INSTITUTE TO ENHANCE PALLIATIVE CARE

PALLIATIVE CARE CASE OF THE MONTH

Huntington's disease: how can palliative care help?

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Case:

A 43-year-old woman, WM was diagnosed with Huntington's Disease (HD) in September 1996. In 1997 WM stopped working due to disability related to her disease progression. Since 2004 she was living at a skilled nursing facility where her care had been stable until 4 weeks prior when her choreiform movements intensified, her oral intake declined, and she was unable to rest or sleep due to incessant body movements. Several medication changes at the nursing facility were unsuccessful and she was transferred to an inpatient hospice unit for end-of-life care. On exam the patient appeared younger than her stated age and cachectic. She was non-ambulatory. She had flailing involuntary movements of her arms and legs, which made a comprehensive physical examination difficult. Her speech was unintelligible and skin on her arms and legs showed bruises and open wounds that she sustained when she hit any furniture around her. The patient's husband and power of attorney's primary goal was to control her movements and allow her to rest.

Brief overview of Huntington's disease:

HD is a neurodegenerative disorder that is inherited in an autosomal dominant fashion so that each child of an affected patient has a 50% chance of developing the disease. Most patients develop symptoms in their forties and fifties. Symptoms are usually a triad of motor, cognitive, and psychiatric issues. The motor symptoms involve impairment in voluntary movements and appearance of involuntary movements called chorea (hence the name Huntington's chorea). Impairment in voluntary movements results in loss of manual dexterity, slurred speech, swallowing difficulties, problems with balance, and falls. The motor symptoms worsen in the middle stages followed by rigidity in the late stages of the illness. Cognitive symptoms manifest in the patient's inability to perform complex tasks with loss of speed and flexibility, progressing to global impairment in the later stages of the disease. Psychiatric symptoms most commonly include depression. Patients may also manifest irritability, anxiety, agitation, impulsivity, apathy, social withdrawal, and obsessiveness.

Treatment:

Medications do not alter the progression of HD. Patients are vulnerable to cognitive side effects of pharmacologic therapy. Motor symptoms are usually managed using either neuroleptic agents such as haloperidol and/or benzodiazepines. There are no medications to treat swallowing difficulties or rigidity that occurs later in the disease. Depression is treated with serotonin specific reuptake inhibitors such as sertraline and paroxetine.

The mainstay of therapy for HD remains non-pharmacologic management. Padding of furniture and weights on wrists and ankles will help reduce injuries and the amplitude of involuntary movements respectively. As swallowing difficulties progress, the food consistency can be softened and pureed. Eating is usually supervised, and the patient is given time to eat with no distractions. Caregivers should be trained in Heimlich maneuver. Cognitive impairment and difficulties in communication can be eased by giving the patient time to answer and asking closed-ended questions. Establishing routines around the patient and activities that adjust to his or her cognitive level help to decrease anxiety and allow cognitive stimulation. Identifying and avoiding triggers for emotional outbursts and providing family and caregiver supports are measures to help manage behavioral manifestations of the disease.

Survival:

Average survival time after diagnosis is 15 to 20 years. Disease course is variable, and every patient is unique, which makes prognostication difficult.

Back to the case:

Initial attempts to sit the patient in a padded wheelchair and feed her proved unsuccessful. A trial of Phenobarbital was initiated to allow her to rest and monitor response. All furniture was removed from her room. Several mattresses were placed on the floor and padding was applied to the walls. Over the course of 3-4 days the patient's movements subsided, and she was able to stay awake. Her appetite improved, and she drank shakes. Feeding times were unique; one staff member would hold WM in her lap while the other would feed her. Over a period of 2 weeks, WM thrived.

Although she was incontinent for bowel and bladder, she would call out to the nursing staff when she needed care. Her room was decorated with her favorite sport team jerseys and family photos. Her husband visited daily and was able to participate in feeding times, and together they watched sports events on TV. WM's only medications were oral fluoxetine, laxatives, antacids and Phenobarbital. While we initiated our treatment plan to relieve distress and attempted palliative sedation therapy as a means to relieve what appeared to be refractory symptoms at the end of life, the patient's response was so dramatic that we now planned for her care for the next several weeks at a nursing facility. Our objective was to achieve a smooth transition for WM, and we encouraged nursing facility staff to visit the hospice unit and witness her personal care and feeding. The patient was eventually transferred and has continued to be stable at the nursing facility. She has not required any medication adjustments.

References:

1. Aubeeluck A, Wilson E. Huntington's disease. Part 1: essential background and management. Br J Nurs. 2008;17(3):146-51.
2. Rosenblatt A, Ranen NG, Nance MA, Paulsen JS. A Physician's Guide to the Management of Huntington's Disease. Second Edition. Huntington's Disease Society of America 1999.