MEDICAL NEUROLOGY

SECOND EDITION

JOHN GILROY · JOHN STIRLING MEYER

NEUROLOGY

Second Edition

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PREFACE TO THE SECOND EDITION

Neurology is a young discipline, only about a century old, and until recently the major challenge was still descriptive, i.e., to define and classify neurologic disorders. Neurology is now becoming a therapeutically oriented specialty. Advances during the past 20 years in diagnostic techniques, neurochemistry, and neuropharmacology have made possible accurate confirmation of neurologic diagnosis during life and the early institution of medical, surgical, and rehabilitative therapy in order to restore neurologic function and minimize disability. This is important, since the incidence of neurologic disorders is high. About 25 per cent of patients dying in general hospitals have lesions of the nervous system. Cerebrovascular disease is now the third most common cause of death and the leading disabler in the United States. About 60 per cent of patients visiting the physician's office complain of functional or organic symptoms relating to the nervous system.

The prevalence of epilepsy in the general population of the United States has been variously estimated as 4.0 to 7.0 per thousand, with the highest incidence during childhood, so that epilepsy is a serious problem in public health. Some 2 per cent of the population are mentally retarded, and about 0.5 per cent have "cerebral palsy." Another significant fraction of the population develops the neurologic complications of diabetes, and numerous individuals succumb to those of alcoholism. Parkinson's disease occurs in about 1 per cent of the population over the age of 50 years. Head injury with damage to the brain is another serious health problem, owing to the increased frequency of automobile accidents.

The authors have extensively revised and updated this second edition to reflect the explosion of new knowledge concerned with the pathogenesis, clinical diagnosis, and, particularly, treatment of neurologic disorders. Recent information on neurotransmitters has been included in several sections throughout the text. The chapters on demyelinating diseases, degenerative diseases, toxic and metabolic disorders, pediatric neurology, and muscle diseases have been extensively rewritten to incorporate the many advances of the last few years.

The techniques of the neurologic examination are described in the first chapter, and the principles of correlation of neurologic signs with neuroanatomic localization of the lesion are discussed.

Chapter 2 provides an up-to-date yet concise coverage of pediatric neurology. Numerous conditions are described that may cause seizures, mental retardation, and so-called "cerebral palsy." Increasing knowledge of the biochemical defects in some pediatric neurologic conditions offers a new approach to treatment or genetic counseling.

Demyelinating diseases are considered in Chapter 3, since they offer the clinician opportunities for observing the development and recovery from neurologic symptoms that may affect almost any part of the nervous system. Various hereditary and nonhereditary metabolic disorders causing dysmyelination are discussed, and current research, particularly on the viral and immunologic aspects of these conditions, is summarized.

Chapter 4, which deals with the degenerative diseases, has been extensively rewritten,

particularly those sections concerned with disorders of involuntary movement, such as Parkinson's disease and Huntington's disease. The importance of recently discovered disorders of neurotransmitter function in these diseases is discussed in detail. It is apparent that, as the biochemical and enzymatic causes become defined, many neurologic diseases will no longer be classified as "degenerative diseases" but as specific biochemical or enzymatic disorders of the nervous system.

Toxic and metabolic diseases of the nervous system, comprising a large group of disorders commonly encountered in clinical practice, are dealt with in Chapter 5. This chapter has also been revised in accordance with the new knowledge that has become available in the past few years, and some new conditions have been included. Provided that the clinician can establish early diagnosis, many of these disorders are remediable.

In Chapter 6 epilepsy, syncope, migraine, and headache are considered together, since they are recurrent, sometimes confused with each other, and sometimes interrelated. New concepts of the etiology, pathology, and treatment of these conditions are discussed. With presently available medications the majority of individuals suffering from epilepsy should have their seizures controlled or reduced to an acceptable minimum. Treatment of syncope, migraine, and headache is also effective, and some recent contributions in these areas have been added.

Virtually all the bacterial and fungal infections of the nervous system described in Chapter 7 are now remediable or preventable. Twenty-five years ago infectious diseases of the brain and spinal cord were common and almost invariably fatal or disabling. A typical example is neurosyphilis, which once provided large numbers of patients for neurologic clinics. It is rarely seen today since it can be treated rapidly and effectively. There is growing evidence that viral diseases of the nervous system either are preventable (e.g., by use of poliomyelitis vaccine) or, if they do occur, may be responsive to serologic and chemotherapeutic agents.

Traumatic disorders of the nervous system are discussed in Chapter 8. With the large number of automobile and motorcycle accidents, traumatic injuries are a sizable public health problem. The emergency nature of epidural and subdural hematoma is emphasized, and methods for rapid establishment of a positive diagnosis prior to surgery are described.

In the past 20 years there have been many advances in our knowledge of cerebrovascular disease. Chapter 9 describes the etiology, incidence, epidemiology, identification, prevention, and clinical diagnosis of stroke, as well as medical and surgical treatment of this common condition.

In Chapter 10 neoplastic diseases of the nervous system are classified, and symptomatology, diagnosis, and the neurologist's contribution to early surgical treatment are discussed. Mention is also made of medical treatment for inoperable lesions, i.e., radiation, steroid therapy, and chemotherapy.

In the two remaining chapters, diseases of the cranial and peripheral nerves (Chap. 11) and muscle (Chap. 12) are classified, and the diagnosis and approaches to treatment are described. New and useful electrodiagnostic and biopsy methods, as well as the evaluation of treatment, are considered.

Many illustrations, both basic and clinical, have been included at appropriate points in the text. The clinical illustrations have been collected by the authors over many years for use in this book, in an effort to clarify the text and add to the practical usefulness and interest of this volume. The neuroanatomic diagrams, schematic outlines of biochemical disorders, and tables have been designed to organize, as concisely as possible, important information for users of this book. Many of the changes incorporated in this second edition have come from constructive advice sought from readers of the first edition.

J. G. and J. S. M.

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CONTENTS

CHAPTER 1

THE NEUROLOGIC EXAMINATION AND FUNCTIONAL NEUROANATOMY

Neurologic History Taking

The neurologic history and examination, in keeping with Hippocratic principles, provide an organized and systematic record from which the diagnosis is established. The astute physician should think in terms of disturbed anatomy and physiology. He should ask himself, "Where is the lesion?" and then be able to localize it from the history and physical examination. After localizing the site of disordered function, consideration is given to the nature of a lesion that could give rise to such a disorder.

History taking is of the utmost importance in arriving at a correct diagnosis. The technique is the same as that used in internal medicine except that special emphasis is placed on the evolution of neurologic symptoms with careful descriptions of any disorder of neuromuscular function as observed by the patient and also by those around him. The chief complaint, history of present illness, past history, family history, and social history are recorded in the usual way. When it is apparent that the patient is unable to supply an adequate history due to a language problem or memory failure, then a close relative who is aware of the problem should be questioned. This should be carried out systematically so that each major category of the history listed above is covered adequately. In addition, the patient or relative is asked a series of direct questions comprising a review of common neurologic symptoms. Inquiry should be made using simple words concerning headache, disturbance of vision, double vision, deafness, ringing in the ears, poor balance, dizziness, vomiting, difficulty with speech, loss of consciousness, seizures, convulsions, blacking-

out spells, brief attacks of paralysis, loss of sensation, numbness, pins-and-needles sensation, and difficulty with control of urine and bowel function. The hint of the presence of such symptoms should prompt more detailed inquiry until all pertinent information has been recorded in detail and in the proper sequence. Neglect of adequate history taking is the most common error for the beginner in neurology.

It is not sufficient to record the fact that headache is present. Headache is a very common complaint and only further questioning will define the seriousness of the complaint and its importance. The following questions should be asked:

When did the headaches start?
When or how frequently do they occur?
Is there any warning of an attack? (scotomas, paresthesias)
What part of the head is involved?
Has the headache undergone any change recently?
What type of pain? (dull ache, throbbing, stabbing, bursting)
Does it occur at any particular time of day?
How long before it reaches maximum intensity?
Are there associated symptoms? (nausea, vomiting, lacrimation, nasal obstruction, conjunctival injection, flushing, pallor, sweating)
Does medication produce relief?
List all medications used in the past for treatment.

In a similar fashion a series of questions should be asked regarding other positive symptoms reported during the neurologic interview. One of the most frequent complaints is "dizziness." This is a nonspecific symptom used by patients to describe vertigo, ataxia, or light-headedness, each of which has a specific meaning to a neurologist. The patient with "dizziness" should be asked:

Do you feel that the room is spinning or that you are moving?
If the answer is "no," are you unsteady when you walk?
Do you have a feeling of unsteadiness in your head?

When a history of syncope or seizures is obtained, the patient and relatives should be questioned, using the technique outlined in Chapter 6.

The examination of the patient begins with a complete general medical examination of all organs and systems, including measurements of pulse, respiratory rate, rhythm, and depth. The blood pressure should be measured in both arms with the patient lying, immediately followed by measurement with the patient standing to detect any postural hypotension.

Neurologic disorders are often a complication of a systemic disease, or a systemic disorder may arise as a complication of a neurologic disease. Examples of the neurologic disorders arising from systemic disease are:

1. Hemiplegia (paralysis of one side of the body) may arise from a septic cerebral embolus due to subacute bacterial endocarditis.
2. Paraplegia (paralysis of both legs) may be due to a spinal cord metastatic tumor arising from carcinoma of the lung.
3. Coma may occur in diabetes mellitus, uremia, or drug intoxication.

Examples of systemic disorders arising from neurologic disease are:

1. Septicemia arising from neglected infection of the urinary tract in a patient with paralysis of the bladder due, for example, to multiple sclerosis.
2. Aspiration pneumonia arising as a complication of stupor in a patient suffering from brain tumor.
3. Generalized dermatitis and granulocytopenia occurring as a complication of anticonvulsive medication in the treatment of epilepsy.

Of all systemic complications arising from neurologic disorders, pulmonary and urinary tract infection in debilitated patients are the most common and should always be suspected when unexplained fever occurs.

When the general examination is completed, a systematic method of neurologic examination should be adhered to; otherwise important signs may be overlooked (Table 1–1).

The Neurologic Examination

The neurologic examination should not be a lengthy and compulsive ritual. In clinical

TABLE 1–1

The Neurologic Examination

Mentation

Awareness
Orientation (oriented to time, place, and person);
level of consciousness (obtunded, stuporous, semicomatose, comatose)

Speech
Normal, dysphasia, dysarthria, dysphonia

Mood
Normal, euphoric, depressed, anxious, agitated

Affect
Normal, flat, inappropriate

Perception
Delusions, illusions, hallucinations

General knowledge
Knowledge of current events, vocabulary

Memory
Intact, recent memory impaired; remote memory impaired

Retention and recall
Recall of objects; digits forward and reversed

Reasoning
Judgment, insight, abstraction (interpretation of proverbs, similarities, and differences)

Praxis
Ideational, ideomotor, motor, and constructional apraxias

Object recognition
Normal, agnosia

Use of symbols
Calculation, reading, writing

Gait, station
Hemiplegic, ataxic, spastic, festinating, hyperkinetic, waddling, apraxia of gait, hysterical gaits, steppage gait, Romberg test

Cranial nerves

Motor system
Atrophy, fasciculations, tremor, dystonia, involuntary movements, palpation, tone, strength

Coordination
Finger-to-nose, heel-to-shin tests; rapid alternating movements

Reflexes
Superficial reflexes; tendon reflexes

Sensation
Touch, pain, temperature, vibration and position sense, tactile localization, two-point discrimination, bilateral simultaneous stimulation, stereognosis, barognosis, skin writing

Head and neck
Bruits over head and neck, scalp and skull tenderness and deformity, signs of head trauma, CSF drainage from ears and nose

Spine, skin
Nuchal rigidity, hairline, shortness of neck, spinal deformity, spinal tenderness, limitation of movement of spine, paravertebral spasm, limitation of straight leg raising, palpation for cervical ribs, pes cavus, peripheral nerve enlargement, adenoma sebaceum, café-au-lait spots, trigeminal hemangioma

practice it seldom requires longer than 30 minutes to provide the examiner with an adequate evaluation of the case. Prior to this, another 30 minutes is usually required to obtain the history.

The outline of examination presented in Table 1–1 is adequate for most neurologic disorders. The examination is modified, however, according to the special problems presented by each case. It is of the utmost importance in carrying out the examination to maintain an orderly approach. It has been found useful to memorize and examine the functions of the nervous system under the headings and in the order outlined in Table 1–1. If such a system is not followed, some vital sign, such as papilledema, may be overlooked.

The instruments used in the neurologic examination need not be numerous or complicated. Standard neurologic instruments include a sphygmomanometer; stethoscope; flashlight; ophthalmoscope; otoscope; material for examining the sense of smell such as tobacco, coffee, and perfume; a red glass for checking double vision; a reflex hammer; two corsage pins with white heads; cotton pledget sticks; a tuning fork (128 cps); and thin-walled containers for ice and for hot water, to test temperature sensation.

Many of these instruments have several applications. The white-headed pins are used for mapping out the visual fields and for testing pain sensation, localization of pin pricks, bilateral simultaneous stimulation, and two-point discrimination. The sphygmomanometer, in addition to its designed function in measuring blood pressure, may be used as a dynamometer to measure strength of muscle contraction. For example, the subject may be asked to repeatedly squeeze the rolled and inflated cuff held in one hand to a recorded pressure of 130 mm every second. In myasthenia gravis, progressive weakness rapidly occurs, and the recorded pressure gradually falls with each squeeze of the hand.

Mentation

The neurologic examination logically begins with assessment of the mental status or "mentation." This really begins as soon as the patient is interviewed, but a detailed evaluation of the mental status should not be overlooked as it may reveal important signs of impairment of brain function. Examination of mental status may be divided into two parts: tests to determine general cerebral integrity and tests that indicate focal disorders of cerebral function.

Awareness

Orientation. The degree of alertness and awareness of the environment is indicated by the patient's orientation to time, place, and person. He is asked, "Do you know what date it is?" "What is the name of this place?" and "Who is that man standing over there?" or "Who am I?"

Levels of Consciousness. When alertness is impaired, the patient's condition can be described accurately, using the following terms and definitions:

OBTUNDITY. The subject can be aroused by stimuli and will then respond to questions or commands. The subject remains aroused as long as the stimuli are applied. During arousal, the subject responds but may be confused.

STUPOR. Spontaneous movements occur, accompanied by groaning, in response to numerous stimuli such as pain, bright light, loud noise, and manipulating a part. Repeated stimulation may lead to brief intervals of responsiveness to questions.

SEMICOMA. Withdrawal of a part in response to painful stimuli is the only response observed.

COMA. There is absence of any observed response to painful stimuli.

In general, disorders of consciousness are usually associated with diffuse cerebral injury.

Speech

Any disorder of speech must be accurately described if the subject is sufficiently alert to respond. If comprehension of spoken words, or the concept of words as symbols, or their motor execution is impaired owing to disordered brain function, the patient is termed "dysphasic."

Dysphasia

Dysphasia may be more briefly defined as a difficulty with comprehension or production of language due to disease of the central ner-

vous system. When the cerebral lesion is so severe that the ability to speak is totally lost, the condition would then be correctly termed "aphasia."

There are two broad categories of dysphasia: *sensory dysphasia,* where the difficulty lies in comprehension of language, and *motor dysphasia,* where there is difficulty in the production of language. In practice the two frequently occur together, but one type usually predominates. The differentiation between these two broad categories of dysphasia is usually not difficult.

The patient with sensory dysphasia fails to comprehend simple questions or requests; he is often voluble and may converse inappropriately and talk in jargon. He may appear to be unaware of his disability since he can comprehend neither his own speech nor that of others. The patient with motor dysphasia understands simple questions or requests but finds difficulty in replying. His vocabulary is reduced, and the speech is slow, broken, and hesitant; he may become exasperated with his efforts to communicate and shows relief when the examiner recognizes his difficulty.

In more than 90 per cent of the population the centers for speech are located in the left cerebral hemisphere; in the remainder, where the speech centers are in the right hemisphere, the individual is usually left-handed. Centers for speech are in the vicinity of the Sylvian fissure (opercular area). Motor dysphasia correlates well with anteriorly located lesions (frontal lobe, Broca's area) and sensory dysphasia with posteriorly placed lesions (temporal, parietal lobes).

Dysarthria

Dysarthria is a deficit in articulation of speech.

Defects in articulation are due to a disorder of neuromuscular control of the muscles involved in articulation and may be classified as lingual, labial, pharyngeal, laryngeal, and cerebellar. The defect may be accentuated by asking the patient to repeat certain phrases, such as "West Register Street," "Fifty-first Artillery Brigade," "Methodist Episcopal." When there are jerkiness and irregularity in volume and rhythm of speech as occur in a cerebellar lesion, which is often called "scanning" speech, this may be brought out by asking the patient to repeat rhythmic sounds such as me-me-me or la-la-la.

Dysphonia

Dysphonia is a condition of disturbed sound, rhythm, or tonal quality of speech. Paralysis of one or both vocal cords may produce hoarseness or reduce the level of speech to a whisper. The rhythm of speech may be disturbed in paralysis agitans and cerebellar ataxia. Tonal quality is disturbed in paralysis agitans in which the speech is characteristically monotonous.

Assuming that the patient is alert and that there is no evidence of dysphasia, the following tests of mentation should be carried out in full.

Mood

At this stage the examiner has taken a full history from the patient and asked him a number of questions regarding the mental status examination. He should now be in a position to assess the patient's mood. Is he depressed? Elated? Fearful? Nervous? Irritable? Labile? He may also be questioned directly about mood, whether he feels happy or sad, worried, anxious, or irritable. Patients with neurologic disorders frequently show changes in mood that may be of help in the final diagnosis; e.g., depression is frequently present in early dementia.

Affect

Affect is the emotional response to a given situation. The patient's response to questions may be accompanied by inappropriate laughter suggesting euphoria often seen in frontal lobe lesions, or he may fail to show any change in emotional response throughout—a flat affect. This is commonly seen in disorders of the basal ganglia. There may be loss of emotional control resulting in pathologic laughing and crying. This is usually associated with pseudobulbar palsy, which indicates bilateral cerebral hemisphere or upper midbrain damage.

Perception

Delusions, Illusions, and Hallucinations

Misconstrued perceptions and ideations are encountered in neurologic practice and should be assessed by the patient's responses during the examination. Direct questioning, particu-

larly concerning delusions and hallucinations, requires tact if the examiner is not to arouse hostility and defensiveness on the part of the patient. Such questions as "Have you had any unusual experiences lately?" "Do you ever get the feeling that you have been in a place before or been through an experience before?" (the *déjà vu* phenomenon) may be asked. It is particularly important to inquire about unusual thoughts, dreamy states, and unusual experiences, sights, tastes, sounds, or smells in the patient suspected of having psychomotor seizures (seizures presumed to arise in the temporal lobes) since these phenomena may form an integral part or the only indication of the seizure pattern.

The following simple definitions of these terms should prove helpful:

Delusion: A false belief.
Illusion: False interpretation of a sensory perception.
Hallucination: A false sensory perception, without basis in fact, resulting from an ideational distortion.

Although a rigid concept of "centers" in the brain subserving highly compartmentalized functions is no longer feasible, certain areas of the brain, which for convenience will be termed "centers," have been found to be associated with certain functions. Involvement of these functions consequently provides information regarding localization of a disease process to certain parts of the brain. Patients with illusions and hallucinations due to organically demonstrable disease are usually found to have lesions located in the temporal, parietal, and occasionally occipital lobes.

General Knowledge

More complex intellectual functions are tested by asking questions of general knowledge such as the name of the president of the United States, the governor of the state, or the mayor of the city and a description of a recent outstanding world event. This latter question will also serve as a test of recent memory and supplement any impression already gained of intellectual function during history taking.

Reduction of general mental capacity usually implies diffuse damage to the cerebral cortex. If the damage has been present since birth or early childhood, the term "amentia" (developmental retardation, etc.) is used; if the damage occurs after learning processes and speech are well established, the term "dementia" is used to define the reduction of general mental capacity.

Vocabulary

Vocabulary is the best indicator of a patient's overall premorbid intellectual capacity when considered against any other single test of mentation. Although it may be diminished in dysphasia, it is relatively refractory to the effects of many cerebral disorders. Testing the patient's vocabulary affords some assessment of his general knowledge prior to any dementia. The patient is asked to define the meaning of words that are presented at increasing levels of difficulty, e.g., winter, assemble, regulate, consume, reluctant, tangible, plagiarize.

Memory

Recent Memory

This usually fails before memory for remote events in organic disease of the temporal lobes; and the degree of impairment may be gauged by specific inquiry about the events of the last two days, preceding the hospital admission.

Remote Memory

Remote memory is tested by questions about the patient's birthplace, parents, schooling, and any important event known to the examiner that may have occurred many years ago. More severe disease of both temporal lobes may be suspected when remote memory is disordered.

Retention and Recall

Recent memory is dependent on the patient's ability to retain information. If this faculty is impaired or lost, no "new" memories will be formed. This process may be tested by having him repeat a number of digits given to him by the examiner beginning with three digits and increasing until a limit is reached. The test is then repeated with the patient reversing the digits, again beginning with three digits. The patient of average intelligence can repeat six digits forward and five digits reversed.

Another test of retention consists of asking the patient to remember the names of three

objects, two similar and the third dissimilar (apple, orange, umbrella). The examination continues, and three minutes later the patient is asked to repeat the three objects. Failure to do so should lead to repetition of the test with another three objects but requesting him to repeat them at two and a half minutes. With reduction of the time span by 30 seconds after every failure, a limit of retention can be defined in seconds. In Korsakoff's psychosis, retention span may be less than 30 seconds.

Reasoning

In general, disorders of judgment, insight, and abstraction correlate well with disordered function of the frontal lobes.

Judgment

This may be defined as the ability to form an assessment of a situation and is commonly tested by asking the patient his interpretation of a simple test story involving judgment.

"What would you do if you were walking along a street and saw a letter lying on the sidewalk which had an address and an unused stamp on the envelope and it was sealed?" A typical reply showing impaired judgment would be "Throw it away."

"What would you do if you were sitting in a crowded theater and you were the first person to notice a fire?" A reply of "Run for the exits yelling, 'Fire, Fire!' " would be considered to show poor judgment.

Insight

The ability to understand the reason for a given situation may be tested by asking the patient why he is in the hospital or why he is undergoing medical examination. If the answer is "I don't know; there is nothing wrong with me," insight may be considered suspect.

Abstraction

This requires understanding and judgment of a fairly high intellectual order and should be used only when the patient has had a Grade 7 education or higher. Three proverbs are given: "A rolling stone gathers no moss." "People in glass houses should not throw stones." "The grass is always greener on the other side of the fence." The patient will usually supply an answer illustrating normal ability for abstract thinking, or his answer will be concrete, or he will fail to answer. An answer to the first proverb such as "Rolling stones don't pick up moss" may be considered concrete. Concrete interpretation and failure to answer are given negative scores. Proverb interpretation can then be scored as so many points out of three, a score of 1 being awarded for each abstraction of a proverb.

An additional test of abstraction may be given by asking the patient to assess similarities and differences between objects. A question such as "How are an apple and an orange the same and how are they different?" or, similarly, "How is a table like a chair and how are they different?" is presented.

Praxis

Apraxia is defined as the inability to execute a planned motor act in the absence of paralysis of the muscles normally used in the performance of the act.

Three stages may be considered in the development of a skilled act: the development of the idea of the movement, the formulation of the plan of its execution, and finally the motor performance of the plan. According to these three stages, three types of apraxia may be recognized.

Ideational Apraxia

Ideational apraxia is often due to a lesion in the supramarginal gyrus of the parietal lobe. The patient is unable to comprehend or formulate a plan of movement in response to a request. The condition resembles extreme absentmindedness. As an example, when requested to light a cigarette the patient may fail to take the cigarette out of the package or, having done so and put it in his mouth, may be unable to take a match out of a box or folder. Given a match, he may attempt to strike it on the cigarette pack or on the smooth surface of the matchbox. There is no impairment of motor movement; the idea of the movement is confused.

Ideomotor Apraxia

Ideomotor apraxia occurs when there is interference with the transmission of the appropriate impulses transcortically to the motor centers to convert the idea into coordinated