A Textbook of
NEUROLOGY

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Preface

NEUROLOGY, a branch of medicine, which for many years has been traditionally allied with psychiatry, has in recent years returned more closely to the fold of internal medicine. This return is an especially fortuitous one because the newer technics used in internal medicine, particularly the application of biochemical methods to the study of disease, offer great promise of solving many of the diseases of the nervous system which have heretofore been labeled as degenerative or heredo-degenerative. For example, it has recently been demonstrated that large amounts of copper are present in the brain and other organs of patients with hepatolenticular degeneration although the copper content of the serum is below normal. This decrease in the copper content of the serum is presumably related to a deficiency of that portion of the serum, caeruloplasmin, to which copper is normally bound. This deficiency in binding, which is probably hereditary, allows ingested copper to diffuse more readily into the living brain and other organs and damage their parenchyma. It is quite probable that with the methods now available many other heredodegenerative diseases also will be found to be due to some inherent or secondary metabolic defect.

The intricacies of neurological disease are so great that they can not be considered fully in a textbook of medicine, and this book is written as a text on neurology for medical students and physicians. In order to conserve space, the traditional sections on anatomy and examination of the nervous system have been omitted. These could at best be inadequately covered and the student is referred to special texts on these subjects. Pertinent aspects of anatomy and the neurological examination, however, are discussed when they are necessary for an understanding of specific disease entities.

Mental symptoms which accompany organic disease of the nervous system are discussed in connection with the appropriate diseases, but the psychoneuroses and the so-called functional psychoses are not considered in this volume. It is my belief that these conditions can be presented better by a physician trained in the methodology of psychiatry and that their consideration is not appropriate to a text on neurology.

An attempt has been made in the text to group diseases according to etiology. Some deviations were necessary in order to expedite the discussion of certain clinical syndromes, such as those resulting from disease of

the peripheral nerves or of the basal ganglia. In addition, it must be admitted that any classification of disease of the nervous system at the present time is inadequate and that changes will be necessary when more is known regarding etiology.

I wish to express my appreciation of the valuable assistance of many of my co-workers: To Dr. Daniel Sciarra for his aid in preparation of the text and the illustrations; to Dr. Lewis P. Rowland and Dr. Robert A. Fishman for editing the proofs and completion of the index; to Miss Virginia Fitzgerald and Mrs. Alice F. Griffin for the typing of the manuscript; and to my wife, Mabel Carmichael Merritt, for her constant assistance in the preparation of the text. My thanks are also due to my colleagues who have furnished many of the illustrations and to the publishers of the various journals and books who have permitted the reproductions of material from previous publications by me and other authors.

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A Textbook of Neurology

Chapter 1

INFECTIONS

INFECTIONS OF THE MENINGES

ACUTE PURULENT MENINGITIS

PATHOGENIC organisms may gain access to the ventriculo-subarachnoid space by way of the blood stream with or without involvement of other organs of the body or by direct extension from a septic focus in the cranial cavity or head. Organisms may enter through compound fractures of the skull or fractures through the nasal sinuses or mastoid. They may be introduced directly into the subarachnoid space by lumbar puncture for the withdrawal of fluid or the administration of sera, air, anesthetics and the like. The pathology, symptomatology and clinical course of patients with acute purulent meningitis are similar regardless of the causative organism. These will be described in connection with the consideration of the most common forms of meningitis. The diagnosis depends on the isolation and identification of the causative organism, and the determination of the source of the infection in the cases of secondary meningitis.

Table 1. Relative Frequency of the Common Forms of Bacterial Meningitis (5,136 Cases Collected from Literature)

Organism	*No. of Cases*	*Per Cent*
Meningococcus	2,052	40
Tubercle bacillus	1,509	29
Pneumococcus	506	10
Streptococcus	403	8
Influenza bacillus	291	6
Miscellaneous	375	7
Total	5,136	100

Acute purulent meningitis is most commonly due to the meningococcus, the influenza bacillus, the pneumococcus or the streptococcus. These four, together with the tubercle bacillus (Table 1), account for the vast majority of the cases. The meninges may be invaded by any of the other path-

ogenic organisms including: B. coli; B. typhosus; B. paratyphosus; anthrax bacillus; B. pyocyaneus; Friedländer's bacillus; diphtheroid bacilli; micrococcus catarrhalis; the gonococcus; brucella, torula and other yeast; and streptothrix.

Meningococcal Meningitis

Meningococcal meningitis or acute cerebrospinal fever was described by Vieusseux in 1805 and the causative organism was identified by Weichselbaum in 1887. It occurs almost constantly in sporadic form and at irregular intervals in epidemic form. Epidemics are especially apt to occur when there are large shifts in population as in time of war.

Pathogenesis: The meningococci may gain access to the meninges directly from the nasopharynx through the cribriform plate or indirectly by way of the blood stream. The fact that organisms can be cultured from the blood or from cutaneous lesions before the appearance of meningitis is strong evidence that the infection takes place through the blood stream by way of the choroid plexus in many, if not all, cases. In addition, it has been shown that in early cases the ventricular fluid may be teeming with organisms before infection of the meninges is present.

Pathology: In acute fulminating cases death may occur before there are any significant pathological changes in the nervous system. In the usual case where death does not occur for several days after the onset of the disease, there is an intense inflammatory reaction in the meninges. The inflammatory reaction is especially intense in the subarachnoid spaces over the convexity of the brain and spinal cord and around the cisterns at the base of the brain and it may extend a short distance along the perivascular spaces into the substance of the brain and spinal cord. In rare cases the inflammatory reaction breaks into the parenchyma. Meningococci, both intra and extracellularly, are found in the meninges and the fluid from the ventricles and subarachnoid space. With progress of the infection the pia-arachnoid becomes thickened and adhesions may form over the convexity of the brain or at the base. Adhesions at the base may interfere with the flow of cerebrospinal fluid from the fourth ventricle and produce hydrocephalus. Inflammatory reaction and fibrosis of the meninges along the roots of the cranial nerves is thought to be the cause of the cranial nerve palsies which occasionally develop. This is not the only mechanism for such paralyses, however. Damage to the auditory nerve often occurs suddenly, and the auditory defect is usually permanent. This may be explained as the result of extension of the infection to the inner ear or to thrombosis of the nutrient artery. In addition, facial paralysis not infrequently occurs after the meningeal reaction has subsided and is best explained on another, perhaps allergic, basis. Signs and symptoms of parenchymatous damage, hemiplegia, aphasia, or cerebellar signs are infrequent and are probably due to the formation of infarcts as the result of thrombosis of inflamed arteries or veins. Damage to the spinal cord, myelitis, is explained on a similar basis. Myelitis or damage to the roots of the cauda equina are rare in the absence of intrathecal treatment with serum or chemicals and it is probable that the latter substances play an important role in the production of these complications.

With efficient treatment, and in some cases without treatment, there is subsidence of the inflammatory reaction in the meninges and no evidence of the infection may be found at autopsy in patients who die some months or years later.

Incidence: The meningococcus is the causative organism in approximately 40 per cent of all cases of meningitis. While both the sporadic and epidemic forms of the disease may attack individuals of all ages, children are predominantly affected. In many large epidemics over 75 per cent of the cases were under ten years of age. Males appear to be slightly more susceptible than females. The normal habitat of the meningococcus is the nasopharynx and the disease is spread by carriers or by individuals with the disease. The normal carrier rate in the general population in non-epidemic periods is approximately 3 to 4 per cent. This incidence increases when there is a chance for close contact of many individuals (as in military barracks), and during the winter months. The carrier rate increases in the presence of upper respiratory infections and it is not uncommon for these to precede the onset of meningitis.

Symptomatology: The onset of meningococcal meningitis, similar to that of other forms of meningitis, is accompanied by chills and fever, headache, nausea and vomiting, pain in the back, stiffness of the neck, and prostration. The occurrence of herpes simplex, conjunctivitis and a petechial or hemorrhagic skin rash is common with meningococcus infections. At the onset, the patient is irritable and in children there is frequently a characteristic sharp shrill cry (meningeal cry). With progress of the disease, the sensorium becomes clouded and stupor or coma may develop. Occasionally the onset may be accompanied by deep coma. Convulsive seizures are not infrequently an early symptom, but focal neurological signs are uncommon. Acute fulminating cases with severe prostration and an extensive cutaneous rash (Waterhouse-Friderichsen syndrome) are considered more fully later.

Signs: The patient appears acutely ill, and may be confused, stuporous or semi-comatose. The temperature is elevated from 101° to 103° F., but occasionally it may be normal at the onset. The pulse is usually rapid and the respiratory rate is increased. Blood pressure is normal except in acute fulminating cases when there may be a severe degree of hypotension. There is rigidity of the neck with positive Kernig and Brudzinski signs. The reflexes are often depressed but occasionally they may be increased. Cranial nerve palsies and focal neurological signs are uncommon. These complications usually do not develop until several days after the onset of the infection. The optic discs are normal, but choking may develop if the meningitis persists for more than a week.

Laboratory Data: The white blood cells in the peripheral blood are increased with counts usually in the range of 10,000 to 30,000 per cu. mm., but they occasionally may be within normal limits or higher than 40,000 per cu. mm. The urine may contain albumin, casts and red blood cells. Meningococci can be cultured from the nasopharynx in practically all cases, from the blood in over one-half of the cases in the early stages, and from the skin lesions when these are present.

The cerebrospinal fluid is under increased pressure, usually between 200 and 500 mm. of water. The fluid is cloudy or purulent and contains a large number of cells, predominantly polymorphonuclear leukocytes. The cell count in the fluid is usually between 2,000 and 10,000 per cu. mm. Occasionally it may be less than 1,000 and infrequently is more than 20,000 per cu. mm. The protein content is increased. The sugar content is decreased, usually between 5 and 30 mg. per 100 cc. The chloride content is moderately or greatly decreased. The colloidal tests may show any type of abnormality. Organisms can be seen intra and extra-cellularly in stained smears of the fluid and they can be cultured on the appropriate media in over 90 per cent of the cases.

Complications and Sequelæ: The complications and sequelæ of meningococcic meningitis include those which are commonly associated with an inflammatory process in the meninges and its blood vessels (convulsions, cranial nerve palsies, focal cerebral lesions, damage to the spinal cord or nerve roots, hydrocephalus), and those which are due to involvement of other portions of the body by meningococci. The latter include panophthalmitis and other types of ocular infection, arthritis, purpura, pericarditis, endocarditis, myocarditis with cardiac enlargement and electrocardiographic changes, pleurisy, orchitis, epididymitis, albuminuria or hematuria, and adrenal hemorrhage. In addition there may be complications arising as the result of intercurrent infection of the upper respiratory tract, middle ear and lungs. Any of the above complications may leave permanent sequelæ but the most common sequelæ are those which are due to injury to the nervous system. These include deafness, ocular palsies, blindness, changes in mentality, convulsions and hydrocephalus.

With the methods of treatment available at the present time, complications and sequelæ of the meningeal infection are rare and the complications due to the involvement of other parts of the body by the meningococci or other intercurrent infections are more readily controlled. In a recent study by Farmer of 300 sulfonamide treated cases, neurological complications were present in less than 9 per cent and a permanent residual in less than 2 per cent. Sixth nerve palsy developed early in the course of the disease in 3 per cent and completely disappeared within a few weeks. Seventh nerve palsy occurred as a late complication in 3 per cent of the cases. It usually developed between the fifth and fourteenth day, at a time when the cerebrospinal fluid was relatively cell-free. Recovery from the facial paralysis, either unilateral or bilateral, was usually complete within a few months. Eighth nerve palsy, usually bilateral, was present in 2 per cent and was followed by permanent deafness. Focal neurological signs—convulsions, hemiplegia and aphasia—were present in only one per cent, and in all of these the signs and symptoms were transient.

Waterhouse-Friderichsen Syndrome: Waterhouse-Friderichsen syndrome, adrenal hemorrhage associated with purpura, is most commonly associated with meningococcic septicemia, but it may occur in the course of other infectious diseases. The incidence of this complication of meningococcic infection is usually very low but in some epidemics it may be as high