

Lance-Adams Syndrome: A Case Report

A rare disorder presenting with myoclonus may be misdiagnosed as Parkinson's disease, dystonia, or chorea.

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Lance-Adams Syndrome is a rarely diagnosed state that occurs after successful cardiopulmonary resuscitation and is likely caused by hypoxic changes. Symptoms such as action myoclonus are accompanied by cerebellar ataxia and appear days or weeks after the event.

In the literature, two types of post-hypoxic myoclonus were described: acute and chronic.

The acute myoclonus usually occurs within 48 hours after a hypoxic insult in a deeply comatose patient and predicts a poor prognosis. The chronic post-hypoxic myoclonus is described as Lance-Adams Syndrome. Worldwide, 122 cases have been reported in the literature. Here, we present a case report of one patient.

HISTORY AND ADMISSION

The patient is a 74-year-old woman admitted to our department because of symptoms that appeared to be a complication of successful cardiopulmonary resuscitation. Patient underwent a cardiorespiratory arrest that occurred during the asthmatic state six months prior. In the following weeks she developed involuntary movement and practically couldn't walk or eat by herself, needing assistance in nearly every daily task. From the patient's personal history we knew she suffered from asthma, type 2 diabetes, and chronic sinusitis. Previously she had experienced non-ST segment elevation myocardial infarction (NSTEMI), pelvic fracture, distal radius fracture, and cholecystectomy. Her treatment history included: haloperidol, biperiden, escitalopram, acetylsalicylic acid, spironolactone, pantoprazole, osseous

hydroxyapatite, salbutamol, budesonide, ipratropium bromide, atorvastatin, and tiapride.

Her primary movement disorder was diagnosed by a general practitioner as chorea and was administered a large dosage of haloperidol. The patient's state grew worse, becoming more rigid and less alert.

At admission she was conscious and fully alert, though she responded very slowly. Other symptoms included: an excessively smooth left nasolabial furrow, reflexes that were asymmetrical and better on right side, tremor in the upper limbs, general rigidity, and action myoclonus.

MRI revealed large ischemic area in the corona radiata and white matter in both hemispheres. EEG was typical for epilepsy and the record changed, especially in the temporal part of the brain. The Lance-Adams Syndrome was diagnosed and valproic acid was administered. Patient was previously treated with levodopa and a large dosage of haloperidol but no effect was achieved. During her stay in our department the patient stabilized and nearly all of her symptoms disappeared.

DISCUSSION

Patients who survive cardiopulmonary insult often suffer from the different disorders that occurred as repercussion of the brain hypoxic injury. Depending on the area of the brain in question and level of the hypoxic damage, different symptoms may appear including: dyskinesia, Parkinson's disease, dystonia, chorea, athetosis, tremor, and Lance-Adams Syndrome.¹

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Pathophysiology of LAS is still not well known, as a result of the acute hypoxemia toxic metabolites, including lactic acid and free radicals emerge and damage astrocytes, glial and mesenchymal cells. It is suggested that a loss of serotonin and gamma-aminobutyric acid (GABA) within the inferior olive may be a cause of the syndrome.² As for treatment, substances like clonazepam, sodium valproate, piracetam, 5-hydroxytryptophan, fluoxetine hydrochloride and carbidopa showed positive influence on the action myoclonus.¹

In the case of our patient, sodium valproate appeared to be significantly effective.

We cannot admit any efficiency of levodopa in monotherapy in our case.

CONCLUSION

Lance-Adams Syndrome is rarely diagnosed. The main reason for this is probably that post-hypoxic myoclonus is diagnosed as a part of post-hypoxic encephalopathy or just epilepsy. With the development of emergency medicine and the increasing number of patients who survive a cardiopulmonary arrest, the amount of LAS diagnosis will gradually grow. The pathogenesis of the syndrome is unknown and further investigation will be needed.

From a practical point of view, the greatest meaning this has is the ability to differentiate action myoclonus that occurs in Lance-Adams Syndrome from other movement disorders, including ticks, chorea or tremor because it influences the treatment. ■

1. Venkatesan A, Frucht S. Movement disorders after resuscitation from cardiac arrest. *Neurol Clin.* 2006;24:123–132.

Korean J Anesthesiol. 2013 October 65(4): 341-344

2. Jae-Young Kwon, Joo-Yun Kim, Eun-Soo Kim, and Hee-Young Kim. Acute onset Lance-Adams syndrome following brief exposure to severe hypoxia without cardiac arrest—a case report. *Ah-Reum*

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3. Zhang YX, Liu JR, Jing B, Liu HQ, Ding MP. *J Zhejiang Univ Sci B.* 2007 Oct;8(10):715–20. Lance-Adams syndrome: a report of two cases. Department of Anesthesia and Pain Medicine, School of Medicine, Pusan National University, Medical Research Institute, Pusan National University Hospital, Busan, Korea