INTERNATIONAL JOURNAL OF EPILEPSY 2 (2015) 90–93

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://www.journals.elsevier.com/international-journal-of-epilepsy

Case Report

Refractory status epilepticus: Febrile Illness Related Epileptic Syndrome (FIRES)

Varun Kumar Sharma *, *Anaita Udwadia Hegde*, *Fazal Nabi*

Jaslok Hospital and Research Centre, Mumbai, India

ARTICLE INFO

Article history:
Received 26 November 2014
Accepted 20 July 2015
Available online 1 October 2015

Keywords:
Refractory epilepsy
FIRES
Ketogenic diet
Lacosamide

ABSTRACT

In recent literature, a new entity has emerged, which focuses on a possible non-encephalitic epileptic encephalopathy precipitated by fever in a previously normal child. We report a typical case of Febrile illness related epileptic syndrome (FIRES) from Indian subcontinent. The index case presented with fever and multiple seizures, which progressed to status epilepticus and encephalopathy. All infectious, metabolic and autoimmune markers were negative. Convulsions were refractory to all possible treatment except thiopentone which achieved burst suppression pattern. Breakthrough seizures were prevented by using lacosamide and ketogenic diet along with multiple anticonvulsants. At 1-year follow-up, patient had a relatively good neurological outcome, however has persistent refractory epilepsy.

1. Introduction

In India, viral encephalitis is an important cause of mortality and morbidity in children. Despite latest diagnostic modalities, no etiological agent is identified in many cases.[1] In recent literature, a new entity has emerged which focuses on a possible non-encephalitic refractory status epilepsy precipitated by non-specific fever in previously normal children. The typical clinical presentation, characteristic seizure semiology, CSF and MRI imaging findings merit it to be listed under the most recent accepted term known as Febrile illness related epileptic syndrome (FIRES).[2,3]

2. Case report

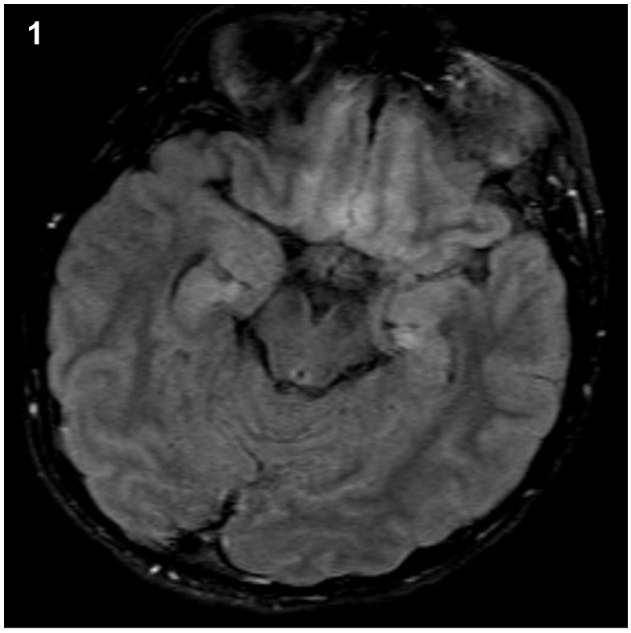
A 13-year-old, previously healthy school-going adolescent girl was admitted to the PICU with two episodes of seizures, which was complex partial in nature. There was history of fever since 3 days and headache since 1 day. On admission in our PICU she was afebrile, conscious, following commands but was slightly irritable. There was no neurological deficit, no signs of meningeal irritation or raised intracranial tension. Past medical, surgical, family, social and environmental exposure history was unremarkable.

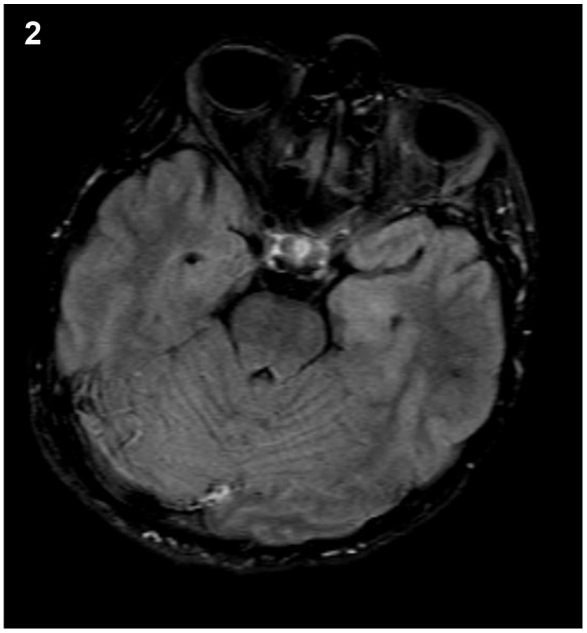
CSF microscopy revealed 8 cells/mm^3 and all were lymphocytes. CSF glucose was 54 mg/dl with a simultaneous blood glucose level of 95 mg/dl. Protein was 45 mg/dl. CSF bacterial culture was negative. CSF for HSV PCR, entire DNA and RNA viral panel (Rapid nucleic acid amplification) was also negative. Anti-NMDA and anti VGKC antibodies were negative. MRI of brain showed very subtle gyriform intracortical hyperintensities on FLAIR images through the lateral perisylvian cortex of the left cerebral hemispheres accompanied by hyperintensities in the left uncus and possibly the left hippocampus (Figs. 1 and 2).

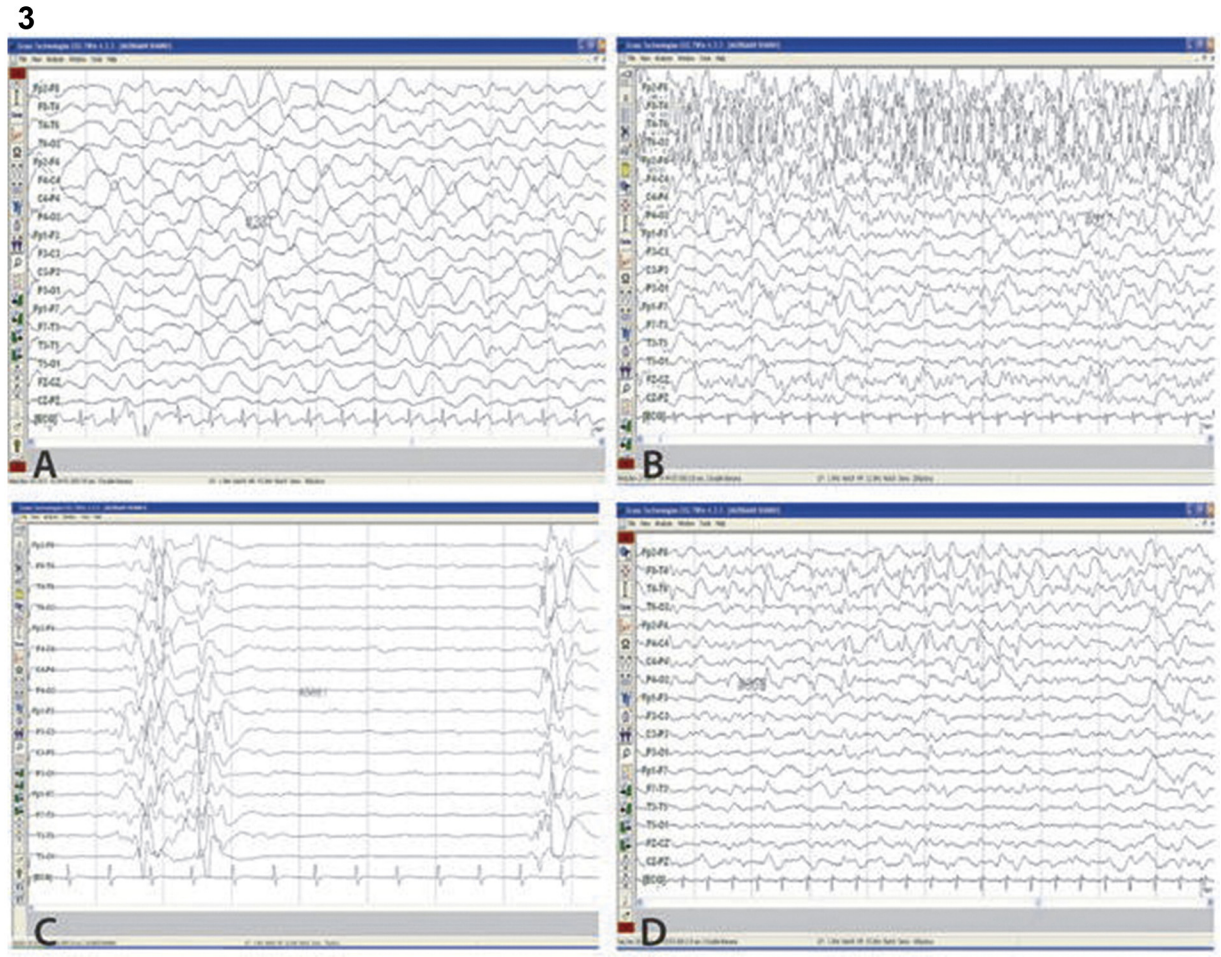
Patient was treated with intravenous ceftriaxone, acyclovir and phenytoin. During the initial hospital course patient

* *Corresponding author at*: 2007/2, 20th Floor, Jaslok Hospital, Dr G Deshmukh Marg, Peddar Road, Mumbai 400026, India. Tel.: +91 09769860225.
E-mail address: drvarun1983@gmail.com (V.K. Sharma).

http://dx.doi.org/10.1016/j.ijep.2015.07.001
2213-6320/







Figs. 1 and 2 – Subtle gyriform intracortical hyperintensities on FLAIR images through the lateral perisylvian cortex of the left cerebral hemispheres accompanied by hyperintensities in the left uncus and hippocampus. *Fig 3*(A) **EEG showing diffuse large amplitude slow waves;** (B) **EEG showing continuous epileptiform discharges;** (C) **Burst suppression pattern on tapering Thiopentone;** (D) **EEG with diffuse slowing along with interictal discharges over both hemisphere but predominantly in right frontal region.**