Posthypoxic action myoclonus (the Lance-Adams syndrome)

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DESCRIPTION

A 62-year-old man was resuscitated after an in-hospital pulseless electrical activity (PEA) cardiac arrest. He awakened after 24 hours but after another 24-hour interval, he had worsening action and stimulus-induced myoclonus.^{1–3} He was able to relax fully without movement but any volitional movement initiated a burst of appendicular arrhythmic jerks (video 1). The disabling myoclonus was significantly muted with levetiracetam and valproate. No cerebellar lesions were found on CT or MRI.

Action myoclonus is exceptionally rare (less than 0.5% in a series of patients who have a cardiac arrest) and is potentially confused with myoclonus status in a comatose patient, yet the examination, imaging, degree of disability, time of onset and prognosis are very divergent.⁴ Typically, there is no electroencephalogram (EEG) seizure correlate.³ The symptoms can improve or persist chronically, and many remain with significant disability from myoclonus. Medication treatment options include

Learning points

- ▶ Myoclonus occurring after hypoxic brain injury from cardiac arrest, characterised by abrupt irregular muscle contractions. (1) Acute: starting within 48 hours after the arrest (when isolated, sometimes terms acute Lance-Adams syndrome). (2) Chronic: Lance-Adams syndrome, which may start from days to weeks after arrest and progressively worsen, with or without other neurological symptoms.
- ▶ Potentially confused with myoclonus status in a comatose patient, yet the examination, imaging, degree of disability and prognosis are very divergent.
- ▶ Typically, no EEG seizure correlates.

levetiracetam, valproic acid and other anti-epileptic drugs (AEDs).

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Post-hypoxic myoclonus

- Action myoclonus is exceptionally rare (less than 0.5% in a series of cardiac arrest patients)
- Potentially confused with myoclonus status in a comatose patient yet the exam, imaging, degree of disability, time of onset and prognosis are very divergent
- Typically no EEG seizure correlates
- Symptoms can improve or persist chronically
- Many remain with significant disability from myoclonus
- Treatment options include levetiracetam, valproic acid and other AEDs

video 1. Pretreatment and posttreatment demonstration of subacute action myoclonus after cardiac arrest, with narration and reference text. The disabling myoclonus was completely muted with levetiracetam and valproate within an additional 72 hours. No acute findings were found on CT or MRI, and had mild confusion but no other focal deficits. Myoclonus occurring after hypoxic brain injury from cardiac arrest, characterised by abrupt irregular muscle contractions. (1) Acute: starting within 48 hours after the arrest (when isolated, sometimes terms acute Lance-Adams syndrome). (2) Chronic: Lance-Adams syndrome, which may start from days to weeks after arrest and progressively worsen, with or without other neurological symptoms.

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