

Periscope

Brain

(Vol. XXXVI, Part I)

1. Parasyphilis of the Nervous System. JAMES MCINTOSH and PAUL FILDES, HENRY HEAD and E. G. FEARNSIDES.
2. The Brain of a Macrocephalic Epileptic. J. WIGGLESWORTH and GEORGE A. WATSON.
3. The Relation of the Myopathies. WILLIAM G. SPILLER.

1. *Parasyphilis of the Nervous System*.—The authors of this paper state that it is a preliminary communication, based upon several lines of work, carried out simultaneously in the wards, and in the bacteriological laboratory of the London Hospital. Each separate research will form later the subject of a paper, to be published in *Brain*, but they have thought it wiser to give a preliminary account of their general conclusions before presenting the various more special and detailed communications.

The results embodied in the paper are based entirely on cases where the serological examinations have been carried out by Dr. Fildes and Dr. McIntosh, and the clinical records compiled by Dr. Head and Dr. Fearnside. Von Pirquet's conception of "allergie" opened a line of thought which led McIntosh and Fildes to formulate the view which the authors hold with regard to the nature of parasyphilis. Von Pirquet showed that all infections led to a change in the character of the reaction of the tissues to the poison. This might take place in two directions; they might respond less and less to infection ("hypoallergie"), until at last they ceased to respond altogether and became "anergic." But, on the other hand, a change in the opposite direction might occur; the reaction might appear both more quickly and with greater violence to a smaller dose of the poison. This he speaks of as hyperallergie and brought into this category the phenomena, described by Richet, under the name of "anaphylaxis."

In their view tertiary and "parasyphilitic" phenomena are manifestations of hypersensitiveness ("hyperallergie"), which reaches its extreme form in anaphylactic shock. As the tissues are hypersensitive, they react more strongly to a minute dose of the poison, and hence some of the difficulty of discovering the spirochete in gummatous and parasyphilitic tissues.

The difference between the anatomical consequences, produced by the impact of the poison of the two cases, depends on the nature of the tissue attacked. Thus, gummatosis is the reaction of hypersensitive vessels and connective tissue, parts which tend to show proliferative changes. On the other hand, the pathological anatomy of "parasyphilis" of the central nervous system is the expression of the response of hypersensitized, highly differentiated nerve-elements which have no power of regeneration. The profound difference between the two forms of disease-process depends, not on the extent to which the tissues have become hyperallergic, but on the fact that, in the one case, proliferative repair can occur at once, whilst, on the other hand, the injury to the nerve-elements results in their death. This

view explains the fact that "parasyphilitic" processes occur in those nerve fibers in which the sheath of Schwann is absent. This structure is not only an essential factor in the process of regeneration, but also, no doubt assumes a nutritive or protective role, to such an extent that a reaction which is sufficient to destroy a nerve-fiber not provided with this sheath may be insufficient to injure to any extent the nerve protected by the neurilemma.

This theory also accounts for the occasional presence in the same patient both of gummatous meningitis and of "parasyphilitic" degeneration; the one is the reaction of a highly sensitized connective tissue, whilst the tabetic changes are the expression of a similar condition in the long fibers of the posterior columns.

On this view we can also explain the peculiar course of the milder forms of parasyphilis. The tendency of many cases of tabes is to run on somewhat acutely to a certain stage and then to stop, frequently for many years. This is particularly well seen where the first manifestations take the form of primary optic atrophy, which leads invariably to complete blindness; but from that time onward the patient may remain free from any further affection until, perhaps many years later, his knee-jerks disappear, or he suffers from some cerebral complication.

The conclusions arrived at in this important paper are as follows: (1) Parasyphilis of the nervous system is a purely clinical conception. It is a diseased state which may affect any part of the brain or spinal cord; the manifestations of tabes and paresis can only be erected into two clinical divisions by an arbitrary selection of signs and symptoms. Some forms of progressive muscular atrophy, lateral and combined sclerosis, primary optic atrophy and periodic epileptiform attacks may be equally definite manifestations of parasyphilis. (2) Parasyphilis is slightly, if at all, amenable to antisyphilitic treatment with compounds of arsenic and mercury, probably because these bodies do not enter the essential structures of the central nervous system. (3) Parasyphilitic states are peculiarly liable to arise after mild syphilitic infection. Sixty per cent. of cases of tabes have suffered from at most a primary sore, and in many cases the whole course of the initial infection was run under cover of a gonorrhea. (4) In paresis, and in active, untreated cases of tabes, and tabo-paresis, the cerebrospinal fluid yields a positive Wassermann reaction, often of great strength. (5) With acute or chronic syphilis of the nervous system, other than parasyphilis, the behavior of the cerebrospinal fluid depends upon the extent to which the spinal cord and its membranes, including those of the brain-stem, are affected. Thus, most cases of meningomyelitis show a strong positive reaction in the cerebrospinal fluid, whilst cerebral lesions tend to give a weakly positive or even a negative reaction. (6) Antisyphilitic treatment has a profound effect on the positive reaction in syphilitic meningomyelitis, and the cerebrospinal fluid may give a negative reaction after a few months. But in cases of parasyphilis no positive change occurs in consequence of such treatment, within any comparable period. (7) Whatever views may be held of the primary and secondary manifestations in syphilis, the authors believe that all tertiary and parasyphilitic manifestations are expressions of the reaction of hypersensitized tissues (hyperallergie). That is to say, during the previous stages of infection, the tissues have been so altered that they react more violently to a smaller dose of the spirochete or its toxins. Gummatosis is the reaction of hypersensitized connective tissues and bloodvessels, whilst parasyphilis is a hyperaller-

gic reaction of the essential nerve elements and neuroglia. (8) The consequence of this hyperallergic reaction in the tissues of the central nervous system is death of any set of fibers or cells which happens to be attacked, and proliferative reaction on the part of the neuroglia within the same territory. Thus, the clinical manifestations of parasyphilis are an expression of the reaction and necrosis of hypersensitized areas of the nervous system, evoked by reappearance of the *Spirochæta pallida*. (9) This hypersensitive (hyperallergic) state of the tissues of the central nervous system is produced, in all probability, by the passage of the spirochetes or their toxins up the nerves from the skin and mucous membranes during the secondary period. But it is also conceivable that it may be due to a slight encephalitis during this stage of infection. The headache and lassitude, unaccompanied by any gross nervous lesions, which so frequently occur during the secondary stage, possibly represent clinically the period during which sensitization occurs. (10) Thus, parasyphilis is a clinical conception which comprises the manifestations of a series of diseased states. From the pathological point of view the term is inadmissible. These states depend on the reaction of hypersensitized tissues to the spirochete or its toxins, and this reaction is as truly syphilitic as the production of gummata. The difference between the consequences of the tertiary and of the parasyphilitic process lies in the nature of the tissues participating in the reaction. In the one case the connective tissue is capable of repair, and the focus is readily reached by the remedial agents. In the case of parasyphilis, reaction of the essential nerve-elements leads to their death, and the anti-syphilitic remedies cannot readily reach the spirochete.

2. *The Brain of a Macrocephalic Epileptic.*—The writers give a short history, and complete notes on the autopsy of the subject of this paper, a male epileptic, who was admitted, at the age of 28, in August, 1901, into Rainhill Asylum and who died there in January, 1910. His occupation was that of a piano tuner. His family history was good. His father was an intelligent man, a Nonconformist minister, and there was no record of psychoses, epilepsy, or allied neuroses in the family. The patient himself had always enjoyed good health and had never had any serious illness except an attack of pleurisy. He was bright and intelligent as a child and early showed promise of great musical ability, but he was always of a nervous, excitable disposition. At the age of 9 years he seems to have had an attack of petit mal, but the first distinct fit occurred when he was 16, and since that time he remained subject to epileptic attacks at variable intervals. For about a year before admission, the parents of the patient had noticed a gradual mental deterioration, which doubtless had been coming on insensibly for some time previously, but no attack of an active psychosis occurred until ten days before admission, when he became delusional, excited and threatening, which necessitated his removal to the asylum. He was on admission a man of good physique, 5 ft. 10 in. in height, and over 168 pounds in weight, with all his viscera apparently healthy. The limbs were well formed and symmetrical; there were no deformities of any kind. The circumference of his head was 25 inches. His mental condition was one of partial dementia. He was dull and weak-minded generally, slow of speech with prolonged reaction time, but was nevertheless fairly intelligent, could give a good account of himself and was an active worker. His history thenceforward was that of epileptics generally. The frequency of his fits increased in spite of treatment by bromides, so that

whereas in the year 1902 his fits had averaged 5.9 per month, these had increased in 1907 to 9 per month. Along with this went steadily increasing impairment of memory and intelligence; he also had periodical attacks of excitement, lasting for a few days at a time, during which he was violent and dangerous, such attacks usually leaving him on their subsidence somewhat duller than before. Even when at his best he was markedly irritable and quarrelsome and showed much emotional instability. The steady mental deterioration was well shown by the fact that when first admitted into the institution in 1901 he was a useful worker in the place, but by the end of 1906 he had become useless as such, and in 1909 he had not sufficient mind to employ himself in any way. Concurrently with this his health gradually failed, and he finally died of an attack of broncho-pneumonia at the age of 37.

On autopsy the case was found to be one of very pronounced macrocephaly, and as these cases are of comparative rarity the authors came to the conclusion that the record may prove of interest even though the origin of the matter still remains obscure. None of the conditions which have assigned a causal relationship in this connection appear to have been present. The internal secretory glands were unfortunately not preserved for microscopical examination, but to the naked eye these all seemed to be quite normal.

The brain, although one of the heaviest on record, was for the most part normal in appearance differing from the average brain mainly in its increased size and complexity of pattern. The microscopical changes found also did not at all necessarily bear upon the question of cerebral hypertrophy, being merely those usually present in the brains of chronic epileptics. On one point, however, the microscopical examination gave definite, although negative, results. There was no general hyperplasia of the neuroglia such as has been considered to form the basis of cerebral hypertrophy in some cases.

Although there were some indications in places of defective development of the sulci and of reverting tendencies, these were not numerous, nor were they pronounced, probably not more so than are such indications in the majority of human brains, even in those which may be described as generally well developed.

There were, however, in the cerebral hemispheres many signs—apart from their large size—of a formative activity much greater than that usually seen. This increased developmental activity, the authors think, proceeded, for the most part, on regular lines, and on the whole the departures from the ordinary in the convolitional pattern of the brain were in the direction of superiority. On the other hand, the tendency to insulation of areas of the cortex, and the marked spurring and forking of certain sulci, such as was seen particularly in the left temporal region, might probably be looked upon as indications of formative activity on irregular or aberrant lines. With this greatly increased developmental activity, whether in the direction of superiority or aberrancy, it seems likely that there would be a tendency to instability.

It is of interest to note, in this connection, that unlike many other recorded cases of this class, the patient was neither idiotic nor imbecilic, but was originally of at least average general intelligence, with special ability in music. Epilepsy, as in this instance, is a frequent complication in these cases, and this suggests the idea that even when the minute structure of

the brain appears to be normally constituted—which it by no means always is in such cases—the usual size of the brain, apart from the increased complexity of pattern, cannot be largely exceeded without introducing a condition of instability which renders its possessor liable to suffer from some form of nervous breakdown, and especially from epilepsy.

3. *The Relation of the Myopathies.*—The author recognizes two groups of muscular atrophy, (1) the congenital, (2) the acquired. The congenital group embraces the cases of arrested growth in certain limited regions of the body, the infantile nuclear arrest ("Kernschwund") of Möbius. This muscular condition may be caused by defect of the muscles with complete integrity of the nerve apparatus, or, on the other hand, the peripheral neurones may be much affected. The congenital arrest usually is not progressive, although it may be progressive in exceptional cases.

The acquired muscular atrophy also consists of two types, the primarily muscular, with intact nervous system, and the nuclear (neuronic), in which the nerve cells of the peripheral neurones are affected. In the former we have the progressive muscular dystrophy, including the bulbar form of Hoffmann; in the latter we have the Werdnig-Hoffmann muscular atrophy, the infantile bulbar atrophy of Fazio, Charcot, and Londe, and the forms of myelopathy occurring later than the period of childhood. These types, as is well known, are progressive.

The various forms are combined in rare cases, and certain of the muscles of the body, as the pectorals and serratus magnus, show a special liability to arrest or atrophy in both forms. It is also well known that reaction of degeneration and fibrillary tremors, as well as changes in the cells of the anterior horns, have been observed in progressive muscular atrophy, so that sharp distinctions cannot invariably be maintained.

The relation of muscular defect to progressive muscular dystrophy has been recognized by a number of investigators. By some the defect has been considered as arrested intra-uterine dystrophy. In persisting portions of arrested muscles the histological findings have been those suggesting intra-uterine muscular dystrophy, and yet we must be cautious in accepting these findings as conclusive, for it is questionable whether a decision can be formed as to the myopathic or myelopathic character of atrophy from the microscopical findings in the muscles alone.

JELLIFFE.

Review of Neurology and Psychiatry

(Vol. XI, No. 12)

The Direct Ventro-Lateral Tract. W. G. Spiller.—The writer first described this tract in 1898. It was traced at the time as far as the first cervical segment. It arises from the pyramidal tract in the pons or medulla oblongata, and when arising in the latter place it is described as passing backward with the anterior external arcuate fibers, or a little interior to these, on the periphery of the medulla oblongata. After reaching the region of Gowers' tract the fibers of the direct ventro-lateral pyramidal tract bend again downward, and pass in this region to the ventral periphery of the lateral column into the thoracic region, or in one case (Bumke) to the lumbar swelling. Ziehen speaks of the tract as the aberrant ventro-lateral pyramidal tract.