Indian Journal of Medical Specialities 8 (2017) 217–220

Contents lists available at ScienceDirect

Indian Journal of Medical Specialities

journal homepage: www.elsevier.com/locate/injms

Case report

Electro-clinical expression and pharmaco-therapeutic options in Jeavons syndrome-case report and review of literature

Sweta Singla[a], Sachin Sureshbabu[a,*], Deepankshi Razora[a], Gaurav Kumar Mittal[a], Sudhir Peter[b], Sobhana Chindripu[b], Priya Gupta[a]

[a] Department of Neurology, St Stephens Hospital, Delhi, India
[b] Department of Pathology, Metropolis Labs, Kochi, Kerala, India

ARTICLE INFO

Article history:
Received 5 July 2017
Received in revised form 15 November 2017
Accepted 15 November 2017
Available online 21 November 2017

Keywords:
Jeavons syndrome
eyelid myoclonia
absence
photosensitivity

ABSTRACT

Jeavons syndrome (JS) is a well known but under reported epilepsy syndrome characterized by distinct clinical and electrographic features. In a 10 year old boy with atypical absences, a videotelemetry was performed and the findings are narrated to illustrate the spectrum of abnormalities in JS, comprising of eye closure induced paroxysms of epileptiform discharges,focal and generalized spike and wave discharges, occipital intermittent rhythmic delta activity and photopraoxysmal response. The report also discusses the therapeutic strategies employed, especially the addition of zonisamide as a very effective first add−on. In conclusion, this case report is an attempt to briefly explore the manifold aspects of this uncommon epilepsy syndrome especially the clinical and electrographic presentations and treatment strategies.

1. Introduction

Jeavons syndrome is a distinct phenotype of childhood epilepsies wherein patient has the characteristic seizure semiology of eyelid myoclonia associated with absence seizures (EMA). The appearance of paroxysms of spike wave discharges in response to slow closure of eyes is the electrographic hallmark. However a host of other abnormalities like focal and generalised interictal discharges, occipital intermittent rhythmic delta waves and a photoparoxysmal response can also be observed in the EEG recordings of these patients. Anti-epileptic drugs like valproate and ethosuximide are commonly used with variable success.

2. Case report

A 10 years old boy, youngest of the four siblings, born out of non consanguineous marriage, had a normal full term vaginal institutional delivery with normal developmental milestones, started having brief episodes (lasting 5–6 s) of retropulsion of neck with uprolling of eyes with repeated blinking of eyes associated with loss of awareness. Daily multiple countless episodes have been occurring since last 6 months. Patient was referred to our Neurology facility for further evaluation. A 24 h long term videotelemetry was performed during which multiple clinical events could be recorded. The interictal record showed the following characteristics:

1) Slow eye closure induced EEG paraoxysms lasting 0.3–12 s which occurred either immediately or few seconds after slow closure of eyes. The corresponding EEG revealed the eye closure artefact followed by a train of 3–4 Hz microV generalized and at times fragmented frontally dominant spike and wave discharges. (Fig. 1A & B)
2) Focal interictal epileptiform discharges (IEDs) in the form of spikes/spike and wave discharges and polyspike/polyspike and wave discharges were observed predominantly in the frontal region but also in the parietal, midline, temporal and occipital distribution. (Fig. 1C & D)
3) Occipital intermittent rhythmic delta waves precipitated by eye closure and often admixed with spikes. (Fig. 1E)
4) Photoparoxysmal response- at 8–20 Hz intermittent photic stimulation with eyes open and closed. The discharges were frontally dominant and generalized. At 20 Hz an admixture of theta-delta rhythm along with few small spikes. (Fig. 1F)

* Corresponding author at: St Stephens Hospital Tis Hazari, Delhi, 110054, India.
E-mail addresses: drsweta79@gmail.com (S. Singla), drsachins1@rediffmail.com (S. Sureshbabu), deepankshi.199@gmail.com (D. Razora), drgkmittal@gmail.com (G.K. Mittal), sudhirpeter@rediffmail.com (S. Peter), consecrated77@yahoo.com (S. Chindripu), priyagutpa@gmail.com (P. Gupta).

https://doi.org/10.1016/j.injms.2017.11.002
0976-2884/

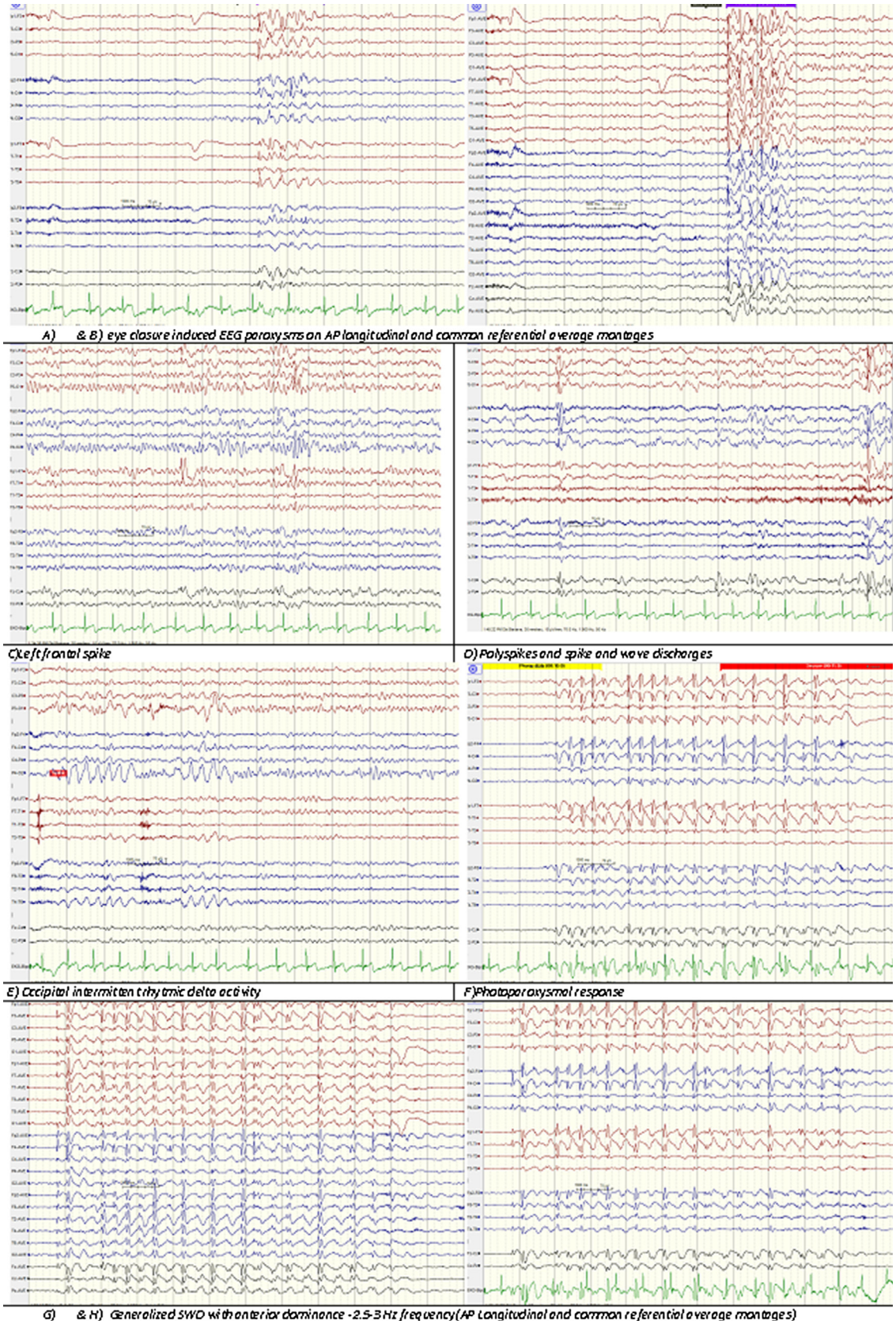

Fig. 1. A) & B)Generalized SWD with anterior dominance −2.5–3 Hz frequency (AP Longitudinal and common referential average montages) C) Left frontal spike D) Polyspikes and spike and wave discharges E) Occipital intermittent rhytmic delta activity, and F) Photoparoxysmal response.