

Case Report

A rare case report of Lance-Adams Syndrome

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Abstract

Lance-Adams syndrome or chronic post-hypoxic myoclonus, is a rare complication of post-cardiopulmonary resuscitation, accompanied by action myoclonus and cerebellar ataxia. It manifests as diffuse action-triggered jerking movements resulting in significant morbidity. A case of Lance-Adams syndrome is presented here in a 60-year-old farmer admitted to the intensive care unit with history of cobra bite, in coma and respiratory failure, revived elsewhere. His EEG and CT brain was unremarkable. This patient's myoclonus was poorly controlled by a variety of anti-epileptic medications. Over 75 days of rehabilitation there was improvement in ambulation and there were no cognitive deficits.

Keywords: Lance-Adams syndrome (LAS), myoclonic jerks, chronic posthypoxic myoclonus.

1. Introduction

With improvements in emergency medical services, the number of survivors of any hypoxic event (respiratory or cardiac) damaging brain has increased. These survivors may develop neurological complications, such as epilepsy, memory disturbances, depression, parkinsonism, dystonia, chorea, athetosis, tremor, akinetic-rigid syndromes and chronic post hypoxic myoclonus (PHM).¹ The latter is also known as Lance-Adams syndrome.² This is predominantly characterized by action myoclonus starting in days to weeks after cardiorespiratory resuscitation (CPR) in survivors. Here we present one case of LAS in a patient during snake bite management.

2. Case report

A 60 year old farmer was brought to emergency room on 4-9-2012, with a reference letter of cardiac resuscitation. 6 hours earlier he was bitten by a cobra over foot in his field, which was initially followed by breathing difficulty and blurring of vision in about 30 min. He was given one antsnake venom injection in a local hospital, was intubated endotracheally before shifting to our hospital. There was no history of nausea, vomiting, paraesthesia, dizziness, sweating, diplopia, weakness of limbs and bleeding manifestations. There was no relevant past medical history and patient had no ill habits.

On examination the patient was moderately built, underweight, unconscious, severe pallor and koilonychia was present. Pulse 90/min. regular, normal volume, blood pressure 160/120 mm Hg, respiratory rate 40/min, shallow, foot had 2 fang marks on dorsum with bleb, purple discoloration, and no bleeding. Cardiac and abdomen examination was unremarkable. Respiratory System showed extensive fine crepitations. Neurologically, Glasgow Coma Score (GCS) 3/15, pupils 4 mm sluggishly reactive to light, generalised hypotonia, hyporeflexia, no fasciculations, power 0/5 and mute plantars.

On arrival, patient had an episode of generalised tonic clonic seizures lasted for 1 to 2 min. for which intravenous diazepam was given. He was paralysed with vecuronium and put on ventilator support. Urinary catheterization revealed frank blood mixed with clots.

Detailed lab investigations included Prothrombin time: 23.3 sec, control: 13.5 sec, INR: 2.08, APTT: 27 sec, control-33.8 sec, Haemoglobin-6.6 gm% TWBC: 19,200 cells/mm³,

DC: N 92%, L 7%, E 1%, platelets: 2L/mm³, ESR 105 mm/hr. peripheral blood smear: dimorphic anemia, blood urea: 17 mg/dl, serum creatinine: 0.82 mg/dl, RBS-120 mg/dl and HIV-non reactive. Electrocardiogram, chest x-ray, renal function test, liver function test, serum electrolytes, arterial blood gas analysis and echocardiogram reports were normal.

A provisional diagnosis of snakebite with severe neurotoxicity, haematotoxicity, respiratory failure and severe anemia was made.

Fig-1: CT brain- normal

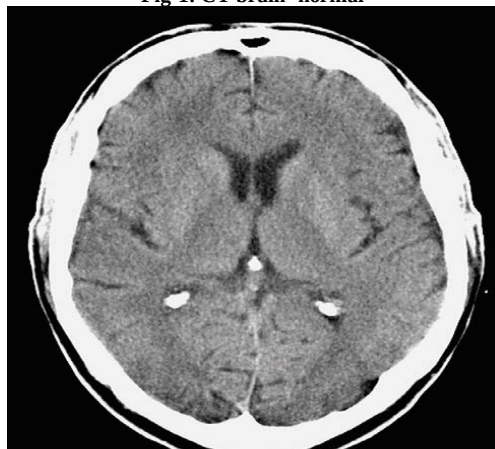
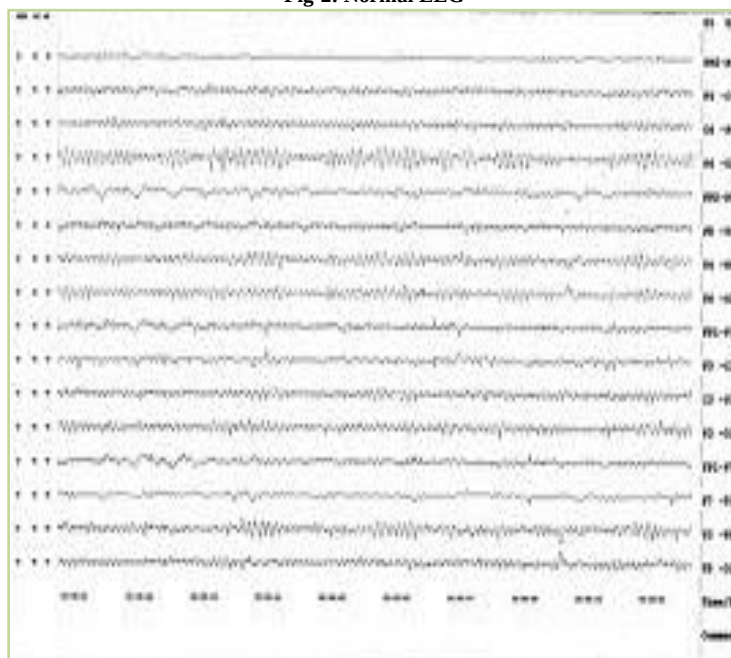


Fig-2: Normal EEG



2.1 Management

He was treated with antsnake venom, fresh frozen plasma, packed red cells, broad spectrum antibiotics, tetanus toxoid, mannitol, inj. phenytoin, metoprolol, supportive measures like intravenous fluids, Ryle's tube feeds, inj. Vitamin B₁₂, bladder catheterization and wound debridement.

3. Results

Patient had multiple ventricular ectopic, self-limited, without hemodynamic disturbance. During next 36 hours patient continued to be comatose GCS-3/15, (not sedated), severe myoclonic jerks appeared involving face, tongue, neck and limbs. Brain stem reflexes were intact (pupillary reflex, deglutition reflex, cough reflex). Clonazepam (0.5mg twice daily), sodium valproate (1.5gm/day in divided doses), infusion of propofol and midazolam temporarily were useful. Serial coagulation profile improved, haemoglobin level improved, platelets and renal function were normal. CT brain was normal.

Course in the hospital- 5 days later brain stem reflexes were found to be preserved, generalised hypotonia, areflexic, unprovoked myoclonic jerks persisted, these were severe whenever patient voluntarily opened eyes and moved limbs. Ventilator support was stopped on 12th day as the patient had adequate respiratory efforts and GCS was 6/15. Temperature was 99-100°F, profuse sweating was noted in between the episodes. Myoclonic jerks varied from time to time, completely absent for 1-2 days and often continuously present during awake hours. His sensorium improved, he gestured of difficulty in swallowing, esophagogastroduodenoscopy showed normal mucosa. Adding phenobarbitone (120mg/day) and piracetam (400mg thrice daily) helped him little. Rehabilitation started with passive limb exercises, ambulation and oral feeds. His blood pressure was high throughout hospital stay and needed antihypertensive drug. Patient started walking with support, over 75 days; he was able to swallow liquids and dysarthria improved. At the time of discharge MMSE score was 23, there were no sensory deficits, bowel and bladder function was normal, clonazepam, sodium valproate, phenobarbitone, piracetam and phenytoin sodium continued.

4. Discussion

Initially, post-hypoxic myoclonic status epilepticus (brainstem origin-and coma is a pre-requisite for the diagnosis), was thought of, since the patient was in deep coma. As he recovered, Lance Adams syndrome (LAS) was considered. The syndrome of intention or action myoclonus as a sequel to hypoxic encephalopathy was originally described by Lance JW and Adams RD in 1963, in the course of primary respiratory failure.³ It is characterized mainly by action myoclonus, associated cerebellar ataxia and very mild intellectual deficit. It was described in four patients after surviving cardiac arrest. These patients initially developed a generalized myoclonus accompanied by dysmetria, dysarthria, and ataxia. Over time, the myoclonus persisted but its character changed to a predominantly action myoclonus involving the limbs. It was hypothesized that this particular constellation of symptoms was due to cerebral hypoxia. Dysfunction of various parts of the central nervous system, including the basal ganglia, thalamus, midbrain, and cerebellum, is implicated in the pathogenesis. The neurotransmitters related to LAS are known to be serotonin and gamma-aminobutyric acid (GABA). Purkinje cells of cerebellum probably being susceptible to anoxic injury, may play a key role.⁴ Purkinje cells are deficient in aldolase C and *Excitatory amino-acid transporter 4* (EAAT4) that allow them to survive pathologically intense synaptic input from the inferior olive after the restoration of blood flow, they have reduced capability to sequester glutamate and reduced ability to generate energy during anoxia. LAS has diverse clinical, electrophysiological, and neurochemical abnormalities and 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 posthypoxic myoclonus.^{3,5}

This patient's myoclonus was poorly controlled by a variety of anti-epileptic medications, i.e. valproate sodium, piracetam, clonazepam, phenobarbitone and phenytoin sodium. Levetiracetam is said to be useful in patients with chronic myoclonus.^{6,7,8} Cranial single photon emission computed tomography (SPECT) and, 18F-fluorodeoxyglucose positron emission tomographic (PET) scan, cranial magnetic resonance spectroscopy (MRS) may show perfusion abnormalities (these tests were not available in our institute).

The probable factors that promoted susceptibility to the development of posthypoxic myoclonus in our patient, were severe anemia, old age, under nutrition and unrecognized hypertension, though snake bite complications were managed adequately.⁹ Over 75 days of rehabilitation there was improvement in ambulation. Deep brain stimulation is tried in severe PHM to interrupt connections of ventrolateral thalamus and pontine tegmentum.¹⁰

5. Conclusion

Worldwide so far about 150 cases of LAS are reported, in various causes like hanging³, myocardial infarction¹¹, bronchial asthma^{12,13,14}, and post surgical recovery.¹⁵ Early recognition and rehabilitation carries long term good prognosis.¹⁶

On thorough search we could not get any case report of LAS from India in journals and we presume that this is the first LAS case report from India.

References

1. Neurology in Clinical Practice, Principles of Diagnosis and Management, volume1, 4th edition,2004,Walter G Bradley, Robert B. Daroff, Gerald M. Fenichel, Joseph Jankovic. pp.1668.
2. Lance JW, Adams RD. The syndrome of intention or action myoclonus as a sequel to hypoxic encephalopathy. *Brain* 1963; 86:111–36.
3. Lee HL, Lee JK. Lance-Adams Syndrome. *Ann Rehabil Med* 2011; 35:939–43.
4. Welsh JP, Yuen G, Placantonakis DG, Vu TQ, Haiss F, O'Hearn E *et al*. Why do Purkinje cells die so easily after global brain ischemia? Aldolase C, EAAT4, and the cerebellar contribution to posthypoxic myoclonus. *Adv Neurol*. 2002; 89:331-59.
5. Welsh JP, Placantonakis DG, Warsetsky SI, Marquez RG, Bernstein L, Aicher SA. The serotonin hypothesis of myoclonus from the perspective of neuronal rhythmicity. *Adv Neurol* 2002; 89:307–29.
6. Frucht SJ, Louis ED, Chuang C, Fahn S. A pilot tolerability and efficacy study of levetiracetam in patients with chronicmyoclonus. *Neurology* 2001; 57:1112-4.
7. Lim LL, Ahmed A. Limited efficacy of levetiracetam on myoclonus of different etiologies. *Parkinsonism Relat Disord*.2005; 11:135-7.
8. Krauss GL, Bergin A, Kramer RE, Cho YW, Reich. Suppression of post-hypoxic and post-encephalitic myoclonus with levetiracetam. *SG Neurology*. 2001; 56:411-2.
9. Goldfrank'stoxicologic emergencies, 8th edition, 2006, Flomenbaum, Goldfrank, Hoffman, howland, Lewin, Nelson. pp. 1643-1654.
10. Frucht SJ, Trost M, Ma Y, Eidelberg D. The metabolic topography of posthypoxic myoclonus. *Neurology* 2004; 62:1879-81.
11. Polesin A, Stern M. Post-anoxic myoclonus: a case presentation and review of management in the rehabilitation setting. *Brain Inj*. 2006; 20:213-7.
12. Werhahn KJ, Brown P, Thompson PD, Marsden CD. Lance-Adams syndrome: a report of two cases. *Mov Disord*. 1997; 12:216-20.
13. Kowalczyk EE, Koszewicz MA, Budrewicz SP, Podemski R, Slotwinski K. Lance-Adams syndrome in patient with anoxic encephalopathy in the course of bronchial asthma. *WiadLek*. 2006; 59:560–62.
14. Werhahn KJ, Brown P, Thompson PD, Marsden CD. The clinical features and prognosis of chronic posthypoxic myoclonus. *Mov Disord*. 1997; 12:216-20.
15. Yan-xing Zhang *et al*. Lance-Adams syndrome: a report of two cases. *J Zhejiang Univ Sci B*. 2007; 8: 715–720.
16. Polesin A, Stern M. Post-anoxic myoclonus: A case presentation and review of management in the rehabilitation setting. *Brain Inj*. 2006; 20:213–17.