Case:

A 57 y/o, previously healthy right handed woman, presents with symptoms of progressive dysarthria which started 10 months ago. Approximately 3 months ago, she noted left arm weakness. Her primary care physician, thinking she may have had a stroke, obtained an MRI of the brain, which was entirely normal. The only medication she takes is a multi-vitamin and a daily aspirin.

The patient denies any pain or tingling with this. She has noticed that the muscles of her left hand have gotten smaller and she has noticed some twitching of muscles of both arms. It takes her a few minutes longer to eat a meal than it used to. She has noted no weakness in the right arm or either leg. She has no shortness of breath.

On exam she has fluent, but mildly dysarthric speech. She is able to name 18 “F” words in 1 minute. Her husband fills out the ALS caregiver’s behavioral screen and results are unremarkable. She performs on the ALS cognitive behavioral screen with 100% accuracy. On cranial nerve testing, she has a weak, fasciculating and mildly atrophic tongue. She has mild weakness of the left arm. Mild atrophy is noted in distal and proximal muscles in that arm. She has fasciculations in muscles of the left arm and left leg.

EMG nerve conduction study is consistent with a suspected diagnosis of ALS. The patient and her husband are counseled on ALS. This includes discussion that in some patients it is associated with cognitive impairment. She is started on riluzole.

She is seen in follow-up every 3 months in the clinic. There is progression of her dysarthria and increasing weakness of the left arm and new weakness of the left leg. She starts to communicate almost exclusively with written communication because of worsening dysarthria. Her husband has noticed over the past month that she is starting to make spelling errors and grammatical mistakes.

It is now a year since she was seen initially in clinic. She is anarthric secondary to profound tongue weakness. She has mild weakness of neck flexors. She has moderate weakness of muscles of the left arm and leg. She has mild weakness proximally in the right arm, but normal strength distally in the right arm. She has diffusely brisk reflexes with bilateral Babinski signs. On cognitive testing, she can write 5 “F” words in 1 minute. On the ALS caregiver’s behavioral screen her husband notes a large change in multiple areas, scoring a total of 22/45.(cutoff score 36 for 86% detection accuracy) On the ALS cognitive behavioral screen, she misses points for attention, tracking and monitoring and initiation and retrieval, scoring a total of 12/20.(cutoff score 17 for 77% detection accuracy)

Discussion with the husband and the patient included acknowledgment of language dysfunction, as a known cognitive and behavioral manifestation that may occur ALS patients. Information is provided about other symptoms of frontotemporal dysfunction that may develop over time.
Rationale:

It is now well recognized that some patient with ALS may have associated cognitive changes, specifically fronto-temporal dysfunction. There is relative preservation of memory, praxis, and visuospatial skills while changes are found in behavior, language, and or personality. Initial features at presentation may include changes in behavior (behavioral variant frontotemporal lobar dysfunction, difficulty with expression of language but with relative preservation of comprehension (primary progressive aphasia or nonfluent progressive aphasia), or impaired language characterized by anomia in conjunction with impaired comprehension (semantic dementia). Another common finding in patients with fronto-temporal dysfunction is executive dysfunction causing difficulties with planning, organizing, abstracting, and prioritizing, along with impaired verbal fluency.

In distinction to Alzheimer’s dementia, the Mini Mental Status Examination (MMSE) is not a good screening tool to detect these changes since memory is generally preserved. Screening procedures for frontotemporal dysfunction include verbal and categorical fluency (if the patient has marked dysarthria, they may be asked to write instead of speak responses) and care giver based behavioral inventories. It is important to exclude underlying depression. Another benefit of using a caregiver questionnaire is that limitations posed by the patient’s physical disabilities are avoided. In addition, because patients with frontotemporal dysfunction may be oblivious to behavioral changes, caregivers are better at identifying them. The Neuropsychiatric Inventory (NPI), Frontal Behavioral Inventory (FBI), Cambridge Behavioral Inventory (CBI), and Frontal Systems Behavioral Scale (FrSBe) were designed by the cognitive neurologists to help screen for frontotemporal dysfunction. There is no consensus as to a preferred screening tool for frontotemporal dysfunction in ALS. The ALS Cognitive Behavioral Screen, developed by Wooley and colleagues, takes approximately 5 minutes to complete. It consists of a 15 item ALS specific behavioral questionnaire filled out by the caregiver, and an 8 item cognitive assessment of the patient that includes verbal fluency. It has been validated in ALS. The Penn State Screening Battery of Frontal and Temporal Dysfunction Syndromes takes approximately 20 minutes to administer. The longer (45 minutes) UCSF screen battery includes an ALS specific version of the frontal behavioral inventory, written verbal fluency, the ALS Cognitive Behavioral Screen, an emotional lability scale, and the Beck Depression Inventory-II. Easy access to the variety of screens described in ALS to evaluate cognitive impairment may be found on an NINDS common data elements website.

A wide range of frequency and severity of cognitive changes in ALS has been reported, likely in part related to the tests used, and how dementia was defined.

Cognitive changes can occur simultaneously, precede, or follow symptoms of motor neuron disease. The most common is cognitive changes to precede recognition of weakness. Screening for cognitive and behavioral change in ALS patients are recommended on a yearly
basis. Cognitive testing is not required if a patient has previously been diagnosed with severe cognitive impairment, declines to be screened, or lacks insurance to cover the testing.

Patients with cognitive impairment have a shorter survival and may be less likely to comply with recommendations concerning treatment such as use of non-invasive ventilation and willingness to obtain a feeding tube.²⁴-²⁵

Evidence Base:

The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:

- Screening for cognitive and behavioral impairment should be considered in patients with ALS (Level B).¹
- Screening tests of executive function may be considered to detect cognitive impairment in patients with ALS prior to confirmation with formal neuropsychological evaluation (Level C).¹


References:


