

096

STATUS EPILEPTICUS SECONDARY TO UNUSUAL ENCEPHALOPATHY

Khalid Ali,¹ Michael Flower,² Charlotte Lawthom¹. ¹*Royal Gwent Hospital;*
²*University Hospital of Wales*

10.1136/jnnp-2014-309236.96

An 18-year-old girl presented urgently with recurrent focal seizures. The seizures start with myoclonic jerking of the left leg, spreading to the left arm and head. She had complained of mild migraine like headaches for the last 4 weeks and had had formal visual hallucinations. She had no past medical history. She was

apyrexial, with continuous myoclonic activity of the left leg, arm and head, left homonymous hemianopia and left sided ataxia.

Routine blood tests were normal or negative, HIV test was negative, MRI brain showed right parieto-occipital gyral swelling, CSF examination was normal with normal CSF lactate. Genetic analysis for the common mitochondrial DNA mutations was negative. Mitochondrial DNA replication and maintenance is governed by nuclear DNA, the patient was found to be homozygous for the 1399G>A base substitution in the polymerase gamma nuclear gene. POLG mutations are thought to be a major human disease gene.

Aggressive antiepileptic treatment is the mainstay of managing this encephalopathy. Initially, levetiracetam, eslicarpazepine and sodium valproate were used to control the seizures. Sodium valproate was stopped due to the high risk of liver failure associated with using it in these patients.

The patient did later present with liver failure. This was managed conservatively.