Case Report

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Intraoperative electrocorticography-guided resection of the epileptogenic zone in an unusual porencephalic cyst: case report and literature review

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Introduction: Porencephalic cysts resulting from perinatal artery infarctions typically manifest as large cysts accompanied by pre-existing neurological deficits. A small porencephalic cyst without any neurological deficit is a rare cause of medically refractory epilepsy.

Case presentation: A 23-year-old female presented with a history of medically refractory epilepsy secondary to a small right parieto-temporal porencephalic cyst. Despite optimal anti-seizure medications, seizures persisted. Surgical intervention was planned, and intraoperative electrocorticography (ioECoG) was used to delineate the epileptogenic zone (EZ), which was found to be two gyri posterior to the cyst.

Discussion: Very focal ischaemia resulting in a small porencephalic cyst from perinatal artery infarction exhibits a distinct organization of the EZ involving wider area posteriorly indicating involvement of arterial territory distal to the cyst. This contrasts with the typical perilesional EZ observed in other lesional epilepsy causes.

Conclusion: Our findings emphasize the need to consider aetiology during interpretation of ioECoG to better define the electrophysiological border between the normal and epileptogenic brain, aiding in achieving a better surgical outcome.

Keywords: Case report, congenital lesions, epilepsy surgery, intraoperative electrocorticography, porencephalic cyst

Introduction

A porencephalic cyst is a rare condition of cerebrospinal fluid (CSF) accumulation within the brain parenchyma[1]. It can manifest with a wide range of clinical presentations, including epilepsy, of which 25% can be intractable[2]. Porencephalic cyst formation is related to pre-and perinatal cerebral artery occlusion with middle cerebral artery (MCA) territory being the most commonly involved[3]. Ischaemic infarction in early uterine life leads to focal loss of cortical neurons and architectural derangement, resulting in polymicrogyria. In contrast, the same accident occurring at a later phase of pregnancy or in the perinatal period can lead to brain cavitation and gliosis[3]. The necrotic brain tissue due to perinatal infarction undergoes extensive resorption due to its high water content and poor density of unmyelinated fibres, allowing the formation of cystic cavities[2].

Most children with large cysts exhibit severe epileptic syndromes, and hemispherotomy is the classic treatment option when congenital hemiparesis and hemianopsia are present[1]. Small cysts with no intellectual impairment or hemiparesis are rare in presentation. However, they can be associated with epilepsy, which may be medically refractory[4].

Focal resection, cyst wall excision and cyst fenestration can be considered as surgical options[5]. However, none of these surgeries can accurately estimate the gliotic epileptogenic cortex, which may extend beyond the cyst and the pericystic margins[6]. Here, we report a case of intractable epilepsy due to a congenital

HIGHLIGHTS

- Rare case of a small porencephalic cyst, deviating from the typical larger cyst presentation.
- Use of intraoperative electrocorticography for precise delineation and resection of the epileptogenic zone (EZ)
- Solely posteriorly organized EZ suggests involvement of arterial territory distal to the cyst, highlighting unique pathophysiology of lesional epilepsy due to congenital vascular malformations.

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small porencephalic cyst in a patient with no motor or cognitive deficits. We utilize intraoperative electrocorticography (ioECoG) to delineate the epileptogenic zone (EZ) extending beyond the pericystic margins.

Case presentation

History

A 23-year-old female patient presented with medically refractory seizures. The seizures began at the age of 9 and were characterized by tingling of the left upper limb, which progressed to focal to bilateral tonic-clonic seizures. The episodes lasted for 2–3 minutes. Over the last 1 year, the frequency of seizures had increased, with an episode occurring every 10–12 days. Despite receiving anti-seizure medications (ASM) at optimal doses, including levetiracetam (1250 mg twice a day) and sodium valproate (500 mg twice a day), the patient's seizures remained poorly controlled.

The patient's mother had a history of twin pregnancies. The patient and the twin were delivered pre-term at 7 months with a low birth weight of 2300 g. The patient spent 3 months in the neonatal ICU (NICU) following delivery, but unfortunately, the twin did not survive. According to the patient's mother, no significant events occurred during the patient's NICU stay.

Apart from a suggestive perinatal history, there was no history of febrile seizures, encephalitis, metabolic disease, head trauma, or CSF rhinorrhoea. The family history was negative for any instances of epilepsy. The patient did not undergo genetic testing as her epilepsy was attributed to a lesional cause, and there were no other clinical characteristics warranting a genetic assessment.

Pre-surgical evaluation

MRI 3 Tesla showed a right parieto-temporal cystic structure communicating with the subarachnoid space and the right sylvian fissure, with the cyst appearance consistent with CSF signal in all sequences. There was no post-contrast enhancement. The features of the cyst were suggestive of a porencephalic cyst. (Fig. 1). Various potential differential diagnoses were considered and ruled out. Post-traumatic cyst was excluded due to absence of

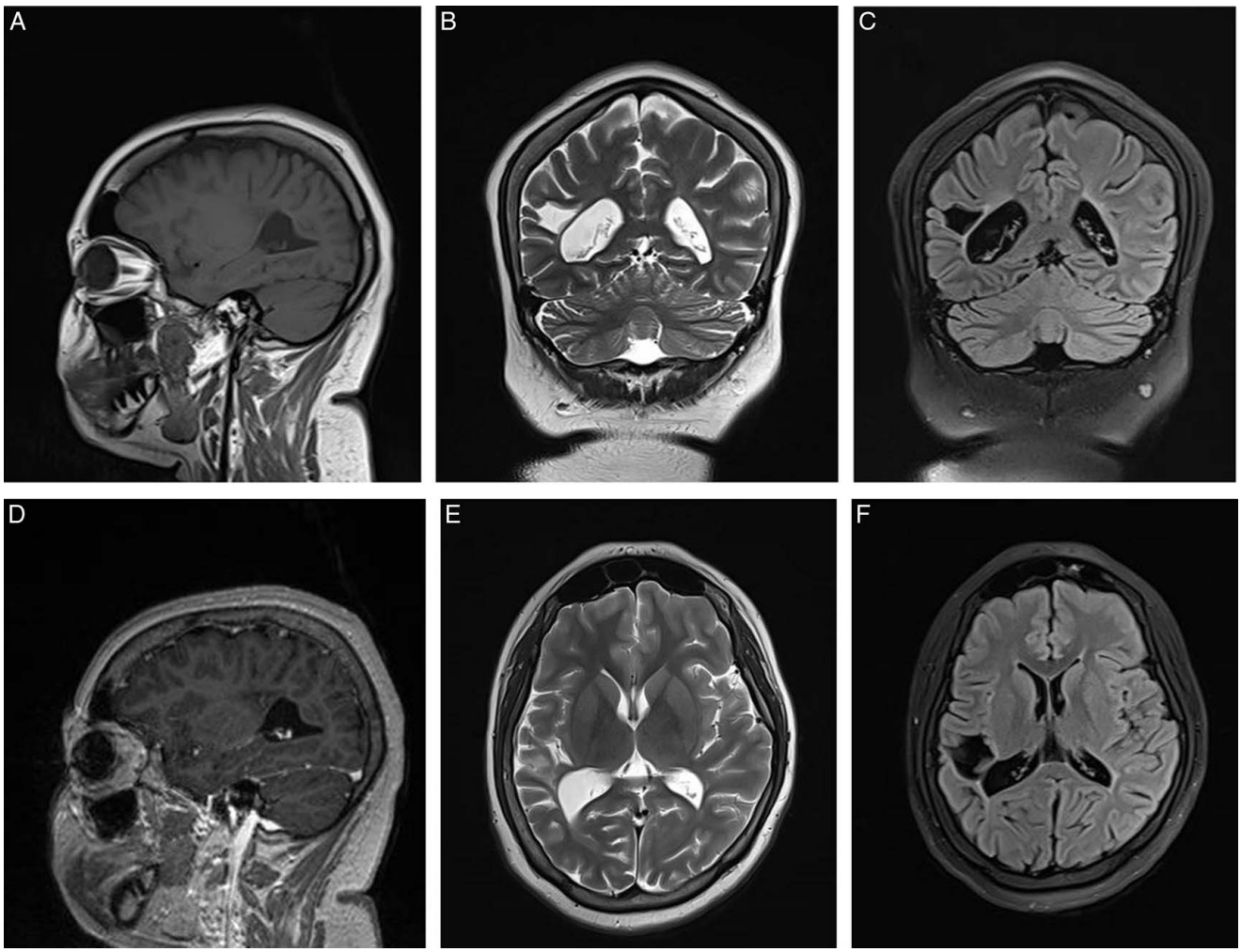

Figure 1. (A) 3 Tesla MRI T1 sagittal, (B) T2 coronal, (C) FLAIR coronal, (D) post-contrast sagittal, (E) T2 axial and (F) FLAIR axial show right temporo-parietal focal cystic structure communicating with the subarachnoid space in the right sylvian fissure following cerebrospinal fluid signal in all sequences with no post-contrast enhancement.

surrounding gliosis and scarring on MRI, coupled with no history of trauma in the patient. Focal cortical dysplasia was ruled out as there was no evidence of blurring of grey-white matter junction or abnormal gyral patterns. Arachnoid cyst was ruled out considering their typical location in the arachnoid membrane, often at the base of the brain. Neoplastic cysts were ruled out based on the absence of a solid component of the tumour and infectious cysts like neurocysticercosis were ruled out given the lack of scolex or enhancing mural nodule. Video electroencephalography (VEEG) monitoring showed right central and parietal (C4, P4) sharp waves during the interictal period and right central (C4) immediately followed by right parietal (P4) spike and wave, followed by rhythmic alpha activity evolving into a generalized seizure pattern during the ictal period.

The concordance between MRI findings, patient's semiology and VEEG in addition to seizures being poorly controlled with two ASM at optimal doses made patient an excellent candidate for epilepsy surgery. Alternatives included adding a new drug, however, literature indicates limited success rate of 3% for achieving seizure freedom with the addition of a third ASM after poor seizure control with two ASM at optimal doses[7]. In contrast, epilepsy surgery presents a significantly higher likelihood of seizure freedom with lesional epilepsy notably demonstrating higher rates of seizure freedom following surgical intervention[8].

Surgery and intraoperative electrocorticography

The patient underwent neuro-navigation guided right parieto-temporal craniotomy under general anaesthesia. The cyst and the surrounding brain were exposed, revealing a thin transparent membrane covering the cyst with the presence of intracystic bleeding. No abnormal cortical tissue was noted around the cyst. Anaesthesia was reduced, and ioECoG was performed around the margins of the cyst (anterior, posterior, medial, and lateral). ioECoG revealed nearly continuous moderate-amplitude polyspikes with paroxysmal fast activity involving the two gyri immediately posterior to the cyst, although it appeared normal in appearance. (Fig. 2). ioECoG performed further posterior did not reveal any epileptogenic activity. Based on these findings, ioECOG-guided gyrectomy of the posterior pericystic area was performed, along with cystotomy of the porencephalic cyst. Post-resection ioECoG showed no epileptogenic activity.

Postoperatively, the patient is seizure free, achieving ILAE IA seizure freedom at 1 year. Histopathology revealed areas of normal-looking brain tissue with mild congested vessels and focal haemorrhage.

Informed consent was obtained from the patient for publication of this case report which is reported in line with the SCARE 2023 criteria[9].

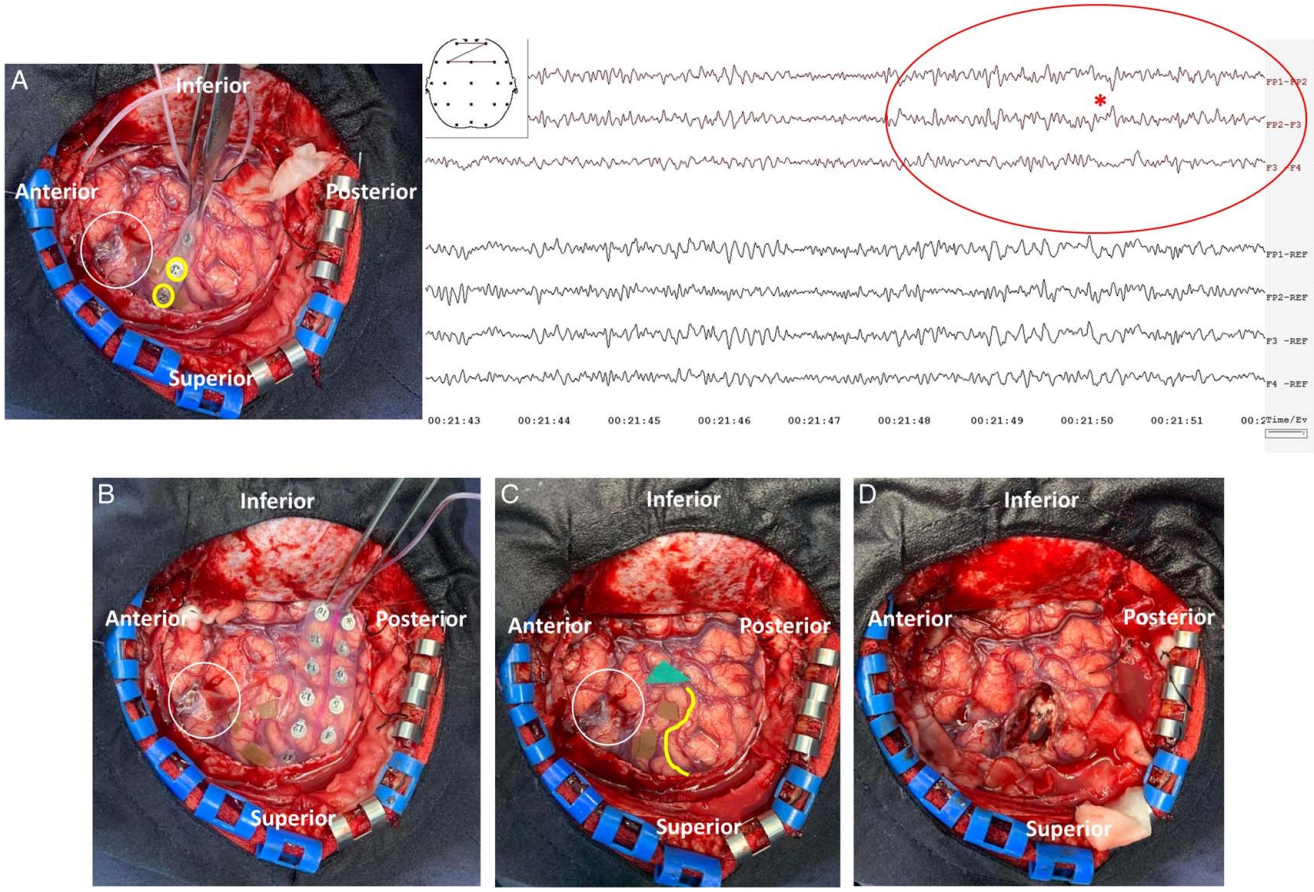

Figure 2. Intraoperative electrocorticography. (A) Epileptogenic activity (red circle and asterix) observed at the posterior part (yellow circles) of the porencephalic cyst (white circle). (B) No epileptogenic activity noted further posteriorly. (C) Brown marks indicate epileptogenic activity and green indicates no epileptogenic activity. Yellow line represents the posterior extent of resection based on intraoperative electrocorticography. (D). Post-resection.

Discussion

The findings of the noninvasive pre-surgical evaluation clearly identified the right centro-parietal region as the location of the epileptogenic activity. However, it was ioECoG that subsequently revealed the extent of the epileptogenic focus.

While the patient did present with a suggestive perinatal history, the nature and presentation of the porencephalic cyst was unusual compared to more commonly encountered cases[3]. The cyst was relatively small, and the patient did not exhibit any neurological deficits. The probable explanation is a highly focal ischaemia within a specific region of the MCA territory, giving rise to this presentation. In spite of the focal nature of the ischaemia, its impact and subsequent epileptogenesis were wide, involving two gyri posterior to the cyst, indicating the involvement of the arterial territory distal to the cyst.

It is not uncommon for the EZ to extend beyond the occluded arterial territory with areas of extensive damage[3,6]. However, in the majority of such cases, this phenomenon is observed in a subset of patients with large cysts and neurological deficits[3]. Our report highlights that even in cases involving small cysts without deficits, the impact of early ischaemic insult can be wide, extending beyond the pericystic margins, especially involving the region of distal arterial territory to the cyst. The extent of gliosis in such instances can remain unknown, emphasizing the necessity of ioECoG to precisely define the border between the normal and epileptogenic brain. The utilization of ioECoG to guide the resection of porencephalic cysts has demonstrated favourable surgical outcomes in cases with larger cysts and more extensive ischaemic damage[6]. Our finding suggests that even in the context of a small porencephalic cyst, the actual EZ may be larger. This stands in contrast to other causes of lesional epilepsy, where, in most cases, the EZ is predominantly found in the surrounding perilesional area[10].

Various surgical approaches are used for the treatment of porencephalic cysts, spanning from simple cyst fenestration to the implementation of stereotactic electroencephalography (SEEG) for precise localization of the EZ and subsequent resection[4,5]. The literature review in our paper on epilepsy surgery for porencephalic cysts indicates that the majority of these cysts are large in size, primarily due to the involvement of a significant arterial territory. Consequently, surgical interventions are often geared towards addressing significant lesions, ranging from functional hemispherotomy to multilobar resection (Table 1)[11,13]. Many patients also presented with pre-existing neurological deficits, such as hemiparesis[13]. Notably, cyst fenestration alone has demonstrated less satisfactory outcomes, frequently requiring re-operation[13]. Epilepsy surgery specifically tailored for small porencephalic cysts without neurological deficits is less frequently documented in the literature, likely due to their rare occurrence.

Several studies have utilized neuroimaging techniques, including magnetoencephalography (MEG) and single-photon emission computerized tomography (SPECT), to localize the EZ in patients with porencephalic cysts , particularly when the electro-clinical findings are inconclusive[11,12]. In addition to MEG and SPECT, SEEG and epileptogenicity index have been utilized

Table 1

Surgically treated case series of porencephalic cysts

Paper	No. patients	Age at surgery	Porencephalic cyst size	Neurological deficit	Localization of EZ	Surgical procedure	Seizure outcome (Engel's class)
Ichikawa *et al.*,[11]	11	4–23	Large in all patients	Present in all patients	Ictal SPECT, FDG-PET	7 hemispherotomy, 3 posterior disconnection 1 occipital disconnection	Engel I and Engel II
Marchi *et al.*,[4]	6	11–39	Large in 5, small in 1	5 patients	SEEG	3 no surgery 1 ATL 2 multilobar disconnection	Engel IA: 1 Engel II: 1 Engel III: 1
Bennet-back *et al.*,[12]	4	1.8–35	Large in all patients	Present in all patients	Intracranial EEG, MEG	2 ATL, 1 multilobar resection	Engel IA: 3 Engel II: 1
Ghatan *et al.*,[1]	9	6 months–6	Large in all patients	Present in all patients	Intracranial EEG	5 functional hemispherotomy 3 multilobar resection	Engel IA: 8 Engel IVA: 1
Guzzeta *et al.*,[3]	12	3 months–2.3		Present in all patients	—	2 cyst uncapping and hemispherectomy 10 Cyst uncapping	Engel I: 5 Engel II: 5 Engel III: 2
Iida *et al.*,[6]	8	10–25	Large in all patients	Present in all patients	ioECOG	8 cortical resection	Engel I: 6, Engel III: 2
Carreno *et al.*,[13]	25	—	Large in all patients	Present in all patients	Intracranial EEG	13 functional hemispherotomy 5 ATL 1 posterior disconnection 3 anterior corpus callosotomy 1 frontal lobectomy 1 mesial parietal resection 1 cyst drainage	Engel IA: 13 Engel IB: 2 Engel IIB: 4 Engel III: 1 Engel IV: 3
Koch *et al.*,[5]	37	3 months–18	Large in all patients	34 patients	—	37 Uncapping and fenestration of the cyst	Engel IA: 9 Engel IB: 14 Engel III: 9 Engel IV: 5

ATL, anterior temporal lobectomy; EZ, epileptogenic zone; FDG-PET, fluorodeoxyglucose positron emission tomography; ioECOG, intraoperative electrocorticography; MEG, magnetoencephalography; SEEG, stereotactic electroencephalography; SPECT, single-photon emission computerized tomography.

to delineate the EZ[4]. In situations where these resources are unavailable, the significance of ioECoG cannot be overstated.

We conclude that ioECOG of lesional epilepsy such as long term epilepsy associated tumours (LEAT), focal cortical dysplasia, neurocysticercosis etc. often have a perilesional epileptogenic zone organized around the lesion. As for small porencephalic cyst, which is a rare cause of lesional epilepsy, the epileptogenic zone was organized only posterior to the cyst, suggesting the involvement of arterial territory distal to the cyst. Our findings suggests that EZ organization in lesional epilepsy due to congential vascular insults differs from other well-known causes of lesional epilepsy and thus interpretation of ioECOG may need to take aetiology of lesional epilepsy into consideration.

Ethical approval

Ethical approval is exempted in case of case reports in our institution. Written informed consent has been taken from the patient and her parents.

Consent

Written informed consent was obtained from the patient and her parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

R.D.: writing the paper, study concept. P.G.: data collection, data analysis or interpretation. J.K.: data collection. S.M.: data collection. T.B.: Study design. P.R.: concept design. B.P.: concept design, data collection.

Conflicts of interest disclosure

The authors have no conflicts of interest directly relevant to the content of this article.

Research registration unique identifying number (UIN)

None.

Guarantor

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Data availability statement

The dissemination of the article data is freely accessed.

Provenance and peer review

The paper was not invited.

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