CASE REPORT

A Case of Lance Adams Syndrome in a Patient with Attempted Hanging

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Abstract

Lance Adams syndrome is the term used to describe late post-hypoxic myoclonus. Here we describe a patient who developed action and intention myoclonus after 7 days of attempted partial hanging. The similarity of Lance Adams syndrome, which is a treatable condition to a cerebellar syndrome, and the diagnostic difficulties have been highlighted.

Keywords: Action myoclonus, Intention myoclonus, Lance Adams syndrome, Post-hypoxic myoclonus.

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Key Messages

Late post-hypoxic action and intention myoclonus can occur in an attempted hanging patient. It is a disabling condition. Since it is a treatable condition, a high index of suspicion is necessary on the part of the treating physician to look out for it.

Introduction

Lance Adams syndrome was first described by James Lance and Raymond Adams in 1963 in 4 patients who had cardiorespiratory arrest.[1] It describes a late post-hypoxic myoclonic disorder that is seen usually after cardiopulmonary resuscitation. Here, we describe a patient who presented with action and intention myoclonus that started 5 days after the attempted hanging episode. He was treated with SSRI, sodium valproate, and benzodiazepines, and was improved dramatically.

Case History

A 30-year-old male presented to casualty with complaints of difficulty in walking and deviation of angle of mouth to the right side, 7 days after he had attempted hanging. His wife gave a history that he had attempted hanging under the influence of alcohol, and they had taken him to a nearby hospital where he was treated for 7 days. The treatment records from outside the hospital revealed that he was unconscious for 2 days after the attempted hanging episode and was intubated and ventilated for 2 days. He had slowly regained consciousness and was weaned off the ventilator in the subsequent 2 days. After 5 days of attempted hanging, he developed jerky clonic movements of the upper and lower limbs whenever he tried to take a glass of water or when he tried to walk. These movements were so disabling that he could not even stand up on his own. He could not eat food or drink water on his own, which would result in significant spillage of water and food if tried to do so. He was diagnosed as having cerebellar ataxia in the nearby hospital. He also had vision loss with only light perception in both eyes with preserved pupillary reflexes. MRI brain with contrast was done and it was found to be normal. He also had a facial deviation to the right side with involvement of the upper half of the face as well, which was attributed to a facial nerve injury at the point of exit from stylomastoid foramen. With these complaints he presented to us, our examination revealed a thin-built individual without any significant general examination findings. He had only light perception in both eyes. There was an lower motor neuron (LMN) facial palsy on the left side. Other cranial nerves were normal. Motor system examination revealed normal bulk and tone, with the power of 4^+ in all four limbs and all muscle groups. Reflexes were present normally. Sensory system examination was unremarkable. But what was striking during the period of his examination was whenever he used to exert force in his limbs (more so in the lower limbs than upper limbs), his entire body would show clonic jerks. These clonic jerks would disappear the moment he was put to rest. There was a disparity between his upper limbs and lower limbs, with lower limbs being disproportionately more involved. While testing for cerebellar functions, the finger-nose test and finger-nose-finger test were done with relatively less jerky movements. Dysdiadochokinesia was also completed with relative ease. But when trying heel-shin-knee tests, the clonic jerks all over the body increased in amplitude. On testing walking, he could hardly stand, and his entire body was thrown into severe clonic jerks the moment he stood on the ground. The relative sparing of upper limbs in cerebellar function tests and the presence of clonic jerks during walking and on putting lower limb muscles into action made us to think of the possibility of action and intention myoclonus. The appropriate clinical setting gave the diagnosis of Lance Adams syndrome. An EEG was done to look for any spike and sharp waves,

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but it was normal. Visual evoked potential (VEP) was done, which was normal. Nerve conduction study (NCS) revealed LMN facial palsy on the left side. The vision loss was attributed to possible hypoxic injury to the occipital cortex. The patient was started initially on sodium valproate 600 mg/day and clonazepam 1 mg/day. He showed dramatic improvement in 3 days in the form of his ability to walk and also used his upper limbs to eat and drink water. By day 4, he was walking without support with minimal jerky movements of his body. He was discharged with sodium valproate, clonazepam, and sertraline.

Discussion

Myoclonus is a brief (most often <100 ms) contraction involving agonist and antagonist muscles, leading to a sudden jerk.[2] Myoclonus can be classified as physiological, essential, epileptic, and symptomatic. It can also be classified according to the site of origin as cortical, subcortical, spinal, or peripheral myoclonus.[3] Post-hypoxic myoclonic syndrome is classified into acute and chronic disorders. Acute post-hypoxic myoclonus manifests within 24 hours of hypoxic injury and is seen as myoclonic status epilepticus and usually has a grave prognosis. Chronic post-hypoxic myoclonus is termed as Lance Adams syndrome and it is seen usually late (days to weeks) after hypoxic injury to the brain. It usually is seen as a multifocal action and intention myoclonus with a good response to drugs.[4] Autopsy of a patient with Lance Adams syndrome showed astrocytic prominence in the midbrain periaqueductal gray, supratrochlear nucleus, the lateral subnucleus of the mesencephalic gray matter, and the cuneiform and subcuneiform nuclei. In other cases, neuronal loss was observed in the thalamus, striatum, mammillary bodies, and brainstem raphe nuclei.[5] Low serotonin states have been noted in patients with post-hypoxic myoclonus, and hence SSRIs have been postulated as a treatment modality. EEG findings commonly seen include 2–4-Hz spike-and-wave discharge, delta-range slowing, and can even be normal. In our patient, the disparity between lower limbs and upper limbs seemed to be a unique presentation in Lance Adams syndrome. EEG was normal, but the dramatic improvement with relatively small doses of sodium valproate and clonazepam confirmed the diagnosis. Treatment options include sodium valproate, clonazepam, SSRI, and levetiracetam.

Conclusion

A high index of suspicion needs to be present in the minds of the treating physician whenever he encounters myoclonic jerks in a patient with recent hypoxic brain damage due to the treatable nature of Lance Adams syndrome and its ability to closely mimic a cerebellar syndrome.

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