Ictal Nystagmus in a Patient with Nonketotic Hyperglycemia: A Rare Clinical Phenomenon of Occipital Epilepsy

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A 45-year-old man with a history of diabetes mellitus and hypertension who had been off medication for the past one year presented with intermittent spells of unresponsiveness characterized by initial giddiness, blurring of vision, a leftward head turn, and eye blinking, followed by involuntary movements of all four limbs that lasted for 3 min for 2 days. He had a total of two spells before admission. He was treated with a levetiracetam loading dose of 40 mg/kg, followed by a maintenance dose of 20 mg/kg. He had further events without involuntary movements while in the hospital with giddiness, visual blurring, a head turn to the left, and ictal nystagmus with a fast component to the left side on day 2 [Video 1]. On day 3, he had visual blurring, a head turn to the left with left upper limb tonic–clonic movement, along with ictal nystagmus with a fast component on the left side.

Video electroencephalogram (EEG) on day 2 demonstrated a clinical event characterized by an initial head turn to the right side, followed by a head turn to the left side, and posturing of the left hand with an ictal event. The EEG graph demonstrated ictal and interictal events, demonstrating spike and wave discharges initially from the left occipital, followed by the right occipital region with adjacent spread anteriorly [Figure 1]. The cause of occipital epilepsy was attributed to hyperglycemia (blood sugar >350 mg/dl). His glycated hemoglobin HbA1C was 14.5%. Serum osmolality was 301 million osmoles, and urinary ketone body was negative. The rest of the metabolic parameters were normal. Magnetic resonance imaging (MRI) of the brain was normal. He continued to have seizures for the next 5 days in spite of adjusting and adding antiseizure medication. He required three antiseizure medications (levetiracetam, oxcarbazepine, and lacosamide) at optimal dosages. His blood sugar and his seizures were controlled. A follow-up visit at 1 month showed well-controlled blood sugar, and he was seizure free.

Ictal or epileptic nystagmus was first described by Gastaut and Roger.[1] It is a rare clinical phenomenon of epilepsy, where the direction of the nystagmus is contralateral to the epileptogenic zone, and it is most often associated with posterior cerebral hemispheric lesions.

Ictal nystagmus with fast nystagmus toward the left side corresponds to epileptic focus in the contralateral posterior hemisphere, which is the right occipital region, as demonstrated in our case. Though the initial origin of the epileptiform discharges was from the left occipital region, as seen by the clinical event of the head turning to the right with EEG discharges, the initial epileptic nystagmus could not be recorded as the eyes were closed. The contraversive fast nystagmus may result from various mechanisms.[2,3] Nystagmus occurs due to the spontaneous activation of cortical saccade regions.

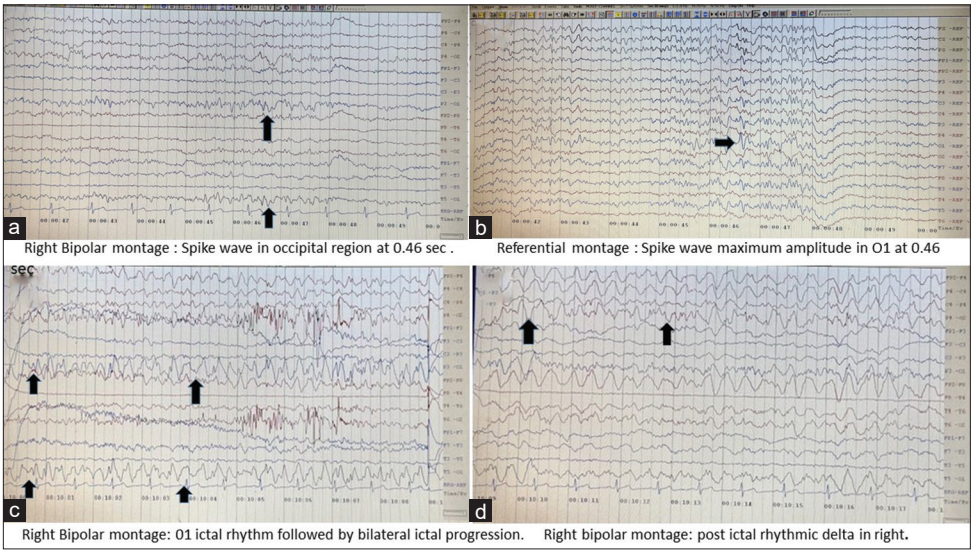

Figure 1: (a and b) Interictal EEG showing left occipital spike in longitudinal bipolar (a) and referential (b) montages; (c) initial ictal rhythm over the left occipital region; (d) late propagation of ictal rhythm over the right hemisphere. EEG = electroencephalogram

Table 1: Clinical manifestation and radiological features in non-ketotic hyperglycemia[10-13]

Clinical manifestation	MRI brain abnormality
Encephalopathy (PRES)	T2 shows symmetrical hyperintensity in bilateral parieto-occipital region and reflects as vasogenic edema
Hemiparesis	Faint diffusion restriction confined to putamen or basal ganglion
Seizure	T2 hypointensity in posterior cerebrum (occipital and parietal involvement)
Chorea-ballismus-athetosis	T1-hyperintense lesion in basal ganglion with variable intensity or hypointensity on T2/FLAIR sequence
Cortical vision loss	Cortical laminar necrosis on DW sequence later on occipital gliosis
Epilepsia partialis continua	T2-hypointense lesion in temporal and insular cortex
Hyperglycemia with hyperviscosity and hyperosmolar state	Diffusion restriction involving bilateral parieto-occipital and basal ganglion region. No contrast enhancement

DW: Diffusion weighted, FLAIR: Fluid attenuated inversion recovery, PRES: Posterior reversible encephalopathy syndrome

Epileptic discharges stimulate cortical pursuit or optokinetic nystagmus mechanisms that build ipsiversive slow-phase eye movements that cross the midline and are accompanied by reflexive contraversive fast phases.

The ictal nystagmus usually originates from the occipital lobe in the majority of cases, followed by the temporo-occipital, temporal, and frontal regions.[3] Seizures are usually symptomatic in 64.9% of cases.[3] Underlying etiologies included stroke, focal cortical dysplasia, head trauma, brain metastasis, metabolic encephalopathy, ketotic hyperglycemia and non-ketotic hyperglycemia (NKH), hypoglycemia, mitochondrial encephalomyopathy, lactic acidosis and stroke-like episodes (MELAS), hypoxia, posterior reversible encephalopathy syndrome (PRES), Sturge–Weber syndrome, arachnoid cyst, and physostigmine intoxication.[4-6]

In the present case, the patient had NKH. Criteria for epileptic seizure in NKH are blood sugar levels of more than 200 mg/dl (11 mmol/l) with hyperosmolality, dehydration, absence of ketosis, and control of seizure with control of blood sugar.[7,8] In NKH, the majority of seizures are focal motor seizures.[9] The pathophysiology of seizures in NKH is due to an increase in the metabolism of Gamma-aminobutyric acid (GABA), leading to a decreased seizure threshold. Another mechanism of seizure is the hyperosmolar gradient occurring in a hyperglycemic state that induces intracellular dehydration and seizure. Occipital lobe seizures are a rare but characteristic manifestation of hyperglycemia. NKH can have a varied clinical presentation[10,11] and radiological features.[12,13] Table 1 shows the clinical and radiological manifestations of NKH.

This case highlights that an uncontrolled hyperglycemia state without ketosis can cause occipital seizures with the rare manifestation of epileptic nystagmus. Strict control of sugar along with antiseizure medication would help to control seizures. The ictal nystagmus is always directed away from the epileptic focus.

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Submitted: 05-Sep-2023 **Revised:** 05-Dec-2023 **Accepted:** 10-Dec-2023
Published: 06-Feb-2024

Video available on: www.annalsofian.org

DOI: 10.4103/aian.aian_800_23