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The Lance-Adams syndrome following cardiopulmonary resuscitation: A report of two cases

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Introduction

It is common knowledge that coma and seizures indicate a significant poor prognosis in the patient resuscitated from cardiopulmonary arrest [1–4]. As a less common complication, myoclonic movements have also been reported following severe global ischemic insult [5].

We herein report two cases of patients who were still comatose after successful cardiopulmonary resuscitation and complicated myoclonic jerks in the intensive care unit. Initially, these movements were thought to be clonic seizures and treated with anticonvulsants. Both patients survived to be conscious, but "action myoclonus" [6] persisted thereafter.

Case histories

Patient 1

A 47-year-old man was transferred to our emergency department with gunshot wounds in his neck and left shoulder. He had had his trachea perforated and the wounds were bleeding freely. Soon after arrival, cardiorespiratory arrest ensued. Prompt resuscitation was successful, and he underwent emergency tracheoplasty and tracheostomy. The patient was then admitted to the intensive care unit and his ventilation was assisted mechanically. Within minutes of admission, he convulsed suddenly with clonic movements of all four limbs. The seizure stopped with either 10 mg of diazepam or 200 mg of thiopental i.v., but for only a short duration. A bolus i.v. injection of pentobarbital 150 mg, every 2 h controlled the seizure and was continued for about 36 h. Antiepileptics including phenytoin, primidone, and phenobarbital were also added. Brain computed tomographic (CT) scans taken on the following day revealed only a slight degree of edema, and EEG showed mostly α- and θ-waves. Toward the end of pentobarbital therapy, no spike discharge was seen and some slow wave components appeared on EEG. After pentobarbital was stopped, however, the patient again developed clonic movements of his limbs, which were initiated especially by the stimulus such as tracheal suction, change of body position or pain. We still thought these clonic movements to be seizures and treated him with anticonvulsants. Four days after admission, it was realized that he seemed to be conscious. Ten days later, a consultant neurologist diagnosed the abnormal limb movements as myoclonic jerks secondary to cerebral anoxia, or Lance-Adams syndrome. The treatment was started with sodium valprorate and clonazepam, resulting in some improvement. EEG taken at this stage showed a spike discharge, which was not detectable 3 days later.

While myoclonus of lesser frequency and severity still continued thereafter, the patient recovered consciousness completely and was discharged from the intensive care unit 22 days after the admission. In the ward, residual myoclonus of tongue prevented him from eating for about 2 weeks. Five months later, he was discharged to another hospital to receive rehabilitation therapy for some residual action myoclonus.

Patient 2

A 47-year-old male had been complaining of dyspnea while working at his office. His condition deteriorated

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and he eventually had a respiratory arrest and severe bradycardia. On arrival at our emergency room, asphyxiation due to an upper airway obstruction caused by acute epiglottis was confirmed when his trachea was intubated under direct laryngoscopy. Prompt resuscitation was performed successfully, and he was transfered in an unconscious state to the intensive care unit where his respiration was supported mechanically. Within several minutes, the patient developed seizure-like clonic movements. The EEG was not taken at this time. A large dose of pentobarbital infusion of 50–100 $mg \cdot h^{-1}$ supplemented by bolus injections of 100–150 mg as needed was used to control the seizure-like activity for about 8 days. A brain CT scan was obtained on the 2nd day which showed only a slight edema. Phenytoin, primidone, and phenobarbital as antiepileptics were started on the 4th day, and with the previous patient in mind, sodium valprorate and clonazepam were also added. Over these days, myoclonic movements were observed in his jaw, tongue, abdomen, and lower limbs. After pentobarbital was stopped, midazolam was administered intravenously to control these abnormal movements, but with transient effect. Twelve days after admission, however, the patient opened his eyes in response to a verbal command. EEG taken a week later showed α- and θ-waves and no spike discharge was found. The diagnosis of Lance-Adams syndrome was confirmed by a consultant neurologist.

The myoclonus improved gradually in its frequency and severity, and the patient was discharged to the ward 25 days after admission. In the ward, he continued to suffer some residual disability such as being unable to feed himself or sit up in bed due to action myoclonus, but his intellectual recovery was completely normal.

Discussion

In 1963, Lance and Adams first described four patients with posthypoxic generalized myoclonic jerks [6]. They were comatose and developed generalized clonic movements for 2–4 days following the hypoxic incident. After recovery of consciousness, the patients continued to have abnormal clonic muscle movements which were initiated by one of many unexpected stimulus such as light, touch, or loud sound, and even by coughing, yawning, or stretching at the later stage. These movements were also elicited more effectively when the patients attempted to perform a finely coordinated, willed action. This fact led Lance and Adams to term these abnormal clonic movements "intention or action myoclonus". Most patients were ataxic and could not stand or walk without assistance. It was particularly interesting that there were no other signs of neurological dysfunction except for myoclonus and cerebellar ataxia.

In our two cases, there are many features which resemble those originally described by Lance and Adams [6]. Following hypoxic events, they both were comatose and had generalized clonic movements which were characteristically elicited by several stimuli given during nursing care. The fits were thought to be seizures until the patients were found to be conscious during the attack of these clonic movements, and at this stage, the diagnosis of posthypoxic myoclonus (Lance-Adams syndrome) was made. The severity and frequency of myoclonus improved with time and with drug therapy, but even after complete recovery of consciousness the patients continued to experience action myoclonus.

The pathophysiology of Lance-Adams syndrome has not been clearly defined. The morphologic studies have not been able to demonstrate any specific destructive lesions [7,8], even in the midline structure of the midbrain, the area in which serotonergic neurons are present [9]. These findings suggest a functional derangement of serotonergic activity rather than structural damage of neurons. Actually, a decrease of the cerebrospinal fluid (CSF) concentration of 5-hydroxyindoleacetic acid (5-HIAA), the acid metabolite of serotonin, was observed in the patients with posthypoxic action myoclonus, and *L*-5-hydroxytryptophan (*L*-5-HTP), a precursor of serotonin [8], clonazepam and valprorate [7] significantly increased the level of 5-HIAA in CSF and improved the symptoms. These facts suggest that a deficiency of serotonin in the brain is a cause of the pathophysiology of the syndrome. Although the CSF concentration of 5-HIAA was not determined in our cases, we administered clonazepam and valprorate with some effects.

Generalized myoclonus, which is a characteristic feature of the early stages of the syndrome, is difficult to be differentiated clinically from posthypoxic seizures or convulsions, but it should be distinguished from seizures as early as possible so that a correct prognosis can be given and correct therapy instituted promptly [10]. Although our cases are not related to anesthesia, we, as anesthesiologists, should also be aware that anesthetic accidents are the most common cause of cerebral hypoxia [7].

We have described two cases of the Lance-Adams syndrome in which generalized myoclonic jerks and thereafter action myoclonus developed following cardiopulmonary resuscitation. Those who practice anesthesia or intensive care should always keep in mind the existence of this syndrome when diagnosing comatose patients resuscitated from cardiopulmonary arrest.

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