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Case Report

Challenging Intensive Care Stimulus-Sensitive Myoclonus Following Prolonged Cardiothoracic Surgery

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Abstract

Stimulus-sensitive myoclonus is a rare condition that can occur following prolonged cardiothoracic surgery. It is similar to Lance-Adams syndrome, which is characterised by post-cardiac arrest myoclonus, and both conditions are related to nervous system hypoxia. In our case, myoclonus developed after a patient underwent an elective replacement of the ascending aorta and total aortic arch due to an aortic aneurysm. The myoclonus primarily affected the right side of the body and was triggered by touch, sound, and light. After a thorough evaluation and comprehensive investigations, we concluded that this case represented post-hypoxic brain injury stimulus-sensitive myoclonus, resembling Lance-Adams syndrome with both cortical and subcortical myoclonus. Eight antiseizure medications were trialled, and significant relief was only achieved with Perampanel.

Keywords: *Lance-Adams; Syndrome; Post-cardiac; Hypoxia; Perampanel; Relief*

Introduction

Myoclonus involves sudden, involuntary muscle jerks and can result from hypoxic brain injury, particularly after cardiothoracic procedures. Stimulus-sensitive myoclonus is triggered by external stimuli and shares features with Lance -Adams syndrome. The underlying pathophysiology involves cortical and subcortical hyperexcitability due to hypoxic-ischemic damage. Managing stimulus-sensitive myoclonus can be challenging due to variable responses to medications such as valproic acid and levetiracetam. However, emerging evidence suggests that perampanel may be beneficial in refractory cases. This report highlights the clinical journey of a patient with severe stimulus-sensitive myoclonus post-surgery, focusing on diagnostic challenges, limitations of standard treatments, and the successful use of perampanel. Our findings contribute to the literature on post-hypoxic myoclonus, underscoring the need for individualized treatment strategies to enhance patient outcomes. (1-5)

Case Presentation

A mid-seventies-year-old female, a right-handed patient, underwent elective replacement of the ascending aorta and total aortic arch due to an aortic aneurysm. Following a prolonged cardiothoracic surgery that lasted over six hours, during which there were periods of hypotension, she was transferred to the intensive care unit.

Twenty-four hours after the surgery, she developed jerky movements on the right side of her body as she was being weaned off sedation. These movements affected her face, limbs, trunk, and abdomen. Despite receiving maximum doses of Levetiracetam and Phenytoin, there was no improvement in her condition. Consequently, she was reintubated and sedated using Propofol and Midazolam.

She was referred to Neurology, where she was initially diagnosed with cortical myoclonus due to prolonged cardiac surgery. Despite trying several anti-seizure medications and undergoing various tests, her right-sided myoclonus persisted. The myoclonus worsened during the daytime and was sensitive to certain stimuli, such as touch, voice, and light.

Investigations:

Five EEGs (Electro-Encephalogram) with recorded video and Electromyography (EMG) showed frequent myoclonic jerks in the face, right arm, and leg, occurring independently, sequentially, or together, with spike activity; one EEG showed burst suppression (Was on Propofol). These movements cause widespread muscle artefacts, making it challenging to identify epileptic discharges.

The MRI head showed a small area of restricted diffusion in the left parietal lobe, indicating an acute infarct and an old infarct in the right cerebellum. Additionally, scattered white matter changes related to small vessel disease and hemosiderin deposition in the bifrontal, pontine, and bilateral cerebellar areas. The MRI spine cervical cord exhibits abnormal signal intensity and expansion from the C2/3 to T1, with low ADC signal change.

- CSF analysis was normal.
- Negative serum and CSF autoimmune encephalitis
- FBC, Electrolytes, RFT, and LFT were all normal.
- Negative vasculitis screening.
- Paraneoplastic and serum ACE were negative.
- EMG: Was inconclusive (Shaking artefact).

The management of seizures has proven to be unsuccessful despite the administration of various anti-epileptic medications. These include a Midazolam infusion administered during intubation, Levetiracetam, Sodium Valproate, Clobazam, Clonazepam, and Phenytoin(All in maximum doses). Additionally, Lorazepam and Diazepam were administered on an as-needed basis.

She was transferred to a quieter environment where all potential triggers were eliminated. We initiated her treatment with Perampanel and gradually tapered her off phenytoin. Following this adjustment, we observed a significant improvement in her myoclonus. Subsequently, we slowly reduced her other anti-seizure medications, and she continued her treatment exclusively with Perampanel at a dosage of 6 mg. After three weeks, the patient made a favorable recovery and was discharged from the intensive care unit, then transferred to the neuro-rehabilitation ward. During her follow-up review eight weeks after hospital discharge, she remained seizure-free, and she continued taking Perampanel at a dose of 6 mg once daily.

Conclusion

The diagnosis of post-hypoxic stimulus-sensitive myoclonus unequivocally relies on a comprehensive clinical assessment alongside MRI, EEG, and EMG findings. Prompt diagnosis and treatment are essential, as they can significantly improve patients' quality of life. Valproic acid, levetiracetam, and clonazepam are widely recognized as the most beneficial antiepileptic medications for the effective management of myoclonus. Moreover, perampanel presents a promising option, particularly for individuals dealing with stimulus-sensitive myoclonus, enhancing the potential for improved outcomes.

Declaration

Authorship Contribution

HMA contributed to the conception, data collection, and drafting of the manuscript and final review. **AS** participated in patient care and critical revision of the manuscript. **RA** supervised the case, provided expert clinical insights, and approved the final version. **KN** participated in patient care and critical revision of the manuscript.

Patient Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the consent form is available for review upon request.

Conflict of Interest

The authors declare no conflicts of interest regarding the publication of this case report.

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