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Case Study

A Rare Case of Adult-Onset Focal Epileptic Spasms Due to Frontal Astrocytoma

Ravish R Keni*, Sreekanta Swamy, M Sowmya, Dhananjaya I Bhat, Ravindra B Kamble, Vivek Rayadurg, M Manjunath

Department of Neurosciences, Aster RV Hospital, Bengaluru, India

***Corresponding Author:** Ravish R Keni, Department of Neurosciences, Aster RV Hospital, Bengaluru, India.

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Abstract

Epileptic spasms (ES) usually present between two weeks and eighteen months of age. The occurrence of ES in adults is rare and usually comprises of patients in whom the seizures started during infancy and persisted into adulthood. We report a rare case onset focal ES as the isolated seizure type in a 30-year-old adult due to frontal astrocytoma. We highlight the electroclinical differences between ES in adults versus infants; and review the factors determining the age of onset of ES.

Keywords: Epileptic Spasms; Focal; Adult-Onset

Introduction

Epileptic spasms (ES) are seizures characterised by brief involuntary muscular contractions, with majority of cases presenting between two weeks and eighteen months of age [1]. They are the prototype seizures in West syndrome which is a triad of IS, hypsarrhythmia in EEG and developmental regression [1]. They can also be seen in other early-onset epileptic encephalopathies such as Ohtahara syndrome and less frequently in elder children with Lennox-Gastaut syndrome [1]. ES may not always indicate generalized epilepsy [2], since they can rarely be the only seizure type in surgically remediable focal and hemispheric epilepsies [3,4] or be associated with other types of focal seizures [5,6].

The occurrence of ES in adults is rare. In previous case reports on ES in adults, the seizures started infancy and persisted into adulthood [7,8]. These patients had ES occurring along with other seizure types. To our knowledge, we report the first case of new onset focal ES as the isolated seizure type in a 30-year-old adult patient. We discuss the electroclinical differences between ES in adults versus infants; and review the factors determining the age of onset of ES. 2.

Case Study

A 30-year-old male, with normal birth and developmental history, presented with recent onset seizures from 1 week. The semiology comprised of intermittent clusters of clonic jerks involving the right hand with retained awareness. He underwent Video EEG for syndromic daignosis. Mutiple seizures characterised by jerking of fingers of right hand, lasting for 270- 300 msec were recorded (Supplementary Video 1). The ictal EEG showed diffuse (predominant left frontal) burst of slow waves measuring 60-80 microvolts followed by electrodecrement suggestive of epileptic spasms (Figure 1). The inter-ictal EEG showed well-formed background intermixed with focal intermittent slowing over the left frontotemporal region (Figure 2).

Video: Ictal video showing brief repetitive focal clonic jerks involving the fingers of right hand lasting for 270-300msecs each.

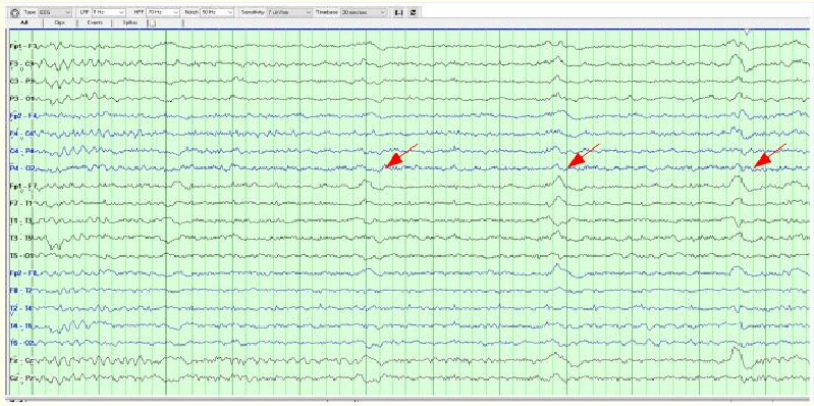

Figure 1: Ictal EEG showing diffuse (predominant left frontal) burst of slow waves (red arrow) measuring 60-80 microvolts followed by electrodecrement.

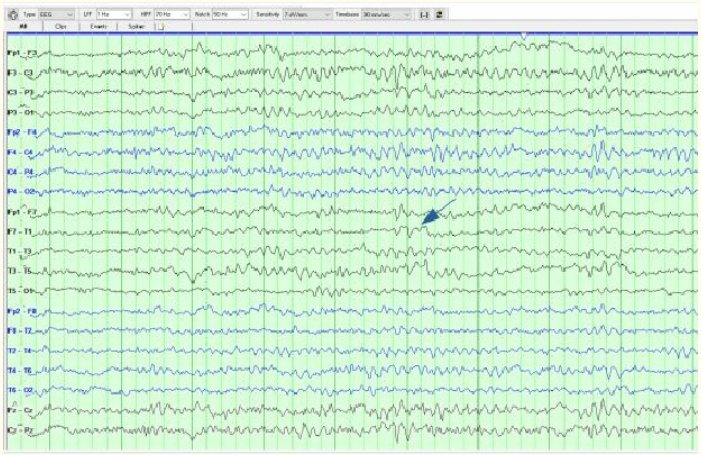

Figure 2: Interictal EEG showing well-formed background intermixed with focal slowing over the left frontotemporal region (blue arrow).

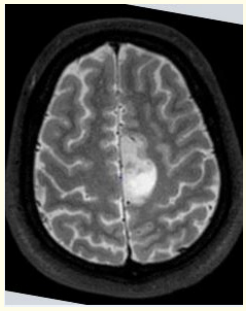

Figure 3: MRI brain showing well-defined irregular T2 hyperintense lesion seen in the left parafalcine frontal lobe measuring approximately 36 x 53 x 20 mm.

His MRI main showed well-defined irregular T2 hyperintense lesion seen in the left parafalcine frontal lobe (Figure 3). He underwent stereotactic biopsy and histopathology showed anaplastic astrocytoma (WHO Grade-III). He was started on Valproate and Zonisamide after which the seizures were well controlled and received chemoradiotherapy for astrocytoma.

Discussion

Maturational changes in the brain with ageing brings about a change in seizure semiology and electrophysiology. The prototype ES in infants are characterised clinically by brief jerking spells of flexion, extension, or combination of flexion-extension of the head, neck, arms, legs, and trunk lasting for two seconds or less and electrophysiologically, have a variety of ictal patterns such as: generalized slow wave, attenuation and increased fast activity occurring alone or in any sequential combination preserving the above order [1].

ES in adults as seen in our case and previous reports, are of shorter duration < 1 second, and of milder intensity compared to infants, although they may also occur in clusters [7,8]. Electrophysiologically, in adult-onset ES, the interictal hypsarrhythmic pattern is replaced by focal abnormalities which may be slowing as seen in our case or focal spikes in previous cases [7,8]. The ictal pattern shows small amplitude slow waves when compared against high amplitude waveforms in infantile-onset spasms [7,8].

The age of onset of ES has been found to depend on three factors in previous studies: i) lesion volume: age of onset being significantly earlier in the hemispheric pathologies than focal lesions [9], ii) type of the lesion: seizures have been found to occur earlier in tuberous sclerosis > cortical dysplasia > glioneural tumors [9], iii) cortical maturation gradient: relative excess of excitative over inhibitive synapses in the cortical network begins early in the posterior parts of the cortex, followed by involvement of the anterior parts later in age [10-12], the variations of activity with age [11]. 4.

Conclusion

The rare occurrence of focal ES in the third decade of life in our case can be plausibly attributable to the three mechanisms discussed above viz. lesion being focal, glioneural tumor and located anteriorly. Our case highlights the importance of video-EEG as a useful tool in the diagnosis in adult-onset ES and many of these patients are potential candidates for epilepsy surgery, particularly when MRI shows unilateral focal abnormality.

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Ethical Compliance Statement

The authors confirm that ethical clearance was obtained from institutional review board. We also confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines. We confirm that all the clinical information including videos of the patient were published after taking informed consent of the patient.

Declaration of Competing Interest

The authors report no declarations of interest.

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