Case Report

Generalized chorea due to delayed encephalopathy after acute carbon monoxide intoxication

Yueh-Feng Sung, Ming-Hua Chen[1], Giia-Sheun Peng, Jiunn-Tay Lee

Department of Neurology, Tri-Service General Hospital, National Defense Medical Center, Taipei, [1]Department of Medicine, Taoyuan Armed Forces General Hospital, Taoyuan, Taiwan

Abstract

Movement disorder due to delayed encephalopathy after carbon monoxide (CO) intoxication is uncommon. Generalized chorea, presenting as an initial symptom of delayed encephalopathy, is extremely rare. We describe a 60-year-old woman, who had completely recovered from acute CO poisoning, developed mental and behavioral changes, urinary incontinence and generalized chorea 2 weeks thereafter. T2-weighted brain magnetic resonance imaging showed extensive hyperintensity of the bilateral periventricular and subcortical white matter and the globus pallidus. Brain single-photon emission computed tomography (SPECT) with technetium-99 ethylene cysteine dimer showed inhomogeneous perfusion in the cerebral cortex, with decreased uptake in bilateral frontal regions. Delayed encephalopathy after acute CO intoxication was diagnosed, and the symptoms gradually improved after hyperbaric oxygen therapy (HBOT). This case report demonstrates that generalized chorea may be one of the initial presenting symptoms of delayed encephalopathy after acute CO intoxication. We hypothesize that the generalized chorea in our patient may have been caused by the subcortical white matter lesions, which most likely interrupted the basal ganglia-thalamocortical circuits and that HBOT may be the treatment of choice for such patients.

Key Words

Chorea, carbon monoxide, hyperbaric oxygen therapy, magnetic resonance imaging

For correspondence:

Dr. Yueh-Feng Sung, Department of Neurology, Tri-Service General Hospital, National Defense Medical Center, No. 325, Section 2, Cheng-Gong Road, Neihu 114, Taipei, Taiwan, R.O.C. E-mail: sungyf@ndmctsgh.edu.tw

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Introduction

The incidence of delayed encephalopathy after carbon monoxide (CO) intoxication is low. Delayed encephalopathy has been shown to occur 14-45 days after complete recovery from the acute stage of CO intoxication and is characterized by mental impairment, motor dysfunction, dementia, or psychosis.[1] Chorea is characterized by a continuous flow of random, brief, involuntary muscle contractions and can result from a wide variety of causes such as hypoxic-ischemic injury, drugs, toxins, infections, autoimmune disorders and endocrine and electrolyte abnormalities.[2] Chorea presenting as a symptom of delayed encephalopathy is extremely rare. We herein report the case of a woman with generalized chorea as one of the first manifestations of delayed encephalopathy following acute CO intoxication. Her symptoms improved gradually after hyperbaric oxygen therapy (HBOT).

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Case Report

A 60-year-old woman, with a history of major depression, attempted suicide by burning charcoal in a closed room 3 weeks before admission. She was found unconscious and was immediately sent to a nearby hospital. She regained full consciousness from the acute stage of CO poisoning within 1 day under high-flow oxygen therapy. She was well after discharge, and no medications were prescribed. One week prior to admission, she began to experience cognitive impairment, behavioral changes, unsteady gait and urinary incontinence. Restlessness and involuntary movements of the limbs were also observed by her family. Due to the persistent symptoms, she was admitted to our hospital.

On examination, she was found to be alert but could not obey or understand orders. Her blood pressure was 128/84 mmHg, heart rate 79 bpm, respiration rate 18 breaths/min, and body temperature 36.5°C. Pupil light reflexes and eye movements were normal. She exhibited akinetic mutism, rigidity of the limbs, and a short-stepped gait. Deep tendon reflexes were normal, muscle strength of the limbs was Medical Research

Council Grade 4, and Babinski's sign was absent. She was found to exhibit persistent, rapid, irregular, involuntary choreic movements of the whole body, which were invisible during sleep. Blood studies, including biochemistry, endocrine and autoimmune tests, were normal.

Electroencephalography showed a poorly sustained alpha background and generalized slow waves, predominantly at the bilateral fronto-temporal lobes, indicating cortical dysfunction. T2-weighted and fluid-attenuated inversion recovery (FLAIR) brain magnetic resonance imaging (MRI) showed bilateral, symmetric, confluent areas of high signal intensity in the periventricular, subcortical white matter, corona radiata, centrum semiovale and globus pallidus [Figure 1a and b]. Diffusion-weighted MRI and the corresponding apparent diffusion coefficient map showed restricted water diffusion at the periventricular white matter, corona radiata, and centrum semiovale, suggesting cytotoxic edema [Figure 1c and d]. Brain single-photon emission computed tomography (SPECT) with technetium-99 ethylene cysteine dimer showed inhomogeneous perfusion in the cerebral cortex, with decreased uptake in bilateral frontal regions [Figure 1e]. Uptake in bilateral basal ganglia and the thalamus was normal.

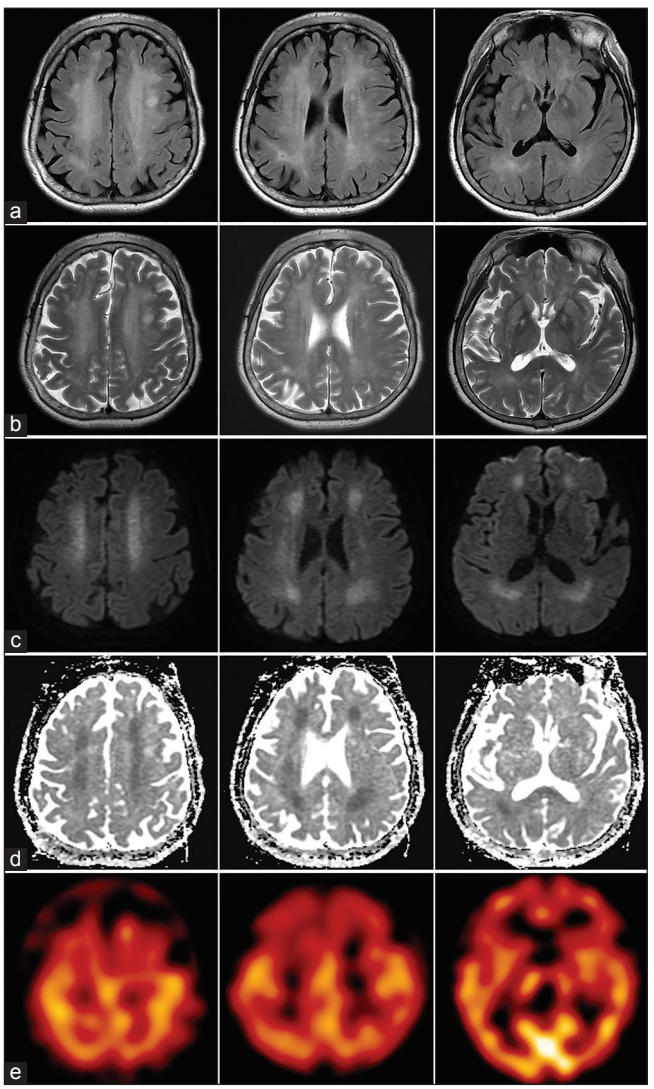

Figure 1: Fluid-attenuated inversion recovery (FLAIR) (a) and T2-weighted (b) brain magnetic resonance imaging (MRI) showing bilateral, symmetric, confluent areas of high signal intensity in the periventricular, subcortical white matter, corona radiata, centrum semiovale and globus pallidus. Diffusion-weighted MRI (c) and the corresponding apparent diffusion coefficient map (d) showing restricted water diffusion at the periventricular white matter, corona radiata and centrum semiovale. Brain single-photon emission computed tomography (e) showing inhomogeneous perfusion in the cerebral cortex, with relative decreased uptake in bilateral frontal regions

The patient was diagnosed with delayed encephalopathy after acute CO intoxication. Neuroleptics for suppression of generalized chorea were not administered, for fear of exacerbating the parkinsonism. She instead underwent HBOT. The symptoms of generalized chorea gradually improved and disappeared completely 2 weeks later. The cognitive impairment improved moderately after 20 sessions of HBOT. However, the urinary incontinence, mild rigidity of the limbs, and short-stepped gait remained unchanged after discharge. Follow-up brain MRI 8 months later showed more cortical atrophy and prominent hyperintensity of the bilateral periventricular white matter seen on FLAIR and T2-weighted image [Figure 2a and b].

Discussion

Delayed encephalopathy after CO intoxication is characterized by mental impairment, psychosis, urinary incontinence, or motor dysfunction, which develops between a few days and a few weeks after acute CO poisoning.[1] Thom hypothesized that CO activates polymorphonuclear leucocytes, which diapedese and cause brain lipid peroxidation, leading to the delayed effects of CO poisoning.[3] The most characteristic neuropathologic findings following CO intoxication are necrosis of the bilateral globus pallidus and progressive white matter demyelination. Involvement of the centrum semiovale is considered to be more related to delayed neuropsychiatric syndrome.[4] Some patients with follow-up MRI have revealed a decrease in signal abnormalities of white matter lesions,

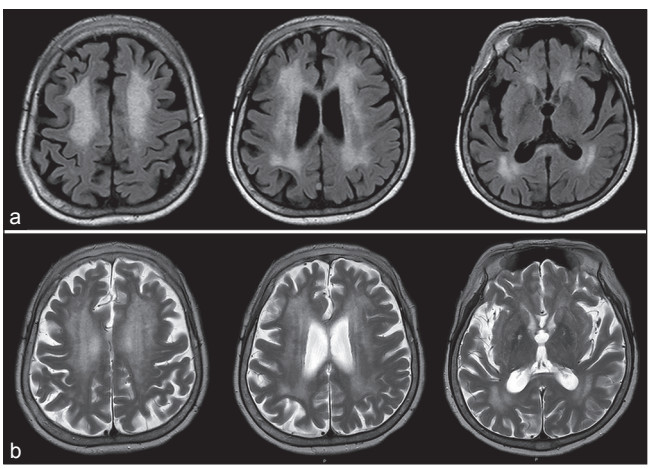

Figure 2: Follow-up brain MRI showing more cortical atrophy and prominent hyperintensity of the bilateral periventricular white matter seen on FLAIR (a) and T2-weighted image (b)

Table 1: The toxic causes of chorea and the radiological findings

Toxins	Radiological findings
Alcohol (intoxication and withdrawal)	MRI: T2 hyperintensity in bilateral globus pallidus
Mercury	MRI: negative
Manganese	CT/MRI: negative
Lead	CT: bilateral and symmetric pallidal calcifications
	MRI: atrophy of bilateral putamen and caudate nuclei (T1 hypointensity and T2 hyperintensity)
Carbon monoxide	CT: bilateral low-density lesions in the basal ganglia and/or in the white matter of the cortex
	MRI: multiple T2 hyperintensity lesions in the subcortical white matter and basal ganglia

accompanied with a lessening in the severity of symptoms, suggesting a reversible process.[4,5]

While delayed encephalopathy following CO intoxication is not normally associated with involuntary movements, it may give rise to chorea in rare cases. Choi *et al*., reported 242 patients with CO poisoning, 32 (13.2%) of whom developed delayed movement disorders. Of these 32 patients, 23 (71.9%) had parkinsonism, five dystonia, three chorea and one myoclonus.[5] Song and Chung reported a patient with delayed encephalopathy due to CO intoxication suffering from chorea involving a unilateral lower limb. There were no basal ganglia lesions, only bilateral white matter lesions, in this patient.[6] In addition to CO, some toxins have been reported to cause choreic movement.[1, 7-11] The toxic causes of chorea and the radiological findings are listed in Table 1.

The probable pathophysiologic mechanism of chorea seems to result from hypoactivity in the indirect pathway from the basal ganglia-thalamocortical motor circuits.[2] We hypothesize that the generalized chorea in our patient may have been caused by the subcortical white matter lesions, which most likely interrupted the basal ganglia-thalamocortical circuits. It remains unclear why similar neuroimaging changes after CO intoxication result in different types of movement disorders.

Some authors have reported that chorea after CO intoxication is usually alleviated by neuroleptics; the fact that the chorea does not relapse after cessation of neuroleptics suggests that these patients may have functional rather than anatomic impairment.[11,12] However, neuroleptics that may aggravate parkinsonism should be of concern for patients with both chorea and parkinsonism.

HBOT administered within 24 hours after acute CO poisoning can reduce the risk of cognitive sequelae. However, the role of HBOT in delayed neuropsychiatric syndrome after acute CO poisoning is still being debated. Chang *et al*., reported 9 patients with delayed neuropsychiatric syndrome following CO poisoning, all of whom showed a good response to HBOT.[13] Our patient experienced complete disappearance of the generalized chorea, followed by moderate improvements in cognitive function after HBOT. We speculate that HBOT had greater benefits for demyelinating leukoencephalopathy than globus pallidus lesions in this patient, based on the clinical observation that she completely recovered from the chorea and mental disturbances, but not parkinsonism. Further study is needed to explore the relationship between lesion sites and involuntary movements, as well as the therapeutic mechanisms of HBOT, in patients with delayed encephalopathy after CO intoxication.

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