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CASE REPORT

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Lance-Adams Syndrome: A Rare Complication Following Cardiac Arrest

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ABSTRACT

Lance-Adams syndrome (LAS) is a rare complication occurring days or weeks following a successful cardiopulmonary resuscitation in which patients develop myoclonus, typically triggered by intentional actions or external stimuli. It can be challenging and confusing to distinguish LAS from acute post-hypoxic status epilepticus, in which patients remain in vegetative or comatose state. Here we report a case of a 71-year-old survivor of cardiorespiratory arrest due to cardiogenic shock with typical features of LAS. Although the pathophysiology of LAS is poorly understood, antiepileptic medications are effective in controlling the myoclonus. Early identification of LAS and distinguishing it from myoclonic status is necessary to start appropriate treatment and provide prognosis.

Keywords: Action myoclonus, Cardiorespiratory Arrest, Lance-Adams syndrome, post-hypoxic myoclonus

INTRODUCTION

The survival rate of out-of-hospital cardiac arrest who received cardiopulmonary resuscitation (CPR) is 12% while for in-hospital cardiac arrest is 24%.[1] Survivors are susceptible to ischemic anoxic encephalopathy and may present with post-hypoxic myoclonus (PHM).[2] It is a rare complication of successful CPR. PHM are of two types: Acute and Chronic. Acute PHM occurs within 24 to 48 hours after cardiopulmonary arrest, the patient remains comatose and has a poor prognosis.[2] Chronic PHM also known as Lance-Adams Syndrome (LAS) was first described in 1960s by Lance and Adams. They described four cases in which patients had involuntary movement disorder after recovering from cardiac arrest.[3] The myoclonus in LAS occurs days to weeks after CPR and the patient regains consciousness and has better prognosis. The myoclonus is triggered by external stimuli and is relieved by rest or sleep.[4,5] Proper distinction between myoclonic status and LAS is important for starting appropriate treatment early and for predicting outcomes.[6,7] Here we present

a case of an older male of 71 years of age with typical features of LAS who developed the myoclonus post-CPR after cardiogenic shock.

CASE REPORT

An older male aged 71 years presented to our casualty with complaints of repetitive, irregular jerky movement of bilateral upper and lower limbs.

The patient is a known case of hypertension under regular medication (Metoprolol 50 mg extended-release formulation once a day) as prescribed by the physician. He had previously presented to another health care center with complaints of epigastric pain and generalized weakness where he suddenly collapsed and was unresponsive. His blood pressure was not recordable. His electrocardiogram (ECG) showed supraventricular tachycardia (ECG unavailable) and he was suspected to have cardiogenic shock. Prompt cardiopulmonary resuscitation (CPR) was started. After 15 minutes of CPR, his vital signs returned (blood pressure of 80/60 mm of Hg). The patient was intubated and was kept under noradrenaline support. The patient developed two episodes of ventricular tachycardia on the 2nd day which was managed with verapamil and lidocaine and had one episode of atrial fibrillation managed with digoxin (doses not mentioned in record from another center). After he was extubated on the 4th day, the patient developed abnormal jerky movements continuously over bilateral upper and lower limbs which aggravated while performing a specific task for which he was started on levetiracetam 500mg twice a day, trihexyphenidyl 2mg thrice a day and clonazepam 0.5 mg once a day. His tremors persisted and he was referred to our center. On presentation, the patient was obeying commands and was verbally oriented. He was wheelchair bound. His pupils were equally reactive to light and vitals were stable. His muscles of all extremities had a Medical Research Council grade of 4. The findings of the other neurological examinations were not specific. The cerebellar function could not be evaluated due to continuous myoclonic jerks. Computed Tomography (CT) of his brain failed to reveal any incidence of acute intracranial hemorrhage, mass effect or infarct (Figure 1). Magnetic Resonance Imaging (MRI) of brain was done to detect hypoxic brain injury; however, it showed no focal lesion except arachnoid cyst present in retrocerebellar region. Electroencephalogram (EEG) was performed to rule out true seizure which showed no significant anomaly (Figure 2). His hematological and biochemical investigations were within normal limits. He was managed conservatively with trihexyphenidyl 2mg thrice a day and clonazepam 0.25 mg twice a day.

His cardiac markers were within normal limits, but his echocardiogram showed severe septal hypertrophy (2.5cm) with left ventricular ejection fraction of 60%. He was diagnosed to have asymmetrical hypertrophic cardiomyopathy and was managed with metoprolol 50mg and furosemide 20mg once a day.

Even after 48hrs of starting treatment, he had persistent myoclonus though they were less severe. So, sodium valproate 500mg twice a day was added, and dosage of clonazepam was increased to 0.5mg in the morning and 1mg at night. This medical regime significantly reduced his jerks, both in severity and frequency. His condition was stable, and vitals were within normal limits. The patient was shifted from intensive care unit to a high dependency unit. He was started on rehabilitative physiotherapy focused on reducing the myoclonic jerks and improving his limb power. During discharge, the myoclonus had decreased but were aggravated by voluntary, coordinated action. He could perform tasks without much assistance, but the myoclonus escalated when he was asked to perform a task within a timeframe. Therefore, to control his jerks, he performed tasks slowly. At 1-month follow-up, the patient was walking by himself with support, he had no myoclonus at rest and only occurred while performing any precise voluntary coordinated action.

DISCUSSION

Early identification of PHS and subsequently distinguishing LAS from myoclonic status can be

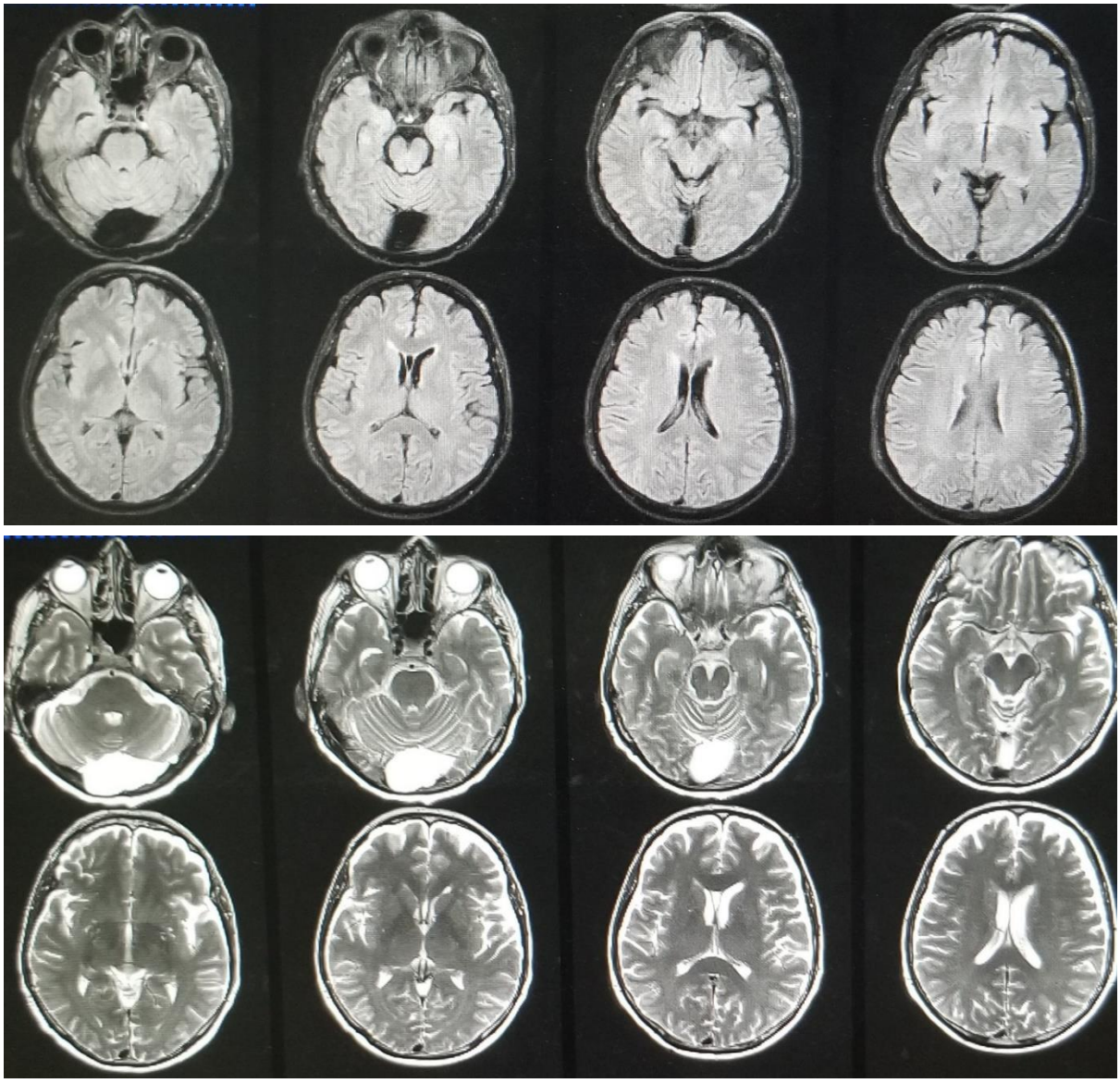

Figure 1. Computer Tomography images showing retro cerebellar arachnoid cyst

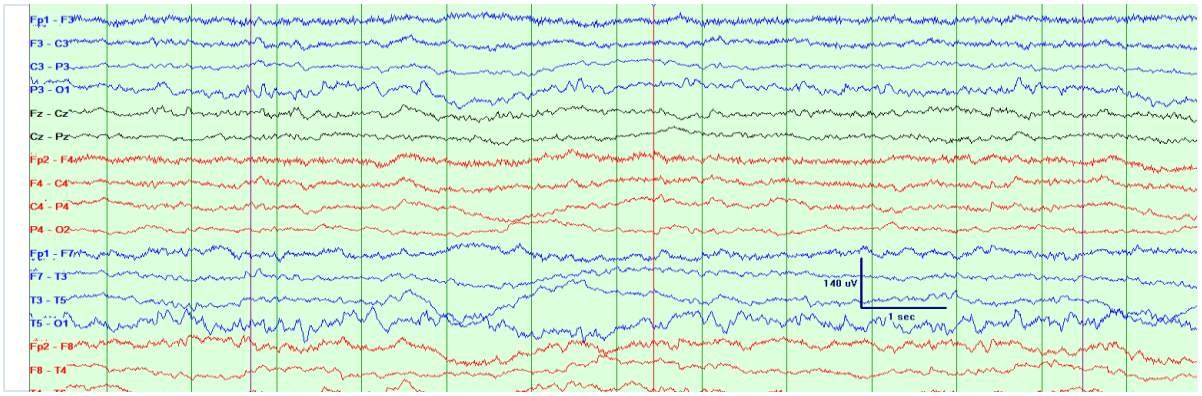

Figure 2. Electroencephalogram showing no significant anomaly

challenging and confusing due to the rarity of this post-CPR complication. The patient in this case had remarkable history and clinical features that were consistent with LAS. The myoclonus started

three days after the patient received CPR and regained consciousness. These myoclonic jerks were triggered voluntary coordinated movements and disappeared during rest and sleep. However, all patients do not meet all the widely accepted criteria to differentiate the two types of PHS. Freund has revealed that 20% of PHM within the first 48 hours after injury could be diagnosed as LAS.8 Reports of myoclonus beginning as soon as the patient was weaned off sedation, suggests that sedation may delay PHM onset and hence, hinder proper diagnosis.[5]

The pathophysiology of PHM is not clear.[9] The levels of serotonin and gamma-aminobutyric acid (GABA) in the inferior olive nucleus are thought to play a role in the pathophysiology of LAS.[10,11] GABA interacts with the serotonin level to keep the post-hypoxic myoclonus suppressed and the loss of serotonin and death of Purkinje cells after anoxia causes the enhanced motor excitability.[12] Investigations like EEG and neuroimaging studies are not consistent and varies with each case. However, most LAS patients show epileptiform waves.[4] Elmer et al (2016) identified two distinct patterns of EEG to distinguish LAS and acute PHM; vertex spike-wave complexes in LAS and suppression-burst background with high amplitude spikes in myoclonic status.[13] Although non-conforming, the EEG of this case showed diffuse theta to delta slowing which is in contrast to previous reports.[3,4,13]

In the present case, both CT and MRI showed no significant abnormality which is in line with other reports which have shown widely variable or insignificant neuroimaging findings.[5,8] However, various studies with newer imaging modalities like Brain SPECT have been reported. A study by Frucht et al (2004) showed that patients with LAS had significantly higher glucose metabolism in the pontine tegmentum, mesencephalon, and ventrolateral thalamus.[14] Another report by Zhang et al. showed mild hypoperfusion in the left temporal lobe in a patient with LAS.[15]

There are no definite treatment guidelines for LAS and medications are prescribed empirically. A study of 100 patients treated with clonazepam, valproate and piracetam showed 50% efficacy while another study has recommended levetiracetam, zonisamide, clonazepam, and valproate as the first line of treatment.[16,17] Intrathecal baclofen has been shown to improve myoclonus in a patient refractory to multiple medications.[18] In our case, sodium valproate and clonazepam were used which lead to control of the myoclonus.

CONCLUSION

LAS remains to be ruled-out in patients regaining consciousness following myoclonus after CPR. It is partly due to the rare presentation of this syndrome and difficulties in distinguishing it from acute myoclonus. However, considering LAS as a differential in survivors of cardiac arrest with movement disorder, can help deliver prompt diagnosis, reduce inappropriate medications, start accurate treatment, and makes prognostication easier.

COMPETING INTEREST

The authors declare that there are no competing interests regarding the publication of this paper.

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