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Case Report

Temporal encephalocele: An epileptogenic focus confirmed by direct intracranial electroencephalography

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ABSTRACT

Several studies have suggested the epileptogenic potential of temporal encephaloceles. However, there is limited literature describing the results of intracranial EEG monitoring for patients with temporal encephaloceles. We describe a 19 year-old right-handed woman with drug-resistant epilepsy who presented with seizure onset at age 16 in the setting of a left temporal encephalocele where the seizure onset zone was confirmed to be the encephalocele via stereo EEG (sEEG). She had focal impaired awareness seizures occurring weekly that would progress to focal to bilateral tonic-clonic seizures monthly. Imaging showed a left anterior inferior temporal lobe encephalocele and a left choroidal fissure cyst that were stable on repeat imaging. Prolonged scalp recorded video EEG recorded seizures that showed either near simultaneous onset in the bitemporal head regions or a transitional left temporal sharp wave followed by maximum evolution in the left temporal region. Invasive monitoring with sEEG electrodes targeting primarily the left limbic system with one electrode directly in the encephalocele captured seizures with onset in the left temporal pole encephalocele. A limited resection was performed based on the results of the sEEG and except for one seizure in the immediate postop period in the setting of infection, patient remains seizure free at her 4 month follow up. This report describes a case of drug-resistant focal epilepsy where sEEG monitoring confirmed a temporal encephalocele to be the seizure onset zone without simultaneous onset at mesial temporal or other neocortical structures that were sampled. Our findings support the potential for epileptogenicity within an encephalocele with direct intracranial monitoring.

Introduction

Temporal encephaloceles (TE) are structural lesions caused by herniation of brain tissue in addition to meninges and cerebrospinal fluid into a skull defect. The exact cause of an encephalocele may be unknown but at times may be secondary to trauma, idiopathic intracranial hypertension, infection or congenital among other causes [1]. Several studies have suggested the epileptogenic potential of encephaloceles and surrounding cortex presumed to be due to local irritation, traction, scarring or associated secondary lesions [1]. Several studies have noted favorable seizure outcomes with limited resections further providing evidence of the potential epileptogenicity of encephaloceles [1–8]. However, to our knowledge there is limited literature providing evidence with chronic intracranial monitoring that an encephalocele is in the seizure onset zone [9,10]. Here we present a case of drug-resistant epilepsy secondary to a left TE who had intracranial stereo EEG monitoring confirmation of the seizure onset zone lying within the encephalocele.

Materials and methods

A 19-year-old right-handed woman presented with drug resistant epilepsy with seizure onset at age 16. Her medical history included depression and anxiety. The most frequent type of seizure were focal seizures with impaired awareness with an aura of rising sensation, followed by flushing, behavioral arrest, oral automatisms, and speech impairment that lasted several minutes and were occurring on a weekly basis. At least monthly, these focal seizures would evolve to bilateral tonic-clonic seizures. She had tried lamotrigine which was ineffective and levetiracetam which caused significant mood issues. At the time of presentation, she was on oxcarbazepine and zonisamide and continued to have seizures. Imaging with skull-based CT and MRI brain showed a left anterior

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inferior temporal lobe encephalocele and a left choroidal fissure cyst that were stable on repeat imaging (Fig. 1A).

Scalp EEG monitoring revealed left temporal intermittent rhythmic delta activity and left temporal sharp waves. Seizures were captured with either near simultaneous onset in the bitemporal head regions or a sentinel left temporal sharp wave followed by maximum evolution in the left temporal region. Neuropsychometric testing was in the low average range, without lateralizing or localizing abnormalities. Stereo-EEG (sEEG) targeting primarily the left limbic region was planned to better localize the seizure onset zone in this dominant temporal lobe epilepsy given the presence of both the encephalocele and choroidal fissure cyst (Fig. 2A). A single electrode (LL) that would directly sample the encephalocele was planned (Fig. 1B). Right temporal coverage was also included given the near simultaneous bitemporal onset seen on scalp EEG for some seizures. In addition, for many temporal cases, we include contralateral temporal coverage given the frequency of bitemporal lobe epilepsy.

Interictal intracranial recording showed spikes independently in contacts corresponding to the left hippocampus, right hippocampus, left posterior insula and the encephalocele. One typical focal to bilateral tonic-clonic seizure and two focal seizures with impaired awareness were captured with onset in the left temporal pole encephalocele contacts (LL 1–4) with subsequent earliest spread to the cingulate, insula, left superior temporal gyrus (LL 11–12) and early spread to left mesial temporal electrodes. (Fig. 2A, 2B, 2C). LA 11–12(left middle temporal) and LC 7–10(left middle temporal) bursts of spikes seen in Fig. 2 were present interictally and did not represent the ictal onset contact. Slower less organized activity was present early in LL 9–13(Fig. 2) which evolved later than LL 1–4 into organized rhythmic discharges indicating that these contacts were not involved in seizure onset and likely represent activation due to volume conduction.

Based on the results of the sEEG, a limited encephalocele resection was planned (Fig. 1C). In the immediate post-surgical period, in the setting of a febrile illness, patient had one seizure. Otherwise, patient remained seizure free at her four month follow-up.

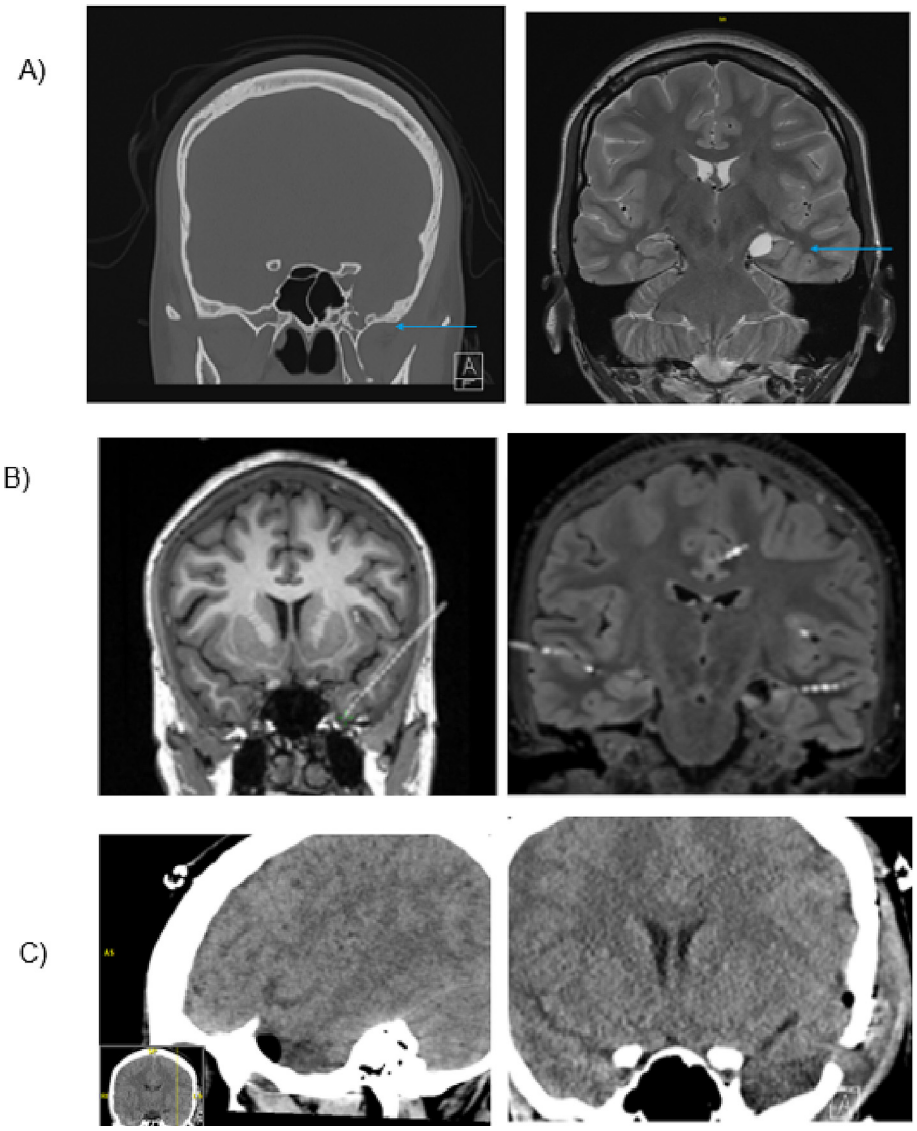

Fig. 1. A) Coronal skull base CT and T2 MRI brain showing a left anterior inferior temporal lobe encephalocele and a left choroidal fissure cyst respectively. B) T1 MRI brain oblique view showing sEEG electrode (LL) with distal contacts in the encephalocele and coronal FLAIR MRI showing sEEG electrode sampling the region around the left temporal choroidal cyst C) Post surgical CT head showing pneumocephalus and extent of resection limited to the encephalocele and surrounding cortex sparing the mesial temporal structures.

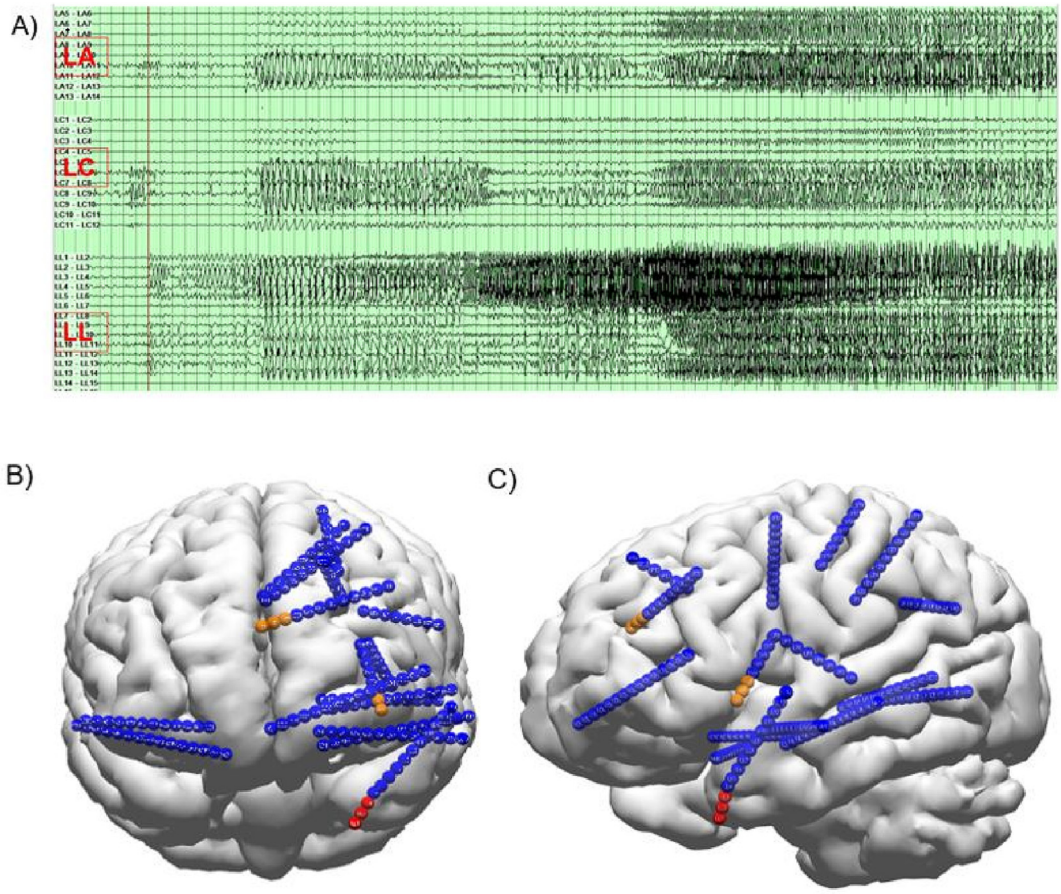

Fig. 2. A) Example of a recorded seizure during sEEG monitoring showing sample recording with 3 leads LA (entry middle temporal gyrus, target left amygdala), LC (entry middle temporal gyrus, target left hippocampal body) and LL (entry superior temporal gyrus, target encephalocele). Onset of ictal activity with low amplitude 32 Hz activity is seen over LL1-4 (contacts in the encephalocele). Activity in LA 11–12(left middle temporal) and LC 7–10(left middle temporal) was noted interictally. Slower less organized activity was present early in LL 9–13 which evolved later than LL 1–4 into organized rhythmic discharges. Sensitivity 50 uV/mm, timebase 5 mm/sec B), C) front and left view of 3D rendering of coregistered sEEG electrodes and MRI/post-op CT imaging using Curry 8.0 software (Neuroscan Compumedics, Charlotte, NC) where contacts corresponding to seizure onset are in red and showing earliest spread to the cingulate and insula in orange. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Discussion

Our case presents evidence of seizure onset within an encephalocele without simultaneous involvement of the mesial temporal electrodes as confirmed by chronic intracranial monitoring for which at present there is limited evidence. This case adds evidence to the utility of sEEG to localize the SOZ and plan a limited resection while sparing mesial temporal structures in the appropriate candidate with TE.

A meningoencephalocele is the herniation of intracranial contents including the brain parenchyma through a skull defect [3,6,10–13]. Encephaloceles may be associated with a secondary lesion which may include heterotopias, focal cortical dysplasia or gliosis [4,6,8,14–16]. In the 1990s, it was first identified that an encephalocele could be a potential epileptogenic lesion in temporal lobe epilepsy [11,17]. However in the last few years, these lesions have been increasingly detected and recognized as the etiology in temporal lobe epilepsy [1–3,8,18,19]. The epileptogenic potential of these lesions may be secondary to cortical irritation, secondary inflammation and gliosis and also associated lesions [1]. Multiple studies provide indirect evidence in support of this theory with reports of limited lesionectomy, corticectomy and bone repair resulting in favorable post-surgical outcomes [5,6,8,9,13,20] without removal of mesial temporal structures or neocortical temporal lobe structures typically resected in a standard anterior temporal lobectomy.

In a series by Panov et al [10], chronic or intraoperative electrocorticography (ECoG) was used to determine the epileptogenicity of encephaloceles and guide surgical resections. In the two patients that had chronic ECoG, subdural grids/strips were implanted in the vicinity or around the encephalocele. Electrical source localization indicated the TE to be the source of ictal activity in both patients with chronic ECoG. Additional evidence was provided in the case report by de Souza et al [9] where stimulation of the temporal pole elicited a typical aura and encephaloceles were discovered during surgery in that area. In the series by Toledano et al [18] four patients with TE had sEEG not necessarily targeting the bulk of the encephalocele; however, in all patients the temporal pole electrodes were in the epileptogenic zone supporting the epileptogenicity of the TE. In a recent series by Samudra et al. [21], sEEG recorded a seizure from a left temporal pole electrode which led to re-interpretation of imaging confirming an encephalocele in the same area.

In our patient, direct sEEG recording data was used to identify the TE to be in the SOZ with subsequent spread to the mesial temporal structures (Fig. 2). It has been noted that although the ictal onset zone may be the TE, in most cases interictal activity is commonly seen in the surrounding cortex, mesial temporal and

neocortical temporal regions indicating a widespread epileptogenic network [9,10]. This was also the case with our patient where sEEG showed frequent interictal activity independently in the bilateral mesial temporal regions.

In our case, the encephalocele was large enough to be sampled directly by a sEEG electrode. This may not be possible in all cases. In this patient, a limited resection of the encephalocele on the defect was planned based on the results. The limitation of our current study is the shorter follow up period of 4 months; however, several prior studies with longer follow-up periods [12,13,15,18,21] have concluded favorable outcomes with limited resections sparing the mesial temporal structures.

It is important to note that although the encephalocele may be the primary lesion of origin for seizures, a wide epileptogenic network may be present. Hence, it may be reasonable to hypothesize that a limited resection of this lesion may still leave the network intact, and there is potential for additional epileptogenic foci. It is also important to note the sampling limitations of sEEG and it is possible that the SOZ was not in the encephalocele, but rather earliest spread was detected in the encephalocele.

The question remains whether sEEG sampling should be performed in all patients with encephaloceles. In our case, there was displacement and distortion of the adjacent parahippocampal gyrus and hippocampus from the choroid fissure cyst which can lead to seizures hence sEEG was planned to additionally sample this region. Recent evidence points to the increased utility of sEEG in cases of encephaloceles to avoid larger resection volumes, permit sparing of mesial temporal structures and decrease risk of further neuropsychological decline [2,4,6,9,10]. However, the extent of resection and surgical outcomes vary and seizure freedom rates of 18–63% are reported [1,2,9,10]. sEEG is an invasive diagnostic test with low but present risks of hemorrhage and infection. Further studies are needed to understand the biomarkers of epileptogenicity of encephaloceles and the utility of sEEG in these cases.

Conclusion

With the help of sEEG, a large encephalocele was identified as the SOZ in a patient with drug-resistant epilepsy. This single case provides direct evidence of the epileptogenic potential of a TE.

Ethics statement

This research was conducted under the approval of the Mayo Clinic Institutional Review Board (IRB: 21-007430).

Declaration of Competing Interest

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