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Anterior Temporal Encephaloceles:
Elusive, Important and Rewarding to Treat

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Abstract

Objective

To investigate the etiology and longitudinal clinical, neuropsychological, psychosocial and surgical outcome profile of patients with medication refractory epilepsy and temporal encephaloceles with a view to highlight diagnostic clues and management strategies.

Methods

The Comprehensive Epilepsy Program databases at two surgical epilepsy centers from January 2000 to October 2018 were reviewed for this observational study, to identify all patients with encephaloceles causing temporal lobe epilepsy and treated with surgical resection. Their clinical, radiological, neuropsychological, psychiatric and surgical data were obtained. Body mass index (BMI) data are also reviewed due to possible correlation between idiopathic intracranial hypertension and encephaloceles.

Results

Thirteen patients (8 female) were identified; only three were recognized on initial MRI report. Temporal encephaloceles were identified on the left in eight patients, on the right in three patients, and bilaterally in two patients. One patient had a strong family history of encephaloceles. The median BMI for patients with seizure onset ≤20 years of age was 22.4, whereas for patients with onset after 20 years median BMI was 32.6 (p = 0.06). Five patients underwent a focal lesionectomy, three patients had limited temporal lobectomy, and five had standard anterior temporal lobectomy. Median post-operative follow-up was 5.5 years. All but one patient were free of disabling seizures. Nine of ten neuropsychologically tested patients had no discernable cognitive decline post-operatively. Postoperative psychosocial adjustment features were present in four patients.

Significance

Genetic factors and a possible association with idiopathic intracranial hypertension (given female predominance and elevated BMI) may contribute to the causation of temporal lobe encephaloceles. Importantly, a targeted surgical approach in the management of patients with TLE associated with encephaloceles has an excellent long-term clinical and neuropsychological outcome. Subtle encephaloceles should be actively searched for in patients with drug resistant TLE because they significantly change surgical strategy and prognostication.

Key words: Epilepsy, temporal lobe; Encephaloceles; Drug resistant epilepsy; Anterior temporal lobectomy; Neuropsychology

Key points

- Small temporal lobe encephaloceles are an underrecognized but important cause of temporal lobe epilepsy.
- Family history and possibly idiopathic intracranial hypertension should be added as risk factors for encephaloceles.
- Targeted surgical resection of these lesions has excellent clinical and neuropsychological outcome.

- Encephaloceles should be carefully and actively searched for because they change surgical strategy and prognostication.

Encephaloceles are herniations of cerebral tissue through a skull defect, and can be classified according to their location or etiology[1]. They occur rarely in neonates as overt birth defects, often associated with spina-bifida. In contrast, inapparent small temporal lobe encephaloceles are a separate clinical entity, and are an underrecognized but important cause of temporal lobe epilepsy (TLE). The clinical relevance of small meningo-encephaloceles has long been recognized in the oto-laryngological literature given the association with cerebral spinal fluid (CSF) leak presenting as CSF rhinorrhea or otorrhea and subsequent CNS infection [1-5]. An association with high body mass index (BMI) and intracranial hypertension has been noted[6-8].

The association of encephaloceles with epilepsy has only been more widely recognized in recent years. Earlier literature on encephalocele-associated TLE predominantly comprise case reports or small case series[9], often with larger lesions. More recent papers, however, enabled by improved MR imaging, have suggested that small encephaloceles are more common amongst patients with refractory TLE than previously suspected[10-13]. Prior to recognizing the encephaloceles, such patients were usually deemed to have 'lesion negative' TLE. Given the good surgical outcome in the majority of cases[10-14] of encephaloceles, accurate detection potentially translates to a significantly improved prognosis in this cohort who might otherwise be denied surgical treatment.

Despite increasing reports of encephaloceles in TLE, there remains notable knowledge gaps in the current literature. Risk factors are unclear, the neuropsychological and psychiatric profiles of these patients have not been well-described, multidisciplinary surgical outcome measures have not been reported, and the ideal surgical approach remains unclear. The aim of this study was to comprehensively characterize the longitudinal clinical, neuropsychological, psychosocial and surgical outcome profile of 13 patients with medication refractory epilepsy arising from temporal encephaloceles with a view to

identifying diagnostic indicators, seizure management strategies, and non-epilepsy associated comorbidities of this entity.

Methods

A retrospective review of the patient database from the Austin Health and St. Vincent's Hospital Comprehensive Epilepsy Programs (CEP) from January 2000 to October 2018 was conducted, to identify patients with encephaloceles causing temporal lobe epilepsy and treated with surgical resection. Hospital medical records were reviewed to extract epidemiological and surgical data, together with pre-and postoperative epileptological, neuropsychological, psychiatric, and psychosocial information.

All patients underwent MRI (1.5 or 3 Tesla) of the brain. MRI sequences were obtained as per the institution protocol for epilepsy patients, including volumetric T1 with axial, coronal and sagittal reformatted images, volumetric or coronal fluid-attenuated inversion recovery (FLAIR); coronal T1 and T2; axial susceptibility weighted imaging, diffusion weighted imaging, and coronal IR. If encephalocele was suspected on MRI, a fine-slice base of skull CT was often performed to confirm its presence. All MRI studies were reviewed with a neuroradiologist with particular expertise in epilepsy imaging to identify the encephaloceles and characterize their imaging features. FDG-PET and CT scans results were also reviewed when performed as part of the surgical workup.

The 10 Austin Health patients underwent a neuropsychological evaluation as a routine component of their pre- and postsurgical evaluation. An estimate of general intellectual background was obtained using the Wechsler Test of Adult Reading (WTAR)[15]. Memory function was assessed using the Rey Auditory Verbal Learning Test, Form 1 (RAVLT: Total Recall Trials 1 – 5 and Trial 6[16]), the Wechsler Memory Scale (WMS) Paired Associate Learning Subtest (PAL: total 'easy' and total 'hard' word pairs recalled over three learning trials[17]), and the Rey-Osterreith Complex Figure Test (RCFT: 30 minute delayed recall[16]). In addition, naming function was assessed with the Boston Naming Test, 2nd Edition (BNT[18]).

All patients were interviewed pre- and post-operatively by a psychiatrist with expertise in psychiatric comorbidities associated with epilepsy. For Austin Health patients, a semi-structured interview (the Austin CEP Interview[19-22]) was employed to address issues pertinent to psychosocial functioning. Patients also received routine post-operative follow up including regular telephone contact with an epilepsy nurse consultant and face-to-face reviews by a clinical neuropsychologist, as per the standard follow-up protocol.

This study was approved by the Austin Health Human Research Ethics Committee (LNR/15/Austin/61).

Results

A total of 13 patients were identified (10 from Austin Health, three from St. Vincent's Hospital), three were reported previously[10]. Eight patients were female. Patient characteristics, encephalocele location, surgical technique and outcomes are summarized in Table 1.

Clinical characteristics

The median age of seizure onset was 20 years (range 12-47 years); the median age at surgery was 39 years (range 17-58 years). All patients had focal seizures with impaired awareness. Some patients had isolated auras (5/13), and the majority had bilateral tonic-clonic seizures (11/13). Semiology was variable but typical for temporal lobe seizures, often involving a combination of symptoms such as déjà vu, olfactory/gustatory hallucinations, intense emotions, speech disturbances, behavioral arrest, oral and manual automatisms.

Temporal encephaloceles were identified on the left in eight patients, on the right in three patients, and bilaterally in two patients. Video EEG monitoring localized seizures to the left temporal lobe in nine patients, and to the right in four patients.

Risk factors for encephalocele development

One patient (Case 3) had a dysembryoplastic neuroepithelial tumor (DNET) associated with the encephalocele. Two patients (Cases 2 and 11) had head trauma prior to the onset of seizures. Case 2 reported onset of seizure two months after an assault with head strike leading to loss of consciousness. Case 11 reported onset of seizure three months after a motor vehicle accident leading to head-strike without loss of consciousness. Neither of these patients had documented intracranial injuries or required neurosurgical intervention as a result of the trauma, so the relationship between these events and encephalocele development is dubious. No structural or traumatic precipitants were identified for the development of encephaloceles in other patients.

One patient (Case 7) had a strong family history of encephaloceles, with a history of right ear CSF leak in her mother and sinusitis in her maternal aunt, which both subsequently led to a diagnosis of encephaloceles in the middle cranial fossa not associated with seizures (Figure 1). There was no family history of epilepsy otherwise. Chromosome microarray was unremarkable, and whole exome sequencing did not reveal a pathogenic variant with particular attention to genes involved with connective tissue.

The median body mass index (BMI) at the time of surgery was 24.8 (IQR: 21.3-30). None of the patients reported significantly different body habitus at the time of seizure onset compared to time of surgery. The median BMI for patients with seizure onset at or prior to 20 years of age was 22.4 (IQR: 20.9-24.8), whereas for patients with seizure onset after 20 years of age was 32.6 (IQR: 23.5-37.6; p=0.06, Student t-test).

Imaging

Encephaloceles were diagnosed in the routine clinical report in only 3/13 cases, although non-specific findings were reported in another three cases. The other cases were identified on neuroradiology review for CEP clinical meetings, in the context of localizing clinical information. Identification of encephaloceles was aided by a systematic search for an alteration of the brain surface contour (13/13 cases), CSF signal around the suspected lesion (9/13) and adjacent parenchymal gliosis (4/13) (Table 2). All encephaloceles arose from the anterior temporal lobe and projected through the floor or anterior pole of the middle cranial fossa, in the greater wing of the sphenoid bone (Figure 2), with the majority (12/13)

in the antero-medial aspect. High resolution CT scanning through the base of skull was helpful in confirming the presence of a skull defect in five patients, one of which had an additional CT venogram to exclude an emissary vein.

Eleven patients had FDG-PET imaging, and all showed temporal lobe hypometabolism in the corresponding side of the seizure lateralization (and side of the encephalocele in unilateral cases).

Surgical approach

Five patients underwent standard anterior temporal lobectomy (ATL), of which one patient (Case 6) had additional limited resection of the hippocampus, part of the amygdala and uncus; and one patient (Case 1) had additional partial resection of the amygdala. The hippocampus and amygdala were spared in all other patients. Three patients had limited temporal lobectomy and five patients had focal lesionectomy of the encephalocele.

Histopathology

All resected specimen showed features consistent with encephaloceles, such as astrocytosis, disturbance of neuronal lamination, and molecular layer gliosis. Case 3 had an associated DNET, and Case 2 had features of mild cortical dysplasia with mild distortion of the gyral profile, with a multinodular surface, and disarray of adjacent underlying neurons.

Surgical outcome

Patients were followed up for a median of 5.5 years (range 1.5-15 years) post-operatively. Ten patients have remained completely seizure free, six of whom have been off all anti-epileptic drugs (AED) (Table 1). One patient is free of disabling seizures and having rare auras only (Case 5). One patient had two seizures provoked by medication non-compliance and lifestyle factors, but has otherwise remained seizure free (Case 8). One had recurrence of medication refractory seizures after 5 years of initial seizure freedom (Case 6, bilateral encephaloceles). Both patients with post-operative seizures underwent standard lobectomy (ATL). All patients who underwent lesionectomy remain seizure-free.

Longitudinal pre and post-surgical neuropsychology findings

For the 10 patients evaluated in detail (Table 3), WTAR confirmed normal background intellect in eight of the 10 patients. In one case (2), significant test-based anxiety limited meaningful psychometric assessment of intellect and cognition, but native intellect impressed as clinically normal. For Case 12, there was a personal and familial history suggestive of a developmental dyslexia, background intellect was otherwise normal.

Relative to population norms, seven patients had normal memory function. Naming function was also normal in five of those same cases. Three patients (Cases 4, 10 & 12) had objective evidence of a mild dysnomia which converged with a subjective complaint. One patient (Case 8) had a mild verbal memory impairment, a finding that converged on his well substantiated account of forgetfulness for verbal information over the preceding decade. One patient (Case 6) had objective evidence of a mild preoperative nonverbal memory deficit (as measured on a 30 minute delayed recall of the RCFT).

Three-month postoperative neuropsychological evaluation of the 10 Austin Hospital cases showed no changes, with the exception of Case 3 (who had a standard left anterior temporal lobectomy) in whom a new mild test-based dysnomia was evident, which was functionally asymptomatic. By contrast, for Case 10 a significant *improvement* in naming function was documented at both an objective and subjective level. In Cases 2 and 6, formal psychometric re-evaluation was not possible. In neither case, however, was there a subjective account of altered cognitive function in daily life. Given the excellent neuropsychological outcomes, no further formal reviews were performed beyond three months post-surgery.

Psychiatric and Psychosocial Features

Eight of the 13 patients presented with an active and/or past history of psychiatric symptoms, notably increased anxiety and low mood. Only one patient (Case 2) was undergoing active mental health treatment at the time of surgery. Three patients

experienced an acute exacerbation of anxiety and mood symptoms in the first three months postoperatively. For all but one patient (Case 2), symptoms settled gradually with intervention including psychotropic medication and psychological therapy. Following surgery, six patients described improved levels of self-confidence, a more positive outlook, lowered anxiety, less avoidance of social gatherings, and an overall increase in life satisfaction and contentment.

In four patients, seizure freedom was accompanied by a pattern of psychosocial adjustment as conceptualized by the model of the "burden of normality"[20-22]. Two patients (Cases 5 & 8) expressed a desire to "make up for lost time" (e.g., take-up mountain climbing, scuba dive, parachute, run a marathon). For one patient (Case 8) a feeling of "invincibility" manifested as sense of 'cure' leading to an isolated episode of non-compliance with his anti-epilepsy drug regime and excessive alcohol consumption with a resultant seizure. By contrast, for a further two patients (Cases 3 & 7), the early postoperative phase was characterized by a period of intense self-reflection and 'grieving' associated with the stigma of epilepsy and sense of social exclusion and non-acceptance. For one of these patients (Case 7), this process was associated with an extended period of recovery characterized by the persistence of a range of somatic complaints over several months.

Nine patients were working at the time of their surgery and all had returned to work post-surgery. One patient (Case 9, age 17 years) returned to school post-surgery. Two patients viewed seizure control as an opportunity to alter their employment outlook (i.e., seeking increased challenges), and one of those patients (Case 4) went onto complete postgraduate qualifications (i.e., PhD).

On direct enquiry, all patients were satisfied with their decision to undergo surgical treatment – many commenting that [surgery] had "changed my life" with a related sense of gratitude for the opportunity to "now feel normal".

Discussion

We describe a series of 13 patients with TLE associated with small temporal encephaloceles treated with surgery. In addition to excellent surgical outcome, a number of clinical

characteristics were analyzed, which have important implications in the diagnostic and management approach to this disorder.

Possible etiology and proposed diagnostic and management approach

Encephaloceles can be a result of brain trauma or be associated with tumours, infections, or inflammatory disorders. However, the majority of cases reported in the literature with small temporal encephaloceles associated with epilepsy do not have an apparent cause and are therefore classified as spontaneous[11-13]. Only one patient in our series of 13 had an identifiable cause (encephalocele associated with a developmental tumour). Although a number of possible pathophysiological models have been suggested[1, 2], the cause for these "spontaneous" cases remains unclear.

Studies on encephaloceles resulting in CSF leaks have shown associations with high BMI and features of idiopathic intracranial hypertension (IIH), including a high incidence of raised CSF opening pressure[5-8]. Similar associations have been suggested in patients with epilepsy in two recently published radiology papers that found a positive correlation between the presence of small temporal encephaloceles and MRI findings suggestive of IIH[23, 24]. In our study, there was a female predominance, which is a known risk factor for IIH. Review of the literature including 194 cases showed that 58% were female[2, 11-14, 23-33]. In addition, our results suggest a trend for patients who developed seizures at an older age to have higher BMI compared to those who developed seizures earlier in life. The two patients with bilateral encephaloceles both developed seizures late in life and had the highest BMI in our cohort (cases 6 and 10).

Additionally, a strong family history of encephaloceles was noted in one patient in our series, consistent with an autosomal dominant pattern. Genetic mutations have been identified for various syndromes associated with congenital and clinically apparent meningoencephaloceles, such as lateral meningocele syndrome (NOTCH3)[34] and ciliopathies, including Meckel syndrome and Joubert syndrome (NPHP1, AHI1, MKS1, RPGRIP1L etc.)[35, 36].   Our literature review, however, did not reveal reports of mutations associated with small temporal encephaloceles or other cases with a similarly strong family history.

Our findings raise the possibility that at least two mechanisms may be present in the development of small temporal encephaloceles. One group of patients may have congenital predisposition to developing encephaloceles, often with early clinical manifestations. The second group presents with seizures later in life, with high BMI or IIH being potential risk factors; at what stage the encephaloceles develop, however, is presently unknown. Given our findings, we propose that the IIH symptoms and its risk factors, as well as family history of non-seizure related symptoms of encephaloceles (such as CSF leak and meningitis) should be explored during assessment of "lesion-negative" TLE patients, particularly those that are medication refractory. The presence of such clinical features should raise suspicion for encephalocele-associated epilepsy, and should prompt careful review of MRI or further imaging such as high-resolution CT of the skull base or high resolution 3D T2-weighted acquisition directed to the anterior temporal lobes; the latter is our preference in recent years.

Targeted surgical approach and seizure outcome

Our series underscores that temporal lobe encephaloceles are an underrecognized but important cause of epilepsy. They often present as subtle lesions on imaging and are difficult to detect. Indeed, half of the patients in our study had an MRI report that was initially reported as normal. Moreover, there appears to be a left-sided predominance and surgical intervention for "lesion-negative left TLE" is usually a cause for concern regarding post-operative language and memory impairment. In our series of 13 patients, all but one patient achieved excellent surgical outcome, while having minimal surgery-related neuropsychological morbidities. Therefore, finding this subtle lesion in patients that are otherwise considered "lesion negative" significantly changes their surgical prognosis.

The optimal surgical approach in these patients remains unclear. While most of the reported cases underwent anterior temporal lobectomy (ATL) with amygdalohippocampectomy (AH), ATL with disconnection, or standard ATL; a smaller number of cases reported in various studies suggest that smaller resections such as tailored temporal pole surgery, lesionectomy or disconnection surgery may be sufficient in achieving seizure-freedom[11, 13, 27, 30, 32]. More recently, minimally invasive approach with MR-guided

laser thermal therapy has also been reported[33]. In our study, five patients underwent lesionectomy only and a further three had a limited resection and all had an excellent outcome. Although our series is small and uncontrolled, their excellent surgical outcome suggests that targeted surgical approach may be appropriate, with dramatically favorable seizure and neuropsychological outcome in comparison to the standard treatment for lesion-negative TLE and conventional ATL.

<u>Cognitive and psychosocial Outcome</u>

Of the nine patients in whom pre-operative psychometric testing was possible, three cases exhibited a mild dysnomia and two cases had evidence of mild lateralized memory deficits. This cognitive pattern differs from that evident in patients with mesial temporal sclerosis in whom a more severe and extensive pattern of cognitive deficit is usually apparent (particularly in left-sided cases[37]), possibly reflecting the very discrete nature of these lesions. Interestingly, an impairment of semantic memory associated with small left temporal polar encephalocele's has been reported[13]. Whether such an impairment represents a characteristic neuropsychological profile feature of this particular patient group, as hypothesized by Toledano et al[13], could not be addressed in our small series, and further exploration with a larger patient cohort and targeted assessment is required.

The most salient cognitive finding in our series is that that resection of the encephalocele appears to have very low neuropsychological morbidity. In only one patient in our series, who notably underwent standard left ATL, was there any psychometrically discernible decline in cognitive status, and even then the patient's post-operative naming decrement was mild and without any significant functional ramifications. For a small group of patients, naming function in fact improved following surgery. These findings stand in contrast to the neuropsychological outcome in other surgically treated TLE patient groups, in particular, lesion-negative cases undergoing standard anterior temporal lobectomy[38, 39].

These patients, however, are not 'immune' to the psychiatric comorbidities that are seen with greater prevalence in the epilepsy population[40, 41]. Postoperative psychosocial adjustment features were present in approximately one third of our sample, being typical of the general epilepsy surgery population[20]. It is well recognized that seizure surgery (and the

related sudden "wellness") can set in train a complex process of adjustment, the symptoms of which can directly inform management strategies and the related clinical outcome for both the patient and the wider family network[20, 21]. Given that seizure freedom is a precursory condition for the 'burden of normality' and that resection of an encephalocele has a high likelihood of rendering the patient seizure-free[11], the presence of postoperative adjustment issues in this surgical subgroup requires particular vigilance and active intervention. Access to a structured seizure surgery follow-up program, as well as access to targeted psychological and psychiatric interventions are crucial if the benefits of seizure relief and psychosocial outcome are to be maximized[20-22].

Patients who underwent lesionectomy have relatively shorter follow-up (1.5 to 3 years) due to a shift to a more targeted surgical approach in recent years. Longer follow up would strengthen the evidence in determining the feasibility of smaller, targeted surgical resection in this group of patients. Targeted studies will be required to further confirm and delineate the risk factors and underlying pathophysiology of the development of these small encephaloceles. One caution in pre-surgical evaluation is that as imaging and image interpretation improves, more small incidental encephaloceles will be found[42]. This reinforces that the principle in pre-surgical localization, of concordance of independent lines of investigation, should be obeyed before regarding an encephalocele as the epileptogenic lesion.

The findings of our study have important clinical implications for the diagnosis and surgical management of encephalocele-associated temporal lobe epilepsy, with data demonstrating excellent surgical outcome including those who underwent limited resections. This would support consideration for a targeted or stepped surgical approach in this patient group, which has lower surgical and neuropsychological risks than standard ATL. Given the significant implication for management and prognostication, it is prudent that clinicians actively search for the presence of these often radiologically subtle encephaloceles routinely in patients with drug-resistant TLE.

Disclosures

None of the authors has any conflict of interest to disclose.

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines

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Table 1. Summary of patient characteristics (in chronological order from oldest surgery date)

Case	Sex	Age onset	Age surgery	BMI	Side of seizure	Initial MRI report	Location of encephalocele	Surgical technique	Pathology	Outcome
1	F	17	24	20.6	R	Right encephalocele	Right anterior temporal pole	R ATL + partial amygdala resection	Encephalocele	Engel 1a. Seizure free 15 years. Off AED
2	F	29	39	21.7	L	Subtle lesion, unclassified	Antero-medial floor of the left middle cranial fossa	L ATL	Mild cortical dysplasia	Engel 1a. Seizure-free 11 years on AED
3	F	13	48	24.8	L	Focal lesion likely dyplastic potential encephalocele	Antero-medial floor of the left middle cranial fossa	L ATL	DNET	Engel 1a. Seizure-free 11 years. Aura when off AED, none since AED restarted
4	F	12	27	17.7	L	Lesion neg	Antero-medial floor of the left middle cranial fossa	Limited L ATL	Encephalocele	Engel 1a. Seizure-free 11 years. Off AED 7 years
5	M	20	27	24.9	L	Possible lesion, unclassified	Antero-medial floor of the left middle cranial fossa,	Limited L ATL	Encephalocele	Engel 1b. Seizure-free 10.5 years, rare auras only. Off AED 1 year

6	F	47	58	38.0	R	Lesion neg	Bilateral encephaloceles R>L. Antero-lateral floor of the right middle cranial fossa	R ATL + partial AH	Encephalocele	Engel 3b. Seizure-free 5 years then seizure recurrence (unclear laterality) on AED
7	F	31	40	36.6	L	Lesion neg	Antero-superior left middle cranial fossa	Limited L ATL	Encephalocele	Engel 1a. Seizure-free 5 years on AED
8	M	41	55	28.7	L	Lesion neg	Antero-medial floor of the left middle cranial fossa	L ATL	Encephalocele	Engel 1c. Seizure-free 5.5 years. Rare seizures only when non-compliant with AED
9	M	16	17	21.3	L	Lesion neg	Antero-medial left middle cranial fossa	L lesionectomy	Encephalocele	Engel 1a. Seizure-free 3 years. Off AED
10	F	37	43	45.7	L	Bilateral encephalocele	Bilateral multiple temporal lobe encephaloceles L>R, mostly anteromedial floor. Resected lesion in lateral floor of left middle cranial fossa.	L lesionectomy	Encephalocele	Engel 1a. Seizure-free 1.5 year on AED

11	F	13	43	30.0	R	Right encephalocele	Antero-medial floor of the right middle cranial fossa	R lesionectomy	Encephalocele	Engel 1a. Seizure-free 2 year on AED
12	M	16	27	22.4	L	Lesion neg	Antero-superior left middle cranial fossa,	L lesionectomy	Encephalocele	Engel 1a. Seizure free 1.5 year. Off AED 0.5 years
13	M	32	35	20.1	R	Lesion neg	Two encephaloceles at right anterior temporal pole, and one antero-lateral meningocele	R lesionectomy	Encephalocele	Engel 1a. Seizure free 1.5 year. Off AED

F: female, M: male; BMI: body mass index; R: right; L: left; neg: negative; ATL: anterior temporal lobectomy; AH: amygdalohippocampectomy; DNET: dysembryoplastic neuroepithelial tumor; AED: anti-epileptic drug

Table 2. Imaging characteristics

Case	Size of encephalocele (Width x Length)	Brain contour abnormality*	CSF signal around encephalocele	Parenchymal gliosis#
1	5mm x 5mm	Y	Y	N
2	12mm x 6mm	Y	N	N
3	14mm x 5mm	Y	Y	Y (associated DNET)
4	20mm x 6mm	Y	Y	N
5	11mm x 10mm	Y	Y	N
6	R: 4mm x 3mm L: 3mm x 2mm	Y	N	N
7	10mm x 5mm	Y	N	N
8	14mm x 9mm	Y	Y	N
9	6mm x 12mm	Y	Y	N
10	Multiple bilateral. Resected lesion 2mm x 2mm Largest lesion (left) 4mm x 3mm	Y	N	Y
11	4mm x 4mm	Y	Y	Y
12	8mm x 12mm	Y	Y	Y
13	3mm x 3mm, 4mm x 4mm	Y	Y	N

* Brain protrusion not conforming to normal gyration pattern; # T2 and/or FLAIR hyperintensity within the lesion; R: right; L: left; CSF: cerebrospinal fluid; Y: present; N: not present; DNET: dysembryoplastic neuroepithelial tumour

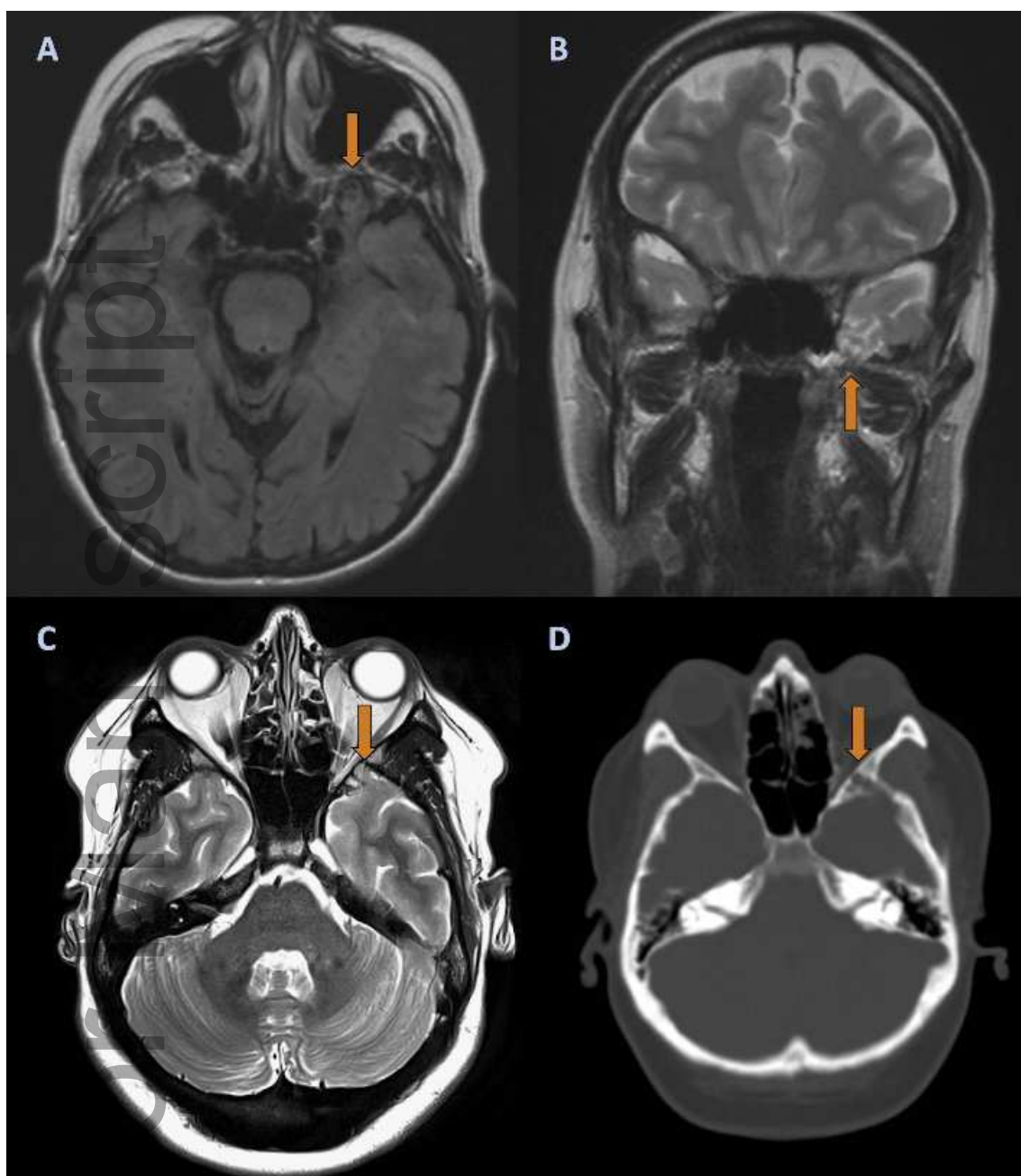**Table 3. Pre-and postoperative neuropsychological findings.**

Case	WTAR (Predicted FSIQ)	RAVLT (Total trial 1 – 5/A6)		RCFT (30' recall)		BNT 60-item		PAL (Total Easy/Hard)	
		T1	T2	T1	T2	T1	T2	T1	T2
2	*	*	*	*	*	*	*	17/7	15/8
3.	107	61/12	-	16.5	-	53	43	18/11	18/8
4.	109	60/12	67/14	21	-	47	50	18/11	18/11
5.	111	53/12	-	18	-	53	54	17/8	18/10
6.	95	59/12	-	12	-	57	-	18/8	-
7.	120	57/12	67/15	30	27.5	55	53	18/9	18/11
8.	102	41/8	43/7	23.5	18	53	56	14/4	17/4

10.	104	54/10	50/11	20	18	44	52	17/9	18/7
11.	111	55/12	58/15	28	24	52	54	18/10	18/11
12.	78 #	54/11	57/10	23	32	41	45	17/9	17/7

Note. WTAR (Wechsler Test of Adult Reading Ability); RAVLT, Form 1 (Rey Auditory Verbal Learning Test: Trial 1 -5 [maximum score = 75], Trial A6 [maximum score = 15]); RCFT (Rey Osterrieth Complex Figure Test: maximum score = 36); BNT, 2nd Edition (Boston Naming Test: maximum score = 60); Wechsler Memory Scale, PAL (Paired Associate Learning subtest: Total Easy Word Pairs Trials 1 – 3 [maximum score = 18], Total Hard Word Pairs Trial 1 – 3 [maximum score = 12]); T1 = Preoperative assessment; T2 = Postoperative review; * Significant test-based anxiety/poor engagement precluded valid psychometric evaluation; # Prominent developmental dyslexia
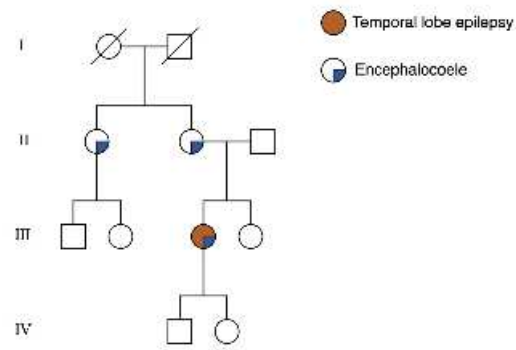
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