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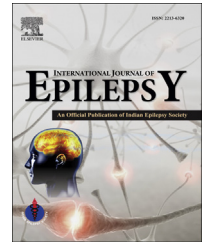
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Image of the Month

EEG in lissencephaly



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ABSTRACT

We report the characteristic EEG findings of an infant with lissencephaly who presented with infantile spasms.

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An eight-month-old male presented with developmental delay and flexor spasms for the last 2 months. He was the first child of non-consanguineous marriage. The antenatal and perinatal periods were uneventful. He achieved social smile at 5 months, partial neck holding at 6 months and was recognizing his mother and cooing at the time of presentation. There was no grasp. There was no regression of attained milestones. He had started having flexor spasms since the last 2 months. The spasms occurred in clusters mainly on waking up from sleep. He also had stiffness in the lower limbs which had been noticed for the last 2 months. This was non-progressive. Vision and hearing were normal. The family history was uneventful.

On examination, the child had normal facies and no neurocutaneous markers. The weight and length were age appropriate. He had microcephaly (OFC 39.5 cm). The cranial nerve examination was normal. The tone was increased in all 4 limbs with brisk deep tendon reflexes and extensor plantar responses. Systemic examination was unremarkable. The MRI of the brain showed lissencephaly with no gradient (Fig. 1). The EEG showed diffuse high voltage theta activity (Fig. 2).

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Fig. 1 – T2 axial MRI image of the brain at the level of the basal ganglia showing thickened cortex with no gradient, suggestive of lissencephaly.

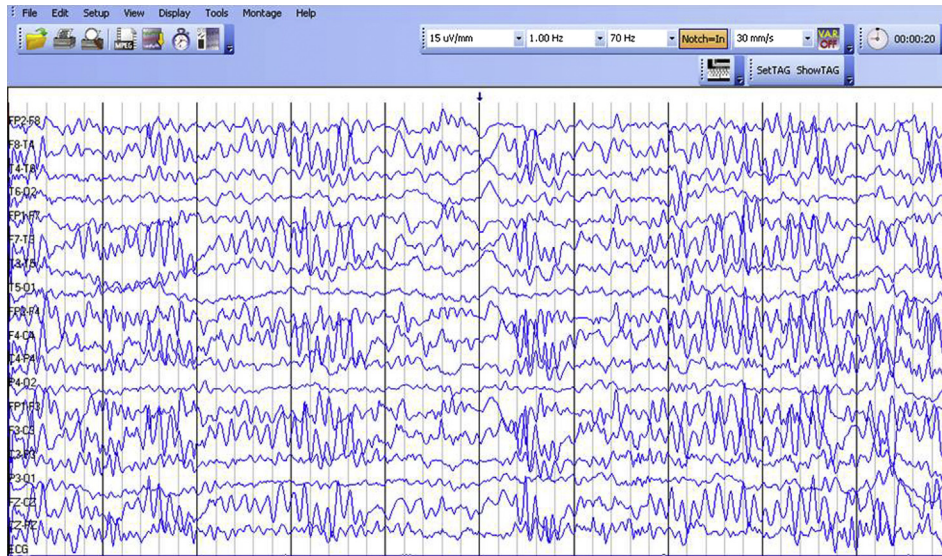


Fig. 2 – EEG showing diffuse high voltage bisynchronous theta activity with amplitude fluctuations of cortical activity attenuation lasting 2–3 s (sensitivity 15 $\mu\text{V}/\text{mm}$).

Lissencephaly is a disorder of neuronal migration characterized by absent or abnormally wide gyri and abnormally thick cortex. Epilepsy is a major presentation of lissencephaly syndromes and is usually drug-refractory.¹ Three EEG patterns have been classically described in patients with lissencephaly.² EEG pattern type I consists of diffuse high amplitude, greater than 100 mV, alpha and beta activity in all cortical regions. EEG pattern II shows alternating high amplitude, up to 300 mV, bursts of sharp and slow waves, followed by short duration periods of attenuation of cortical activity that last up to 3 s. EEG pattern III is characterized by high amplitude spike waves or sharp wave activity, without alpha or beta frequencies, or attenuations of the cortical activity.³ The current case showed EEG pattern II. The characteristic EEG patterns along with the clinical findings may help neurologists to consider the diagnosis of lissencephaly.

Conflicts of interest

All authors have none to declare.

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