Lance-Adams Syndrome: A Rare Case of Post-hypoxic Myoclonus, Developing After a Snake Bite

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ABSTRACT

Lance-Adams syndrome (LAS) is a rare complication of a successful cardiopulmonary resuscitation (CPR) and is often accompanied by post-hypoxic action myoclonus. Less than 200 cases have been reported in medical literature till date. A 50-year-old female presented to the Emergency Department in a state of unconsciousness. Urgent intubation and CPR resulted in stabilization of vitals over a period of few days. On regaining consciousness, the patient developed myoclonus, which was characteristically present only on activity and absent at rest or during sleep. This action myoclonus was troublesome to the patient and interfering in the day-to-day activities of the patient. Patient was started on a combination therapy with levetiracetam and clonazepam, which resulted in marked diminution of myoclonus over a period of 15-20 days. Though LAS is a rare complication, proper diagnosis and prompt management may significantly reduce the morbidity and improves the quality-of-life.

Keywords: Lance-Adams syndrome, cardiopulmonary resuscitation, myoclonus, levetiracetam, clonazepam

Lance-Adams syndrome (LAS) is a rare complication of a successful cardiopulmonary resuscitation (CPR). LAS is known to present as action myoclonus, days to weeks after a successful CPR, due to hypoxic injury to brain. Post-hypoxic myoclonus (PHM) is divided into two types:

- The *acute type*, which is called "myoclonic status epilepticus," occurs within 12 hours in most cases after hypoxic brain damage in patients who are deeply comatose.
- The *chronic type*, called "the Lance-Adams syndrome," is characterized by action myoclonus beginning days to weeks after a successful CPR and persists in patients who have recovered consciousness after CPR.

LAS is a rare complication and less than 200 cases have been reported in medical literature till date. We present here a patient who was diagnosed as LAS after CPR due to cardiorespiratory arrest following a neurotoxic snake bite.

CASE REPORT

A 50-year-old female reported to our Emergency Department in a state of unconsciousness. Following a snake bite, she slipped into unconsciousness, as reported by the husband accompanying her. Her oxygen saturation on admission was 40% as measured by a finger oximeter, and respiratory movements were almost absent. After 10 minutes of vigorous CPR at the Emergency Department, her vital signs started to return. She was shifted to intensive care unit (ICU), and was given 20 vials of antisnake venom in total. She regained consciousness the next day, after being on intermittent positive pressure mechanical ventilation and vasopressor support for a day. Meanwhile, a computed tomography (CT) scan of the brain was done, which showed no significant abnormalities (Fig. 1). All routine investigations like complete blood count, liver function tests, renal function tests, urine routine and microbiological examination and serum electrolytes were within normal range as given in Table 1.

When she was tapered from sedation (midazolam) and muscle relaxation (atracurium), on the 3rd day of ICU, she developed a generalized seizure and subsequently myoclonic movements were continuously observed

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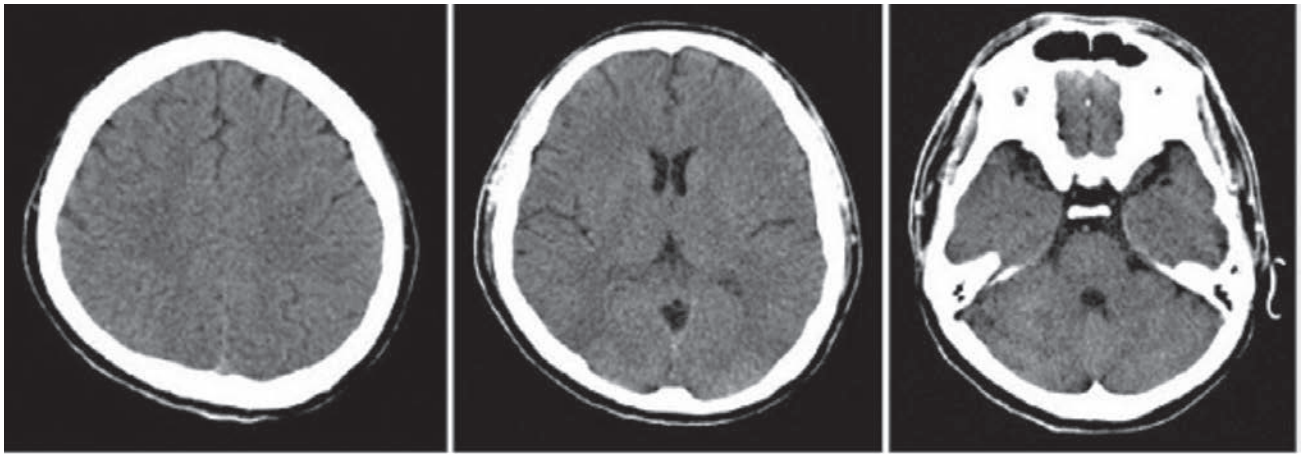

Figure 1. CT scan of the brain showing no significant abnormalities.

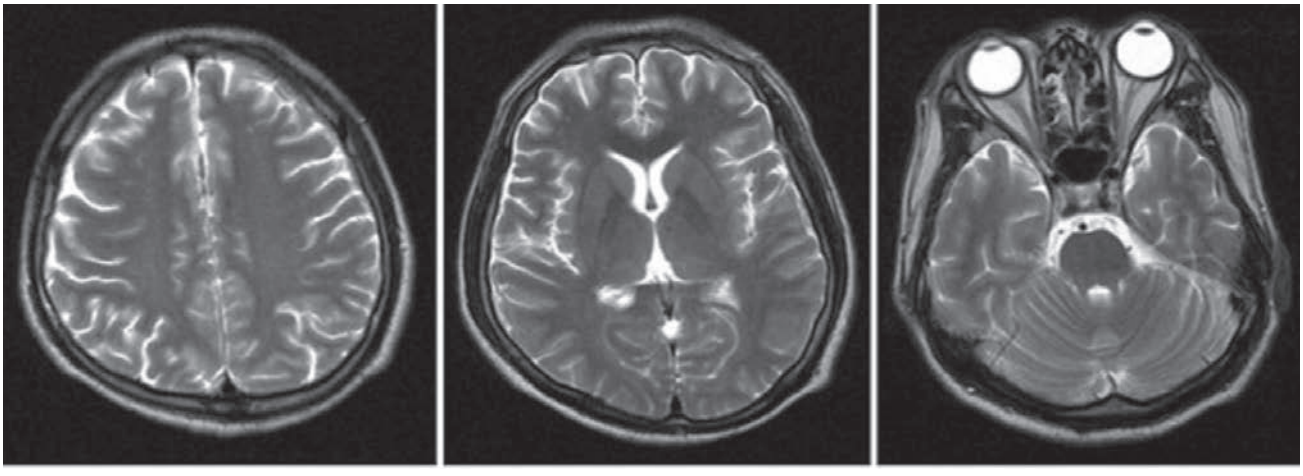

Figure 2. T_2-weighted MRI showing mild diffuse cerebral atrophy.

throughout her body, including face. At that time, the myoclonic movements were considered as generalized myoclonus secondary to hypoxic brain insult and were empirically treated with sodium valproate. However, these were not controlled with sodium valproate. The myoclonic jerks ceased with a single bolus dose of midazolam but the effect was transient. A repeat CT scan was meanwhile done, which showed no abnormalities and the T_2-weighted magnetic resonance imaging (MRI) showed mild diffuse cerebral atrophy (Fig. 2). On Day 5, the patient's mental status improved, she was now intermittently able to obey simple commands, but the myoclonic jerks continued. The rest of the central nervous system (CNS) examination was normal except higher functions, where the patient had dysarthria, dyscalculia and some attention deficits.

Subsequently, on Day 9, the patient was shifted to ward and started on levetiracetam (500 mg b.i.d.) and subsequently clonazepam (1 mg t.d.s.) was added. With these medications and physiotherapy, the patient started showing improvement, in the form of reduced frequency of myoclonic jerks, and also some improvements in higher functions. Patient was discharged on Day 21 when

Table 1. Routine Investigations

Test	Value
Hemoglobin	10.8 g/dL
Total WBC count	15,200 cells/mm^3
Differential count (N/L/M/E/B)	66/20/12/02/00%
Platelet count	1,64,000 cells/mm^3
PT/INR	14.4 sec/1.1
Serum bilirubin (total/direct/indirect)	1.2/0.4/0.8 mg/dL
SGPT	32 IU/L
Serum creatinine	0.8 mg/dL
Blood urea	28 mg/dL
Serum Na^+	138 mEq/L
Serum K^+	4.6 mEq/L

the frequency of jerks reduced to 4-9 per action, when she used to do actions slowly, but marked escalation of jerks was found on trying to speed up actions. Mild cognitive defects and dysarthria persisted though.

On regular follow-up, the patient slowly improved, though some dysarthria persisted. On 6 monthly follow-up, patient showed mark improvement, she could walk about 30 meters without support and there was marked diminution in the frequency of myoclonic jerks.

DISCUSSION

Lance-Adams syndrome (LAS) was first described in the 1960s by Lance and Adams, who described 4 patients who developed myoclonic jerks within a few days following an episode of anoxia. After recovery of consciousness, the patients continued their abnormal clonic movements, which were triggered by intentional action or external stimuli and they were relieved at resting or during sleep. Although the pathophysiology of LAS remains undetermined, the prognosis is good. It is important to distinguish LAS from post-hypoxic seizures, so a correct prognosis can be provided. One of the important clinical features is consciousness, in the acute type of post-hypoxic seizures, the patient's mental status persists as comatose, but in LAS, the patient later regains consciousness. Intentional myoclonus develops several days after the hypoxic brain insult and persists thereafter, but in post-hypoxic seizures, generalized myoclonus usually occurs within 48 hours after CPR. The myoclonus in LAS has no consistent correlation with electroencephalography (EEG) abnormalities. The patient in this case had remarkable features that are consistent with LAS.

Diagnostic imaging tests such as CT or MRI of the brain are not helpful to make a diagnosis of LAS. Neuroimaging, such as brain single-photon emission computed tomography (SPECT) or brain positron emission tomography (PET), has recently showed the anatomical and pathophysiological basis of LAS. Some studies have reported that compared with control groups, patients with LAS had significantly increased glucose metabolism in the pontine tegmentum, mesencephalon and ventrolateral thalamus. In our case, brain CT and brain MRI of the patient demonstrated no abnormalities and EEG was normal too.

The neurotransmitters related to LAS are known to be serotonin and gamma-aminobutyric acid (GABA). The loss of serotonin within the inferior olive nucleus has been thought to play a certain role and GABA may interact with the serotonin system to suppress PHM.

The treatment of LAS has not been established and a combination of medications based on the neurotransmitters has been reported. Studies have found that valproate, piracetam, levetiracetam, zonisamide, clonazepam, etc. are effective.

In this case, the patient was treated with clonazepam and levetiracetam, which were effective in controlling the PHM. Failure to recognize LAS may result in inappropriate anticonvulsant therapy and delayed treatment.

Therefore, when a patient develops uncontrolled myoclonus on regaining consciousness after CPR and the myoclonus is unsuccessfully treated with traditional anticonvulsants for a certain period, the possibility of LAS should be considered. This can lead to minimizing the patient's disabilities and improving the prognosis.

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