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Essay

Lance-Adams Syndrome: A Case Report

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Introduction

Hypoxic injury to the brain may cause unexplained jerking in some patients, a phenomenon dubbed post-hypoxic myoclonus (PHM). This condition may be divided into acute or chronic delineations. The former generally occurs within twelve hours of hypoxic injury, often following cardiac arrest. The latter, also known as Lance-Adams Syndrome, can occur within days to months following cardiopulmonary resuscitation (CPR) and persists following recovery of consciousness.[1] With fewer than 200 reported cases since its description in 1963, we report a case of Lance-Adams Syndrome after CPR following PEA cardiac arrest.[1,2]

Case Presentation

The patient is a 60 year-old gentleman with a PMH of ESRD (on Tuesday/Thursday/Saturday dialysis since 2012), hypertension, type II diabetes mellitus, hyperlipidemia, hypothyroidism, sleep apnea, and peripheral vascular disease. EMS was called after the patient complained of shortness of breath and collapsed. Shortly after the paramedics arrived, the patient went into witnessed PEA arrest. CPR was immediately initiated. After 12 minutes of chest compressions and 1 mg of epinephrine, the patient had return of spontaneous circulation. He was intubated and transferred to the ED.

Upon arrival to the ED, the patient was immediately started on the hypothermia protocol. Chest x-ray revealed diffuse pulmonary edema and bilateral pleural effusions. CTA was negative for pulmonary embolus. Initial VBG was 7.18/53/73/20 with a lactic acid of 6.0. WBC count was 16.6. A loading dose of vancomycin and cefepime were given for possible sepsis and the patient was transferred to the ICU. In the ICU, the patient was started on his scheduled hemodialysis. Continuous Renal Replacement Therapy (CRRT) was also initiated for volume overload. On day 1, rewarming was performed and sedation titrated down. On day 2, the patient was awake and responsive, but unable to follow commands. CRRT was discontinued at that time. On day 3, he continued to improve and was able to follow commands. CT revealed no acute intracranial abnormality. He was extubated on day 5, and transferred to a non-intensive care unit on day 7. He remained afebrile since day 3; leukocytosis resolved on day 6. On transfer, he was awake, alert, and oriented to person, place, and time without any residual neurologic deficit.

While on the floor, the patient continued to improve with daily PT/OT sessions, restarting of home medications, and continuation of his scheduled hemodialysis. However, on the afternoon of day 8, shortly after returning from dialysis, the primary team was called as the patient became ataxic, complaining of involuntary “twitching,” and began to slur his speech. On reexamination, the patient was found to have a resting tremor, which worsened with intention, and myoclonus in the bilateral upper extremities. The patient also had suspected

mild dysarthria, but was later determined to be dysphonia secondary to the mechanical effects of a new white, asymmetrical, loculated lesion on the tip of the tongue. An MRI was obtained which revealed no recent cerebral infarction or hemorrhage, and was significant only for an old left cerebellar infarction and chronic microvascular ischemic changes. The EEG performed was abnormal, displaying epileptiform discharges in the midline and parasagittal regions suggestive of cortical dysfunction. Given the patient's history of ESRD and exam findings akin to previous patients seen by our attending physician, aluminum levels were ordered to rule out an alternative explanation for his myoclonus. Aluminum levels returned within normal limits. On day 10, the patient was started on levetiracetam, and the following day, after three days of symptoms, was noted to have complete resolution of his myoclonus. To potentially aid in diagnosis of Lance-Adams syndrome, subsequent positron emission tomography/single photon emission computed tomography (PET/SPECT) was ordered but revealed no intracranial abnormalities. The patient continued to improve and was discharged home on day 14 with scheduled follow-up with neurology in three months.

Discussion

Chronic post-hypoxic myoclonus was first described in 1963 by Lance and Adams[2] and was thus named in their honor. In their article, they describe four patients who developed myoclonic jerks (greater than 24 hours) following brain anoxia. The jerks continued following return of consciousness and worsened with intention or external stimuli. The myoclonus disappeared at rest and with sleep.[2] LAS can often be mistaken for post-hypoxic seizures (PHS), however in the latter, the patient often remains comatose, has myoclonus that begins within 12-24 hours and ceases after 24 hours, and has abnormalities on EEG. LAS is associated with late onset myoclonus, days to months after resuscitation, and no targeted EEG abnormalities.[3] Given the presentation of intention myoclonus eight days following CPR and the resolution of symptoms with sleep, our patient fit the clinical picture of LAS more than PHS. Though EEG findings were abnormal, the readings were not suggestive of a seizure disorder.

Diagnosis of LAS is primarily clinical. Imaging with CT and MRI are ineffective in the diagnosis of LAS.[1] However, some groups have suggested the use of brain PET and SPECT to aid in the diagnosis. In a report of seven patients with LAS, one group described enhanced glucose metabolism in the pontine tegmentum, mesencephalon, and ventrolateral thalamus when compared to controls.[4] A separate study with two patients, performed by another group, found mild bilateral decreased glucose metabolism in the frontal lobes of one of the patients. This group also performed brain SPECT in this study, noting mild hypoperfusion in the left temporal lobe of the other patient.[5] The literature discussing the use of brain PET and SPECT is currently limited. Further research would need to be performed to warrant the use of either as a primary diagnostic tool in LAS. Brain PET and SPECT performed on our patient revealed no abnormalities.

There is no established standard treatment for LAS, but serotonin and gamma-aminobutyric acid (GABA) are believed to be involved in the pathophysiology of myoclonus. Thus, medications targeting serotonergic and GABAergic pathways have historically been used.[6,7] Clonazepam, valproic acid, and piracetam were found to be effective in approximately half of LAS cases.[8] Levetiracetam, utilized with great results in our patient, has also been documented as effective. Positive results with levodopa, intrathecal baclofen, and bilateral pallidal deep brain stimulation have also been reported.[9,10,11] Prognosis for LAS is favorable; however, patients may require chronic treatment for persistent myoclonus and cerebellar dysfunction.[3]

Conclusion

Lance-Adams Syndrome, though rare, remains an important clinical entity to consider when evaluating a patient with intention myoclonus following hypoxic brain injury. Our patient developed symptoms 8 days after PEA arrest and successful CPR. Treatment with levetiracetam controlled his symptoms and he returned to his baseline level of functioning

prior to the event. Greater awareness and education about LAS may lead to faster identification and avoidance of inappropriate care for post-cardiac arrest patients.

Reflections from the Primary Author

This research experience provided me a great opportunity to learn more about this rare condition and take an opportunity to write my first case report. This patient, who I saw during my third year Internal Medicine clerkship, fascinated me. His unique symptoms were all things I couldn't explain with one diagnosis, but that didn't deter our team from trying. The random and rare diagnoses we discussed during our differential reminded me again just how fascinating and exciting medicine can truly be. The curiosity and intellectual pursuit we experienced helped push me towards pursuing a residency in Internal Medicine. I would like to thank all members of the team, Dr. Brandon Gentry, Dr. Min Lee, Dr. Aron Simkins, Kayla Riggs, Kirsten Haack, and our attending, Dr. Kevin Finkel, for providing the intellectual spark I needed to choose my future specialty. From this experience, I hope to continue to add to my medical knowledge and supplement it with the research and writing skills I have acquired. That feeling of wonder and curiosity only further excites me to treat patients and perform more research in the future.

References

1. Lee HL, Lee JK. Lance-Adams Syndrome. Ann Rehabil Med 2011; 35: 939-943

2. Lance JW, Adams RD. The syndrome of intention or action myoclonus as a sequel to hypoxic encephalopathy. Brain 1963; 86: 111-136

3. English WA, Giffin NJ, Nolan JP. Myoclonus after cardiac arrest: pitfalls in diagnosis and prognosis. Anaesthesia 2009; 64: 908-911

4. Frucht SJ, Trost M, Ma Y, Eidelberg D. The metabolic topography of posthypoxic myoclonus. Neurology 2004; 62: 1879-1881

5. Zhang YX, Liu JR, Jiang B, Liu HQ, Ding MP, Song SJ, Zhang BR, Zhang H, Xu B, Chen HH, et al. Lance- Adams syndrome : a report of two cases. J Zhejiang Univ Sci B 2007; 8: 715-720

6. 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: 307-329

7. Matsumoto RR, Truong DD, Nguyen KD, Dang AT, Hoang TT, Vo PQ, Sandroni P. Involvement of GABA(A) receptors in myoclonus. Mov Disord 2000; 15 Suppl 1: 47-52

8. Frucht S, Fahn S. The clinical spectrum of posthypoxic myoclonus. Mov Disord 2000; 15 Suppl 1: 2-7

9. Coletti A, Mandelli A, Minoli G, Tredici G. Post-anoxic action myoclonus (Lance-Adams syndrome) treated with levodopa and GABAergic drugs. J Neurol 1980; 223: 67-70

10. Birthi P, Walters C, Ortiz Vargas O, Karandikar N. The use of intrathecal baclofen therapy for myoclonus in a patient with Lance Adams syndrome. PMR 2011; 3: 671-673

11. Asahi T, Kashiwazaki D, Dougu N, Oyama G, Takashima S, Tanaka K, Kuroda S. Alleviation of myoclonus after bilateral pallidal deep brain stimulation for Lance-Adams syndrome. J Neurol 2015; 262: 1581-1583