**Abstract**

New onset refractory status epilepticus (NORSE), is a rare, neurological condition characterised by prolonged periods of refractory epileptic seizure with no readily identifiable cause in otherwise healthy individuals. Anatomical imaging like MRI and serology is usually unremarkable. In patients who have underlying etiology as auto-immune encephalitis without any evidence of auto-antibodies FDG PET may help in early diagnosis and treatment response as it tends to accumulate in the neuronal tissue whenever there is increased blood flow, metabolic demand or increased electrical activity which reverts back with clinical recovery.

**Keywords:** $^{18}$F-FDG PET, NORSE, MRI

We present the case of a 14-year-old male patient who presented with fever of 5 days’ duration followed with refractory status epilepticus (SE). His electroencephalography showed generalized epileptiform discharges. Contrast-enhanced magnetic resonance imaging (MRI) brain [Figure 1] and cerebrospinal fluid (CSF) examination were unremarkable except for mild pleocytosis. Blood tests including viral markers, HIV, scrub typhus, dengue, herpes, mycoplasma serology, onco-neuronal antibodies, and autoimmune profile were unremarkable. In view of clinical findings, it was suspected that the patient had occult autoimmune encephalopathy which was preceded by fever. The patient then underwent whole-body $^{18}$F-fluorodeoxyglucose positron emission tomography-computed tomography ($^{18}$F-FDG PET-CT) brain scan to confirm the diagnosis and rule out other diagnostic possibilities. The whole-body $^{18}$F-FDG PET-CT scan was unremarkable, however $^{18}$F-FDG PET-CT scan of the brain revealed [Figure 2] multiple focal areas of hypermetabolism in bilateral fronto-parieto-temporal and cingulate cortices with globally reduced $^{18}$F-FDG uptake in rest of the bilateral cerebral, cerebellar, and subcortical regions, without any obvious abnormality on the CT images that were likely to be of inflammatory etiology. Three-dimensional stereotactic surface projection (3D-SSP) analysis with Z score estimation of the $^{18}$F-FDG PET brain images was done with Cortex ID software (Illinois, Chicago, USA) (GE Healthcare) using age-matched controls [Figure 3], which...
showed multiple areas of hypermetabolism (red-yellow color) and hypometabolism (blue color) in the brain parenchyma. Follow-up \textsuperscript{18}F-FDG PET study [Figure 4] done at 6 months after the initial presentation and after a complete course of intravenous immunoglobulin, showed normalization of the cerebral glucose metabolism.

NORSE is a rare form of super-refractory SE where no etiological factor is identified in a patient with no prior history of epilepsy.\textsuperscript{[1]} The common clinical features include super-refractory SE following a mild febrile illness with possible initial CSF pleocytosis and unremarkable anatomical imaging such as MRI as seen in our case.\textsuperscript{[1-4]} The outcome is frequently fatal with severe neurological sequelae as optimal management for this devastating condition remains unclear.\textsuperscript{[2,3]} Recently, few case series in literature have suggested an underlying etiology of autoimmune encephalitis without any evidence of autoantibodies and for such patients, immunotherapies could be a given treatment of choice, which could improve the patient outcome.\textsuperscript{[5-7]} In our case, the patient made complete recovery following treatment with immunotherapy. \textsuperscript{18}F-FDG PET-CT not only helped in the diagnosis by documenting hypermetabolic areas in the brain suggesting cerebral inflammation, but
Figure 3: 3D-SSP analysis with Z score estimation of the \(^{18}\)F-FDG PET brain images was done with Cortex ID software (GE Healthcare) using age-matched controls, which showed multiple areas of hypermetabolism (red-yellow color) and hypometabolism (blue color) in the brain parenchyma.

Figure 4: Comparative \(^{18}\)F-FDG PET study showing initial baseline axial \(^{18}\)F-FDG PET brain images (a-h) done at the time of initial presentation of symptoms, and follow-up \(^{18}\)F-FDG PET brain images (i-p) done at 6 months after the initial presentation and after a complete course of intravenous immunoglobulin, showing normalization of the cerebral glucose metabolism.
also helped in monitoring response to immunotherapy and prognosis of the patient.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Wilder-Smith EP, Lim EC, Teoh HL, Sharma VK, Tan JJ, Chan BP, et al. The NORSE (new-onset refractory status epilepticus) syndrome: Defining a disease entity. Ann Acad Med Singapore 2005;34:417-20.
2. Gall CR, Jumma O, Mohanraj R. Five cases of new onset refractory status epilepticus (NORSE) syndrome: Outcomes with early immunotherapy. Seizure 2013;22:217-20.
3. Van Lierde I, Van Paesschen W, Dupont P, Maes A, Sciot R. De novo cryptogenic refractory multifocal febrile status epilepticus in the young adult: A review of six cases. Acta Neurol Belg 2003;103:88-94.
4. Baxter P, Clarke A, Cross H, Harding B, Hicks E, Livingston J, et al. Idiopathic catastrophic epileptic encephalopathy presenting with acute onset intractable status. Seizure 2003;12:379-87.
5. Ishikura T, Okuno T, Araki K, Takahashi MP, Watabe K, Mochizuki H. A case of new-onset refractory status epilepticus (NORSE) with an autoimmune etiology. Rinsho Shinkeigaku 2015;55:909-13.
6. Gupta P, Patel S, Ranjan R, Agrawal CS. An interesting case of super-refractory status epilepticus. Neurol India 2015;63:628-9.
7. van Baalen A, Häusler M, Boor R, Rohr A, Sperner J, Karlmann G, et al. Febrile infection-related epilepsy syndrome (FIRES): A nonencephalitic encephalopathy in childhood. Epilepsia 2010;51:1323-8.