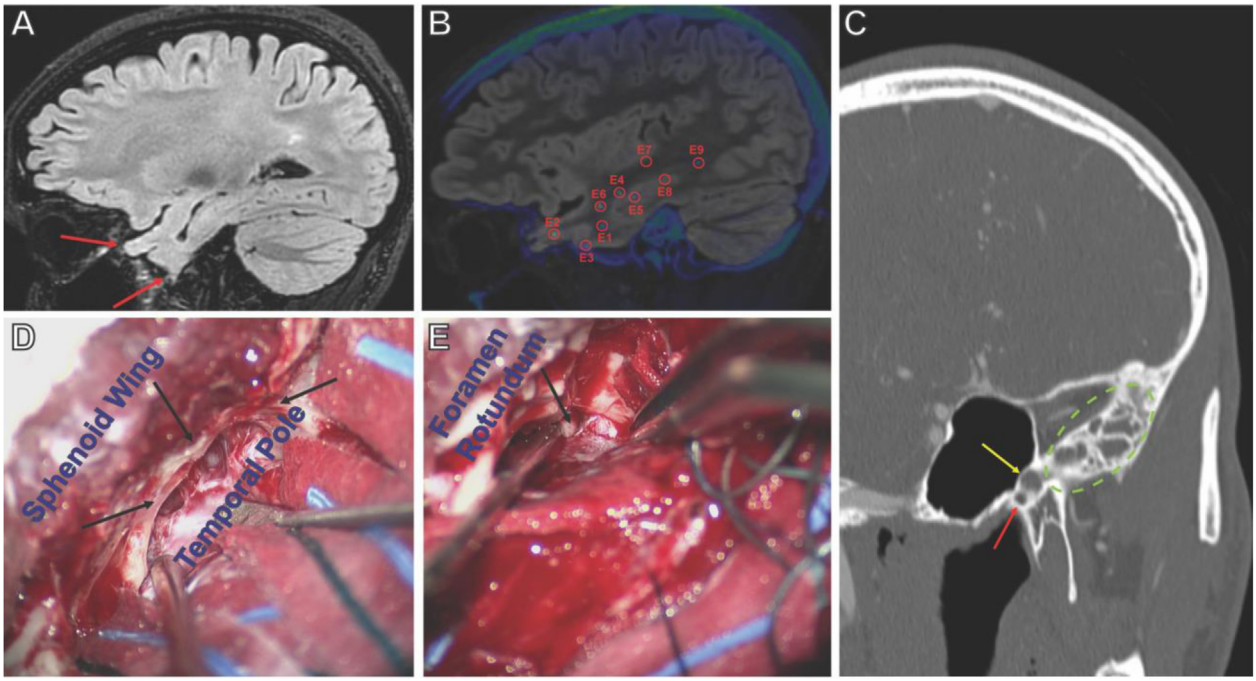

Fig. 2. (A) Sagittal view of the left temporal pole encephalocele (red arrows) as demonstrated by pre-operative MRI. (B) Sagittal view of anatomical position of the sEEG electrodes. (C) Pre-operative coronal-oblique reformatted CT image demonstrating the spatial relationship between the vidian canal (red arrow), foramen rotundum (yellow arrow), and temporal pole encephalocele (green circle). (D) Intra-operative extradural exposure showing defects in the sphenoid bone and encephalocele observed during resection. (E) Uncovered V2 segment of trigeminal nerve passing through foramen rotundum in extradural subtemporal exposure. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

(SPECT) imaging showed a corresponding area of focal hyperperfusion for a seizure with bilateral upper extremity stiffening. PET scan did not show focal changes in cerebral metabolism.

2.4. Pre-surgical sEEG mapping

The identification of the left temporal encephalocele that was concordant with the electrographic onset of seizures that localized well on scalp EEG provided a promising surgical target for lesionectomy. However, the patient was hesitant to proceed with epilepsy surgery due to the potential for ongoing seizures with lesionectomy or verbal memory deficits with left anterior temporal lobectomy and amygdalohippocampectomy (ATLAH). To better localize seizure onset zone, the patient underwent phase 2 epilepsy surgery workup with sEEG. Nine depth electrodes were implanted in the left temporal lobe to determine whether mesial-temporal structures were part of his seizure onset zone (Fig. 2B). Nine spontaneous seizures were recorded on sEEG with no involvement of the mesial temporal structures (Fig. 1B). Extra-operative electrocortical stimulation was also performed to map language and stimulate typical seizures. Stimulation was delivered in five second trains at 50 Hz, beginning with electrode nearest the encephalocele. On stimulation of the second contact pair with relatively low current (2 mA), the patient experienced severe 7/10 burning, lancinating pain in the left cheek for the duration of the stimulus train, accompanied by subtle rapid twitching of the left lower eyelid. The patient understandably refused to continue unless providers could explain the cause of his discomfort and guarantee that it would not recur.

After some thought, the team explained that the most likely cause of his pain was unintentional stimulation of the trigeminal nerve, which lies just inferior to the encephalocele (Fig. 2C). Stimulation of the temporal cortex is not associated with pain, and in our experience electrocortical stimulation of electrodes adjacent to the dura at much higher currents has not resulted in pain. Thus, we reasoned his pain was likely to arise from stimulation of another adjacent structure. The quality of the patient's pain was akin to that of trigeminal neuralgia, defined as a sudden, usually unilateral, severe, brief, stabbing, recurrent pain in the distribution of one or more branches by the trigeminal nerve [12]. Although his pain that was time locked to electrocortical stimulation differs from trigeminal neuralgia in that it is not an ongoing disorder, the similar character of pain implicated the trigeminal nerve as the likely source of the patient's discomfort. While the trigeminal nerve does not provide motor innervation to the eyelid, the patient's unilateral eyelid twitching was thought to result from stimulation of the left orbicularis oculi via connections between distal branches of the trigeminal and facial nerve. The patient was satisfied with this explanation and completed mapping in contacts remote to the encephalocele.

2.5. Intraoperative findings

Based on the sEEG findings, the patient later underwent resection of the left temporal pole sparing the mesial temporal structures, as well as repair of the skull base defect with autologous temporalis fascia graft. Intraoperatively, the V2 segment of the trigeminal nerve was visualized passing through the foramen rotundum adjacent to the encephalocele (Fig. 2D and E). Intra-op electrocorticography showed frequent spikes in the left anterior temporal region with no spikes at post-op resection margins.

2.6. Post-operative course

The patient recovered well following the surgery and was discharged on postoperative day three. He described left-sided facial numbness beneath his eye during the three months following sur-

gery which subsequently resolved. He remained seizure-free at his last follow-up one year after surgery.

3. Discussion

One of the most common localizations of focal epilepsy is the temporal lobe [13]. Individuals with TLE may present with a wide variety of symptoms, including visceral symptoms (rising epigastric sensation), olfactory or gustatory hallucinations, autonomic symptoms, psychic sensations (déjà vu, jamais vu, depersonalization, panoramic visions), affective symptoms (dysphoria, euphoria, fear, terror, impending doom), behavioral arrest with loss of awareness and motor automatisms, or better lateralizing motor symptoms such as dystonic posturing of contralateral limbs [13]. Temporal lobe seizures may be of structural, genetic, autoimmune, or infectious etiology. As structural causes are most amenable to treatment with surgical resection, imaging plays an essential role in the workup of patients with TLE.

Improved resolution of neuroimaging has led to increased recognition of temporal encephalocele as a potentially curable cause of drug-resistant TLE [3,10]. Temporal encephaloceles are easily overlooked, and a high index of suspicion should be maintained in cases of non-lesional TLE [4,5,14]. This case shows that even with 3T MRI, a temporal encephalocele may not be initially identified, and underscores the utility of repeat MRI imaging in cases of drug-resistant focal epilepsy. When identified, radiologic features of the encephalocele do not predict the seizure onset zone, which may not be adjacent to the encephalocele [14]. Additionally, a recent study suggests that the majority of patients with temporal encephalocele may not have epilepsy at the time of imaging [15]. Therefore, it should not be assumed that a temporal encephalocele identified on MRI is the underlying cause of TLE. Instead, clinical electrophysiology studies such as sEEG should be used together with neuroimaging findings to localize the seizure onset zone and to determine an appropriate surgical approach.

Temporal encephalocele has been treated with a variety of surgical approaches ranging from lesionectomy to ATLAH [3], which have different risk/benefit profiles. The utility of sEEG in surgical planning has been previously described [10]. In this case, there was concern that, given this patient's excellent verbal function, resection of the dominant mesial temporal structures could lead to deficits in verbal memory. sEEG established that the seizure onset zone was proximal to the encephalocele and did not include the mesial temporal structures, which were consequently spared to reduce the risk of postoperative deficits in verbal memory. The patient was very concerned about verbal memory deficits; it was only because this risk was minimized that the patient chose epilepsy surgery as a treatment option.

In this case, stimulation of cortex adjacent to encephalocele elicited ipsilateral left-sided facial pain for the duration of the stimulation. Pain is not evoked by electrocortical stimulation with a few exceptions: painful sensations have been described with stimulation of primary sensory areas or posterior insula [16]. Mechanical or electrical stimulation of dura has been shown to cause pain, but to our knowledge has not been described in the sEEG literature despite routine stimulation of electrodes proximal to dura at much higher amplitudes [17–19]. Additionally, dural stimulation does not explain associated unilateral eyelid movements that were time locked to stimulation. Thus, given the distribution and lancinating quality of the pain, it likely was caused by inadvertent stimulation of the V2 branch of the left trigeminal nerve through the adjacent temporal encephalocele (Fig. 2).

Finally, it is important to consider the risk of injury to the trigeminal nerve when planning a surgical approach for temporal encephalocele. While there have not been reported cases of trigeminal nerve damage specifically from temporal encephalocele resection, a recent report described two cases of trigeminal neuropathic pain resulting from anterior temporal lobectomies for drug-resistant epilepsy [20]. Additionally, cases of trigeminal neuralgia secondary to an unresected temporal lobe encephalocele have previously been reported, highlighting the anatomical proximity of temporal encephaloceles and the trigeminal nerve [21]. Recently a novel case series was published that described the ablation of temporal encephaloceles with magnetic-resonance guided laser interstitial thermal therapy (MRgLITT) and reported excellent post-operative seizure outcomes [9]. The two patients described in this series were similar to the patient in this report: they presented with drug-resistant focal impaired awareness seizures secondary to an anterior temporal encephalocele and underwent epilepsy surgery during adolescence. While MRgLITT is an attractive option that is less invasive than lesionectomy or ATLAH, this case highlights the proximity of the trigeminal nerve to the encephalocele and the potential for thermal damage to the nerve during the procedure.

4. Conclusion

Temporal lobe encephaloceles are an important and potentially unrecognized cause of drug-resistant temporal lobe epilepsy that is treatable with epilepsy surgery. Lesionectomy, ATLAH and MRgLITT have all demonstrated favorable post-operative seizure outcomes. This case highlights the anatomical proximity of the trigeminal nerve to the temporal encephalocele, an important consideration in the selection of a surgical approach.

5. Ethical statement

LCS, DH, RA, JB and ATK have no relevant financial or non-financial relationships to disclose. Informed consent was obtained by the individual described in the case report.

CRediT authorship contribution statement

Laura C. Swanson: Investigation, Writing - original draft. **David Hsu:** Data curation, Formal analysis, Writing - review & editing. **Raheel Ahmed:** Data curation, Formal analysis, Writing - review & editing. **Justin Brucker:** Data curation, Formal analysis. **Andrew T. Knox:** Conceptualization, Data curation, Formal analysis, Supervision, Writing - review & editing.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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