

The ventral precuneal-posterior cingulate region as a site of epileptogenicity

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Introduction

- Ventral precuneus-posterior cingulate (VP-PC) region is anatomically sheltered
 - Difficult to study by noninvasive means
- VP-PC distant connections
 - Dorsal stream: ventromedial prefrontal cortex
 - Ventral stream: parahippocampal gyrus
- VP-PC functions
 - Default Mode Network

Introduction

- Epilepsy arising from the VP-PC region has variable semiology
 - Connectivity provides multiple routes of ictal spread
 - Semiology may be characteristic of spreading pattern
- VP-PC sites of epileptogenicity can masquerade as other, well-characterized forms of epilepsy
 - Mesial temporal lobe

Case 1: 14yo M w/ epilepsy since 6mo age

- Semiology

- Epigastric discomfort w/o rising or experiential sensations
- Behavioral arrest with staring and rightward versive eye motion
- Intentional walking in a particular direction
- Tonically flexed posture at waist with extension of right arm backward w/ upturned palm
- Inarticulate or nonsensical vocalization
- Postictal word finding difficulty and fatigue

- Medical management

- Lamotrigine, carbamazepine, topiramate, perampanel, and clonazepam

Case 1: 14yo M w/ epilepsy since 6mo age

- Imaging / Electrographic studies
 - MRI: no overt pathology
 - Phase 1 scalp EEG: parasagittal / centroparietal activity at ictal onset
 - MEG: unremarkable
 - fMRI: left hemispheric speech dominance
 - Phase 2 surface EEG: VP-PC and posterior parahippocampal gyrus sites of epileptogenicity (figure 1)
- Other studies
 - Neuropsychological: FSIQ -19pts; VCI -9pts (between ages 10 and 14)

Case 1: 14yo M w/ epilepsy since 6mo age

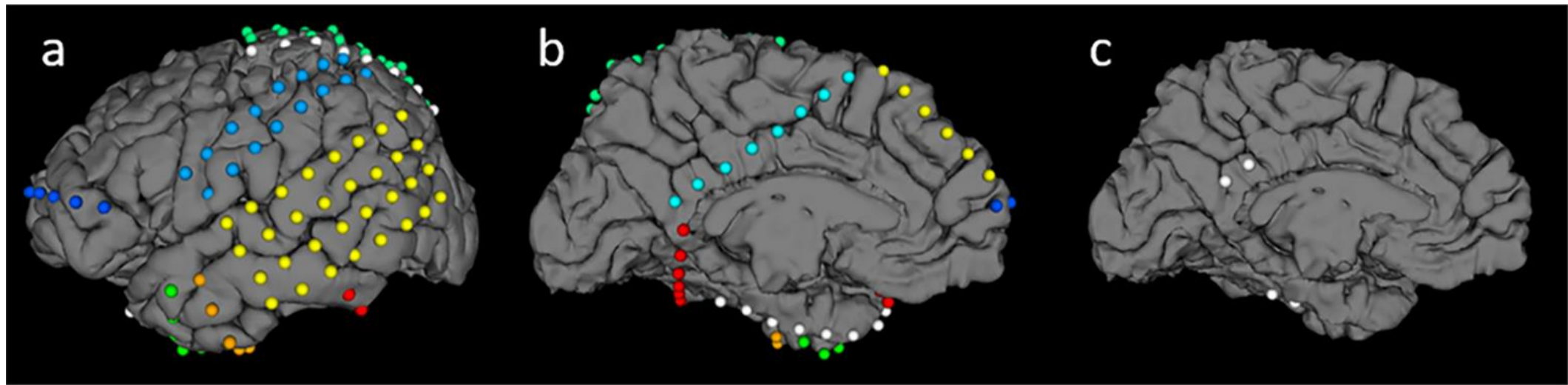


Figure 1. Case 1: phase-2 surface electrodes.

Case 1: 14yo M w/ epilepsy since 6mo age

- Management
 - Responsive Neurostimulation (RNS) implantation targeting sites of epileptogenicity
 - Discharge medications: lamotrigine, carbamazepine, topiramate, levetiracetam
- Outcome
 - Clinically seizure-free x 5yrs
 - RNS recordings continue to show interictal spike activity
 - Medication tapered: lamotrigine, brivaracetam
 - Improvements in FSIQ, visual spatial index, working memory index
 - Employed as mechanic

Case 2: 21yo F w/ 1-year history of focal unaware seizures

- Semiology

- Aura: nausea, malaise
- Tendency to walk with intent
- Mumbling, spitting
- Rubbing, pinching nose w/ R hand
- Postictal fatigue

- Medical management

- Lamotrigine, oxcarbazepine, levetiracetam, clonazepam

Case 2: 21yo F w/ 1-year history of focal unaware seizures

- Imaging / Electrographic studies
 - Phase 1 scalp EEG: R anterior/mid temporal ictal onset
 - MRI: no lateralizing or localizing features
 - PET: no lateralizing or localizing features
 - Phase 2 surface EEG: appeared to confirm R mesial temporal epilepsy (figure 2)
 - Note: R mesial frontoparietal cerebral surface not sampled
- Other studies
 - Neuropsychological: no significant discrepancy between VCI and PRI, average scores

Case 2: 21yo F w/ 1-year history of focal unaware seizures



Figure 2: Case-2 phase-2 surface recordings revealed R mesial temporal spike activity

Case 2: 21yo F w/ 1-year history of focal unaware seizures

- Initial management
 - R mesial temporal resection
- Initial outcome
 - Three generalized seizures at 7mo
 - Subsequent return to original seizure pattern w/ decreased intensity
- Further studies
 - MRI: slowly evolving enhancing lesion in R posterior VP-PC region (figure 3a-c)
 - Intraoperative depth electrocorticography: interictal discharges arising from lesion (figure 3d)
 - Lesion pathology: pilocytic astrocytoma w/ adjacent cortical dysplasia

Case 2: 21yo F w/ 1-year history of focal unaware seizures

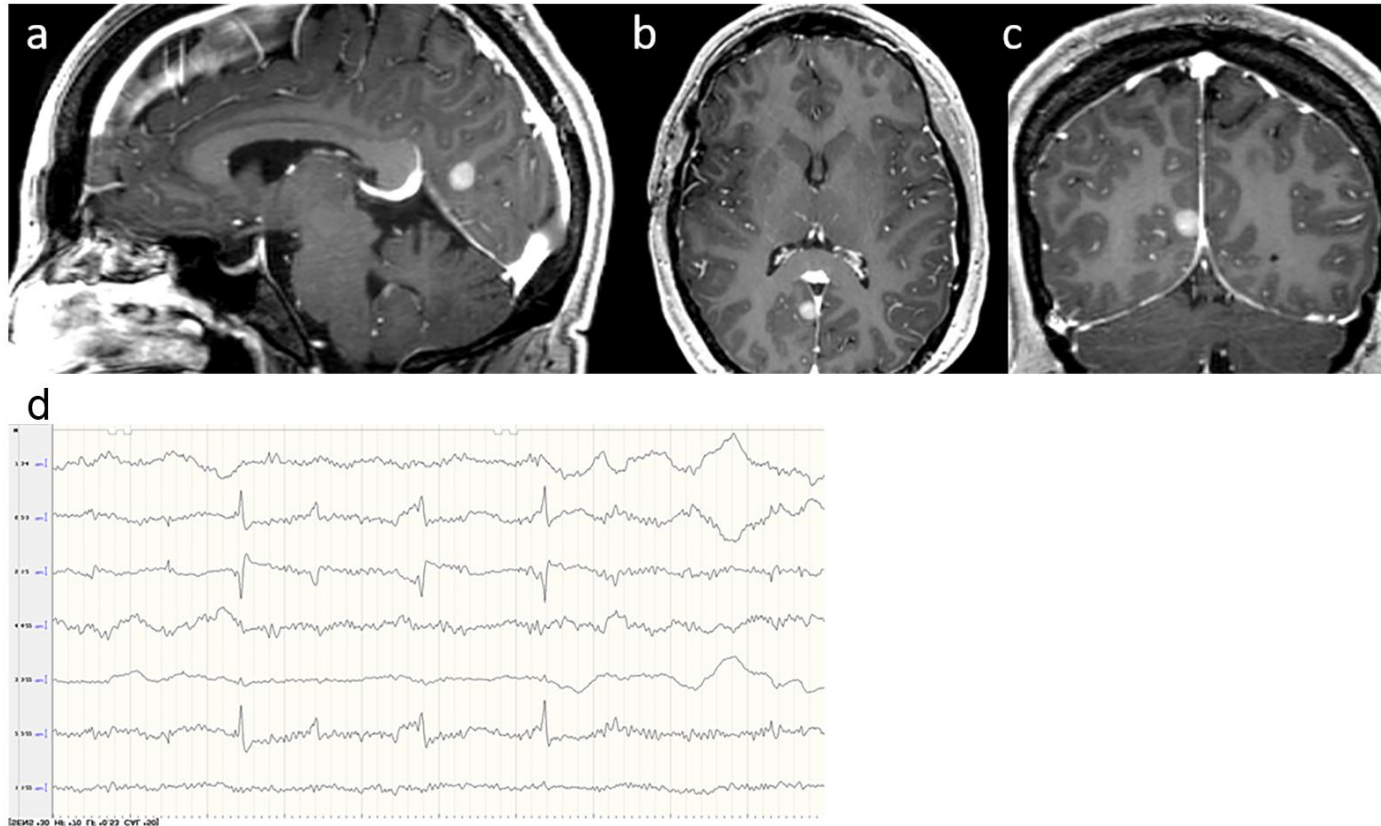


Figure 3. Case 2: post-resection MRI (a, b, c) showing R posterior VP-PC lesion and subsequent intraoperative depth electrocorticography (d) with interictal discharges

Case 2: 21yo F w/ 1-year history of focal unaware seizures

- Further management
 - VP-PC lesion ablated via stereotactically-guided laser interstitial thermal therapy (LITT)
- Outcome
 - Clinically seizure-free x 3yrs
 - Medication tapered: clobazam
 - Employed part-time
 - Subsequent analysis of tractography showed projections from VP-PC lesion to region of temporal lobe initially thought to be the site of epileptogenicity (figure 4)

Case 2: 21yo F w/ 1-year history of focal unaware seizures

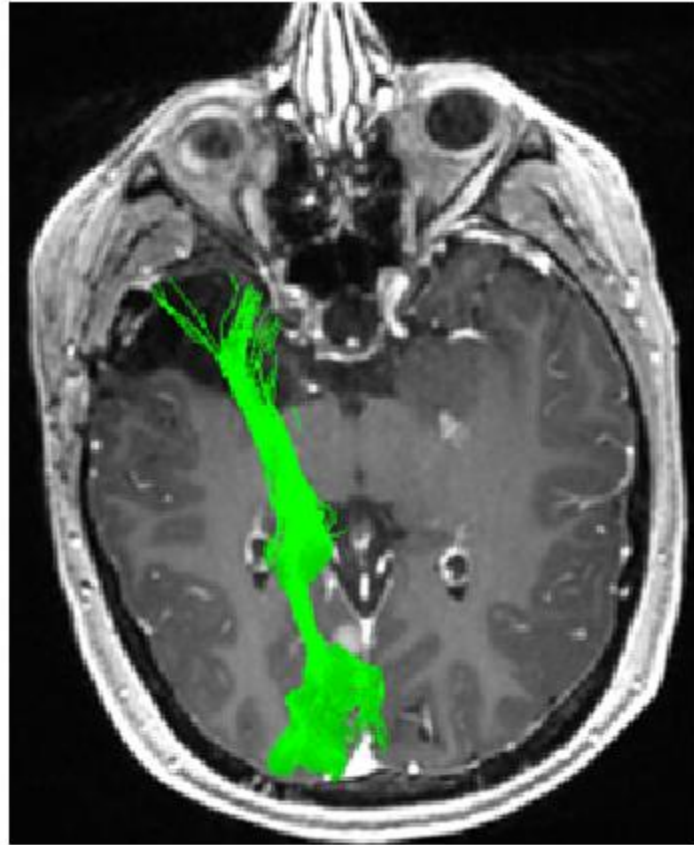


Figure 4. Case 2: tractography demonstrating projections from VP-PC lesion to ipsilateral temporal lobe.

Key Points

1. The ventral precuneal-posterior cingulate area (VP-PC) represents a salient hub within the default mode network with extensive connectivity along two primary streams – dorsal and ventral – targeting both frontal and temporal lobes, respectively.
2. Ictal expression may manifest both intrinsic features characteristic of local attribution or instigate remotely-activated semiologies suggestive of distant spread.

Key Points

3. Of particular concern, may be a propensity for expressing a mesial temporal epileptogenicity which may be difficult to discern from the more common primary condition.

4. The deep location of the VP-PC within the mesial ventral parietal surface, in the absence of an overt lesion, makes the diagnosis problematic. Reliance on clinical acumen to reconcile the variability in ictal expression with such a location and quantitative neuroimaging measures that may discount a primary temporal lobe epilepsy must guide appropriate decision-making.