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Extraoccipital photoparoxysmal response in a case of focal encephalitis

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ABSTRACT – Photoparoxysmal response (PPR) is commonly associated with idiopathic generalised epilepsies. Most of the focal events induced by intermittent photic stimulation (IPS) are reported to be of occipital origin. Only six temporal lobe epilepsy patients have been reported in the literature with focal PPR at extraoccipital sites. We report a four-year-old girl with possible encephalitis who presented initially with epilepsia partialis continua of limbs on the right side. Interictally, she had left centro-parietal periodic lateralized epileptiform discharges (PLEDs). She responded to medical treatment and was free of seizures and motor and cognitive deficits at six months follow-up. Repeat EEG at follow-up showed left centro-parietal spikes accentuated by IPS. This is the first report of an extraoccipital, extratemporal focus showing PPR. The possible mechanism of PPR from this fronto-parietal epileptogenic focus is discussed.

Key words: Focal photoparoxysmal response, intermittent photic stimulation, focal encephalitis, extraoccipital

Photoparoxysmal response (PPR) is characterized by the occurrence of spikes or spike-and-wave discharges on the electroencephalogram (EEG) in response to intermittent photic stimulation (IPS) and has a strong genetic component. Visual sensitivity (VS), on the other hand, is defined as the susceptibility to experience seizures triggered by visual stimuli (Kasteleijn-Nolst Trenité *et al*., 2001). PPR is a common feature of idiopathic generalised epilepsies (IGE) and may be observed in 40-90% of IGE patients (Lu *et al*., 2008). There are several reported cases of focal seizures of occipital origin induced by IPS (Ricci and Vigevano, 1993). Interestingly, only five cases of VS and one of PPR of extraoccipital origin, all originating from the mesial temporal lobe, have been reported so far (Benbadis *et al*., 1996; Fiore *et al*., 2003; Isnard *et al*., 1998; Seddigh *et al*., 1999; Thomas and Zifkin, 1998). We report an unusual case of focal epilepsy due to focal encephalitis with left centro-parietal interictal discharges activated during PPR.

Case report

A four-year-old female child, with normal birth and development and

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development and no family history of epilepsy, presented with a one-month history of recurrent right focal motor seizures associated with postictal dysphasia and right-sided weakness lasting one to two hours. Three weeks into the illness, she developed epilepsia partialis continua (EPC) and persistent weakness involving right-sided limbs. There were no occipital symptoms and visual fields were normal. MRI of the brain revealed left caudate nucleus T2 and FLAIR hyper intensity without diffusion restriction or contrast enhancement *(figure 1A)*. Cerebrospinal fluid (CSF) showed 30 cells (98% lymphocytes) with normal sugar and protein. CSF was sterile and DNA PCR for HSV 1 and 2 was negative.

EEG showed left centro-parieto-temporal slowing *(figure 1B)* and left parietal periodic lateralized epileptiform discharges (PLEDs) *(figure 1C)*. Focal encephalitis of probable viral or autoimmune aetiology was considered. She was treated with intravenous (i.v.) acyclovir followed by i.v. methylprednisolone. Her EPC and right hemiparesis gradually improved over two weeks. On discharge, she was seizure-free. On review, after six months, she remained seizure-free. There was no cognitive or motor deficit. Repeat EEG showed spikes over the left centro-parietal region along with slowing over the same region *(figure 1D)*. IPS was performed with a stroboscope placed 30 cm from the nasion for photic frequencies ranging from 1-40 Hz. The flash stimulus was given for 10 seconds for all photic frequencies (five seconds with eyes open and five seconds with eyes closed). We observed consistent appearance of rhythmic bursts of spikes lasting for 1-1.5 seconds mostly when eyes were closed at a frequency ranging from 6-35 Hz *(figure 1E-G)*. There was variable delay ranging from 0.3-6.5 seconds from the onset of photic drive to the appearance of the

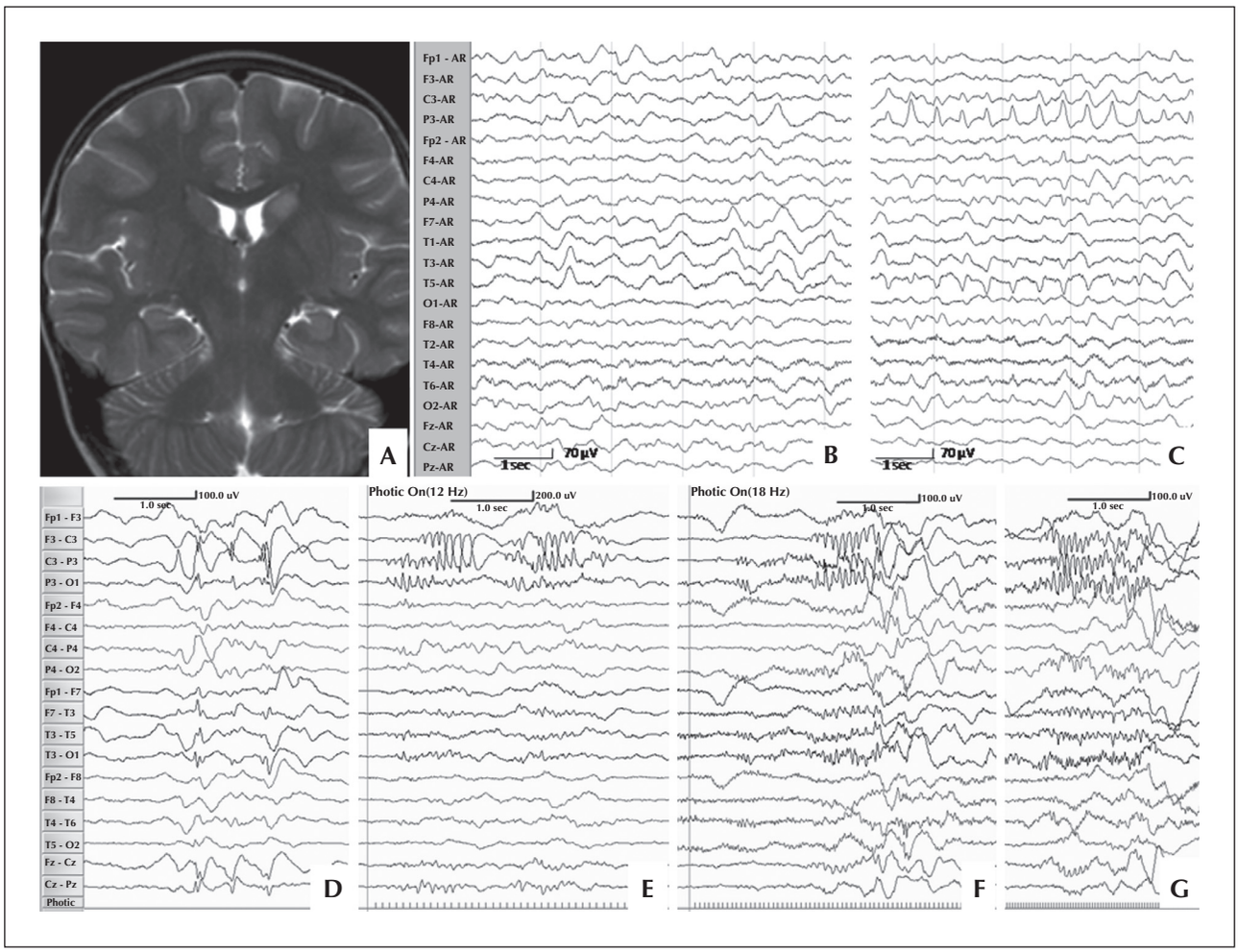

Figure 1. A) Coronal T2 MRI showing left caudate hyperintensity during acute phase of the illness.
B) Initial EEG showing left parieto-temporal slowing (**B**) and (**C**) left parietal PLEDs.
D-G) EEG at six months follow-up showing (**D**) left centro-parietal spike-and-wave discharges and focal photoparoxysmal response at the left centro-parietal region with 12 Hz (**E**), 18 Hz (**F**) and 30 Hz (**G**) photic frequencies. See text for further details.

spikes. The initial response was noted over the left parietal region *(figure 1E-G)* followed by rapid spread (400-600 ms) to the central region. This response was self-limiting and did not precipitate any seizures.

Discussion

Until 1996, all the case reports of photic-induced focal seizures were of occipital origin resulting in the common belief that photosensitive focal seizures originate only from occipital region (Ricci and Vigevano, 1993). It is pertinent to note that the atypical features observed in some of these seizures were explained by the clinical variability of ictal symptoms of occipital seizures. Thomas and Zifkin (1998) and Seddigh *et al*. (1999) reported complex partial seizures provoked by photic stimulation in patients with temporal lobe epilepsy (TLE) and believed that repetitive stimulation of the occipital cortex resulted in seizures due to infrasylvian spread to the hyper-excitable temporal lobe.

Meanwhile, more and more reports supporting the photosensitivity of the temporal lobe, independent of the occipital lobe, were published. Benbadis *et al*. (1996), using subdural recordings, and Isnard *et al*. (1998), using stereo EEG, demonstrated the photosensitivity of mesio-temporal seizures in the absence of occipital cortex involvement. Fiore *et al*. (2003) demonstrated temporal hyperperfusion in ictal SPECT performed during IPS in a case of photosensitive TLE. Our case would therefore appear to be the seventh report of an extraoccipital focus, and the first report of an extraoccipital focus outside the temporal lobe, demonstrating PPR.

Our patient was probably suffering from focal encephalitis, either viral or autoimmune. Despite making a good recovery, she now requires further follow-up before any conclusion can be drawn regarding the nature of the current illness as well as the possibility of coexisting IGE syndrome. The electroclinical data consisting of right EPC and EEG spikes and slowing in the left centro-parietal region favoured the left posterior frontal and the adjacent parietal cortex as the epileptogenic focus. There was radiological evidence supporting involvement of subcortical structures in the form of left caudate hyperintensity. There was no electrical, clinical or radiological evidence for occipital involvement at initial presentation or at follow-up.

Whether the photosensitivity of the fronto-parietal focus in our case is independent of the occipital cortex is uncertain. One possibility is that the expression of epileptiform abnormality over the centro-parietal region is dependent on the occipital cortex. Parietal to central spread of the focal PPR suggests posterior to anterior propagation and would favour this

Table 1. Characteristics of the reported cases of extraoccipital epilepsy with focal photoparoxysmal response.

Investigator	Age of onset (in years)	Semiology	Neuroimaging	Interictal EEG	Visual phenomenon demonstrated	Proposed hypothesis in relation to occipital lobe
Benbadis *et al*. (1996)	21	Temporal	Normal	Bitemporal spikes	VS	Independent
Isnard *et al*. (1998)	20	Temporal	Normal	Right temporal discharges	VS	Independent
Thomas and Zifkin (1998)	7	Temporal	Right mesial temporal sclerosis	Normal	VS	Dependent
Seddigh *et al*. (1999)	11	Temporal	Normal	Spikes with phase reversal over F8	VS	Dependent
	17	Temporal	Normal	Spikes with phase reversal over T6	VS	Dependent
Fiore *et al*. (2003)	13	Temporal	Right hippocampal sclerosis	Right temporal sharp waves	PPR	Independent

VS: visual sensitivity; PPR: photoparoxysmal response.

hypothesis. However, the widely variable time lag between the onset of photic drive and focal PPR would either suggest a differential propagation from the occipital cortex to the left centro-parietal region or the presence of an independently functioning extraoccipital focus. We have evidence from recent functional MRI (fMRI) studies supporting the possibility of an independent focus. In one EEG-fMRI study, PPR-associated activation was identified in the parietal cortex adjacent to the intraparietal sulcus in five and in the frontal eye field region of the premotor cortex in all six subjects in whom PPR was observed (Moeller *et al*., 2009). The frontal eye field and the intra-parietal cortex are involved in the generation of saccades. Surprisingly, a unilalateral predominance of the fronto-parietal activation was seen in five subjects thereby favouring a unilateral generation of PPR in either the left or right hemisphere. Another fMRI study demonstrated BOLD signal decreases in perirolandic areas during IPS (Hill *et al*., 1999) and a TMS study showed altered excitability of the motor cortex during IPS when compared to controls (Siniatchkin *et al*., 2007).

Overall, six cases of temporal lobe epilepsies are available in the literature *(table 1)* which demonstrate the presence of focal PPR in extraoccipital epilepsies. Three (50%) of these cases have been suggested to occur independently of the occipital lobe based on invasive recordings or functional imaging. For want of such investigations, we are unable to throw light on the dependence or otherwise of the centroparietal focus on the occipital lobe in our case. However, these cases emphasise the need for further studies using invasive monitoring with intracranial electrodes and functional neuroimaging to better understand the pathophysiology of photoparoxysmal responses. □

Disclosure.
None of the authors has any conflict of interest or financial support to disclose.

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