

When the Pieces of the Puzzle Don't Quite Fit: Assessing for Non-Alzheimer-type Dementias

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Introduction

If four million people have a dementing illness, and about half of those illnesses are caused by Alzheimer's disease, as many as two million people have non-Alzheimer dementias. There are dozens of non-Alzheimer type (atypical) dementias, yet diagnosis is often overlooked after an initial evaluation for whether the disease is reversible. Differences in care needs are also not identified.

Because of its prevalence, Alzheimer's-type dementia has been the focus of care and program development. Assumptions have been made that people with atypical dementias have the same or similar needs, thus are candidates for Alzheimer's care programs. Individualizing care often only considers the patient's premorbid personality or cultural preferences. This results in poor fit.

Families of people with atypical dementias are often mainstreamed into Alzheimer's support groups. They report frustration with having their questions answered or not receiving understanding support from professionals or other support group members. It is therefore important that health providers understand and assess for those behaviors that may indicate atypical presentations of dementia. The following article describes how the non-physician can assess for atypical dementias in order to suggest that a family seek diagnosis from a behavioral neurologist.

Guidelines for Professionals when Encouraging Families to Pursue a Diagnosis of Dementia

Often aged people with memory loss come to service providers having had no formal diagnosis. Families may have been unsure about whether to seek a diagnosis, feel that memory loss is normal for older adults, that the memory loss is not severe enough to warrant an evaluation, or are generally fearful of what may be found. The following list offers guidelines for professionals to discuss with families when encouraging to seek a diagnosis for dementing illness.

- Memory impairment severe enough to impair function
- Episodes of confusion
- Problems with finding or substituting words
- Change in abilities in day to day functions
- Personality change
- Changes in motor skills

Diagnosis of dementing processes

The three critical questions that are posed during the diagnostic process:

1. Is memory loss present?

The answer to the first question comes from a combination of data accrued by the primary care provider or health professional including the following:

A. History from the patient and their family

- Changes in memory and function from previous abilities
- Changes in behavior patterns
- History of mental health problems/substance use/OTC and prescribed medications
- Other symptoms
- Medical history and medications
- Social history
- Functional level
- Onset and duration of the symptoms
- Waxing and waning of symptoms

B. A mental status screen

- Mini-Mental Status Examination
- Clock Drawing
- Figure copying

C. Observation of behavior and interaction with others

- Language Function
- Ability to cope with noise or groups
- Motor Functions
- Evidence of paranoia or psychosis
- Ability to navigate the environment and see

D. Barriers to diagnosing memory loss

- Some patients can pass mental status tests despite functional limitations
- Patients who are either very bright at baseline or who have strong personality characteristic may be able to “bluff” their way through the evaluation
- The physician or diagnostician has a strong relationship with patient and some difficulty seeing decline
- The providers are concerned about the family motives in pursuing a diagnosis, especially in light of a history of family conflict
- Some conditions take years to diagnose. ALS with frontal lobe presentation, diffuse Lewy body disease, and other frontal lobe dementias frequently are misdiagnosed until several years after the family reports changes.

Once the provider is certain that cognitive loss is present, issues of cause must be addressed. The usual medical evaluation answers the second question using the following test results.

2. What could be causing it?

A. The goal is to rule out all other causes of progressive dementia

- History & thorough physical examination

- Imaging - should be negative
 - Rule out tumor, strokes, trauma, NPH
 - May see areas of localized atrophy
 - Can be CT scan, MRI, PET
- Neuropsychological testing
 - Reconfirm memory loss
 - Defines areas of weakness and therefore areas of brain affected
 - Begins the care plan
- Laboratory evaluation
 - CBC - anemia and blood dyscrasias
 - TSH - hypothyroid
 - Electrolytes - imbalances, renal disease, dyhydration
 - Blood sugar - diabetes
 - B12 & Folate - B12 deficiency
 - Urinalysis - infection, signs of renal disease
 - HIV, STD testing

B. Other tests as indicated

- Lumbar puncture - Tumor, infection, NPH
- EEG - Seizures
- Chest X-ray - Tumor
- Cardiogram, Holter monitor - arrhythmia
- EMG - ALS
- Overnight oxymetry - sleep apnea
- Sleep studies - restless legs
- Arterial biopsy - arteritis

All of the diagnostic data are compiled and interpreted to answer the last question....

3. Are the symptoms and course of the disease consistent with what we know as Alzheimer's disease?

A. If the symptoms are consistent with Alzheimer's Disease, the following may be noted:

- The history and neuropsychological tests will reflect losses in cognition.
- Imaging studies and laboratory tests will be negative.
- The symptoms and behavioral presentation will be consistent with AD
 - Slow onset, insidious progression
 - Global losses in cognition, planning, language, memory, and visual-spatial perception
 - Changes in short-term auditory and/or visual memory
 - Subtle intensifying of negative personality characteristics
 - Decreased ability to inhibit plus increased self-absorption
 - Decreased tolerance for noise, crowds, change
 - Increased symptoms with fatigue
 - Uneven symptoms presentation
 - Episodes of depression

- Gradual loss of functional abilities because of problems with planning and sequencing

B. You might suspect other diseases if the following symptoms are present (especially early in the disease process). This is a partial list of some of the more common atypical syndromes.

Presenting Symptoms	Possible Syndrome
Early onset of language problems, ataxia, incontinence	Normal pressure hydrocephalus
Early loss of language	Progressive aphasia, Frontal lobe degeneration, Corticobasal ganglionic degeneration (CBGD)
Severe disinhibition as evidenced by making poor decisions coupled with hyperactivity, or socially inappropriate behavior	Frontal dementia syndromes including Pick's disease, frontal lobe degeneration, ALS
Abulia (failure to initiate activities) Apathy	Frontal dementia syndromes including Pick's disease, frontal lobe degeneration, ALS
Changes in motor function, tremor, spasticity, weakness, uncoordination.	ALS, CBGD, Diffuse Lewy Body disease, Progressive supranuclear palsy
Psychosis	Bitemporal dementia, Diffuse Lewy body disease
Loss of vision	Visual variants of Alzheimer's disease, Progressive asimultanagnosia, Agnosia, CBGD
"Spells"	Seizure disorder, stroke disorders, amyloid angiopathy
Muscle wasting	ALS

Reasons for Pursuing a Diagnosis of an Atypical Dementia

Once the diagnosis of an irreversible dementing illness is established, responsibility for planning and providing daily care is assumed by family and allied health professionals. Enormous strides have been made in both the understanding of pathophysiology of chronic dementing illnesses and the development of research-based techniques for providing care to victims of Alzheimer's disease and related disorders (ARD) and their families. Yet, while neuroscientists have identified multiple variants of dementing illnesses, care improvements still focus primarily on typical presentations of Alzheimer's disease, assuming people with dementia have similar symptoms and therefore, similar care needs. Thus, care programming for people with dementia has become essentially a "one size fits all" strategy.

This approach falls short with patients presenting with atypical presentations, rare, or multifactoral dementias -- perhaps as many as 20% of the patient population with irreversible dementias. Identifying atypical presentations, whether histologically linked to Alzheimer's disease or not, can only help practitioners and caregivers to understand observed behaviors, distinguish between primary and secondary symptoms, and plan care accordingly. For example, people with Lewy body disease have special medication needs that, if not understood, can worsen their symptoms dramatically. Or, expecting a person with a frontotemporal dementia to respond to environmental modifications in the same way as a person with Alzheimer's Disease, will result in frustration and disillusionment in caregivers.

Care of the person with an atypical cortical degenerative syndrome may be modified to accommodate subtle differences in disease presentation or an entirely new strategy may have to be developed -- especially early in the disease. Caregiving professionals need to understand both the functions of the areas of the cortex affected and how these areas influence symptom presentation in order to make decisions about care.

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