POSTERIOR CORTEX SEIZURES – PEDIATRIC CHALLENGES

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SUMMARY – Posterior cortex seizures have a complex semiologic presentation that is especially challenging in the pediatric population. Therefore, using clinical presentation in localizing ictal involvement is not sufficient in children, thus making this type of epilepsy quite under-recognized. As most of the ictal symptoms are subjective and could well be overshadowed by symptoms arising from adjacent cortices, primarily temporal and central ones, it is necessary not to overlook this large source of pharmacoresistant epilepsies. The parietal lobe as part of an extensive synaptic network is a great imitator, thus quite often producing inaccurate localization readings on scalp electroencephalography (EEG) due to very scattered interictal discharges and uninformative ictal recordings. Using direct cortical recordings in delineating the epileptogenic zone is helpful in some cases but even highly experienced epileptologists may erroneously interpret some features as arising from other localizations, especially the frontal lobe. Epilepsy surgery from the posterior quadrant is still quite rare and relatively unsuccessful, especially in non-lesional epilepsies due to elaborate mechanisms of connectivity, misleading semiology, and non-localizing EEG recordings, possibly due to insufficiency of parietal cortex synchronicity. Applying the aforementioned to the pediatric age makes it perhaps the most difficult challenge for a pediatric epileptologist.

Key words: Posterior cortex epilepsy; Semiology; Pediatric epilepsy; Epilepsy surgery; Connectivity

Introduction

Posterior cortex seizures primarily originate from the parietal and occipital lobes, as well as from the posterior part of the temporal lobe1. They account for 5%-10% of all focal seizures, but their frequency might be underestimated especially in children due to diagnostic difficulties2. The challenge of identifying posterior cortex epilepsy is even greater as the seizure semiology is often a result of very fast ictal propagation and peculiar connectivity of posterior cortex that gives rise to the highly variable ictal patterns such as focal tonic or clonic seizure activity and seizures with automatisms that might mimic frontal or temporal lobe semiology. Polymorphism of ictal manifestations in addition to the inability of children to accurately describe auras and ictal sensations accounts for the potential misdiagnosis of posterior cortex seizures and potentially gives rise to the increasing number of diagnosed psychogenic non-epileptic events. As one of the rarest localizations of epilepsy surgery, the posterior cortex has been examined less extensively in terms of anatomo-electro-clinical features, thus potentially leaving many inadequately treated pharmaco-resistant children3,4. For this reason, profound understanding of epileptic networks, semiology propagation and treatment options is especially crucial in this group of patients.

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Classification of Posterior Cortex Epilepsies

In addition to the conventional classification of epilepsies defined by the International League Against Epilepsy, posterior cortex epilepsies could be classified by the anatomic structures involved, especially for surgical series. This type of classification has a more outcome oriented view as the key principle is to determine the percentage of epilepsies from a certain lobe, or beyond in reference to the postsurgical seizure freedom. In pediatric patients, the most frequently performed surgeries are multilobar resections. A more difficult way of classifying posterior cortex epilepsies is based on identifying the propagation pathways by using invasive procedures such as intracranial depth electrodes. This classification aims to improve our understanding of ictal phenomena observed in posterior cortex epilepsies by differentiating ventral propagation to the temporal/mesotemporal structures and dorsal propagation to the frontal and parietal structures. Even though some semiologic features are unspecific, more occipital semiology is related to dorsal propagation, whereas ventral propagation yields more temporal semiology. Therefore, the presence of oroalimentary automatisms typical for children with posterior cortex seizures could mostly be linked to the secondary involvement of mesial temporal lobe structures, probably via the multiple anatomical connections existing among the parietal lobe and the hippocampal formation. Bipedal automatisms and some other automatic motor movements seen more frequently in adult patients might point to involvement of the frontoparietal system by disinhibition of innate activities controlled by the central pattern generators. Overall, the need for different classifications of posterior cortex epilepsies due to the less defined boundaries and peculiar seizure propagation underlines the complexity of this entity.

Auras

As the first ictal symptoms, auras could be a very valuable piece of information in localizing and lateralizing the lobe of seizure origin. Recognizing, describing, and recalling auras is a challenge in itself, needless to say how difficult it may be for a child, or even impossible for a non-verbal child. The polymorphic character of posterior cortex auras emphasizes diagnostic uncertainty. Somatosensory auras are highly suggestive of parietal seizures, even though they could be found in other posterior cortex epilepsies, but could also be elicited with stimulation of the supplementary motor area (SMA). Somatosensory auras normally include tingling, numbness, electrical shock-like feelings, thermal sensations, and pain. Somatosensory illusions such as movement of body parts and sensations of altered body size are most likely elicited from the non-dominant inferior parietal lobe or temporo-parieto-occipital junction. Visual auras could be simple such as flashing lights, or moving colors arising from the primary visual cortex, as opposed to more complex ones arising from the basal temporal cortex or temporo-occipital junction involving scenes and objects. Perhaps most widely reported visual auras are blurry vision and visual motions from the mesial parieto-occipital region. Like complex visual auras, complex auditory auras are produced by activation of auditory association areas in the temporo-occipital cortex. Both visual and auditory auras are often associated with vertiginous auras such as sensation of rotation or falling arising from the parietal operculum and temporo-parietal area.

The well-established role of the parietal cortex in the processing of polysensory information could be related to unfavorable outcome of seizures related to polymorphic auras. However, the ability to recall different details of auras may imply that the initial discharge remains within the parietal lobe, without spreading to deeper structures or to the contralateral side, which might cause loss of awareness and amnesia for the auras. As expected, not many children report an aura, while parents observe behavioral arrest in more than half of the cases. Using the aura could somewhat focus the search for epileptogenic zone in cases where the patient is aware of the aura and old enough to explain it properly. However, using solely the aura and the semiology could never be specific enough to suggest the posterior cortex seizure without further investigations.

Seizure Semiology in Posterior Cortex Epilepsies

Posterior cortex epilepsy published series are scarce, especially in children. Perhaps the most common notion of posterior cortex epilepsies is the fact that they mimic epilepsies from other brain regions.
ing the large volume of the posterior cortex lacking clear anatomic boundaries, as well as its peculiar functional connectivity via fast propagation pathways, it is no wonder that focal seizures arising from this area will be of quite variable semiology. Most of the posterior cortex seizures are short, up to three minutes, and nearly half of them start in sleep. Seizures from the parietal lobe usually start in young children around the age of 6 years, and in multilobar involvement even younger, between 1 and 2 years. Seizure frequency is usually quite high (even daily), and the occurrence of status epilepticus is not uncommon.

As per semiologic presentations seen in posterior cortex epilepsies, the presence of manual and oral automatisms with loss of consciousness indicates a more temporal involvement, whereas tonic posturing with hypermotor behavior indicates frontal lobe involvement. In the very young patients, posterior cortex epilepsies might even present as epileptic spasms which are pharmacoresistant and may require surgery. Vestibular symptoms, vertiginous sensations, and visual illusions might be the most common symptoms reported in seizures of the posterior cortex. The superior parietal cortex, especially the temporal-parietal junction and the intraparietal sulcus can produce vestibular symptoms, whereas visual illusions such as object motion and blurred vision might be most likely evoked by stimulation of the basal temporal-occipital, mesial parietal-occipital, or temporal-parietal-occipital junctional region. The ambiguity of certain posterior cortex symptoms, especially when not coupled with secondary generalization or clear-cut epileptic signs, underscores the misdiagnosis of posterior cortex seizures with non-epileptic events, especially in pediatric population where the incidence of parietal lobe epilepsy is very high.

Motor manifestations are mostly myoclonic (especially with the head nod) as opposed to the adult patients, tonic seizures (elevation of arms) indicating the spread to the SMA, and epileptic spasms, while the rest are non-motor seizures (psychomotor and hypomotor seizures) indicating a possible spread to the temporal lobe. Oculomotor features related to posterior cortex seizures are nystagmus, eye deviation, lid myoclonia, and repetitive blinking. Ictally, these seizures might also generate vocalization, laughing, smiling, or flushing. Some of these manifestations could only be seen by very careful video-electroencephalog-
and their transient character might not be sufficient for a child to report with confidence. Localizing the hemifield with visual symptoms might point to involvement of the contralateral lobe, and recording epileptic nystagmus, or eyelid flutter could aid in confirmation of the occipital lobe involvement. However, pure eye deviation (regardless of head deviation) should be considered as related to the parietal eye field, located in the lateral intraparietal subregion, but also to the presence of a second eye field in the medial parietal area.

**Electroencephalography in Posterior Cortex Seizures**

Unfortunately, like seizure semiology, encephalography in posterior cortex seizures is also far from clear and uniform. Most of the studies have shown multifocal, bilateral, or sometimes falsely localizing activity. Furthermore, studies have not even found the prognostic relevance of one or another type of EEG findings. A large diagnostic problem poses uninformative EEG, as there is often a tendency of multiple EEG spread patterns and non-localizing scalp EEG findings. In fact, the presence of large association areas in the parietal lobe and its elaborate connectivity to various distant regions reasonably accounts for these EEG features, and probably accounts for a relatively higher incidence of invasive procedures in parietal lobe epilepsy compared with other focal epilepsies. Certainly, the recommendation is to use the invasive stereo-EEG method in cases where there is justified suspicion of posterior cortex epilepsy and indication for surgical treatment.

**Conclusion**

Childhood and adulthood posterior cortex seizures share many common clinical features but there are important distinct age-dependent differences. As a significant number of posterior cortex epilepsies occur among children with a high number of seizures and the likelihood to develop pharmacoresistant epileptic spasms, it is of utmost importance to recognize them early in the course of the disease. This phenomenon could be related to the early development and maturation of the posterior cortex but also to the presence of focal cortical dysplasia, which may delay maturation and distort the widespread networks early on. A distinct pediatric problem is the immaturity of patients and their inability (sometimes related to associated disabilities) to report auras, which may be the only symptom of a seizure arising from the posterior cortex. Poor localization of the EEG coupled with very short and frequent seizures, the lack of aura description, and heterogeneous semiology that reflects secondary spread rather than primary seizure onset makes this group of epilepsies very challenging. Detailed description of auras (by detailed history-taking) followed by video analysis of the chronology of ictal semiology and electrophysiologic features (interictal and ictal scalp EEG) together with neuroimaging (high-resolution magnetic resonance imaging) should aid in creating a complete electroclinical pattern to provide topographic orientation for posterior cortex epilepsy. In cases where these data should fail to discover the epileptogenic zone, a stereo-EEG evaluation with stereotactically placed intracerebral electrodes arranged according to a pre-defined localization could explore the areas suspected in seizure onset and early propagation towards the most effective rate of surgical success of posterior cortex epilepsy, especially in pediatric population. In addition, neurological and neuropsychological evaluation with standardized tests applied according to the patient age should complete the work-up in these challenging epilepsies. Identifying posterior cortex epilepsies by understanding the whole network upon completing the extensive diagnostic investigations should provide the best possible seizure and functional outcome.

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