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Epileptic nystagmus in a patient with nonconvulsive status epilepticus

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Epileptic nystagmus (EN) is a rare form of rhythmic eye oscillations occurring during seizure activity. Not only convulsive states, but also nonconvulsive status may represent with EN and this phenomenon may be the only motor manifestation of seizure activity. Epileptic activation of a cortical saccade region may be distinguished from activation of a cortical pursuit region clinically as activation of pursuit regions results in nystagmus slow phases that bring the eyes across the midline. Horizontal EN results most commonly from seizure activity involving the occipital cortex. In this report, horizontal EN in a patient with nonconvulsive status epilepticus (NCSE) is described with clinical, radiological and electrophysiological findings that occur probably due to posterior leukoencephalopathy syndrome.

Key words: epileptic nystagmus; nonconvulsive status; electroencephalography; magnetic resonance imaging; posterior leukoencephalopathy syndrome.

INTRODUCTION

Nonconvulsive status epilepticus (NCSE) represents an epileptic state of more than 30 minutes with change of mental status or behaviour together with seizure activity on electroencephalography (EEG). The clinical correlates of such states involve a number of higher cortical functions. Anterograde and retrograde memory, affect and speech may be disturbed. Motor manifestations may be present such as subtle twitching of the face and fingers, peroral and eyelid myoclonus[1], intercurrent limb jerks, and eye deviation with nystagmus[2,3]. The latter finding, epileptic nystagmus (EN), is a rare form of rhythmic eye oscillations that occur only during epileptic seizures[4]. There are few reports in the literature about epileptic nystagmus in NCSE[3]. This is the first report of a patient with epileptic nystagmus, probably due to posterior leukoencephalopathy syndrome (PLES).

CASE REPORT

A 62-year-old woman, who had hysterectomy for endometrial carcinoma 2 years ago, was admitted to the emergency department with the findings of intestinal obstruction. Computerised tomography (CT) of abdomen revealed multiple hypointense lesions in liver and a recto-vaginal fistula. In diagnostic laparatomy, multiple metastases in liver and a pelvic solid tumour were observed and biopsy from small bowel serosa revealed adenocarcinoma infiltration.

She was followed in the intensive care unit in postoperative period. Although her neurological and physical status were normal following the operation, generalised tonic–clonic seizure developed on the third hospital-day. She had semipurposeful arm withdrawal to painful stimuli. Pupillary, corneal, vestibular and gag reflexes were intact. She had bilateral increased deep tendon reflexes and bilateral extensor plantar responses. Routine blood tests were unremarkable. Cranial CT showed bilateral multiple periventricular hypodense lesions and cerebral atrophy. Antiepileptic medication consisting of phenytoin was started with following drug blood levels. Postictal EEG revealed generalised slow background activity. She was then seizure-free until the fifth hospital-day, but still unconscious.

On the fifth hospital-day, twitching on the right side of her face was observed. On neurological

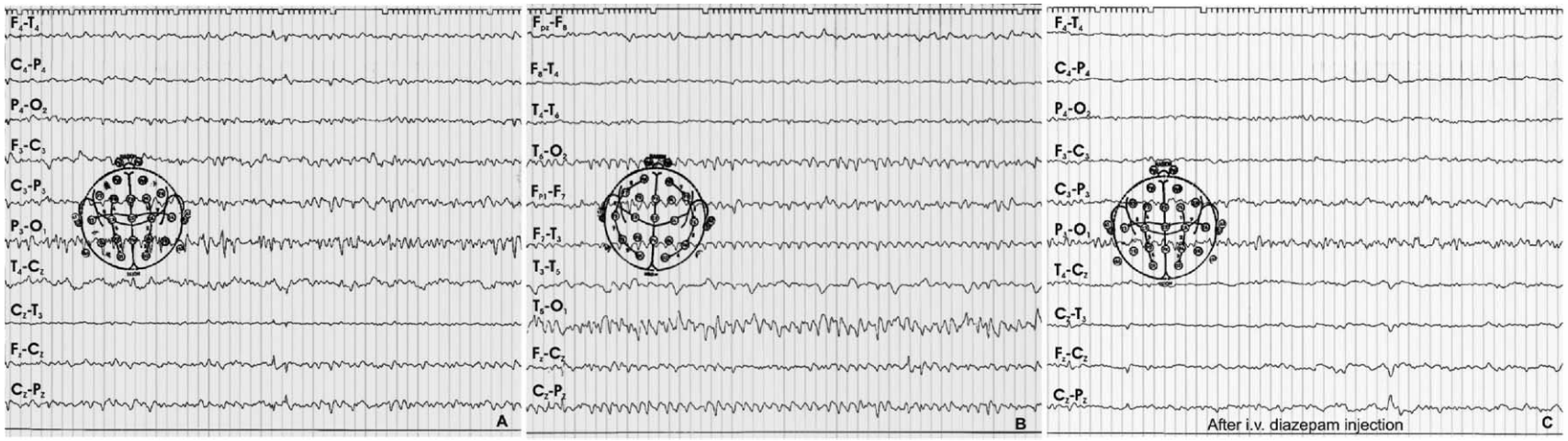

Fig. 1: (*A*) EEG recording of the patient illustrates the seizure activity on the left occipital lobe during nystagmus. (*B*) Discharges were spreading to the contralateral occipital lobe and other parts of the left hemisphere, but with the highest amplitude in the left occipital lobe. (*C*) After administration of intravenous 10 mg diazepam, suppression of ictal activity and cessation of nystagmus with no clinical improvement were observed.

examination, she had no response to painful stimuli. Pupillary reflex was normal, but deep tendon reflexes, plantar responses, corneal and gag reflexes were unresponsive. She had continuous right beating nystagmus that did not cross the midline. Her blood pressures did not exceed 140/80 mmHg. Blood tests were notable only for hyponatremia, with serum sodium of 130 mmol/l (normal 135–145), thrombocytopenia (25×10^3 mm^{-3}, normal 150×10^3–450×10^3 mm^{-3}) and elevated blood leukocytes to 18.5×10^3 mm^{-3}. Her serum sodium had decreased from the basal level of 143 to 130 mmol/l. EEG showed rhythmic 5–6 Hz sharp theta activity on the left occipital lobe (Fig. 1A). During the recording, sometimes-rhythmical discharges were appeared over the left occipital, left temporal, midline and contralateral occipital areas without any changes of nystagmus, and discharges with highest amplitude were recorded over left occipital lobe (Fig. 1B). After administration of intravenous 10 mg diazepam, suppression of ictal activity and cessation of nystagmus with no clinical improvement were observed (Fig. 1C). However, after 7 minutes, continuous nystagmus was repeated and treated successfully with midazolam therapy at a loading dose of 0.2 mg/kg, followed by a maintenance infusion dose of 0.1 mg/kg/hour. A cranial MRI performed on the same day revealed hyperintense lesions in both cerebellar hemispheres (Fig. 2A) and the left middle cerebral peduncle on T2 weighted (T2W) series. In the supratentorial compartment, bilateral lesions were found in the corpus callosum, basal ganglia and deep and subcortical white matter (Fig. 2B and C). None of these expansive lesions enhanced on postcontrast T1W images. Superficial hyperintensity along the sulci with a moderate effacement of the subarachnoid spaces, more prominent on the left posterior side on FLAIR images (Fig. 2B and C) and leptomeningeal enhancement on contrast enhanced T1W imaging (Fig. 2D) were observed. As the patient was thrombocytopenic, lumbar puncture was not performed. The patient died from sepsis on the same day. Further evaluation of aetiology with autopsy could not have been done, as patient's relatives did not give consent.

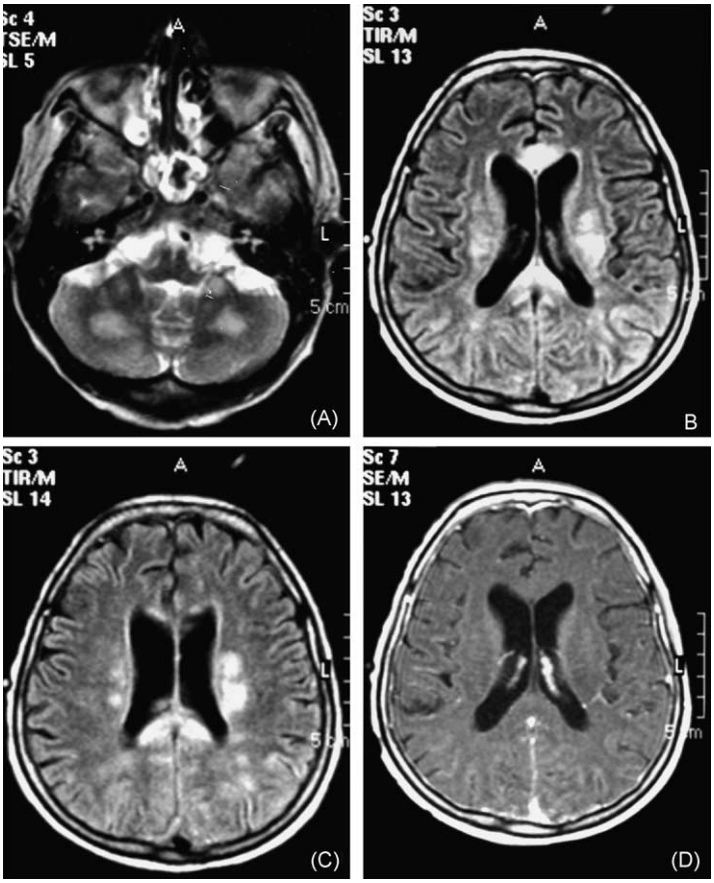

Fig. 2: (*A*) T2 weighted image (T2 WI) (TR/TE, 4000/100) shows hyperintense lesions in both cerebellar hemispheres. (*B*) Oedematous lesions in the corpus callosum and (*C*) white matter are seen on FLAIR (TR/TE/TI, 5000/100/1900) images. (*D*) Lesions do not enhance with contrast material (Gd-DTPA) on T1W SE (TR/TE, 550/15) imaging. Note that diffuse superficial hyperintensity with an effacement of the sulci, more prominent posteriorly on FLAIR (*A*, *B*) and leptomeningeal enhancement on postcontrast T1W SE images (*D*).

DISCUSSION

EN is an uncommon phenomenon characterised by repetitive and rapid saccades, in association with epileptic discharges. EN is caused by an epileptic focus near ocular motor system. Horizontal EN results most commonly from seizure activity involving the occipital cortex, although participation of adjoining portions of the parietal and temporal cortexes is possible[5,6]. There are three postulated mechanisms for the eye deviation in generation of EN. Epileptic activity of a cortical saccade region or a cortical pursuit region can produce nystagmus of different characteristics. Seizure discharge from cortical saccade regions of the temporo-occipital or frontal cortex induces a contraversive quick phase. After this phase, the defect of gaze-holding system (leaky integrator) causes slow drift of the eyes to midline[2]. Slow phase velocity decreases, as the eyes approach to midline and the eyes never cross midline[7]. Epileptic activity of cortical pursuit region in temporo-occipital cortex generates ipsiversive slow phase followed by a reflexive quick phase. Unlike EN induced by stimulation of saccade regions, those induced by stimulation of pursuit regions results in linear slow phases in which the eye movements are likely to cross the midline[8]. Epileptic activation of a cortical optokinetic region causes EN similar to the one caused by epileptic activation of cortical pursuit region[1].

In our patient, horizontal nystagmus was thought to be caused by the epileptic activation of the contralateral cortical saccade region of the left temporo-occipital cortex, since quick phases were generated away from the epileptic focus. MRI lesions and ictal EEG confirmed occipital involvement contralateral to the direction of nystagmus. Additionally, slow phase not crossing midline, can be explained by altered mental status disturbing the gaze-holding system.

In the clinical and laboratory setting of this patient, lesions were against infectious or paraneoplastic aetiology; hence, the lesions suggested acute demyelination or oedematous lesions similar to those occurring in PLES. PLES is an acute, progressive syndrome characterised by headache, nausea and vomiting, visual disturbances, altered mental status, and seizures. It is predominantly associated with white matter oedema in the posterior parietal–temporal and occipital brain regions, which may spread to basal ganglia, brainstem and cerebellum[9]. Although malignant hypertension, toxaemia of pregnancy, immunosuppressive treatment and interferon alpha therapy are the evidence based aetiologic factors in PLES, severe hypertension is not mandatory for PLES to develop, either radiological or clinically. But, postoperative hyponatremia especially in women was suggested as a risk factor for the development of cerebral oedema in recent reports[10]. Although, several seizure types have been reported in PLES, according to our knowledge our case is the first report that suggests an association between EN and PLES.

As a conclusion, in patients with NCSE, very subtle motor phenomena like nystagmus can easily be missed. Careful examination, EEG and MRI are helpful to detect the epileptic nystagmus and involvement of occipital cortex and as a result will lead to early diagnosis of this rare and interesting ictal phenomenon.

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