

# New-Onset Refractory Status Epilepticus Syndrome (NORSE), Early-Onset Schizophrenia and Possession: A Clinical Dilemma in the Multicultural Asian Emergency Department Setting

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## Abstract

The association between epilepsy, possession and psychiatric symptoms has long been appreciated. NORSE for example, is a diagnostic challenge since its presentation is atypical. One clinical presentation can be wrongly interpreted for another disease. It is still an ambiguous issue. This has resulted in varying and controversial attempts at classifying their associations. This paper discusses how a young patient with NORSE was initially treated for possession and later diagnosed as early onset schizophrenia. The ambiguous nature of the clinical presentation of NORSE and the Asian traditional attributes affected the delay of this young girl's accurate diagnosis and treatment.

## INTRODUCTION

We present a young girl with NORSE initially treated for possession and later diagnosed as early onset schizophrenia.

## CASE REPORT

We present a young girl with NORSE initially treated for possession and later diagnosed as early onset schizophrenia. Her illness began when she experienced a sudden acute confusional state after returning from school one day. She spoke irrelevantly for a few hours and was well the next day. A few days later she developed generalized tonic-clonic fits (GTC) lasting for 10 minutes after which she vomited. She was never febrile. She was seen and stabilized in Emergency Department. CT brain was normal and subsequently she was admitted for 2 days. She was thoroughly investigated including lumbar puncture which reveals negative results.

A Magnetic Resonance Imaging (MRI) of the brain was done and it also showed normal findings. Fortunately the electro-encephalogram (EEG) showed focal slow waves over the left temporal region and spike waves over the right temporal region. She remained seizure free for the duration of admission and was discharged well. A day later she developed GTC fits lasting for 10 minutes with urinary incontinence and was admitted for a period of 4 days.

Sodium Valproate 3.5ml twice daily and Clonazepam 2 mg daily was administered orally. On discharge she was well for 2 weeks before she became confused for 2 days followed by episodic left focal clonic jerks. She was subsequently seen in Emergency Department and was readmitted in neuro-medical ward. Her condition deteriorated. She was unable to walk or talk. There was continuous lip smacking and chewing movements with bilateral left eye deviation. There was also occasional up-rolling of eyeballs with drooling of saliva. There was poor and erratic sleep. She frequently bit her hands and scratched her skin until it bled.

A clinical diagnosis of New-onset Refractory Status Epilepticus Syndrome (NORSE) was made and intra-venous antiepileptic treatment was given. Further investigations including cerebro-spinal fluid (CSF) analysis, and also culture ruled out all infective or auto-immune causes. The repeat MRI Brain showed ventriculomegaly with mild cortical atrophy. The repeat EEG was suggestive of Epileptic encephalopathy and there was no improvement in the subsequent EEG or behaviour despite intra-nasal midazolam.

After 1 week, the neuro-medical team decided to refer to psychiatry and a diagnosis of early onset schizophrenia has been made.

The clinical presentation was so ambiguous that she was sent to faith healers and to a psychiatrist with the diagnosis of Early Onset Schizophrenia. She appeared better after two days but developed basal ganglia/extrapyramidal signs like dystonic rigidity, tongue tremors and bradykinesia on the third day.

The onset of extrapyramidal symptoms indicated possible irreversibility of her condition. However her parents were convinced that she was possessed and insisted on discharge. She never returned to the hospital.

## **DISCUSSION**

“With respect to epileptic diseases, the fits of which, it was affirmed, constantly returned every new and full moon. Galen says the moon governed the periods of epileptic case, hence epileptics were what Greeks and Romans called lunatics. The epileptic mad was not considered to be mad or vexed with the demon at all times, but only under the paroxysms of his epileptic disorder, which returned at the changes of the moon [1]. This illustrates how much we do not understand about epilepsy, the great mimicker of medical conditions.

Status epilepticus (SE) is a clinical condition characterised by an epileptic seizure or a series of seizures that lasts for at least 30 minutes without consciousness being regained. After 30 to 60 minutes of continuous seizures, there is an increased risk of neuronal damage due to systemic, metabolic disturbances and the direct excitotoxic effect of neuronal discharges during the seizure. Some authors have added a time line of 60 minutes. The incidence of generalised convulsive status epilepticus (GCSE) is between 40 and 80 per 100,000 [2]. The associated mortality rate has been estimated to be as high as 22% varying from 7.6% to 19% within the first 30 days [3, 4]. Refractory status epilepticus (RSE) is a life-threatening condition, characterized by the failure to respond to first- and second-line anticonvulsant therapy. Risk factors for RSE include delays in treatment, central nervous system (CNS) infections, metabolic encephalopathy and hypoxia [5].

## **POSSESSION**

The syndrome of possession, which is relevant to some belief systems, is still very much encountered in Malaysia and many Asian countries. This would suggest that cross-cultural syndromes, which share a common belief, may have that supporting and maintaining factors of religion. Epileptic seizures have a historical association with religion, primarily

through the concept of spirit possession. This can be explained within the context of the Voodoo belief system [6]. In Malaysia, with a multi-ethnic, multi-cultural population, there are numerous conditions that are attributed to possession and traditionally accepted conditions among others, Latah and Amok [7]. In this young patient, the supernatural beliefs played a detrimental role as it delayed treatment which in irreversible brain damage

## **CHILDHOOD-ONSET SCHIZOPHRENIA (COS)**

This patient was referred to the psychiatrist as a case of early-onset Schizophrenia. The prevalence of COS is very low with rates of 0.14 in 1000, with a higher rate among boys with a ratio of 2.5 to 1 [8, 9]. The onset of COS is almost always insidious [10]. This patient presented with an acute confusional state followed by visual and auditory hallucinations. Interestingly, the ventriculomegaly and cortical atrophy seen in this patient was similar to what is often seen in COS [11].

## **CONCLUSION**

The NORSE syndrome is a serious and life threatening condition that requires immediate attention. A delay in treatment will result in rapid deterioration and possibly death. The awareness of how epilepsy can mimic a psychiatric condition is important to avoid misdiagnosis and inappropriate treatment during the presentation to Emergency Department. Finally, the traditional and cultural beliefs of Asians can lead to negligence purely due to the lack of awareness.

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