

CHAPTER 14

Neurobehavioral Disorders (Organic Brain Syndromes)

The neurobehavioral disorders are a group of conditions in which brain damage or dysfunction produces primarily intellectual (cognitive) and behavioral change. The behavior change can be in intellectual functions, such as memory impairment or language dysfunction or emotional, as seen in personality and behavioral change accompanying frontal lobe lesions. In widespread diseases such as dementia and delirium, many aspects of behavior will be affected, whereas with focal lesions e.g. strokes, tumors more restricted abnormalities such as inadequate verbal comprehension and expression (aphasia) may be primary neurological dysfunction that is demonstrated on examination. These syndromes are encountered in everyday medical practice and can usually be diagnosed with careful history, brief mental status examination, and some basic knowledge of the clinical syndromes.

EVALUATION

The clinical evaluation of the patient with behavioral change requires history and mental status examination tailored to elicit specific organic symptoms and signs. The history should be taken from both the patient and someone who knows the patient well because the patient is frequently unable to give an accurate history of the illness because of their memory and cognitive impairment. In the history of the current illness, it is important to ask exactly how the patient's behavior has changed and over what period of time this occurred. From the patient with behavioral change in whom organic brain disease is suspected, specific historical information should be obtained as outlined in Box 14-1.

BOX 14-1 Pertinent General History

1. *Age.* Behavioral change in the elderly is most likely organic and not functional psychiatric disorder.
2. *Education and vocation.* This information establishes level of expectation on mental status examination.
3. *Handedness.* Knowledge of handedness is important in assessing laterality of focal lesion in aphasic patients.
4. *Review of medical history.* Endocrine, vascular, renal, hepatic, pulmonary, and many other medical diseases can affect brain and behavior.
5. *Review of neurologic history.* Strokes, head trauma, seizures, and parkinsonism are important causes of brain disease affecting behavior.
6. *Family history.* There are many familial forms of dementia: Huntington's disease, Alzheimer's disease, Pick's disease.
7. *Medications.* Many medications or combinations of medicine can produce delirium and sometimes dementia-like state.
8. *Alcohol or illicit drug abuse.* May cause psychoses, depression, anxiety.
9. *Toxic exposure.* Insect sprays, hydrocarbon vapors, and other toxins affect the brain.
10. *Psychiatric history.* Depression is well known to masquerade as dementia in the elderly (pseudodementia). Any significant psychiatric illness will effect cognitive functioning.

Specific Review of Systems for Organic Behavior Change

During review of systems for organic behavior change, several changes must be observed (Box 14-2).

After careful history mental status examination such as that outlined in Chapter 1 should be performed. Neurologic and physical examinations are also necessary. In dementia (global impairment of intellectual-cognitive capability occurring with clear sensorium and which interferes with activities of daily living) and delirium (agitated confusional inattentive state occurring with impaired sensorium), neurologic examination is frequently normal with exception of mental status exam, but elicitation of localizing neurologic signs suggests that focal lesion is responsible for behavior change. Such signs as papilledema or stiff neck suggest specific intracranial disease (e.g., mass lesion, intracranial hypertension, meningitis). Physical examination can reveal cause of behavior change such as underlying medical conditions (see chapters 18 and 22).

BOX 14-2. Organic Behavior Change

- 1 Memory difficulty
- 2 General loss of intellectual ability
- 3 Word-finding problems in speech
- 4 Geographic disorientation (e.g., getting lost)
- 5 Personality change
- 6 Inattention or difficulty concentrating
- 7 Bizarre behavior (e.g., nocturnal wandering, improperly clothed)
- 8 Lethargy or excessive sleepiness

ACUTE CONFUSIONAL STATE (DELIRIUM)

An acute confusional state (variously labeled delirium, metabolic encephalopathy, or acute brain failure) is as the name implies—rapidly developing mental change in which patient looks and acts confused. More specifically, clinical picture is one of altered and fluctuating alertness, incoherent or clouded mental processes, marked inattentiveness, and usually change in activity level featuring either agitation or lethargy. In addition, hallucinations, most often visual or tactile (e.g., spiders crawling on the skin); emotional changes such as suspiciousness, irritability, euphoria, or violence; indistinct speech; and autonomic changes such as dilated pupils, tachycardia, and diaphoresis can be present. The course of the condition is typically rapid in onset (hours to days) with considerable fluctuation in symptoms during the day and distinct propensity for nocturnal exacerbations. If cause is not determined and proper treatment instituted, mental state will deteriorate, and patient will become stuporous and comatose and can eventually die.

Attention is the patient's capability to focus on a specific mental task without distraction. Attention has two components – attaining attention, which is function of ascending reticular activating system and sustaining attention (concentration) which is combined function of both cerebral hemispheres. To test whether patient can attain attention, observe the patient during the evaluation and note if there is distractibility. To test whether patient can sustain attention, evaluate digit repetition in which examiner asks patient to repeat series of numbers. Keep in mind that normally patient should be able to remember up to seven digits and this is why telephone numbers never exceed this number. Also, utilize random letter test in which series of

random letters is given and patient is told to raise their hand which examiner says the target letter, for example “A”. Lastly, the serial “seven” or “three” subtraction test is another test of sustained attention. Unilateral inattention or extinction to double simultaneous sensory stimulation is usually caused by focal right parietal lesions and usually occurs on left side of body (see Box 14-4).

The causes of confusion are many and closely parallel those of stupor and coma. Predisposing factors include increasing age, early dementia, previous brain damage, and previous history of delirium. The cause of confusional state is almost always medical or neurologic rather than psychiatric (e.g., mania, schizophrenia); therefore physician's first obligation is to review patient's medical status with particular attention to medications. Older people are especially sensitive to medications and can become confusional on even small doses of standard prescription drugs. Common offenders are psychotropics, antiparkinsonism drugs, diuretics, sedatives, strong analgesics, and of course alcohol and illicit drugs. Frequently the combination of drugs produces organic behavioral change.

The next area to consider is systemic illness. Some of the most common are generalized metabolic dysfunction; organ failure (e.g., cardiac, renal, pulmonary, or hepatic); sepsis; marked hypertension; endocrine disease; or electrolyte disturbances. Neurologic disease such as increased intracranial pressure, brain tumor, meningitis, or acute cerebral infarction (particularly in frontal lobe or inferior parietal region of non-dominant hemisphere) can cause confusional state. In many cases, however, combination of factors is involved. Nowhere is this truer than in postoperative patient, in whom medications, electrolyte fluctuations, fever, and primary disease combine to produce abnormal metabolic milieu for brain. Alcohol withdrawal, which can itself produce classic delirium, can also be factor in any patient who stops drinking on entrance to the hospital. Sleep loss is another very important factor that is well known for its capacity to produce and perpetuate confusional behavior.

The evaluation of confused patient is similar to that for coma and involves complete history, physical and neurologic examination, after which blood counts, chemical screens, and drug screens are obtained. The remaining evaluation is largely dictated by what is found at this point. Spinal tap or brain imaging (CT/MRI) is indicated if primary neurologic disease is suspected or patient does not respond to medical management within 24 to 48 hours.

Mild Cognitive Impairment (MCI)

These patients report memory complaints which can be confirmed by an observer who knows the patient and objective memory impairment can be confirmed by mental status testing. This memory impairment does not interfere with patients activities of daily living. The patient has normal cognitive-intellectual testing and shows no evidence of dementia. This may represent the initial symptomatic stage of Alzheimer's disease and progression to dementia occurs at rate of 12% per year. Neuropathological studies of patients with MCI show similar changes to those patients with Alzheimer's disease. It is important not to confuse the cognitive inefficiency of depression in which concentration and thinking is subjectively impaired (pseudodementia) from MCI. The patient with MCI has objective memory abnormalities on mental status examination whereas the patient with depression merely reports perceived difficulty with memory and concentration, but can report no instances of memory failure and objective memory testing is normal. Also, as patients “age” and develop medical conditions e.g. hypertension, dyslipidemia, diabetes, heart disease, renal impairment and take medication which may affect the CNS, memory may be affected and patients forget names, misplace objects and occasionally forget to pay a bill; this is probably “normal cognitive aging” and is quite similar to MCI. Both types of patients utilize compensatory strategies (e.g. keeping calendars and lists to allow them to

continue normal functioning). MRI and CT usually show no brain atrophy or shrinkage unless there is progression of MCI to dementia.

DEMENTIA

Dementia is very common clinical entity in which there is progressive deterioration in intellectual and social adaptive functions; there is usually associated behavior and personality change. There are multiple cognitive deficits including memory impairment, aphasia, apraxia, agnosia, disturbances in executive functioning, impaired problem solving associated with behavioral and personality deficits, which interfere with activities of daily living. Dementia represents decline from higher level of functioning such that patient can no longer carry out previously performed level of functioning. This distinguishes dementia from mental retardation in which cognition is impaired, but there has been no declining course. Although dementia was previously divided into presenile and senile varieties, this distinction seems to be unjustified; the dementing illnesses seen before age 65 appear identical to those seen thereafter. It is realized that some dementia can be reversed. In general, it is younger patients (40s or 50s) who will prove to have reversible conditions; nevertheless, full evaluation should be made in all demented persons.

Alzheimer's Disease [AD]

Alzheimer's disease is by far the most common cause of dementia, probably constituting more than two thirds of dementia cases. Four million patients have this disorder and this will increase to 7 million by early in this century. Risk factors include age, genetic factors, apolipoprotein E status (homozygous patients who carry two APOE E-4 alleles), female gender, and prior head injury. Onset of cognitive decline is insidious, and its course is long (10 years average with some patients living longer). Initial symptoms are usually a subtle personality change and recent memory difficulty. The personality change is usually characterized by apathy, lack of interest, hypochondriasis, and often an accentuation of previous personality traits. If examined carefully, patients can show difficulty with abstract reasoning on proverb interpretation, constructional impairment on drawings, and subtle word finding difficulty. There are no gait, motor, or coordination, abnormalities in the early stage. The standard neurologic examination is usually normal except for primitive reflexes e.g. grasp reflex, palmomental reflex, snout reflex. As disease progresses, aphasia, agnosia, apraxia, and inattention occur along with marked worsening of memory and other intellectual functions. The patients, however, remain alert and can be somewhat hyperactive.

No laboratory test can confirm diagnosis during life or identify individuals at risk for development of AD. The purpose of laboratory evaluation is to exclude alternative etiologies. Blood work should evaluate treatable conditions – thyroid dysfunction, B12 level, CBC, ESR, biochemical profile, urinalysis, toxicology, heavy metal screen, syphilis and HIV serology, chest radiogram, electroencephalogram and CSF exam. The following clinical features make diagnosis of AD unlikely –sudden onset and rapid progression, focal neurological deficit, seizures. Of patients who are demented, 10 to 20% have a potentially treatable etiology. Brain imaging (CT/MRI) excludes vascular or mass lesions and hydrocephalus. CSF shows infectious etiologies. Typical CT/MRI findings in AD include enlarged ventricles and subarachnoid spaces; however, in early stages, CT/MRI may be normal and single photon emission computed tomography (SPECT) might show reduced perfusion in temporal and parietal regions. SPECT is of value when CT/MRI shows no abnormality.

Treatment is largely directed toward symptomatic enhancement of memory by affecting the cholinergic and glutaminergic systems. Currently available centrally acting cholinesterase

inhibitors which enhance the memory system include: 1) Aricept (donepezil) with daily dose of 5 or 10 mgm dosed once daily; 2) Galantamine (Reminyl) with daily dose of 8 to 24 mgm dosed twice daily; 3) Rivastigmine (Exelon) at dose of 3 to 12 mgm dosed twice daily. They have modest effect on memory, cognition and daily living activities. Major side effects are gastrointestinal (e.g., nausea, vomiting, diarrhea, anorexia), insomnia, leg cramps, vivid dreams and these medications should be avoided if patient has glaucoma or urological symptoms. Memantine (Namenda) is an N-methyl D-aspartate receptor antagonist, which is used at dose of 5 to 10 mgm twice daily and may be used in combination with Aricept or alone. The role of neuroprotective strategies e.g., antioxidants, anti-inflammatory drugs, statins, and possibly selegine drug previously used in Parkinson disease is not clear.

Pathological changes in AD are initially seen in temporal and parietal regions. Brain shows marked atrophy with enlarged ventricles, widened basal cisterns and copious cortical sulcal spaces; there is shrinkage of cortical gyrus and cortical ribbon. Due to early atrophy of hippocampus and amygdala, temporal horns of lateral ventricles are markedly enlarged. There is microscopic evidence of neural dropout, loss of synaptic connections, loss of dendritic arborizaion, neuritic plaques consisting of beta-amyloid and neurofibrillary tangles. As there is no specific biologic marker or laboratory test for AD, definitive confirmation of this diagnosis can only be established by neuropathologic findings.

Vascular Cognitive Impairment (Arteriosclerotic Dementia)

Cerebrovascular disease can produce progressive cognitive decline, but the prevalence of disorder has been overestimated. Approximately 15% to 20% of all demented patients have vascular dementia, and another 10% to 15% have mixed dementia -- Alzheimer's changes plus brain damage from strokes. Clinical picture of vascular dementia is usually one of step-wise deterioration, with multiple acute exacerbations and gradual accumulation of positive neurologic findings such as pseudobulbar paralysis, labile affect, reflex asymmetries, motor deficit and aphasia. These are due to the multiple stroke episodes. Memory function is usually less impaired, in vascular than in Alzheimer dementia, but executive function is impaired to greater degree in vascular dementia. Patient with vascular dementia usually show multiple stroke risk factors.

There are several mechanisms by which cerebrovascular disease can result in cognitive impairment. In hypertensive patients, multiple small lacunar infarcts most prevalent in white matter and basal ganglia, produce most common form of vascular cognitive impairment—multi-infarct dementia (50%). A second type of vascular dementia is produced by multiple emboli to brain cortex. These emboli can be of cardiac origin or from lesions in major vessels leading to brain. A large single infarct or hemorrhage can also cause dementia. Another mechanism is chronic fluctuating hypoxia. Patients with hypotension, poor cardiac output, or severe stenosis of major cerebral vessels will experience decreased cerebral blood flow. This hypoxia leads to confusion and can eventually produce ischemia to brain such that cells in temporal lobes can be damaged (memory circuits) and areas of ischemic demyelination can be seen in hemispheric white matter. In patients with vascular dementia, there should be focal signs on neurological examination consistent with stroke as visualized by brain imaging (CT, MRI). The latter include multiple basal ganglia and white matter lacunar infarcts or periventricular white matter ischemic lesions. Temporal pattern of dementia should develop abruptly or with fluctuating step-wise progression of cognitive decline, or develop within 3 months of clinically recognized stroke. In some patients with no clinical stroke episodes who show cognitive decline, MRI shows extensive white matter periventricular high signal lesions. This is called leukoariosis. These are most commonly seen in patients with extensive vascular risk factors. There MRI hyperintense lesions are not specific for cerebrovascular disease and may be seen in AD and even in some cognitively

intact elderly patients. Treatment of vascular dementia depends on cause, control of hypertension, prevention of cerebral emboli, or raising blood pressure in those patients with significant hypotension. Cholinesterase inhibitors are beneficial in this type of dementia.

Frontotemporal Dementia

This is cause of dementia in which cerebral atrophy is localized to frontal and temporal lobes (Pick's lobar atrophy). Atrophy is frequently asymmetric with left hemisphere more commonly affected than right. In patients with primary frontal-temporal atrophy, there is decline in personal appearance and social appropriateness. The frontal patients are withdrawn, talk less, and eventually become mute. If temporal atrophy is marked, memory loss and language problems will predominate clinical picture. Usually clinical picture in Pick's disease with more prominent personality and behavioral disturbances is sufficiently different from Alzheimer's disease such that clinical diagnosis can be made. Pick's disease patients show loss of executive function and attentional deficits in excess of memory deficit as contrasted with Alzheimer disease. Focal temporal-frontal atrophy on CT or MRI scan further increases accuracy of diagnosis (Figure 14-1) and SPECT shows reduced perfusion in frontal and temporal regions in Pick's disease. This contrasts with pathologic findings in Alzheimer's disease in which cerebral atrophy is more diffuse and most marked in parietal and temporal region. In frontal-temporal dementia, genetic mutations in microtubule associated protein "tau" are detected on chromosome 17. Treatment with cholinesterase inhibitors is ineffective.

Dementia with Lewy Bodies

These patients present with cognitive decline with early and prominent attentional deficits, impaired visual-spatial skills, difficulty understanding multi-step instructions and poor problem solving skills. These patients are slow in performing mental tasks. Visual hallucinations and sleep disturbances (e.g., excessive daytime sleepiness, rapid-eye movement behavioral, and violent dreams and nightmares disorders are prominent). Motoric dysfunction suggestive of Parkinsonian syndrome is frequently seen. Clinical course fluctuates. When behavioral disorders are severe enough to require medication, patients are very sensitive to neuroleptics and extrapyramidal features frequently worsen. Clinically, differentiation of dementia with Lewy bodies from Alzheimer's and Parkinson disease may be difficult. Constellation of clinical features-cognitive decline, hallucinations, fluctuating course, delirium, and Parkinsonian features, sleep disturbances suggest Lewy body dementia. Treatment of behavioral disturbances with non-neuroleptic medication is indicated and cholinesterase inhibitors are ineffective. Neuropathology shows intracytoplasmic eosinophilic neuronal inclusion bodies with dense hyaline core and halo of radiating filaments; these contain the protein alpha-synuclein.

Alcoholic Dementia

Chronic alcoholism produces dementia as well as some of the more well-known alcoholic behavioral syndromes of delirium tremens and Wernicke-Korsakoff syndrome (see chapter 23). The dementia is usually mild and is characterized by mild memory loss and apathy. Any alcoholic patient with severe memory loss probably has Alzheimer's disease plus the effects of alcohol. The exception is when the memory loss was sudden and occurred after a period of confusion (Wernicke-Korsakoff syndrome).

Huntington's Disease (see chapter 20)

Mass Lesions

Neoplasms, abscesses, subdural hematomas, or other mass lesion can produce gradual deterioration in mental capacities. The clinical picture, however, shows focal neurologic findings, altered level of consciousness, seizures, papilledema, or focal behavioral change e.g. aphasia, frontal dysfunction. Certain neoplasms cause tissue infiltration and not cause mass effect. In these cases, patient can develop progressive dementia without other focal neurologic signs. Isolated vasculitis of CNS may cause confusion, disorientation and cognitive decline. Brain imaging in this disorder may show multiple mass lesions. CSF may show pleocytosis and elevated protein content. Angiography may show multifocal stenotic lesions, but if small vessels are involved vascular imaging may be negative. Brain or meningeal biopsy is necessary to confirm this diagnosis. This condition may be treated with corticosteroids or immunosuppressive medication.

Infectious-inflammatory etiologies (see Chapter 18)

Hydrocephalus

In adults, the condition called "normal pressure hydrocephalus" (NPH) has been identified as correctable cause of dementia. Communicating hydrocephalus (often with normal intracranial pressure) can produce characteristic clinical picture of which mental deterioration is one feature. Mental change can be intermittent confusion, memory difficulty, frontal lobe personality change, or psychiatric symptoms. However, more prominent and usually appearing as initial symptoms are a very unsteady gait and urinary incontinence. The gait disorder is called gait apraxia; it is characterized by a magnetic quality in which the patient does not pick the feet off the ground.

Pathogenesis is due to deterioration of reabsorptive surface of arachnoid villi in subarachnoid spaces. This leads to progressive ventricular dilatation but with normal intraventricular and lumbar cerebrospinal fluid pressures. CT/MRI shows dilatation of all ventricles without enlarged subarachnoid spaces (Figure 14-2). Most valuable test for diagnosing and predicting response of shunting is spinal puncture with removal of 25 to 50 ml of CSF. If removal of CSF produces significant improvement in patient's clinical symptoms, then diagnosis is established, and shunting is highly likely to improve patient's mental and neurological signs.

Head injury (see Chapter 12)

Other Causes of Dementia

The effects of the other causes of dementia can be either potentially reversible or irreversible. Box 14-3 lists these diseases.

The evaluation of a patient with dementia should include the following:

- CT or MRI scan
- Complete blood cell count
- Chemical profile
- Thyroid studies including anti-thyroid antibodies
- Vitamin B₁₂ and folate levels
- Syphilis and HIV serology
- Urine screen for drugs and toxins (if indicated)

- Lumbar puncture with cerebrospinal fluid analysis

Electroencephalography is sometimes useful particularly if Creutzfeldt-Jakob disease is suspected (see chapter 18). Lumbar puncture is not required in every patient, although it should be considered if diagnosis is not established by other studies. Neuropsychologic evaluation can be useful when the diagnosis of dementia is in doubt and depression (pseudodementia) is considered or if the mental competency level needs to be evaluated.

Two words of warning need to be issued concerning evaluation of dementia. The first is that CT scan can show atrophy in normal elderly persons and conversely no atrophy in some patients with significant Alzheimer's disease. The second point is that depression can often mimic dementia. The depressed patient often has apathy and memory complaints, so physician must be very cautious making diagnosis of dementia in potentially depressed patient. Most dementia is irreversible and untreatable, whereas depression is quite treatable and reversible. The implication of erroneous diagnosis of dementia in a depressed patient cannot be overstressed.

BOX 14-3

Potentially Reversible

1. Chronic meningitis (Cryptococcus and Mycobacterium)
2. Thyroid (Hashimoto) encephalopathy
3. Vitamin B₁₂ deficiency
4. Pituitary or parathyroid disease
5. Wilson's disease
6. Meningovascular syphilis

Degenerative Irreversible

1. Pick's disease
2. Huntington's disease
3. Parkinsonism
4. Lewy body dementia

Infectious Irreversible

1. Creutzfeldt-Jakob disease
 2. General paresis
 3. Acquired immune deficiency syndrome (AIDS)
 4. Viral (herpes simplex, arbovirus, and West Nile)
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A full neuropsychologic test battery and psychiatric consultation are often necessary to make correct diagnosis in complex cases. Depressed or anxious patients can perform poorly on mental status testing yet return to normal functioning when their emotional problem is treated (cognitive inefficiency of depression). Such patients are diagnosed as having pseudodementia. Such conditions usually appear rather suddenly after emotional crisis and show fluctuating and inconsistent findings from history and mental status examinations.

NEUROBEHAVIORAL SYNDROMES SECONDARY TO FOCAL LESIONS

Aphasia

Aphasia is language disturbance that is secondary to brain damage. Improper syntax, word choice, and imperfect comprehension are the principal features. The demonstration of aphasia is unequivocal evidence of brain dysfunction and most frequently indicates left-hemisphere

involvement. Almost all right-handed persons have their speech in left hemisphere; but left-handed persons, particularly those with family history of left-handedness, frequently have bilateral speech representation. The evaluation of language includes listening to spontaneous speech and evaluating comprehensive, repetition, and naming of objects. In acute aphasia following stroke patient can also show confusional behavior, so it is often difficult to assess aphasia fully early in its evolution.

Global Aphasia. Global aphasia is caused by large lesion involving entire middle cerebral artery territory. Spontaneous speech consists only of explicative or stereotyped sounds. Comprehension, repetition, naming, reading, and writing are similarly profoundly affected. Hemiparesis and hemianesthesia are usually present.

Mixed Aphasia. Many patients exhibit some problems in all language functions without any single function being relatively spared or impaired. This is not surprising, particularly in vascular disease in which there is such a great variation in patterns of infarction and hemorrhage.

Broca's Aphasia. This type of aphasia is usually produced by lesion in frontal lobe that involves Broca's region 44. The patients have markedly reduced spontaneous speech and dysarthria. They produce only a few single words and do so with great effort. Usually the words are highly meaningful nouns or verbs; this is referred to as "telegraphic" speech. Because temporal and parietal lobes are spared, comprehension is near normal. Repetition is impaired to the same degree as spontaneous speech, and naming errors are common. The patients are usually hemiparetic. Reading for comprehension is good, but writing is impaired.

Wernicke's Aphasia. Wernicke's classic aphasic syndrome is caused by a lesion in the posterior temporal area and contiguous parietal lobe. The patient's spontaneous speech is fluent, circumlocutory, surprisingly devoid of content, and paraphasic. Comprehension is severely impaired, as is repetition. Naming is paraphasic. No hemiparesis is present, but visual field defect is sometimes observed. The patient may be initially and incorrectly considered to have a primary psychiatric disorder because the most prominent feature of the syndrome is bizarre language output. That the patient has had a stroke or other focal lesion is overlooked because of the lack of classic motor signs usually associated with stroke or mass lesion.

Conduction Aphasia. Conduction aphasia is an uncommon but interesting type of aphasia in which spontaneous speech, comprehension, and naming are quite good (paraphasia is often present), yet repetition is severely impaired. The lesion causing this syndrome is in the posterior sylvian fissure region and extends into the deep white matter in that area.

Transcortical Aphasia. Transcortical aphasia is an unusual yet not uncommon aphasia. It is the direct linguistic opposite of conduction aphasia. In these patients repetition is excellent, whereas spontaneous speech, comprehension, and naming are nil or severely involved. The lesion causing this interesting syndrome is a border zone infarction caused by carotid stenosis, hypoxia from decreased perfusion, or carbon monoxide poisoning. The lesion has the appearance of backwards "C" and involves border zone cortex between anterior and middle cerebral arteries anteriorly and middle and posterior cerebral arteries posteriorly. If lesion is predominantly anterior, comprehension can be good and speech production absent (transcortical motor aphasia); conversely, if lesion is posterior, speech can be present (albeit aphasic), but comprehension is poor (transcortical sensory aphasia).

Anomic Aphasia. Some patients develop aphasic syndrome in which ability to name objects is only significant defect. Speech is fluent yet halting as patient searches for specific nouns or verbs. Comprehension and repetition are good, but naming is very poor and often grossly paraphasic. There is no specific lesion localization for this syndrome, but inferior temporal lobe or parietal lobe lesions are the most common.

Frontal Lobe Syndrome

In patients with destruction of frontal lobes, rather characteristic behavioral syndrome occurs. Patients develop apathy and lose motivation and goal direction. They do not initiate activities unless encouraged by others, including washing and dressing themselves. They are often euphoric and indifferent, at times to point of inappropriate jocularity or childishness. Irritability that is short-lived, perseveration, and tendency to act inappropriately in social situations are frequently present. They have difficulty sustaining attention, and they cannot make mental shifts in their thinking (perseveration). In some cases depressive symptoms are prominent. Causes include tumors (glioma, metastatic, meningioma, and pituitary adenoma), hydrocephalus, Pick's disease, alcoholic dementia, general paresis, head trauma, and Huntington's disease. To develop the syndrome bilateral damage must be present. The prognosis for normal social integration and productive employment is unfortunately not good after significant frontal damage even when intellectual functions are spared.

Organic Amnesia

Amnesia, as term is used in neurology, refers to defect in recent memory or, more properly, in ability to learn new material. Learning requires limbic structures of hippocampi, mammillary bodies, dorsal medial thalami, and perhaps other limbic structures. When brain disease specifically damages these structures *bilaterally*, severe organic amnesia or recent memory deficit results. There are several common causes: thiamine deficiency in Wernicke-Korsakoff's syndrome (mammillary bodies and dorsal medial thalami), herpes encephalitis (hippocampi), head trauma (hippocampi), bilateral temporal lobe strokes, and transient form called transient global amnesia in which the mechanism is not known. In transient global amnesia, cases, patients constantly repeat same questions and usually appear rather confused during the attack. This disorder develops abruptly and may simulate transient ischemic attack; however, it is usually solitary and resolves completely without permanent memory loss. Another differential diagnostic consideration for transient global amnesia is partial complex seizures; however, EEG is normal. In Alzheimer's and senile dementia it is the hippocampi and the cholinergic neurons in the basilar nuclei that are involved earliest and most severely. In Korsakoff's syndrome, usually seen in alcoholics, the amnesia is usually the only significantly abnormal cognitive function; IQ and other functions can be normal. In other conditions mentioned above the cortical involvement is more widespread, and other mental status changes can be demonstrated. In psychiatric amnesia (dissociative or fugue states) the patient blocks out periods of time, yet during that time (during which the patient later claims not to remember) the patient is able to learn adequately if tested. Behavior during that time is frequently normal and not noticed to be abnormal by others. In organic amnesia recent memory is impaired, but remote memory is spared; whereas in psychogenic amnesia remote, recent and immediate memory are equally impaired.

Transient global amnesia is a unique type of organic memory disturbance. In this disorder, there is acute but short-lived confusional state in which patients become amnesic. They are disoriented to time and place. This amnesia usually clears within several hours and rarely recurs. Occasionally, SPECT shows reduced perfusion in the temporal lobes but usually brain imaging and EEG are normal to exclude partial complex seizures. The mechanism of this disorder is not clearly determined; however, migraine, transient ischemic attack and focal epilepsy need to be excluded.

Parietal Lobe Syndromes

Damage to parietal lobes produces host of interesting signs, many of which can easily be overlooked on cursory examination. Box 14-4 outlines signs that can be seen due to parietal lobes lesions.

BOX 14-4

1. Signs that can be seen with lesions in either parietal lobe
 - a. Drawing and other constructional tasks are frequently impaired; lesions of the right parietal give more dramatic abnormalities, however (constructional apraxia)
 - b. Right-left disorientation
 - c. Finger agnosia (inability to recognize fingers and their relative position on the hand)
 - d. Calculation errors (dyscalculia). In left parietal lesion this can be a loss of basic arithmetic processes, whereas with right lesions calculation failure is usually on the basis of spatial errors (inability to keep numbers in their proper alignment).
 - e. Contralateral astereognosis or graphesthesia (inability to integrate sensory stimuli for identification of objects placed in the hand or writing on the skin).
2. Left parietal lesions
 - a. Reading and writing problems. With lesions in the inferior parietal lobule (angular and supramarginal gyri, areas 39 and 40) there is a defect in the translation of verbal language into written language. The resultant syndrome is called an alexia with agraphia; the syndrome is often accompanied by an anomic aphasia.
 - b. Gerstmann syndrome. On rare occasions left superior parietal lesions produce this syndrome in which a combination of finger agnosia, right-left orientation, dyscalculia, and dysgraphia is present. Constructional impairment apraxia is also present but is not part of the original syndrome,
 - c. Ideomotor apraxia. This fascinating inability to carry out rather complex motor acts such as flipping a coin, drinking through a straw, or using a hammer is often seen with parietal lesions, usually the left. Left frontal lesions can also produce apraxia,
3. Right parietal lesions
 - a. Unilateral neglect. Lesions in the right parietal lobe frequently produce an interesting condition in which the patient neglects both the left side of the body and the left side of the environment. The patient may shave only the right side of his face, completely ignore people to the left, and fail to use the left arm even though it has normal strength. Occasionally, the patient will frankly deny having any defect at all. This syndrome is occasionally seen with left-hemisphere lesions but not with the frequency that it is seen with right lesions.
 - b. Geographic disorientation. Patients often lose their orientation in their environment and can even get lost in their own houses.

EVALUATION

Any person showing evidence of a focal lesion on higher cortical function testing should be evaluated in same fashion as one would be if patient had any other focal neurologic deficit (electroencephalography, CT/MRI scanning, and angiography).

SUMMARY

Neurobehavioral disorders, previously called organic brain syndromes, represent myriad ways in which patient's behavior can be changed by physical disease affecting the brain. Sudden events will produce global changes described as acute confusion, global behavioral abnormality that usually produces altered level of alertness. If focal lesion has been cause of the change, then there will also be evidence of a specific cognitive loss that is dependent on location of lesion. The slowly developing changes represent dementia syndrome whose cause can usually be determined with full medical and neurologic evaluation.

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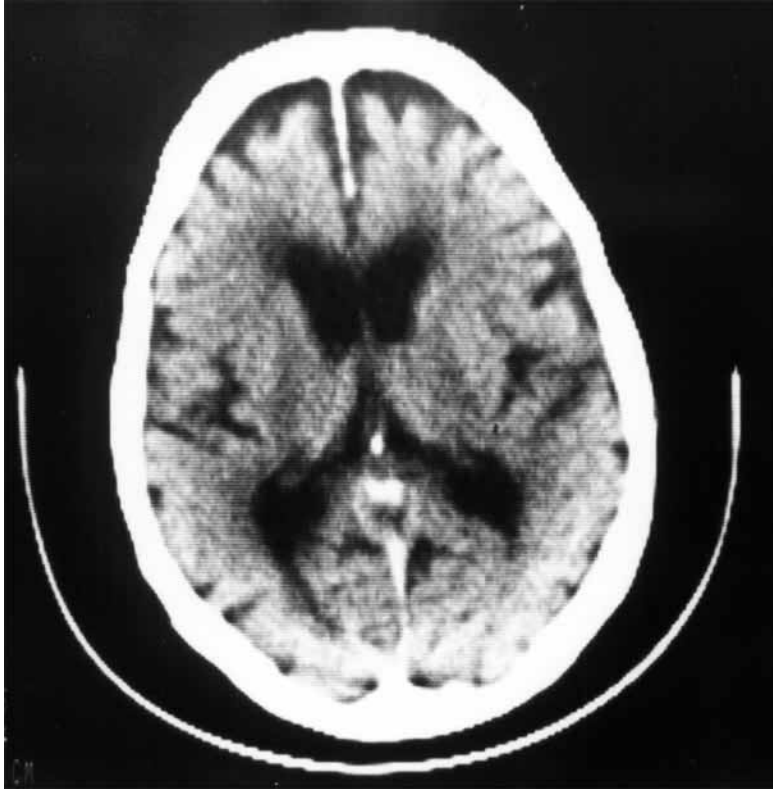


FIGURE 14-1. CT scan in a patient with Alzheimer's disease. Cortical atrophy is prominent, and the ventricles are enlarged. This ventricular enlargement, often called hydrocephalus ex vacuo, is secondary to atrophy and not pressure. A word of caution about the interpretation of atrophy seen on the CT scan: Many normal elderly persons will demonstrate cerebral atrophy; therefore the diagnosis of dementia must be made on clinical grounds and not from the CT scan. Conversely some patients with unequivocal clinical evidence of Alzheimer's type dementia will have a normal-appearing CT scan.

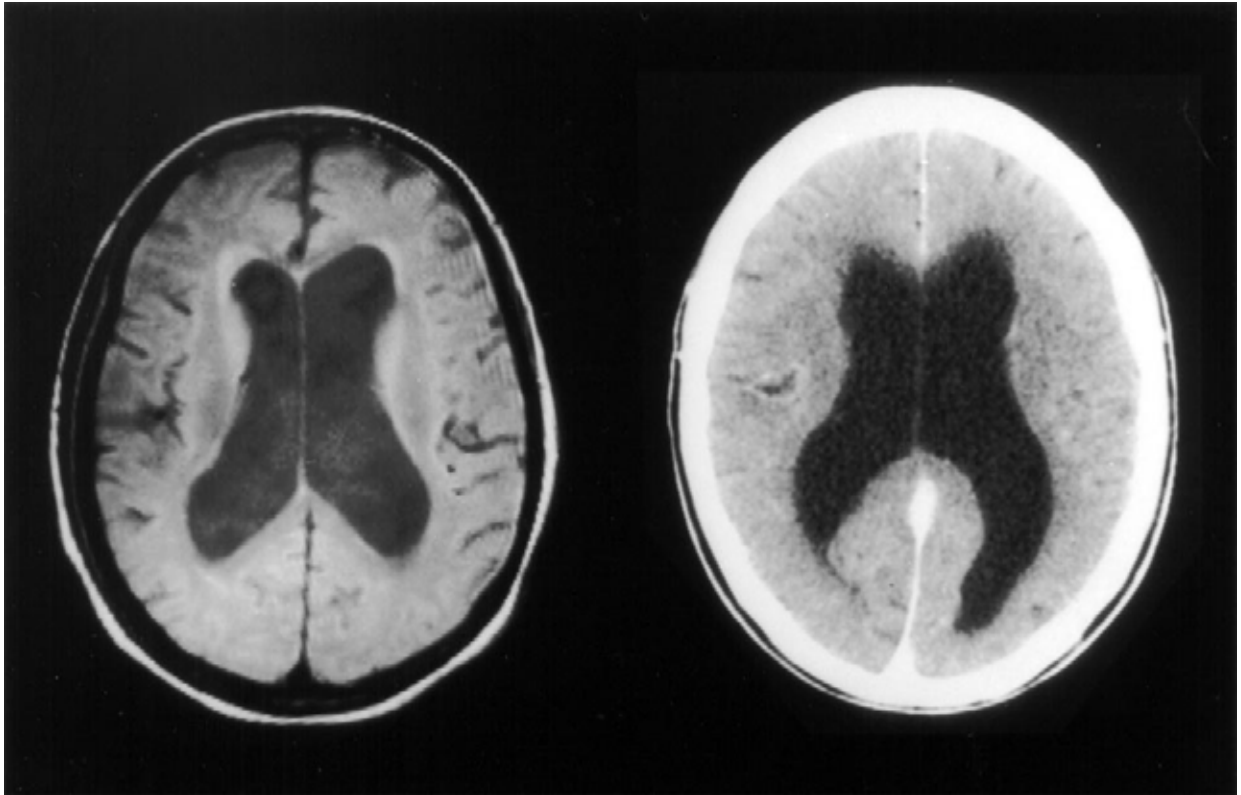


FIGURE 14-2. MRI (left) and CT scan (right) of a patient with communicating hydrocephalus. Note the enlarged ventricles and small cortical sulci for the patient's age (75 years old).