Frontotemporal Dementia

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KEYWORDS

• Dementia • Behavioral disturbance • Abulia • Inattention • Disinhibition • Impulsivity • Aphasia neurodegenerative disorder

KEY POINTS

- The initial description of this disorder is attributed to Arnold Pick in 1892.
- Involvement of the dominant hemisphere is characterized by primary progressive aphasia.
- The primary manifestations of the behavioral variant, reflective of nondominant hemisphere involvement, include the following: cognitive impairment including loss of initiative with apathy, abulia reflective of this loss of initiative including impaired executive function and lack of spontaneity, impulsivity, disinhibition, oral fixation, lack of sympathy and empathy, and repetitive behavior.
- There is well-recognized heterogeneity of clinicopathologic correlations.

OVERVIEW

A major challenge in the detection of frontotemporal dementia (FTD) is the potential for subtle presentation, which can result in serious consequences for patients unless the confounding early manifestations are correctly identified.[1–3] This point is illustrated in the following 3 case presentations:

Case Presentation 1

A 56-year-old right-handed man is brought to your office accompanied by his wife of 30 years. He is an accountant who is on administrative leave from his work because of increasing errors over the past several months. He has been described as a mild-mannered salt-of-the-earth type who regularly attends church and serves as a deacon. However, over the past year, he has made suggestive comments to several of his female coworkers, which is highly atypical for him according to his wife, and began looking at porn sites on his computer. This behavior is also viewed as highly atypical. He comes across as a bit reserved initially, and his wife provides much of

Disclosures: None.
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Neurol Clin 34 (2016) 171–181
http://dx.doi.org/10.1016/j.ncl.2015.08.007 **neurologic.theclinics.com**
0733-8619/16/$ – see front matter

his history. Generally, he has been in excellent health outside of hypertension of several years' duration and recently placed on a statin for dyslipidemia.

He has a college education, never smoked, drinks an occasional beer or glass of wine, and has never used illicit drugs. He has no history of sexually transmitted disease and has no history of significant head trauma. There is the question of dementia affecting his maternal grandfather in his 60s as well as a maternal uncle, but the specifics are not well known. There was an interesting recollection of a great uncle, described as a religious teetotaler, who ended up being killed in a brothel under mysterious circumstances.

The general examination is unremarkable. He does not come across as overly depressed and seems to cooperate fairly well with testing, often glancing over at his wife. On neurologic testing, he is somewhat delayed in his responses, with 2 mistakes on serial 7 subtractions, difficulty reproducing a design, and scores 27 out of 30 on the mini-mental state examination. The neurologic examination is otherwise quite normal. You order routine laboratory tests along with a serum vitamin B12 and folate level and thyroid profile with all coming back normal. A computed tomography (CT) brain scan and MRI brain scan both reveal fairly prominent frontotemporal atrophy, especially on the right. An electroencephalogram (EEG) reveals some bifrontal slowing but no epileptiform activity.

Case Presentation 2

A 62-year-old right-handed woman comes to your office with her female partner. They have lived together for the past 12 years and both have worked as elementary school teachers. The patient has been noted to having some communication issues in recent months, and it is beginning to be picked up by other teachers as well as the students. She has word-finding difficulty, with speech hesitancy, and uses inappropriate word choices at times. She has complained of some reading difficulty as well, and this is of particular concern to her as she has been an avid reader.

She has been quite healthy all her life and was a marathon runner at one time as well as a state tennis champion. She has been on no medications and continues an active daily exercise program but is noticing diminished exercise tolerance in recent months. She is quite selective in her diet and has maintained a weight of 120 lb during her adult life with a height of 5 ft 3 in. However, her weight is recorded at 114 lb in your office. The patient is surprised by this, although her partner has noticed a reduction in her typically finely toned musculature over the past half year. She has never smoked, drinks a glass of wine each evening, and does not use recreational drugs. The family history is noncontributory as she is adopted.

On examination, she looks quite healthy but has some obvious speech hesitancy and the suggestion of a mild bulbar type of dysarthria at times. The general physical is quite normal. She has some difficulty with naming and repetition, although comprehension seems to be intact. Her palate elevates symmetrically, and her gag reflex is intact. Her tongue is not of full bulk with suggestion of fasciculations. There is fair rapid alternating movement of the tongue. Strength and tone seems to be normal on motor testing, with some atrophy of the shoulder muscles; scattered fasciculations are noted. She does well with cerebellar testing and with gait, and no sensory loss is noted. Deep tendon reflexes are 3+ in both biceps and triceps and 3+ to 4− at both knees with a crossed adduction response, and there are several beats of clonus at both ankles, with the Babinski response positive on both sides and with a 2+ jaw jerk.

Brain imaging reveals the suggestion of frontal lobe atrophy, more prominent on the left. Blood tests come back fine. The cervical spine MRI is normal. Electromyography