■ Original article

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Epileptic Disorders

Temporal encephalocele: a rare but treatable cause of temporal lobe epilepsy

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ABSTRACT

Objective. Although rare, temporal encephalocele is an important causative agent in surgically remediable drug-refractory epilepsy. The ideal treatment for temporal encephalocele remains unclear with a variety of resective surgeries recommended. Here, we analyse patient data on temporal encephalocele with a view to highlighting diagnostic clues and management strategies.

Methods. Comprehensive databases at Deenanath Mangeshkar Hospital, Pune from January 2015 to June 2019 were reviewed for this observational study. Of 107 temporal lobe epilepsy surgery patients, nine individuals with temporal encephalocele were identified, who formed the study cohort. Their clinical, neuropsychological, EEG, imaging and long-term outcome data were analysed.

Results. The study cohort consisted of seven males and two females with a mean age of 22 years. Epilepsy onset age varied from 4.5 to 19 years. Seven patients had focal non-motor seizures with impaired awareness, while two patients had focal motor seizures. Temporal encephalocele detection by MRI was reported in only two patients, and was missed in seven individuals. Three patients underwent standard anterior temporal lobectomy while the remaining six underwent resection of the temporal encephalocele with surrounding temporal pole. Eight patients showed Engel Class I outcome and one showed Class IIa outcome after a mean follow-up duration of 27 months (17-44 months). Histopathology confirmed gliosis in seven, hippocampal sclerosis type I in one and suspicious dyslamination with prominent gliosis in one patient. Six of eight patients reported an improvement in their psychological state (mood, anxiety and motivation) over time.

Significance. A careful review of MRI in patients with temporal lobe epilepsy is necessary, followed by investigations for the presence of an encephalocele. When temporal lobe epilepsy is associated with encephalocele, tailored resection of the encephalocele and the surrounding temporal pole, sparing mesial temporal structures, demonstrates excellent long-term clinical and neuropsychological outcome.

Key words: temporal encephalocele, temporal lobe epilepsy, epilepsy surgery

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Although rare, temporal encephalocele (TE) is an important causative agent in surgically remediable drug-refractory epilepsy. To date, very few case reports have highlighted the importance of identifying and treating encephaloceles despite a reasonable amount of documented success in alleviating seizures [1-4]. Less than 70 cases in total, predominantly comprising case reports or small case series of women and individuals >30 years, have

been reported in the literature [1-15]. Although various theories have been put forward, the exact mechanism of epileptogenicity is not known. Similarly, the ideal treatment for TE remains unclear, and a variety of resective surgery strategies have been proposed (*i.e.*, tailored resection of encephalocele, resection of encephalocele with the removal of the surrounding temporal pole, standard anterior temporal lobectomy, *etc.*) [7, 8, 12, 16, 17]. Here, we analysed TE patient data with a view to highlighting diagnostic clues and management strategies.

Materials and methods

We performed a retrospective chart review of patients with temporal lobe epilepsy who underwent pre-surgical evaluation followed by surgery from January 2015 to June 2019, through our electronic database at Bajaj Allianz Comprehensive Epilepsy care centre, Deenanath Mangeshkar Hospital, Pune, India. Pre-surgical evaluation, included prolonged video-EEG, 3 Tesla MRI brain epilepsy protocol, brain positron emission tomography (PET), and neuropsychological evaluation. Video-EEG was performed using a 32-channel EEG system (Nicolet) with T1 and T2 scalp electrodes placed according to the standard 10-20 system. Antiseizure medication (ASM) tapering was implemented with 30% reduction per day. Two or more seizures were recorded for each individual over 2-7 days. After data acquisition and patient counselling, decisions were made regarding the need for further investigations, invasive monitoring of patients, or the type of surgery to be performed.

A total of 107 patients underwent surgery for temporal lobe epilepsy, of whom nine had TE. Their pre-surgical clinical, video-EEG, neuropsychological and imaging data were obtained. Information on long-term outcomes was obtained through follow-up appointments regarding seizure-free periods and multiple telephonic interviews to gauge the individuals' subjective experience of cognitive changes post-surgery. For paediatric patients, responses were provided by caregivers. The questionnaire consisted of 17 items, covering domains such as attention and memory (n = 5), language (n = 4), executive skills (n = 5) and psychological state (n=3). This observational study was approved by the institutional ethical standards committee on human experimentation. Written informed consent was obtained from all participants (or guardians of participants).

Results

Of 107 temporal epilepsy surgeries performed during the period January 2015 to June 2019, nine patients underwent surgery for TE; seven males and two females with a mean age of 22 years (age range of 8 years to 44 years). The age of epilepsy onset was from 4.5 years to 19 years (mean age: 14.5 years). Two patients had febrile seizures in childhood. Birth and development were normal in all patients without any history of perinatal insult. No patient had a history of head injury or any trauma. All nine patients were on a minimum of two ASMs (range: 2-4). All patients were right-handed with body mass index (BMI) ranging from 16.4 to 26.4 (mean: 21.7). Based on BMI categorisation, two patients were overweight but none were obese.

Semiology and EEG

Seven patients reported a subjective onset characterised by an unspecified aura in four, a non-specific pulling sensation or tingling in the head in two, and fear in one. Seven patients had focal non-motor seizures with impaired awareness and two had focal motor seizures. Six patients had clustering of seizures. Five patients experienced a focal to bilateral tonic-clonic seizure after drug withdrawal. One patient had a gyratory seizure, as described by Jagtap *et al.* [9] Interictal epileptiform discharges (IEDs) were confined to the ipsilateral temporal lobe; predominantly the anterior temporal lobe in all subjects (*table 1, figure 1*). Two patients showed additional, interictal, frontally dominant generalized discharges, suggestive of focal epilepsy with a genetically generalized epilepsy (GGE) trait (*figure 1*). The ictal pattern was temporal with a 6-8-Hz rhythm in seven patients and a 3-4-Hz rhythm followed by 5-7-Hz rhythm in the remaining two.

Imaging and surgery

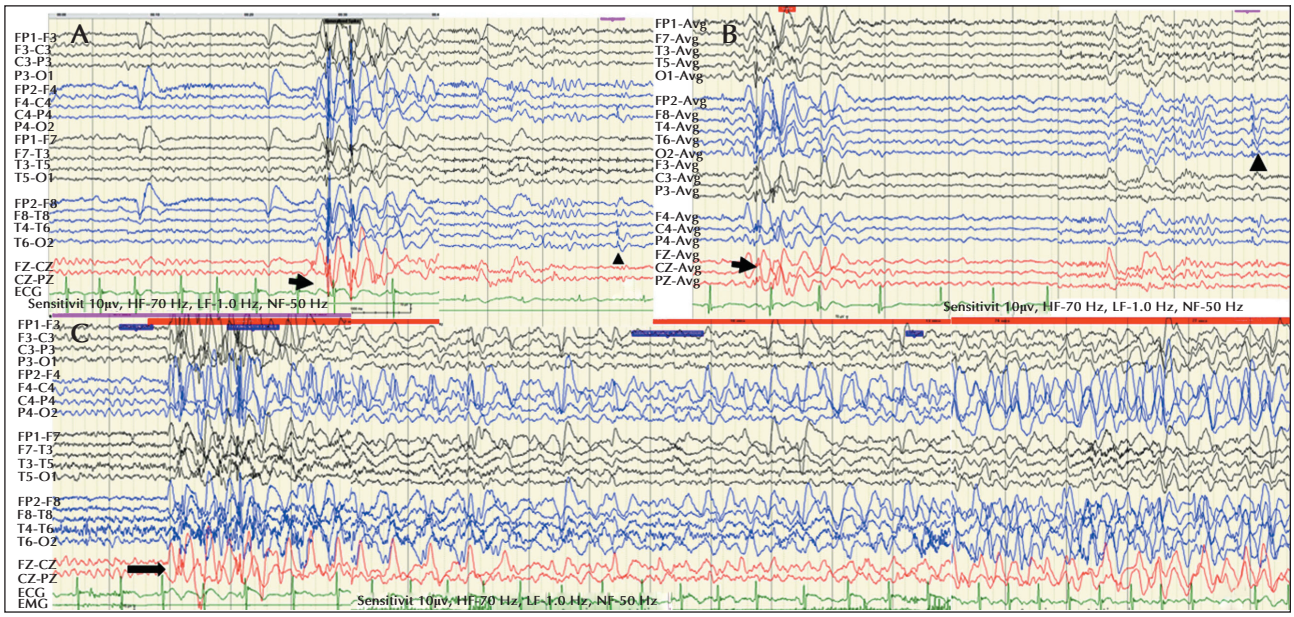
TE was initially reported in only two patients, and was missed in seven patients. The first patient with TE was diagnosed intra-operatively, while other cases were diagnosed during a patient management conference. The size of the encephalocele varied from 3.2×2.4 mm to 8.9×4.8 mm, with CSF (cerebrospinal fluid) signal abnormality around the encephalocele noted in five patients (*figures 2, 3*). PET was performed during the interictal period in all patients and showed unilateral temporal hypometabolism in all cases (*figure 2*). Seizure semiology, IEDs and ictal onset were lateralised to the MRI lesion.

Three patients underwent standard anterior temporal lobectomy while resection of the encephalocele with surrounding temporal pole under electrocorticography guidance was performed in the remaining six patients. Electrocorticography from mesial structures did not show any abnormal discharges in the first

▼ **Table 1.** Patient demographics, EEG findings, surgery, pathology and outcome.

Case No. Handedness	Age at onset	BMI	IEDS	Semiology	Ictal EEG	Surgery	Pathology	Follow-up	Engel outcome
1 Right	12	23.5	Left temporal	Unspecified aura, BA, stare, gyrates to left five times	Left anterior temporal polymorphic 6-8-Hz rhythm	Left ATLAH	Gliosis	44	Ia
2 Right	15	23.5	Left temporal	Unspecified aura, BA, rocking movement of both legs, right hand dystonia	Left temporal 3-4-Hz rhythm.	Left ATLAH	Gliosis	32	Ia
3 Right	17	26	Right temporal and frontally dominant generalised SWD	Pulling sensation in head, BA, stare, right hand automatism, eye deviation to left side, left hand posturing	Right temporal 5-7-Hz rhythm	Right ATLAH	Hippocampal sclerosis type I with reactive gliosis in amygdala with normal temporal cortex	30	Ia
4 Right	15	26.4	Left temporal	Unspecified aura, BA, irrelevant talk, chewing automatism	Left temporal polymorphic 6-7-Hz rhythm	Left ATL	Gliosis	19	Ia
5 Right	14	16.4	Right temporal	Tingling in head, BA, stare, right hand automatism	Right temporal 5-7-Hz rhythm	Right ATL	Gliosis	32	Ia
6 Right	4.5	18.2	Frontally dominant generalised spike and wave and right anterior temporal spikes	BA, stare, right hand automatism	Burst of 2.5-3-Hz generalised spikes followed by right fronto-temporal delta	Right ATL	Suspicious dyslamination with gliosis	34	IIa
7 Right	15	17.9	Left temporal and bifrontal generalised 3-3.5-Hz SWD	Unspecified aura, BA, stare	Left temporal polymorphic 5-6-Hz rhythm	Left ATL	Gliosis	17	Ia
8 Right	15	19.6	Right temporal	Fear, BA, Stare, left hand dystonia, right hand automatism	Right temporal 5-7-Hz rhythm	Right ATL	Gliosis	17	Ia
9 Right	17	22.7	Left anterior temporal spike-and-wave discharges	BA, stare, chewing automatism	Left temporal 6-7-Hz rhythm	Left ATL	Gliosis	20	Ia

ATL: anterior temporal lobectomy; ATL + AH: anterior temporal lobectomy with amygdalo hippocampectomy; BA: behavioural arrest; SWD: spike-and-wave discharges.



■ **Figure 1.** Interictal EEG. (A) Bipolar longitudinal montage. (B) Average montage showing frontally dominant generalized spike-and-wave discharges (arrow) and right temporal spikes (arrowhead). (C) Ictal EEG showing a burst of frontally dominant generalized spike-and-wave discharges, followed by right fronto-temporal temporal 2-3-Hz spike-and-wave discharges, evolving into right fronto-temporal polymorphic 5-6-Hz rhythm (Case 6).

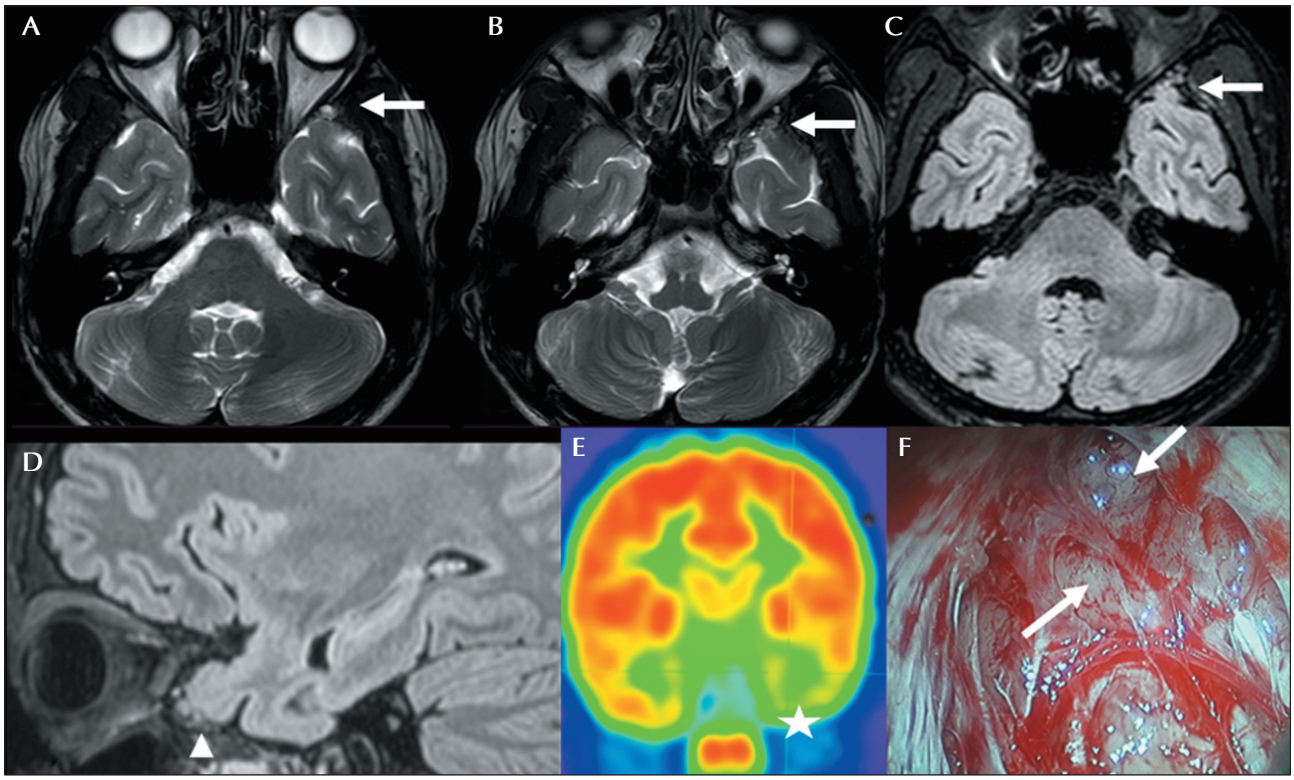
three patients, which led us to modify our surgical strategy with preservation of mesial structures. All patients had Engel Class I outcome, except one who had Class IIa outcome after a mean follow-up of 27 months (17-44 months). Histopathology confirmed gliosis in seven cases, hippocampal sclerosis type I with reactive gliosis in the amygdala with normal temporal cortex in one, and suspicious dyslamination with prominent gliosis in one (*table 1*).

Subjective follow-up of cognitive status

A total of eight patients were followed by telephone to gain information pertaining to subjective changes in their cognition and psychological state (*table 2*). Half of the patients (4/8) reported a subjective decline in attention and memory, while the other half felt that functioning in this domain remained stable or improved. Language skills declined in 50% of individuals with a left TE, with no decline reported by those with a right TE. No clear trends were apparent with regards to executive functioning. Interestingly, 6/8 individuals reported an improvement in their psychological state (mood, anxiety and motivation), while the remaining two reported a minor increase in anxiety.

Discussion

TE is a rare but important causative agent in medically refractory epilepsy. The exact prevalence of TE in temporal lobe epilepsy is not known, with estimates in the literature varying from 1% to 13% [12, 18]. Of 107 temporal lobe surgeries, nine showed TE; a prevalence rate of 8.4% compared to 12.5% by Campbell *et al.* [11]. Our first case was initially thought to be MRI-negative, PET-positive temporal lobe epilepsy. The patient underwent amygdalohippocampectomy (ATLAH), and during surgery, encephalocele was discovered which was evident on MRI, retrospectively [9]. Surgeons should cautiously investigate for encephalocele, especially during standard anterior temporal lobectomy for MRI-negative, PET-positive temporal lobe epilepsy patients. Encephaloceles are mostly spontaneous but may also be congenital or related to trauma, infection, inflammation or surgery [13]. In this series, all TEs were of spontaneous origin. Also, most patients were in the adolescence age group (mean age: 14.5 years) and the majority were male (77.7%). In previous studies, however, all patients were middle-aged (mean age: 34) with the majority being female (78%), pointing towards a genetic nature of encephalocele [13]. An association between higher BMI and

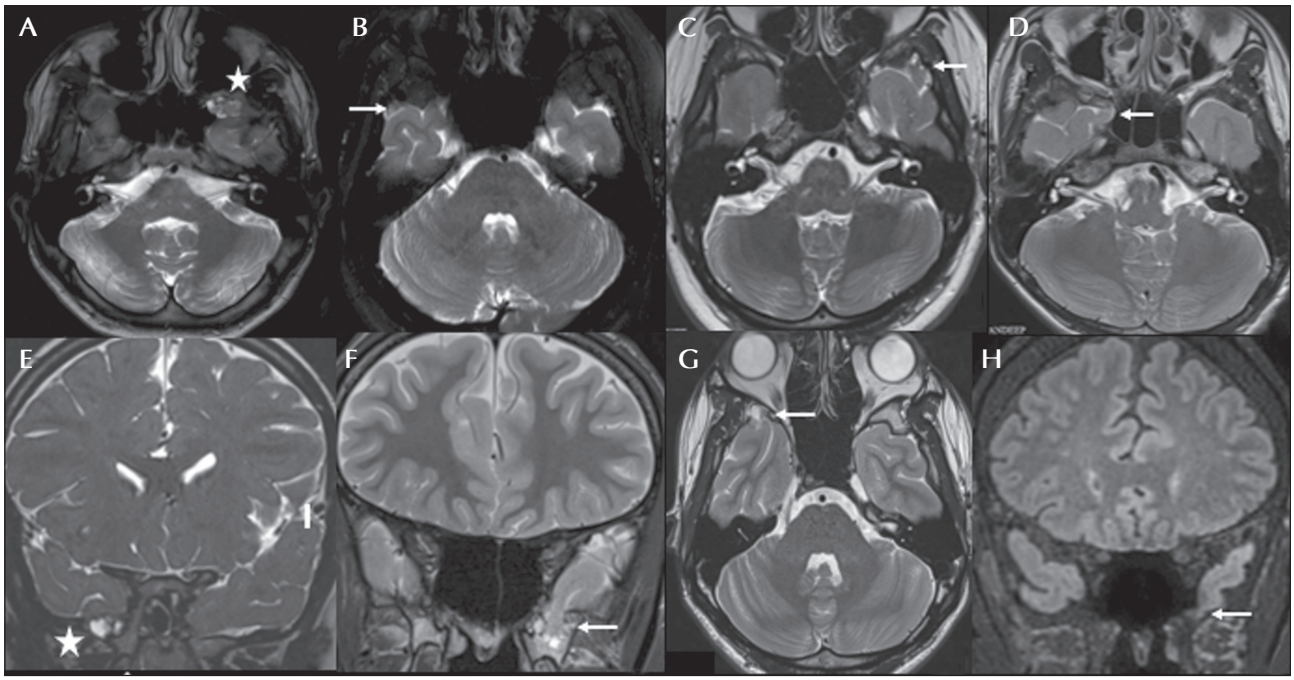


■ **Figure 2.** (A, B) Axial T2 image. (C) Axial FLAIR image showing an encephalocele (arrow) protruding through the left middle cranial fossa. (D) FLAIR sagittal image of encephalocele protruding through the left middle cranial fossa (arrowhead). (E) PET image showing left temporal hypometabolism (star). (F) Intraoperative photograph of encephalocele showing deficient dura and gliotic brain, terminating into bony pits (arrow) (Case 1).

encephalocele has been reported in prior studies [11, 16, 18]. One of the postulated causes for the development of encephalocele is higher BMI which is associated with idiopathic intracranial hypertension (IIH). In this study, mean BMI was 21.9 and the maximum was 26.4; only two patients were in the overweight category. In the study by Tse *et al*. [16], higher BMI correlated with late onset of epilepsy (>20 years). In our cohort, with lower BMI, epilepsy onset was earlier.

Video-EEG

The presence of non-specific or absence of aura, as well as clustering of seizures, points towards neocortical epilepsy rather than mesial temporal lobe epilepsy [19]. Video-EEG showed interictal epileptiform discharges (IEDs) confined to the temporal lobe, although two patients showed bifrontal and generalised IEDs, suggestive of focal epilepsy with a GGE trait. In the absence of focal spikes or sharp waves leading to bilaterally synchronous spike-wave discharges (BSSW), these frontally dominant generalized discharges, as secondary synchronous bilateral discharges, were less likely. Temporal lobe epilepsy with a GGE trait has been described in the literature although not with TE [20, 21]. In this study, the ictal pattern was temporal with a 6-8-Hz rhythm in seven patients and a 3-4-Hz rhythm followed by 5-7-Hz rhythm in the remaining two (*table 1*), comparable to the study by Panov *et al*. [15]. This points towards a focal temporal neocortical generator in two and a mesial temporal generator in seven patients. Mesial temporal epilepsy onset in most patients can be explained by functional connectivity between the mesial and neocortical temporal lobe. In the study by Panov *et al*. [15], depth electrode recording in two patients showed onset of TE with a rapid spread to the hippocampus, pointing towards TE as the source of epileptogenesis and later involving a network of mesial structures. Two patients showed poor lateralisation or localisation of ictal patterns, of whom one



■ **Figure 3.** (A) T2 axial image showing a left encephalocele (star) (Case 2). (B) T2 axial image showing a right encephalocele (arrow) (Case 3). (C) Axial T2 image showing a left TE (arrow) (Case 4). (D) Axial T2 image showing a right TE (arrow) (Case 5). (E) Coronal T2 image showing a right TE (star) (Case 6). (F) Coronal T2 image showing a left TE (arrow) (Case 7). (G) T2 axial image showing a right TE (arrow) (Case 8). (H) FLAIR coronal image showing a left TE (arrow) (Case 9).

underwent surgery with good outcome [15]. In our study, all patients had electroclinico-radiological concordance.

Imaging and histopathology

Of nine cases in this study, encephalocele was reported in only two, accounting for a 22.2% detection rate, as compared to 13.5% by Campbell *et al.* [11]. Here, we suggest careful review of all "lesion-negative" temporal lobe epilepsy cases for encephalocele, as subtle abnormalities may be present at the temporal tip [1, 2]. Also, critical analysis of MRI, before labelling it as normal, is required. Coronal reformation image analysis with CT, to look for skull defects, is an additional tool for the confirmation of encephalocele diagnosis [1].

The exact mechanism of epileptogenesis associated with TE is not known. It may be due to the dysmorphic tissue in the encephalocele. Another possibility is

▼ **Table 2.** Post-operative neuropsychological outcome.

	Left (*n* = 4)			Right (*n* = 4)		
	Decline	Improved	No change	Decline	Improved	No change
Attention	1	2	1	1	2	1
Memory	2	2	0	1	2	1
Language	2	2	0	2	1	1
Executive function	1	0	3	0	2	2
Psychological state	1	3	0	1	2	1

stretching of neural tissue in encephalocele resulting in gliosis with development of an epileptogenic network [7]. In our study, histopathology confirmed gliosis in seven cases, hippocampal sclerosis type I in one, and suspicious dyslamination with prominent gliosis in one patient. The mechanism of development of hippocampal sclerosis is not clear but it might be secondary to repeated seizures from encephalocele. The patient with hippocampal sclerosis had nine years of epilepsy before she underwent surgery.

Surgery

In patients with TE, an important question is whether the source of epileptogenesis is derived from the encephalocele itself or the surrounding cortex with a widespread network involving the hippocampus. Available surgical strategies for TE include resection of encephalocele with or without the removal of the surrounding temporal pole and standard anterior temporal lobectomy. Focal resection has been widely successful in ameliorating epilepsy, pointing towards a preference for tailored resection rather than ATLAH, to avoid deficits [1, 8, 16]. In our study, the first three patients underwent standard ATLAH while resection of encephalocele along with the surrounding temporal pole was carried out in six patients with good outcomes. Desirable surgical outcome following resection of encephalocele, with or without additional removal of the surrounding temporal pole, in present as well as previous studies, points towards encephalocele or immediately adjacent cortex as the epileptogenic network rather than mesial structures [1, 4, 7, 8].

Subjective follow-up regarding cognitive status

A notable finding in our cohort, albeit a very small sample, was that the majority of individuals (6/8) reported post-surgical improvement in their psychological state. Our observations are in concordance with those of Tse *et al*. [16]. Further, 50% of the patients reported a decline in attention and memory, regardless of the side of surgery. This is important when counselling patients for surgery with such manifestations. Follow-up with a larger cohort would allow for subgroup analysis (e.g., left vs right-sided surgery, tailored vs wider resection) and help identify factors that predict stability/decline in cognition.

The retrospective nature of the study and the small sample size are certainly restrictive in terms of the analyses that can be conducted. Nonetheless, the study provides preliminary support for the notion that all patients with MRI-negative temporal lobe epilepsy should be carefully investigated for the presence of encephalocele. If detected, a tailored resection of the encephalocele, along with the surrounding temporal pole while sparing mesial temporal structures, can be beneficial with good clinical outcome. ■

Key points

- All patients with MRI-negative temporal lobe epilepsy should be carefully investigated for the presence of encephalocele.
- Tailored resection of the encephalocele along with the surrounding temporal pole is associated with a good surgical outcome.

Supplementary material.
Summary slides accompanying the manuscript are available at www.epilepticdisorders.com.

Disclosures.
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TEST YOURSELF

(1) In a case of typical temporal lobe epilepsy thought to be MRI-negative, what other possibility should you consider before establishing that it is truly MRI-negative?

(2) What is the aetiopathogenesis of temporal encephalocele?

(3) What is the ideal treatment for temporal encephalocele?

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