A case of alcoholic pellagra encephalopathy presenting with spinal myoclonus

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Practical Implications

Consider multiple vitamin therapy including niacin to treat patients presenting with myoclonus and chronic alcoholism.

Video

Neurology.org/cp

A 56-year-old man with a history of heavy alcohol consumption (100 g/day for 2 years) had disorientation and memory disturbance for 4 months. He was unable to walk because of instability and then experienced visual and auditory hallucinations. He was admitted to a local hospital and became fully oriented and able to walk after IV administration of thiamine and multivitamin preparations without niacin. One month later, however, his mental status fluctuated and deteriorated again. The patient developed dysuria, gait disturbance, and lower extremity myoclonus and was referred to our hospital. He was emaciated (height 170 cm; weight 47.6 kg; body mass index 16.4), and he had a sacral decubitus ulcer. His face was darkened and hyperkeratotic. He was disoriented and irritated, with flapping tremors. Lower extremity myoclonus was observed every 20–30 seconds (see video at Neurology.org/cp), which was triggered by a touch on the legs and worsened in the supine position, even during sleep. Deep tendon reflex was exaggerated (more evident at knee jerks than ankle jerks), and Babinski sign was positive bilaterally. Finger-nose-finger and heel-knee tests revealed cerebellar ataxia. Romberg sign was positive and joint position sense of the great toes was disturbed. He had stocking sensory impairment. The patient also had orthostatic hypotension, constipation, and urinary retention and incontinence.

He was not cirrhotic. Routine blood laboratory data, including albumin (3.5 g/dL), ammonia (23 μ g/dL), thiamine (179.8 mg/mL), niacin (5.4 mg/mL), and thyroid hormone levels, were normal, and CSF including culture was normal. The chest CT revealed pulmonary emphysema. MRI of the brain and spinal cord showed no abnormal findings. EEG showed generalized delta waves intermittently and regional delta waves in the bifrontal and bioccipital areas. Evoked response potentials from median nerve stimulation were normal but from tibial nerve stimulation were poor. Nerve conduction study revealed sensorimotor axonal polyneuropathy. Surface EMG revealed that the myoclonus originated from the lumbosacral spinal segment, spreading rostrally up to the T7 level (figure). The bursts lasted for 140–200 ms, suggesting spinal origin.¹ Because of worsening delirium despite receiving high doses of thiamine, pellagra encephalopathy was suspected and oral administration of niacin (1,500 mg/day) was initiated 16 days after admission. The patient's mental status improved dramatically within 48 hours, and his face became smoother and whiter several days after the treatment. Spontaneous lower extremity myoclonus vanished 4 weeks later, and cerebellar ataxia and dysautonomia resolved in 3 months. Sensory ataxia also improved gradually over the course of 1 year and he could walk unassisted, but numbness by sensorimotor polyneuropathy and pyramidal tract sign still existed. Although

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Figure Surface EMG recording of myoclonus

Electric discharges started in the tibialis anterior and spread toward the rostral muscles and up to the T7 level (cal: 200 μ V; 200 ms).

myoclonus that had been diminished by clonazepam reappeared several times when his general condition was not good, other symptoms did not recur after cessation of alcohol consumption.

DISCUSSION

Pellagra due to niacin deficiency is rare in developed countries, except in cases of chronic alcoholism.² Making a diagnosis of pellagra is challenging because patients do not always show all of the “3 D’s”: dementia, diarrhea, and dermatitis.³ Dermatitis is often limited to subtle changes, as was the case in our patient. Alcoholic pellagra often lacks typical dermatitis because it develops too rapidly for the dermatologic changes to occur.⁴ In addition, as evidenced in our patient, serum niacin levels are not always helpful in confirming diagnosis because they are often normal. In alcoholic pellagra encephalopathy, confusion, hallucinations, clouding of consciousness, oppositional hypertonus, ataxia, and myoclonus are frequently observed, and myoclonus is observed in 50% of patients.⁵ However, it is uncommon in other types of alcohol-related encephalopathy that often coexist with alcoholic pellagra. In this context, myoclonus is a key feature of alcoholic pellagra. The reported classification of the myoclonus in alcoholic pellagra is startle myoclonus that is thought to originate in the brainstem.⁵ In the current patient, hyperreflexia, positive Babinski sign, dysuria, and severe sensory ataxia were consistent with the involvement of the spinal cord, and electrophysiologic findings suggested that myoclonus was of spinal origin. The anterior horn cells of the spinal cord, along with Betz cells, other large cortical neurons, and the pontine, dorsal vagal, and vestibular nuclei, were shown to have severe chromatolytic changes.⁶ Pellagra encephalopathy can present with spinal myoclonus when it involves the spinal cord.

One final important consideration is that alcoholic pellagra can develop after the administration of thiamine without niacin because relatively excessive thiamine levels indirectly elevate the demand for niacin.⁷

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AUTHOR CONTRIBUTIONS

Kwiyoung Park: drafting/revising the manuscript. Tomoko Oeda: drafting/revising the manuscript. Hideyuki Sawada: drafting/revising the manuscript.

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