Case Discussion in Neurology

Dealing a Patient with Dementia : Some Basic Concepts

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It is a common belief that patient with dementia presents with forgetfulness only. Dementia encompasses deficits in multiple domains of cognition. Presently there are very few treatment options for dementia. Few are coming out. Anatomical localisation is the basic requisite for diagnosis. Symptom exploration still is the best way to reach a diagnosis. Details of the deficit as well as the relative strength in different cognitive domains are the cornerstone of dementia classification. Conceptual approach is more needed than specific skill set when obtaining a history. This conceptual framework is needed for eliciting specific information about the patient's cognition, behavior and daily function. Age, handedness, education, occupation is important consideration. Onset, duration, progression and chronology of the symptoms better characterize the dementia. History of other neurological, medical diseases, family history is important as well. Neurological and neuropsychological tests may give important clues. Everyone of us should be very careful using these tools, as often they are overlapping in eliciting results. At the same time, multiple tests are needed frequently to elicit a single domain. Neuroimaging adds to the armamentarium of dementia diagnosis. Needless to say, it is a comprehensive approach to deal cases of dementia.

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Common belief is that dementia means some problem in memory and people complain of forgetfulness. In DSM IV, memory problem is a compulsory criterion for defining dementia. In DSM V, the term "Dementia" has been changed to "Neurocognitive disorders" and memory problem is no more compulsory but one of the criteria. So, dealing a patient with dementia we have to know about the domains of cognition. They are executive function, attention, memory, visuospatial function, language and behavior. Before we go into the details, let's explore some cases.

Case 1 :

A 78-year-old right handed man with 14 years of education was brought into the clinic urgently for assessment of the acute onset of "confusion". He had been evaluated 4 days prior to assess his 2-year history of the insidious onset of progressive anterograde memory loss that affected his usual instrumental activities of daily living. Neuroimaging at that initial visit revealed bilateral hippocampal and parietal atrophy. At the initial visit, he was diagnosed with Alzheimer's disease. He had been well over the intervening 4 days but in the morning, his family noted that he appeared confused and requested an urgent reassessment. His medical history was remarkable for hypertension, type 2 diabetes mellitus and hypercholesterolemia.

Editor's Comment :

- Symptom elaboration is very crucial, as deficits in most of the other domains usually present as memory deficit erroneously by the patient.
- Deficit as well as the relative strength of the cognitive domain are to be handled with equal importance, as dementia can be characterized best by this approach.
- Conceptual approach is more needed than specific skill set.
- Real life incidences are of more values than results of the tests done in clinics.
- Dementia approach is a comprehensive one encompassing all the neurological, psychological, behavioral symptoms along with necessary neuropsychological tests, imaging.

On cognitive testing, he scored 17/38 on the Short test of Mental status[1,2]; he had scored 25/38 at the initial evaluation 4 days earlier. The elements of each test and his scores for each area are listed below. His elemental neurologic examination was normal on both visits.

Short Test of Cognitive Domain	Score at initial visit	Score at follow-up visit
Total score	25/38	17/38
Orientation	6/8	5/8
Attention	6/7	4/7
Learning (number of trials)	4/4 (1 trial)	4/4 (2 trials)
Calculations	4/4	0/4
Similarities	2/3	2/3
Construction/drawing	3/4	0/4
Information	4/4	¾
Delayed recall	0/4	0/4

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Given the acute clinical change and the results of the clinical assessment at the second visit, he was clinically diagnosed with an acute left parietal stroke and sent to the emergency department for urgent neuroimaging. Brain CT revealed an acute left parietal intracerebral hemorrhage.

This case illustrates that cognitive testing can aid with the localization of brain lesions. When comparing the two test results, a marked and focal difference is seen in this patient's ability to perform calculations and draw items, and both functions localize to the left parietal lobe. Given the acute change reported, the clinical conclusion was an acute stroke affecting left parietal lobe.

Conceptual approach is more needed than specific skill set when obtaining a history. This conceptual framework is needed for eliciting specific information about the patient's cognition, behavior, and daily function. History from collateral informant is very important. A patient's cognitive profile includes not only the patient's weakness but also his relative strengths. History from collateral informant is very important to clarify the symptoms, to determine the chronologic progression of the signs and symptoms and to determine the course of the progression (gradual, fluctuating or stepwise) (Table 1).

Identifying Patient Features : Depending on the age when the first symptom develops, the dementia can be early onset (before age 65) or late onset (age 65 or older). Although not exclusionary, older age of onset likely predicts that a neurodegenerative process is the etiology, while genetic, vascular, or metabolic causes are more often found in younger patients. It is important to note that ischemic cerebrovascular disease is commonly associated with neurodegenerative disorders, such as AD. Handedness is a marker for lateralization of cognitive functions in the brain. In 95% of right-handed patients, language function is lateralized to the left hemisphere. In contrast, 22% of left-handed patients have language centers either in the right hemisphere or both hemispheres. Change in use of preferred hand can imply either weakness or apraxia on the dominant side. Education andoccupational history provide information about the patient's premorbid level of intelligence and function. It is also helpful for the interpretation of cognitive test results.

Table 1 — *Comments to Consider by Presenting Symptoms*

First Symptom Noticed	Affected Cognitive Domain	Disorders to consider
Repeats him/herself; rapidly forgets conversation	Anterograde memory loss	Alzheimer's disease
Cannot recall people he/she sees on the street; does not recognize familiar people at a party; cannot recognize his or her own house	Prosopagnosia	Semantic dementia variant of frontotemporal dementia
Cannot align things; has problem seeing, reading, blurry vision; cannot fill out a form; cannot find things in the refrigerator; cannot read a map; misplaces items; gets lost/geographic disorientation	Visuospatial dysfunction	Alzheimer's disease (posterior cortical atrophy variant), dementia with Lewy bodies
Inability to fix things	Apraxia, executive dysfunction, visuospatial dysfunction, attentional dysfunction	Corticobasal syndrome, Alzheimer's disease, dementia with Lewy bodies, vascular cognitive impairment
Forgets words; describes words; talks around them; mixes up words; mispronounces words; forgets what a word means	Language (anomia)	Primary progressive aphasia (nonfluent, logopenic, or semantic variants)
Sometimes able to do things and sometimes appears more confused and cannot do things	Attention (fluctuations)	Dementia with Lewy bodies
Cannot plan, multitask, or stay on task; must do everything in single steps; cannot combine tasks	Executive dysfunction	Alzheimer' disease, vascular cognitive impairment, behavioral variant of frontotemporal dementia (behavioral abnormalities must also be presentfor this diagnosis), dementia with Lewy bodies

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Case 2 :

A 55-year-old right-handed man with17 years of formal education presented with the insidious onset of progressive word-finding difficulties of 3 years in duration. In conversations, he knew that he should know some words, but he had forgotten what they meant. Although he could recognize colleagues, he did not recall their names. He had no change in personality or behavior.

On cognitive testing, he scored 30/38 on the Short Test of Mental Status[1, 2], with the following breakdown of the subitems: orientation 8/8, attention 5/7, learning 4/4 words in three trials, calculations 4/4, construction/ drawing 4/4, information ¾, and delayed recall ¾. His elemental neurologic examination was normal.

Additional cognitive testing revealed normal performance on the Rey-Osterrieth Complex Figure Test[3] (a test that involves copy and recall of a complex figure and measures several functions, including visuospatial ability, planning, and visual memory), Logical memory subtest of the Weschsler Memory Scale[4] (a test of memory in which the patient learns and recalls elements of a story), and Rey Auditory Verbal Learning Test[5] (a test of memory in which the patients learns and recalls a list of unrelated words). However, he obtained 5/30 on the Boston Naming test, generated seven animals for semantic fluency, and generated 25 words on verbal letter fluency (C, F, L).

Brain MRI revealed focal left anterior temporal lobe atrophy. Brain single-photon emission computed tomography (SPECT) revealed left more than right anterior bitemporal hypoperfusion. A diagnosis of semantic variant primary progressive aphasia was made.

This case illustrates the limitation of certain bedside screeningtests to adequately evaluate a patient to reach a diagnosis. The patient had scored reasonably well on the Short Test of Mental Status, losing points for difficulties in learning words as additional points are deduced for learning after one trial. This test also does not adequately evaluate language; therefore, additional language tests were administered as language deficits were the patient's presenting symptom. Furthermore, his deficits were clinically localized to the left temporal lobe, which was confirmed on MRI.

It is important to say that, no single test can cover the entire aspect of cognitive domains. On the contrary, there is usually no one is to one relationship in a test (that means no single test can measure a single domain only). Therefore, we have to depend on history crucially. We usually try to evaluate the predominant cognitive domain affected first. At the same time, the spared cognitive domains are evaluated with equal importance. Because both these deficit and strength gives the actual picture. Systematically we explore the cognitive domain involved first, then we ask for the behavioral aspect. Next, we look for any other neurological, medical illness. We search for family, occupational history next.

With this back-up we start clarifying symptoms and the course of the illness. Requisites for this are the information about the onset (insidious or acute), nature of the symptoms, course of the condition (gradually progressive, stepwise, fluctuating, or improving) and duration. Correct assessment of these information leads us nearer to provisional diagnosis and differentials. Like other streams of neurology, our endeavor is to localize anatomically, then pathology of the lesion and lastly the etiology. What is unique in cognitive neurology, anatomical localization is not very concrete. One single anatomical site is responsible for may cognitive domains and multiple sites may involve in a single cognitive processing. The organization of brain adds more complexity. A single cognitive domain has different level of processing depending on different anatomical sites. For example, visual activity in brain starts with occipital cortex subserving the elemental function (brightness, contrast, colour, size etc). Then gradually, depth, movement perception comes in through different levels of occipital cortex (V1, V2, V3, V4…..V8 etc.). As it proceeds further, ramification of that domains takes place. If it moves to the (ventral path) temporal cortex, particularly the inferior temporal cortex ("Whatpathway") object recognition results. If it moves to the (dorsal path) parietal cortex ("where pathway") spatial processing results[6]. On top of this different cognitive modalities interact among themselves through association cortex. For these reasons, we are gradually shifting to the "network" concept[7].

Considering all these complexities and paradigm shift, we still bang on the basic approach. Depending on onset, progression and duration we can deal with the pathology and etiology.

Onset and Duration

Acute (seconds to days) : stroke, infection (viral bacterial), metabolic.

Subacute (weeks to months) : metabolic, infection (Creutzfeldt-Jakob disease, fungal, spirochete)

Chronic (years) : neurodegeneration, chronic cerebrovascular disease

Progression

Improving : stroke, infection (viral bacterial), metabolic, delirium

Static : stroke (fixed deficit)

Fluctuating : epilepsy, paraneoplastic, metabolic, dementia with Lewy bodies (DLB).

One of the important aspects of dementia diagnosis is to explore the chronology of symptom appearance. Memory, executive dysfunction, language, behavior all are affected in both Alzheimer's disease (AD) and behavioral variant of frontotemporal dementia (bV-FTD), but the sequence is different. Certain Neurodegenerative syndromes may evolve to incorporate other distinct clinical syndrome, and this is being reflected in revised diagnostic criteria.[8, 9] For example, patients presenting with progressivenon-fluent aphasia (PNFA) can progress over time to develop corticobasal syndrome (CBD), progressive supranuclear palsy (PSP), or bV-FTD[8].

In AD the usual sequence of affected domain is: memory, executive function, language, visuospatial and lastly behavior.

In bV-FTD the usual sequence of affected domain is: behavior, executive function, language and lastly memory.

The chronologic approach can also determine if differing pathologies are developing in the same person. It has been reported that the patient develop the core features of DLB after developing typical AD many years earlier.[10, 11]

The ability of the patients to perform their activities for daily living (ADL) [grooming, managing hygiene, bathing, eating, dressing etc.] should be determined. Changes in the basic ADLs range from fully independent to needing reminders, requiring help, and fully dependent.

Major deficit seen on testing	Pattern	Example conditions
Orientation, delayed word recall	Amnesic	Mild cognitive impairment (amnestic) [MCI], Alzheimer's disease.
Planning and monitoring, attention, sequencing (eg. Three-step command) word list generation for letters	Executive dysfunction, frontal-subcortical dysfunction	Dementia with Lewy bodies (DLB), Parkinson disease dementia, vascular dementia.
Drawing	Visuospatial impairment	Posterior cortical atrophy (PCA), DLB, corticobasal syndrome (CBD).
Naming, repetition, writing	Aphasia	Primary progressive aphasia.
Normal testing	Not applicable	Can be seen in bV-FTD, subjective cognitive impairment.

Examination element	Examination finding	Disorders to consider
Extra ocular movements	Initiation of saccades (indicates an ocular apraxia): PCA, CBD. Slow saccade velocity: PSP.Limitation of extra ocular movement: PSP	CBD, PSP, PCA.
Upper motor neuron signs	Pyramidal distribution weakness (weakness pattern in arms: extensors >flexors; in legs: flexors >extensors), hyperreflexia, extensor plantar response.	Stroke, CBD, intracranial mass lesion.
Assessment for parkinsonism	Bradykinesia, bradtphrenia, masked facies, limitations/absence of downgaze: PSPRigidity with or without cogwheeling[distinguish between axial (PSP) versus appendicular (Parkinson's disease [PD], DLB, CBD) rigidity], rest tremor(not typically seen in DLB, PSP, CBD); fatiguing (rapid alternating movements eg. Finger tapping, opening/ closing hands, festinating gait, progressive hypophonic speech); early postural instability: PSP.	Parkinson disease with dementia (PDD), DLB, CBD, PSP, Stroke.
Assessment for early focal cortical dysfunction	Balint syndrome (some or all components): simultanagnosia [inability to see objects simultaneously]; optic ataxia (inability to reach a target under visual guidance); ocular apraxia (inability to move the eyes to a target purposefully). Gerstmann syndrome (some or all components): right-left disorientation, finger agnosia, acalculia, dysgraphia Dressing apraxia Ideomotor apraxia Language (anomia)	PCA, CBD (bilateral parietooccipital area kesion) PCA, CBD, Logopenic progressive aphasia. (Inferior parietal lobule, Angular gyrus) PCA, Corticobasal syndrome (CBS). CBS, AD. Primary progressive aphasia (PPA).

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We have to keep it in mind that, first we have to ascertain the true cause of the symptom or functional decline. Suppose a patient who is no longer able to use to use a mobile phone could be demonstrating difficulty with executive function (affecting task setting and monitoring to track the appropriate steps), visuospatial deficit (to recognize the mobile itself or its parts), language impairment (deficit in the meaning of the number engraved in the mobile), apraxia (problem I organizing or sequencing of movements) or simply physical weakness. A combination may play role.

Medication history, medical and neurological past history that could affect cognition like endocrinopathies, chronic organ failure, and chronic neurologic disorders like Parkinson disease, stroke, epilepsy, multiple sclerosis (MS) should be noted. A history of concussion or traumatic brain injury (TBI), confusion following recent surgeries, alcohol, smoking, and illicit drug use should be sought for.

A few words about the cognitive tests must be included here.

Lastly, to discuss something about neurologic examination findings in neurodegenerative disorders, the following table would help.

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The post-operative fart is music to the surgeon's ears.

— Moshe Schein