Landau-Kleffner Syndrome : A Case Report

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Abstract— A 6-year-old boy known sickle cell trait developed generalized tonic- clonic seizures , one month later he suffered from decreased attention , hyperactivity , lack of response to his family, loss of understanding , aggressive and oppositional behavior. Brain CT and MRI were normal . EEG showed partial epileptic activity . He was diagnosed as Landau-Kleffner syndrome (LKS) . His condition improved on corticosteroid regimen .

Index Terms— Landau-Kleffner syndrome, Epileptic aphasia , Landau-Kleffner syndrome in Jazan- Saudi Arabia.

1 Introduction

Landau-Kleffner syndrome (LKS) is an age-related syndrome of childhood. Its main features are loss of language skills,and seizures.

The language skills and speech may improve over time. LKS usually starts before the age of six years and affects twice as many boys as girls. The underlying cause of LKS is not yet known [1].We are presenting this case of Landau-Kleffner syndrome as a first reported case in Jazan region-KSA to increase the awareness about early diagnosis and to highlight the importance of appropriate and early management for a better patient outcome.

2 Case report:

6-year-old boy, known case of sickle cell trait , he was doing well until 6 months prior to presentation when he started to has generalized tonic clonic seizures, proceeded by facial twitching and lasts for 5 minutes then stop-spontanously followed with postictal sleep for two hours. He has no history of fall down. The patient went to peripheral hospital , basic investigations were done , CT- brain was done and was normal , he was discharged on carbamazepine. One month later , the mother noticed her child behavior was changed in form of decreased attention , hyperactivity , loss of understanding , looks like he can't hear or respond to family orders , aggressive and oppositional behavior. His speech was affected gradually started with stutter , stammer and ended with aphasia after 2 months.They went to other hospital , EEG was done and showed partial seizure,and MRI brain done and no abnormality was detected , and started him on sodium valproate with no improvement. The family sought medical advice in another hospital and was referred to Jizan king Fahd central hospital to pediatric neurology clinic. He was started on oral prednislon course for 6 weeks .After 3 weeks from this regimen , the patient's attention understanding & speech was improved dramatically. On examination: the patient looks well, conscious , vitally stable with normal growth parameters for his age. Neurological examination revealed aphasia , hyperactivity , and inattention .Cranial nerves were intact with normal tone , power , reflexes and sensation . In our hospital thePatient was diagnosed as Landau-Kleffner syndrome and started him on steroids therapy with tapering over 6weeks. After 3 weeks of steroids the patient's condition improved. Now the patient has 4 relapses in the last three years since the diagnosis was made with the last relapse on April-4 ,2016.

3 Discussion

Landau-Kleffner syndrome (LKS) is an age-related epileptic syndrome of childhood. Its main features are a loss of speech and language skills, with seizures[1].

Landau-Kleffner syndrome is a syndrome characterized by acquired aphasia and convulsive disorders , and sleep-activated EEG paroxysms predominating over the temporal or parieto-occipital regions. Other symptoms include behavioral or psychomotor disturbances and epilepsy with a favorable outcome for seizure control. The prevalence is not clear. The syndrome has a male predominance, with an approximately 2:1 ratio. This regressive syndrome affects children between 3 and 9 years of age at the time of presentation [2]. LKS has an unpredictable outcomes and varying severity with a relapsing remitting course, requiring constant adaptation and resourcefulness from parents, speech/language therapists, neuropsychologists, and neurologists[3]. The Outcomes range from complete recovery to severe permanent aphasia, with most experiencing improvement and residual moderate language deficits [4]. The etiology remains unknown, and it is thought to be due to diverse causes. Encephalitis has been postulated, but not verified [5]**.** Other etiologies reported include genetic predisposition [6], [7] ,[8] ,toxoplasmosis[9], neurocysticercosis {10} , temporal astrocytoma [11],temporal ganglioglioma [12] , hemophilusinfluenzae meningitis[13] , subacute sclerosing panencephalitis{14}, inflammatory demyelinating disease[15], [16] and abnormal zinc metabolism[17].

Investigations have not delineated sufficient evidence to explain the pathophysiology. CSF [18],[19],[20], computed tomography[21] ,[22] ,[23] and magnetic resonance imaging (MRI) [22] [8] [24] are normal. Various single photon emission computed tomography (SPECT) and positron emission tomography (PET) studies on small numbers of patients have shown temporal lobe abnormalities in brain perfusion and glucose metabolism [25],[26],[27].

EEG findings in LKS are variable, but striking. Bilateral independent temporoparietal or temporal spikes, bilateral 1–3 Hz slow-wave maximally temporal activity, generalized sharp- or slow-wave discharges, and unilateral or multifocal spikes are described [28].

LKS is difficult to treat. Treatment modalities include antiepileptic drugs , corticosteroids, intravenous immunoglobulin, ketogen-

ic diet , multiple subpial transections [29] and temporal lobectomy [11]. The patient was started on corticosteroids regimen , and after 3 weeks of steroids he had significant improvement in language , attention and comprehension.He had4 relapses in the last three years since the diagnosis was made with the last relapse on April-4 ,2016 and he is on regular follow up.

4 REFERENCES

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