Clinicopathological Conference / HMS Dementia: A Comprehensive Update

June 9, 2016, 10:00 a.m., Fairmont Copley Plaza Hotel
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PRESENTATION OF CASE

A 70-year-old woman was seen in the Memory Disorders outpatient clinic of this hospital because of gradually progressive cognitive difficulties involving word-finding. She was interviewed with the assistance of her daughter.

The patient reportedly was well until approximately 8 years before this evaluation, when gradually progressive word-finding difficulties developed, associated with confusion about the meaning of some words and increased “egocentric behavior”, in which she more frequently spoke about herself. For example, during a conversation about a recent family event, she did not understand what the word “punchbowl” meant. She had a good memory for recent events of her life, recounting multiple stories (at times to an excessive extent during history-taking, requiring redirection) and an excellent sense of direction. She exhibited an uncharacteristically "matter of fact" reaction to the death of a sibling and her dog (she was less upset than her family expected). She occasionally exhibited mildly inappropriate behavior (would say "I love you" to people to whom she was not particularly close).

The patient was right-handed. Approximately 10 years earlier, she had had a tick removed and was treated with antibiotics after a positive test result for Lyme disease, without further complication. At age 7 years, she had fallen out of a tree and hit her head, without loss of consciousness or hospitalization. There was no history of recent head injury, stroke, seizure,
transient ischemic attack, meningitis, encephalitis, HIV exposure, thyroid disorder, heart disease, hypertension, high cholesterol, diabetes mellitus, liver disease, kidney disease, cancer, pulmonary disorders, exposure to heavy metals, or learning disability. Her only surgical history included three cesarean sections. Her medications were a multivitamin, vitamin C, ginkgo, biloba, calcium, magnesium, and zinc, and she had no allergies. Since the death of her husband 10 years earlier, she had lived alone in another state and was able to perform all instrumental activities of daily living independently, including managing her money, paying bills, and volunteering in the community (visiting senior centers to play piano and attending regular community choir practices). She drank alcohol once or twice per month and did not smoke or use illicit drugs. She was a former teacher, who later worked in business with her husband until his death. A brother in his 60s had Parkinson’s disease and a sister in her 80s had Parkinson’s disease and dementia. Her daughter and her other five children and grandchildren were healthy. On examination, she was alert, attentive, well-groomed, cooperative, and pleasant. The blood pressure was 120/70 mm Hg and pulse 78 beats per minute and regular, and the weight was 68.6 kg. On cognitive assessment with the Blessed Dementia Scale (a scale with scores ranging from 0 to 27, with higher scores indicating more severe cognitive impairment), she did not know the name or the street of the hospital (of note, she was from another state) and received a score of 2 errors. The Clinical Dementia Rating (a scale with scores ranging from 0 to 3) was rated as 0, indicating very little or no cognitive impairment. The neurologic examination was normal, including the second through twelfth cranial nerves, power, bulk, tone, and coordination, stance, and gait, with deep tendon reflexes 1+ throughout. The platelet count was 362,000 per cubic millimeter (reference range, 150,000-350,000) and the level of folate was greater than 20.0 ng per milliliter (reference range, 3.1-17.4); the hematocrit, hemoglobin, white-cell count, and blood
levels of vitamin B12, electrolytes, glucose, calcium, total protein, albumin, globulin, thyrotropin were normal, as were tests of renal and liver function. A rapid plasma reagin test was nonreactive, and testing for antibodies to Borrelia burgdorferi was negative. The patient was referred for neuropsychologic testing and a brain MRI scan and for participation in a clinical research program.

On neuropsychologic testing, results indicated average premorbid intellectual abilities based on a screening measure of nonverbal abilities. Language impairment was evident on tasks of confrontation naming, fluency (predominantly semantic fluency), verbal abstract reasoning, and comprehension of individual words. She had fluent speech, but with significant word finding difficulty and occasional paraphasic errors, including difficulty comprehending words such as “misplace,” “bored”, or “pessimism.” Performance on tests of attention and executive functioning were variable, with her greatest impairment evident on language-mediated attentional tasks. Performance on attentional tasks that were not verbally mediated was generally normal. Similarly, memory testing was notable for normal performance on a measure of nonverbal memory and for significantly impaired performance on a measure of verbal memory. Verbal memory storage was difficult to assess because of her impaired acquisition and anomia. Nonverbal memory storage and performance on tasks of visuospatial ability were normal, and she did not endorse symptoms of depression or anxiety on self-reported measures of emotional functioning.

Speech pathology evaluation revealed fluent, articulate speech with word-finding pauses, vague word substitutions, and circumlocutions. She was somewhat tangential in her speech and was fixated in telling stories from her life, requiring frequent redirection to the topic at hand. A test of naming revealed substantial impairment with superordinate responses and impairment on
word-picture matching. Rare phonemic paraphasias were present. Auditory comprehension was reduced for words she no longer recognized and for sentences with complex syntax. Sentence repetition was intact. Written language assessment revealed normal reading and mild agrammatism in written language samples. A semantic picture-picture matching test revealed no evidence of visual agnosia.

A clinical diagnosis was made, and additional diagnostic imaging tests were ordered.