Consultant360 Multidisciplinary Medical Information Network

CASE IN POINT

PEER REVIEWED

Pseudobulbar Affect

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A 75-year-old man presented to the emergency department (ED) with constant, unremitting vocal outbursts and confusion. The vocal outbursts had been present for the past 4 months and had progressively become more frequent over the past 24 hours. On further history-taking in the ED, the patient's wife reported that her husband had become more confused over the past 24 hours and did not respond to her questions despite her increasing her tone of voice.

Results of urinalysis done in the ED were significant for 10 to 15 red blood cells per high-power field (HPF) (reference range, 0-2 per HPF), 100 mg/dL protein (reference value, no protein), 5+ bacteria per HPF (reference value, no bacteria), and positive nitrites; large leukocytes also were seen in the urine. The patient was empirically treated with antibiotics in the ED for symptomatic urinary tract infection (UTI) and was admitted to the internal medicine service for further evaluation.

A consulting neurologist diagnosed Alzheimer disease accompanied with agitation/aggression and recommended an *N*-methyl-D-Aspartate (NMDA) receptor antagonist. A consulting psychiatrist diagnosed generalized anxiety disorder and recommended lorazepam, 0.5 mg every 12 hours, and quetiapine fumarate, 300 mg once daily at bedtime. The patient was discharged from the hospital to continue care with close monitoring at a nursing/rehabilitation facility.

Over the next week, the patient's response to medical therapy was monitored. Signs and symptoms of the UTI resolved, the number of vocal outbursts decreased, and the confusion improved. Nevertheless, despite improvement, his vocal outbursts returned at the beginning of the second week after discharge. Nursing staff at the rehabilitation facility reported that the outbursts returned abruptly within 12 hours of administration of a dose of lorazepam.

At this point, the patient was seen by our on-call team, where he presented with loud, constant, unremitting vocal outbursts ongoing for 10 to 12 hours straight. It was difficult to assess the patient's mental status as the shouting became more frequent and seemingly undirected at anyone after 24 hours. Expert advice from physicians with experience in treating patients with similar presentations led us to consider pseudobulbar affect (PBA), a rare neurological disorder with unknown prevalence.

Using the criteria developed by Poeck published in 1969,[1] the diagnosis of PBA is largely clinical. According to the Poeck criteria, a cerebellar disease must be present, and 4 specific criteria must be met.[2] The emotional response of the patient to stimuli is situationally inappropriate; the patient's internal feeling and the respective affect are not closely related and pose a challenge to assessment; the patient cannot control the duration and severity of the episodes on his or her own in respect to the howling behavior; and the expression of howling or yelling behavior has no bearing on a patient's feeling of relief, and so patients of this complexity must be medically managed for relief.[2]

In our patient's case, underlying Alzheimer disease was present. He also met the first criterion, in that an inappropriate response was stimulated by nonspecific stimuli—in this case, the patient's symptomatology was observed to increase when someone suddenly approached him. Next, there appeared to be no close relationship between the emotional expression and the patient's internal mood at the time of an episode. The patient's blank facial emotional expression accompanied with vocal outbursts did not reflect his internal mood, thereby meeting the second criterion. The patient's paroxysmal, ritualistic episodes of vocal outbursts were extremely difficult for him to control; thus, the third criterion was met. And finally, no signs of emotional relief were seen despite the increasing intensity of the outbursts. All of the Poeck criteria were met, and a diagnosis of PBA was made.

The patient was started on dextromethorphan-quinidine, 20 mg/10 mg capsules, twice daily. This

combination medication is the first and only drug approved in the United States by the Food and Drug Administration for the treatment of PBA.[3]

As per the patient's wife's request to pursue hospice care, the patient's dextromethorphan-quinidine dosage was increased to 3 times daily. Additionally, lorazepam, 0.5 mg, was discontinued and a higher dose of 1 mg every 12 hours was begun. A 100-mg quetiapine fumarate tablet during the day was added to supplement the 300-mg tablet at bedtime. This therapy regimen proved to be effective in controlling the patient's symptoms, and he was observed to have a more than 90% decrease in vocal outbursts for the following 2 weeks of monitoring.

Shortly thereafter, the patient died in hospice care. Laboratory test results before the time of death showed a serum sodium level greater than 160 mEq/L (reference range, 135-145 mEq/L). It is theorized that underlying cerebral disease in this patient deteriorated midline brain structures, including the osmoreceptors in the hypothalamus, thus causing an unnoticeable dangerously high serum osmolality that proved to be silent in nature and led to sudden death in this patient.[4]

References:

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