CASE REPORT

Late relapse myoclonus in a case of Lance-Adams syndrome

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SUMMARY

A 66-year-old man had a history of an anterior myocardial infarction followed by a successful cardiopulmonary resuscitation and a posthypoxic myoclonus, also known as Lance-Adams syndrome (LAS). Eight years ago, he was admitted to the emergency department with the same myoclonic jerks during an intercurrent respiratory infection. After treatment with clonazepam and resolution of the infection, the myoclonus promptly disappeared. This case report suggests that relapses of successfully treated LAS can occur in vulnerable patients, as it's a chronic form of myoclonus.

BACKGROUND

In 1963, James Lance and Raymond Adams described a syndrome of intention or action myoclonus, seen in four survivors of cardiorespiratory arrest. After a period of cerebral hypoxia they developed myoclonic jerks which were often accompanied by a cerebellar syndrome and mild cognitive impairment, typically occurring days to weeks after the event.¹ Here we present a unique case of a patient with a history of Lance-Adams syndrome (LAS) who was successfully treated 8 years before the recent hospital presentation, but was now referred having a relapse with similar symptoms.

CASE PRESENTATION

A 66-year-old man was referred to the emergency department of a district general hospital with jerky movements of the upper and lower limbs, trunk and face. The 'shakiness' started 2 h before presentation and was progressive since the beginning. Furthermore, he told us that he was coughing for almost 6 weeks and his general practitioner prescribed him prednisone 3 days before the hospital presentation.

Eight years ago, he survived an out-of-hospital cardiac arrest due to an anterior myocardial infarction followed by successful cardiopulmonary resuscitation. At that time, 4 days after the hypoxic insult, he developed a posthypoxic myoclonus which was controlled with sodium valproate. His EEG showed only waveforms suggesting hypoxic brain injury and no epileptic activity. The medication was stopped after a month and besides a mild cognitive impairment he experienced no other symptoms for almost a decade.

INVESTIGATIONS

On neurological examination, the patient was conscious and alert, but sweating and exhausted. Myoclonic jerks were seen diffusely, more on the left side of the body and aggravated by voluntary

action (see videos). There were no focal neurological deficits and the reflexes were normal. The same pattern of myoclonic movements appeared after his cardiac arrest 8 years ago and was at that time diagnosed as a complication of postanoxic encephalopathy, also known as LAS.

Blood examination showed elevated levels of C reactive protein (25 mg/L) and leucocytes ($23.5 \times 10^9/L$), but metabolic screening was negative. A CT scan of the brain did not reveal any abnormalities.

DIFFERENTIAL DIAGNOSIS

Myoclonus is a brief contraction involving agonist and antagonist muscles, leading to a sudden, jerky, involuntary movement. Although it may be a normal phenomenon, myoclonus can be seen in a broad range of neurological and systemic diseases.

Myoclonic movements can be positive, as a contraction of a single (or a group) agonist and antagonist muscles, or negative, as a transient loss of tonic muscle tone (asterixis).

In patients who survive hypoxic brain injury, two types of myoclonus can occur: the acute posthypoxic status epilepticus which is associated with a poor prognosis, and the chronic form (LAS), which prognosis is known to be much better with adequate treatment. LAS is predominantly characterised by an action myoclonus starting days to weeks after cerebral anoxia.^{2,3}

As the involuntary movements in our patient were equal to the pattern of the posthypoxic myoclonus that he had experienced 8 years ago, we considered the possible relapse of myoclonus during intercurrent pulmonary infection as a possible cause of his current condition.

Other known causes of myoclonus were also considered, such as metabolic disease or toxic agent. However, the patient had no signs of metabolic disease and he was not exposed to any drugs or toxins which can cause myoclonus.⁴ Also, he had no history of chronic or metabolic disease, in which a transient myoclonic state with asterixis can be seen.⁵

TREATMENT

We admitted the patient for observation. The myoclonic movements disappeared promptly after treatment with intravenous clonazepam 1 mg. The next day he had no myoclonus anymore and with a dose of clonazepam 0.5 mg two times a day he returned home.

OUTCOME AND FOLLOW-UP

The patient has remained on clonazepam for 2 months at last follow-up and had remained stable with no further myoclonic jerks.

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DISCUSSION

Worldwide, almost 150 cases of LAS have been reported in literature so far, but the exact pathophysiology of chronic posthypoxic myoclonus is still not clear.^{6–7} Although MRI and positron emission tomography studies showed involvement of the cerebellum, ventrolateral thalamus and pontine tegmentum, imaging tests are not helpful to make a diagnosis of LAS.^{8–9} The loss of the neurotransmitter serotonin (5-hydroxytryptamine) has been thought to play an important role, because its interaction with γ -aminobutyric acid normally suppresses the posthypoxic myoclonus.¹⁰ The treatment of LAS, however, is still based on expert opinion. According to clinical practises, the myoclonus can be treated with clonazepam, sodium valproate or levetiracetam.¹¹

A relapse of LAS, which was seen in our case, is unique in literature and the cause remains speculative. The possibility of drug-induced relapse should be considered, however the only medication in our patient was prednisone, which seems not to be associated with aggravation of myoclonus. Because the evident respiratory infection was the only significant alteration in the patient's condition when relapse occurred, one could assume that, as in many other neurological conditions, intercurrent infection can aggravate the symptomatology of LAS. Also the fact that clonazepam, similar treatment used for LAS, was effective in our patient, implicates that this relapse was an expression of chronic posthypoxic myoclonus.

Learning points

- Lance-Adams syndrome (LAS) is a chronic form of myoclonus that can occur after hypoxic brain injury.¹
- Clonazepam, sodium valproate and levetiracetam have shown excellent outcomes.
- This report suggests that relapses of successfully treated LAS can occur in vulnerable patients, as it is a chronic form of myoclonus.

Contributors TL wrote the first draft and prepared the manuscript. AW participated in the manuscript preparation and revisions and helped to bring the manuscript to its final version.

Competing interests

Patient consent Obtained.

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