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RESEARCH ARTICLE

POST-HYPOXIC BRAIN INJURY (LANCE ADAMS SYNDROME); A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Action myoclonus is a rare complication that may follow after a successful cardiac resuscitation event. Lance Adams Syndrome, also known as chronic post-hypoxic myoclonus is an action myoclonus seen in patients who have regained consciousness after undergoing a cardiopulmonary arrest. It develops after days to weeks following the event. There are less than 155 reported cases in the world. Here we present the case of an 18 year old female who had complains of jerky movements in her upper limbs bilaterally after suffering two episodes of cardiac arrests. This case was diagnosed as Lance Adams Syndrome taking into accord a detailed history of hypoxic brain injury, clinical presentation, and appropriate neuroimaging studies. A correct and early diagnosis is essential and directly related to a positively related functional outcome of the patient. Recognition of post-hypoxic myoclonus depends on the awareness that such a syndrome exists, and the patients' outcome relies on early diagnosis and appropriate management.

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INTRODUCTION

Post-hypoxic myoclonus (PHM) is a neurological complication that rarely occurs after a successful cardiopulmonary resuscitation (CPR). It is characterized by uncontrolled myoclonic jerks which may be associated with cerebellar ataxia. PHM can be subdivided into two categories, acute and chronic. Acute PHM occurs within 12 hours of the hypoxic brain injury. Acute PHM patients are usually in a deep comatose state which implies poor prognosis. Chronic PHM on the other hand, (also known as LAS), is identified by a myoclonus that appears in a patient after a successful CPR within a few days to weeks (English *et al.,* 2009; Lance and Adams, 1963). Chronic PHM patients have regained consciousness. Here we present the case of a young female who suffered with bilateral jerky movements of her upper limbs after she survived 2 separate events of cardiac arrest spaced almost a year apart.

CASE REPORT

An 18 year old female presented to the hospital with three days history of repetitive, irregular brief jerks in both her face and upper limbs bilaterally.

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These jerks were present throughout most of her day, without associated frothing, tongue biting or urinary incontinence. The jerks had progressively increased in intensity and frequency for the past 24 hours which had prompted her to visit the emergency department. She had a previous history of two cardiac arrests in 2012 and 2014, each followed by successful CPR. In her childhood, she had a history of episodes of sudden loss of consciousness during periods of emotional stress. Following each episode, she always regained consciousness in less than a minute. In April 2012, she was sitting with her family when she had an acute loss of consciousness. Her parents tried to revive her by splashing water onto her face. With no relief evident, she was driven to the hospital by her parents. She remained unconscious for a total of 10 minutes before cardiopulmonary resuscitation efforts were initiated in the ED. She was intubated after a 10 minute successful CPR. She remained on the ventilator for 4 days before she was extubated, and was subsequently discharged on LAMICITAL (100mg twice daily), in a vitally stable condition in the following days.

She suffered her second cardiac arrest 2 years later in March 2014. She was in a hotel room and was emotionally distressed due to family matters and lost consciousness. Hotel management was called and the CPR initiated after attempts at revival with water splashing by her family failed. The time from losing consciousness till the initiation of CPR during this

cardiac arrest was approximately 15 minutes. Emergency medical services were called and took over the resuscitation from the hotel employees. After a successful CPR that lasted almost 20 minutes, the patient was intubated and taken to the hospital. She was administered Levophed (norepinephrine) and amiodarone during resuscitation attempts by the EMS. The patient had ventricular tachycardia upon arrival in the ED. She was cardioverted twice before her return to a normal cardiac rhythm. She was fitted with an Automatic Implantable Cardiac Device (AICD) after initial workup revealed a prolonged QT syndrome. She was loaded with phosphenytoin (1000mg) and kept under sedation with midazolam and fentanyl. Electroencephalogram (EEG) revealed diffuse slowing of all brain waves secondary to sedation, along with some sharply contoured activity originating in the paracentral and right parietofrontal area, suggestive of cortical irritative seizure activity. Her echocardiogram (ECHO) showed a Left Ventricular Ejection Fraction of 30%, presumably secondary to severe metabolic derangements. Computed Tomography (CT) of her brain failed to reveal any incidence of acute intracranial hemorrhage, mass effect or infarct. The patient was kept in phenobarbitol induced coma for 4 weeks. Her hospital course was complicated by Abdominal compartment syndrome, upper extremity deep venous thrombosis (DVT) secondary to a peripherally inserted central catheter (PICC line), and clostridium difficle infection. Upon extubation, she was well oriented to time, place and person but had some residual speech impediment and left hemiparesis. Physiotherapy was initiated and her motor dysfunction gradually improved. A few weeks later she developed jerky movements, generalized in nature, occurring intermittently once an hour and lasting for almost 30 seconds. The progressively increasing myoclonus and hemiparesis made it impossible for her to stay a seat, ambulate, or perform simple daily tasks without assistance. These myoclonic jerks were triggered by emotional stress and voluntary coordinated movements, and disappeared upon mental relaxation, abatement of mental or emotional stress, and sleep. Magnetic resonance imaging (MRI) of her brain showed a subtle and nonspecific symmetric increase in signals in her basal ganglia, principally, in her posterior putamen, suggestive of hypoxic injury. The patient was started on Lamotrigine (150mg, once daily), Gabapentin (300mg, trice daily), Levetiracetam (500mg twice daily), Lorazepam, and Duloxetine. This medical regime significantly reduced her jerks. Regular physiotherapy and rehabilitation improved the hemiparesis and the patient was able to walk and perform daily activities without any assistance.

DISCUSSION

Myoclonus is described as sudden contraction of a muscle or a group of muscles, resulting in an involuntary body movement known as a myoclonic jerk or twitch. Myoclonic jerks may present with a particular pattern, i.e. focal, multifocal or they may be generalized.

Post-Hypoxic Myoclonus (PHM) is a forms have a myoclonus that occurs after hypoxic brain injury, and it may be acute or chronic. It is important to accurately differentiate between the acute and chronic form, as both forms have a different prognosis. Acute PHM occurs within 24-48 hours of the hypoxic brain injury and carries a poor prognosis. Chronic PHM, also known as Lance Adams Syndrome, can occur days to weeks after the hypoxic brain injury, and carries a good prognosis. It was first described in the 1960s by Lance & Adams, who described the cases of 4 patients that experienced a form of myoclonus (known as action or intention myoclonus) within a few days following an episode of anoxia. Action or intention myoclonus occurs when an attempt is made to touch a target with the index finger, and the motion is interrupted along the line of movement by side-to-side oscillations that increase towards the end of the movement. Action myoclonus is specific to LAS, whereas patients with acute PHM usually have generalized multifocal myoclonus. Another important differentiating clinical feature is consciousness; patients with LAS regain consciousness and are aware, whereas patients with acute PHM remain comatose. It is important to note that heavy sedation can mask these effects in LAS patients. The patient in this case had remarkable clinical features that are consistent with LAS. The pathophysiology of LAS is not clear, but it has been stated that it is associated with neurochemical abnormalities, particularly involving the neurotransmitters serotonin and GABA. In LAS there is a loss of serotonin within the inferior olive, and this is thought to play an important role in the pathogenesis of LAS. GABA is also thought to be involved in the pathogenesis, as it interacts with the serotonin system and suppresses the PHM. The loss of GABAnergic inhibition due to ischemia can lead to enhanced motor excitability and myocolonus. The lack of clear understanding about the pathophysiology of LAS has limited the treatment options. The current treatment regime is based on the neurotransmitter hypothesis, and often a combination of drugs is required. Frucht and Fahn reviewed more than 100 patients with LAS and found that clonazepam, sodium valporate, and piracetam were significantly effective in treating approximately 50% of the cases. Recent studies also show that levetiracetam is effective, and it may be used in combination with clonazepam, sodium valporate, and piracetam as a first-line agent.

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