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Lance-Adams Syndrome (LAS). chronic post-hypoxic myoclonus in consultation-liaison psychiatry: Case report

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CASE STUDY

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ABSTRACT

Post-hypoxic myoclonus is a rare finding after cardiorespiratory arrest. Clinical Case: We present a case of a 40-year-old woman with toxic neuropathy due to hyperthyroidism. She was under prolonged resuscitation due to a neural cardiorespiratory attack. Four days later, she developed rhythmic and involuntary movements of high frequency and low amplitude. Video-electroencephalographic monitoring was normal. Cerebral NMR showed extensive and diffuse hyperintensity in multiple encephalon areas and microhemorrhages in both cerebral hemispheres. She was managed with valproic acid. The treatment of this condition must be multidisciplinary; it includes medication (valproic acid, levetiracetam, and clonazepam), physical and occupational therapy with close collaborative management alongside the neurology service.

Key Words
Lance Adams Syndrome, Hypoxia, Brain, Myoclonus, Neuropsychiatry, Cardiopulmonary resuscitation, Consultation-Liaison Psychiatry

Implications for Practice

Myoclonus is a rare finding after heart attacks; its prevalence is about 1.5 per cent. Fewer than 150 cases have been reported in the literature. It has been hypothesized that transient cerebral hypoxia causes permanent synaptic modification of motor neuronal networks. It is an entity that is underdiagnosed by most doctors with the subsequent delay in treatment. Treatment and diagnosis must be multidisciplinary with close collaborative management alongside Neurology and Consultation-Liaison Psychiatry.

Background

Myoclonus is defined as a sudden muscle contraction or lapse in muscle tone. They are brief, involuntary, and shock-like events.[1,2] Myoclonus is a descriptive term for heterogeneous phenomena. Most myoclonic cramps result from a short burst of muscular activity, resulting in positive myoclonus.[3] Myoclonus is a rare finding that occurs after heart attacks; its prevalence is about 1.5 per cent.[4,5] Fewer than 150 cases have been reported in the literature.[6] Two types of myoclonus have been described: Post hypoxic acute or epileptic myoclonus and Lance Adams Syndrome (LAS) or chronic post-hypoxic myoclonus. Acute Post-hypoxic myoclonus, termed post-hypoxic myoclonic status epilepticus (MSE), occurs soon after a hypoxic insult in patients who are profoundly comatose, and it implies a poor prognosis. On the other hand, LAS begins days or weeks after cardiopulmonary resuscitation (CPR) and it may persist over time in patients who have recovered consciousness.[3,7,8] We present the clinical case of a patient that was diagnosed with LAS in the Consultation-Liaison Psychiatric service of our general hospital.

Case Details

The patient is a 40-year-old female, originally from the urban area of Amazonas (Brazilian-Colombian Border), who

had a loss of strength, most likely because of toxic neuropathy by hyperthyroidism. She had a cardiorespiratory attack and received prolonged cardiopulmonary resuscitation. She, just four days later, developed rhythmic and involuntary movements of both high frequency and low amplitude in the lower facial region, as well as in the right limbs (both upper and lower.) The electroencephalographic layout presented discrete slow activity in the frontocentral bilateral regions and discharged over the sensorimotor cortical areas (Figure 1). Video-electroencephalographic monitoring was normal. Cerebral NMR showed extensive and diffuse hyperintensity in the cerebellar cortex, the posterior arm of the internal capsules, thalamus, hippocampus, mesial cortex of the occipital lobes, and multiple areas of white subcortical substance, as well as microhemorrhages in both cerebral hemispheres (Figure 2). The liaison psychiatry service diagnosed LAS; she was treated with valproic acid (19mg/kg/day.) We also implemented this management in our Psychiatric Consultation Service. The patient had a slow motor recovery in the first year of follow-ups.

Discussion

LAS were initially described as inattention or action myoclonus in 1963 by Lance and Adams.[3] The myoclonic jerks are triggered explicitly by an action, startle, or a stimulus such as touch, tracheal suctioning, and loud handclaps. They usually disappear with body and limb relaxation or with sleep. LAS occur in patients after they have regained consciousness, specifically days to weeks after CPR. LAS diagnosis is clinical, and a careful history, including the anoxic event and detailed physical assessment, should be done.[5] Possible anatomical areas involved for LAS remain unknown. Diffuse damage and neuronal loss in the thalamus, striatum, mammillary bodies, and brainstem raphe nuclei have been described.[1] It is believed that transient cerebral hypoxia provoked a permanent synaptic rearrangement of the neuronal networks involved in the pathogenesis of LAS.[9] Nevertheless, LAS can reach a functional outcome with appropriate treatment.[4]

Notably, it is a typical presentation of LAS. However, the diagnosis was delayed until Consultant-Liaison psychiatry service assessment, done in order to rule out a possible functional motor disorder. As a result, there were no unusual or atypical neurological symptoms in this patient. Other clinical entities, including functional or psychogenic movement disorders, were ruled out. Herein lays the academic importance of this clinical case. Interdisciplinary work and collaborative search for early and differential diagnoses will improve the prognosis of this condition.

Functional (psychogenic) myoclonus (FM), serotonin syndrome, parkinsonism, dystonia, chorea, tics, athetosis, tremor, prion diseases, celiac disease, and other toxic-metabolic disorders as well as epilepsy, alcohol abuse /withdrawal must be ruled out. FM and SS are of particular interest In Consultant- Liaison Psychiatry. Functional or psychogenic movement disorders are common and disabling, and sometimes challenging to diagnose. FM accounts for 5–20 per cent of active movement.[2]

Conclusion

LAS begin a few days after hypoxic brain damage. The diagnosis of LAS is clinical. It is typically manifested as myoclonus of action, predominantly of the extremities, with varying degrees of disability and a preserved cognitive function. Treatment includes medication (valproic acid, levetiracetam, clonazepam, among others) and physical and occupational therapy with close collaborative management alongside Neurology and Consultation-Liaison Psychiatry. Consultation-Liaison Psychiatrists play an essential role in diagnosing (early and differential) and guiding neurologists toward proper treatment.

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PEER REVIEW

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CONFLICTS OF INTEREST

The authors state that they have no competing interests.

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PATIENT CONSENT

The authors, Bejarano-Pedraza EM, Barrera-Forero S, Gutiérrez-Segura JC, and Oviedo-Lugo GF, state that:

1. They have obtained written, informed consent for the publication of the details regarding the patient(s) mentioned in this report.
2. All possible steps have been taken to safeguard the identity of the patient(s).
3. This submission complies with the requirements of local research ethics committees.

Figures

Figure 1: (a) Slow activity in frontocentral bilateral regions. (b) Discharge over the sensorimotor cortical areas.

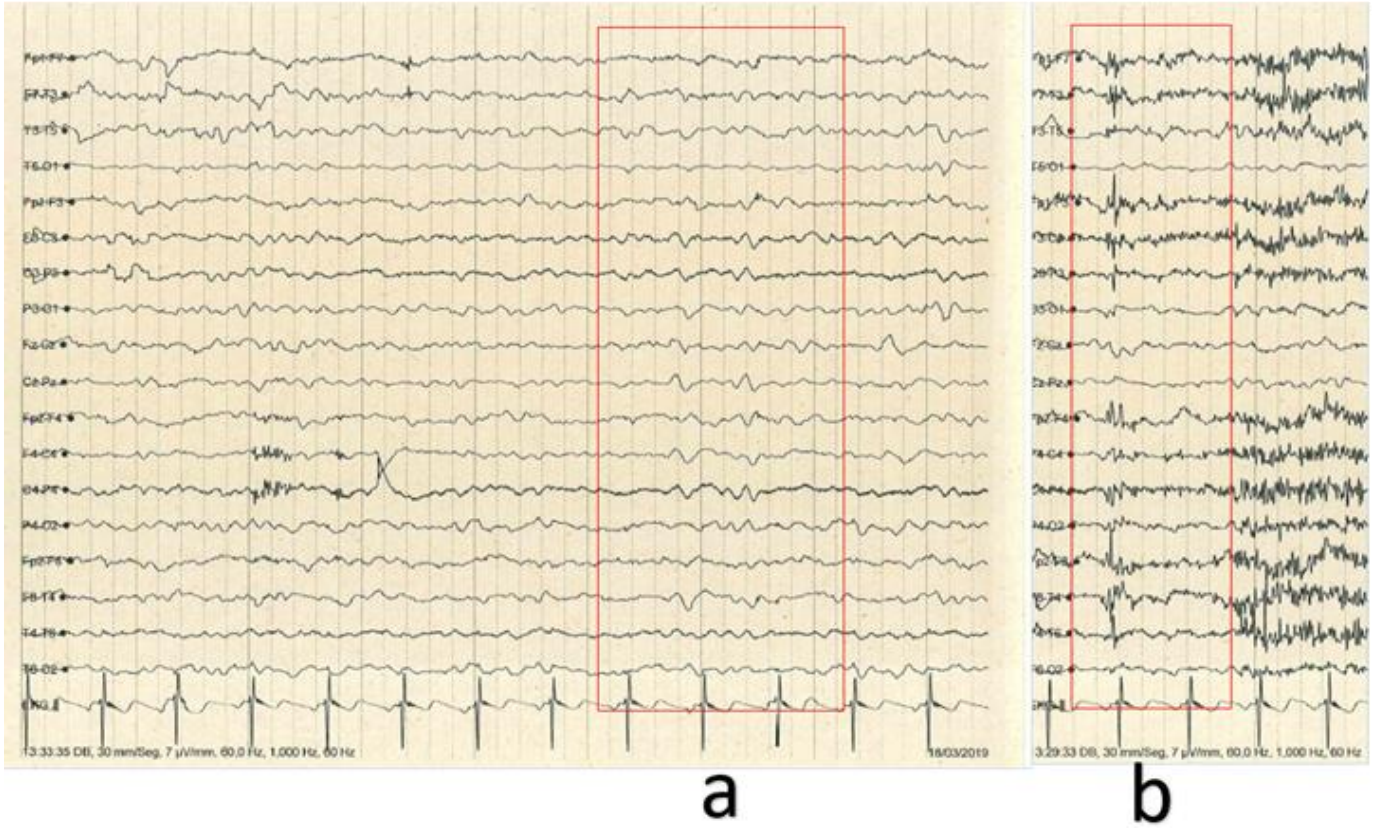

Figure 2: Extensive and diffuse hyperintensity in the cerebellar cortex; the posterior arm of the internal capsules, thalamus, hippocampus, and the mesial cortex of the occipital lobes, as well as multiple areas of white subcortical. In addition, there are microhemorrhages in both cerebral hemispheres.