FAMILIAL TABES DORSALIS.

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THAT heredity is an important factor in the incidence of nervous disease cannot be gainsaid, though the discoveries of modern bacteriology have shed the light of knowledge in many dark places where conjecture was lacking in material proof of an acquired infection. Modern thought turns with increasing respect to the ultra-microscope for evidence of organisms as a cause of disease, and present-day theories anticipate much that will most surely become commonplace fact when protein chemistry advances far enough to simplify many 'reactions' which now rest upon mere supposition. In our concentration upon the study of parasitic organisms as disease-producing agents, we are apt to thrust into the background the obscure factors of tissue-peculiarity, 'sensitiveness', and selective action, or in other words, the state of the soil in which the seeds of disease are sown. Neurology has always recognized the potent influences of family tendency and selective action in the causation of chemical and microbic lesions in the various highly specialized units of the nervous system. 'Sensitiveness', tissue-affinity, and cell defence are almost incalculable factors in disease. We cannot form any exact estimate of how much damage will result with a known dose of toxin in any individual, and a repeated dose will have an effect differing at least in its intensity from that of the first—a phenomenon now well recognized in the solution of all problems of acquired and artificially produced immunity. If the source and virulence of any infective agent be known, there are still disturbing reactions, such as tissue sensitiveness and other biochemical reactions of defence, which will confound the clinician in his estimate of any resulting lesion.

Family tendencies to certain diseases have been much discredited by the findings of modern bacteriology. There are notable exceptions, such as hæmophilia, pseudohypertrophic paralysis, and so on; but tuberculosis and other microbic diseases are known to fail in their attempts to pass through a healthy placenta to the foetus. It is easy to believe that certain strains of bacteria acquire a selective action upon particular tissues. Cases have been reported where several patients, friends not related by tissue-continuity, have acquired syphilis from a single source, and where the late effects have been general paralysis in all. An attractive way of explaining such clinical histories would be to associate a certain type of virus with a

definite selective action upon certain tissues, a course made all the more credible by recognized precedents—for example, tetanus, etc. Since the demonstration by Noguchi of the *Spirochaeta pallida* in the brains and cords of general paralytics and cases of tabes, findings confirmed by most other neuropathologists to the extent of about 80 per cent of all cases of these diseases, there is a tendency to discredit any other factor except mechanical and other influences such as vascular and lymphogenous portals of entry to the site of the lesions.

When, however, several members of a family born of healthy parents develop tabes—following upon a clear history of acquired syphilis in four cases, and all from widely different sources—then surely an explanation must be sought in the realms of speculation, such as family- or tissue-peculiarity; clearly a similar strain of spirochaete cannot be held accountable in this set of circumstances.

GENEALOGICAL CHART OF THE X FAMILY.

It may be remarked that in all these patients there has been not only the same disease, but a similar type of tabes, uncomplicated in all by paralysis, marked optic atrophy, gummata, or aneurysm of the aorta. The following abbreviated notes form a record which may be allowed to speak for itself.

Family History.—The father, a member of the medical profession, was a healthy man until he reached the age of 70 years, when, one cold winter, he succumbed to an attack of acute bronchitis with heart failure. The mother is a delightful old lady, now alive, and active in all her faculties at the age of 92. She suffered from scarlet fever as a young girl, but since then she has never ailed more than with an occasional winter catarrh. She is a little deaf to-day, but takes her daily walk and does her household shopping on foot. I had the opportunity of examining her, and could find no physical signs of disease in any organ from a superficial examination of her chest and the usual routine examination of her nervous system. The arteries were very healthy for her age, the pulse good in volume, and

the sphygmomanometer read 160 mm. of mercury for her systolic pressure.

Details of Family of Mrs. X.—

1. Daughter (Mrs. A. T.), now dead, aged 67, 'stroke'. Examined in 1914 at age of 63 and found to be definitely tabetic.

2. Son (W. H. X.), now 67, first examined in 1914 at age of 61, a chronic tabetic.

3. Twins, one still-born; the other lived twenty-four hours, weakly at birth, no rash.

4. Daughter (Mrs. W.), now 63, examined in 1914 and again recently; definitely tabetic.

5. Daughter, died at age of six months of bronchopneumonia.

6. Son (G. X.), the original patient, examined in 1914, age 51, and pronounced to have tabes. Re-examined November, 1920, now 57.

7. Son (C. X.), age 55, healthy.

8. Son (R. L. X.), age 46; suffers from early tabes.

There were no other children and no miscarriages.

CLINICAL NOTES UPON THE INDIVIDUAL CASES.

Case 1.—The first member of the family to come to my notice was No. 6, G. X., admitted to the General Infirmary, Leeds, in April, 1914, under the care of Dr. Barrs, to whom I am grateful for permission to publish the clinical notes.

The patient, whose age on admission was 51, a well-nourished man, complained of stabbing pains in his legs, and inability to stand for long without support, or to walk without assistance on either side. He spoke in a natural manner, with some lack of facial expression, and demonstrated that he was not paralyzed, but drew attention to the unsteadiness of his legs, which he said were out of his proper control.

History.—Had measles and scarlet fever as a child. Acquired syphilis when 27, primary sore with secondaries. Treated for about a year with mercury and iodide; no further manifestations.

He first noticed the beginnings of his present complaint ten years ago, at the age of 41. He had some difficulty in walking downhill or downstairs. At dusk he was uncertain of the ground. On shutting his eyes he at once became unsteady, and would fall unless there was some support handy. A few years later he felt a curious tight sensation across the upper part of his body and the lower ribs, which has persisted since, but he has ignored it of late. About five years ago he had trouble in starting micturition, but this has passed off lately because he has always been careful to urinate at regular times, finding that he could not tell when his bladder was full. On a few occasions the urine has dribbled and wetted his clothes, but there has been no retention. There have been 'rheumatic pains' in his legs, especially at night-time, sharp darting pains which come and go, rarely disturbing sleep. He has gradually lost the proper control of his legs in walking, and latterly in standing. He says that his difficulties are due to numbness of his feet and uncertainty of knowing where to place them.

Objective Examination.—No external scars except a doubtful one on the glans penis, no nodes in the long bones or skull.

NERVOUS SYSTEM.—Memory, attention, and ideation good. No headaches, no fits. Speech excellent; writing a little tremulous, but does not differ in any other way from that of early years.

Gait is wildly ataxic; the legs are awkwardly pulled up, and he steps high, bringing the feet down heavily in attempting to walk. He requires support on either side, and leans heavily upon his helpers. The arms are almost normal until the eyes are shut, when he is mildly inaccurate in finger tests.

Cranial Nerves.—I: Normal. II: Normal in acuity, fields, and fundus; the latter is a little pale, but definite atrophy is not diagnosed. III, IV, and VI: Pupils small, slightly unequal, react on accommodation but fail to react to light; no ophthalmoplegia, no nystagmus. V: Corneal reflex sluggish, no motor change. VII: Expression rather wooden, no paresis. VIII: Slight nerve deafness, no vertigo. IX, X, XI, XII: All normal, tongue steady.

Motor Functions.—Marked hypotonia in both legs, none in arms, inco-ordination as above. Deep reflexes: Jaw-jerk present. Biceps-, triceps-, supinator-jerks obtained on re-enforcement. Knee-jerks absent, ankle-jerks absent. Skin reflexes: Scapular obtained, abdominals sluggish (adipose patient). Gluteals, cremasteric, and bulbocavernosus reflexes not obtained, probably absent. Plantar reflexes very sluggish, but flexor response obtained on warming foot.

Sensory Functions.—Upper limbs: Full sensation except deep-pressure pain, which is practically absent; joint and muscle sense to passive movements, etc., good; no astereognosis; tuning-fork vibration not felt. Lower limbs: Total loss of joint and muscle sense below knee; marked analgesia on deep pressure over nerve-trunks; some anæsthesia of soles; hypo-æsthesia to rough tests with hot and cold tubes in lower leg on both sides.

OTHER SYSTEMS.—Healthy. Arteries generally a little thickened; systolic blood-pressure, 150 mm. of mercury.

Wassermann reaction with blood-serum negative.

Lumbar puncture not required for diagnosis, and not done.

Subsequent History.—During the late war I was unable to keep in touch with him, but I re-examined him on Nov. 1, 1920. He is now much worse than he was six years ago, and does not leave his chair except for essential duties. His arms are now inco-ordinate, and his writing is worse. He has a Charcot's shoulder affection, and also an enlarged painless elbow, both on the left side. These joints grate on movement, and are easily dislocated. His sight is good, and the pains are much better. His speech and mentality are normal. All signs are much as they were on the first examination, except for the arm condition, which came on quite suddenly about a year ago. The joint sense is quite good in the fingers, even those of his left hand. Pallanæsthesia is noted in all four limbs. There are no trophic ulcers.

Case 2.—W. H. X., elder brother of above. Examined June 9, 1914, when his age was 61. The patient states that he has 'locomotor ataxia'.

History.—Has had measles, scarlet fever, influenza. At the age of 27 he acquired syphilis from a different source than in the case of G. X. He was a medical student at the time, and was quite *au fait* with his disease and its treatment. He was carefully treated for fully a year and a half with perchloride of mercury, black wash, and iodide afterwards. No further symptoms.

About the age of 46 he found that in writing his hand occasionally went off at a tangent. He noticed unsteadiness in standing to wash his face, and felt very awkward at a concert about that time when he had to find his seat with the lights turned low. He gradually developed a difficulty in crossing a road when he had to look up and down the street to notice if vehicles were passing; on other occasions he found that his eyes were most important help to his walking. Since the age of 56 he has been unable to walk in the dark. For the last four or five years he has had pains in his legs. For some years he confesses to a girdle sensation, and his sight has not been so good lately.

Objective Examination.

NERVOUS SYSTEM.—Memory good. Speech perfect, higher centres unimpaired. Mask-like expression a feature. No fits. Sexual appetite retained. No bladder or rectal trouble, no stomach symptoms.

Cranial Nerves.—Pupils small, regular, centric, fail to react to light but move on convergence. Fields normal to rough tests. Discs: slight pallor, but no definite atrophy. All other cranial nerves healthy. Tongue steady.

Motor Functions.—Power good, joints perfect. Hypotonia in arms and legs on passive movement. Deep reflexes: Biceps- and triceps-jerks much reduced, but obtained on re-enforcement. Knee-jerks and ankle-jerks absent. Abdominal reflexes sluggish, cremasteric reflexes obtained. Plantar reflexes doubtful; any response is flexor.

Sensory Functions.—Great impairment of deep sensibility in lower limbs; the patient cannot imitate the positions of joints passively moved. Deep analgesia. Total pallanæsthesia in arms and legs. Some preservation of joint sense in hands; no gross cutaneous anæsthesia. Gait ataxic, but not remarkably so. He at once becomes unsteady when his eyes are closed.

OTHER SYSTEMS.—Healthy. No aneurysm.

Wassermann with serum negative.

Subsequent History.—Re-examined November, 1920. The patient's condition is little changed in the last six years. All functions are about the same; he thinks, however, that he can walk a little better than he could a few years ago. There has been no alteration in the optic discs, which are pale without definite atrophy.

Case 3.—Mrs. A. T. Examined June 9, 1914, her age then being 63.

History.—This patient was perfectly well until her marriage at the age of 18. She contracted syphilis from her husband. There were definite specific lesions, chancre, condylomata, falling of the hair, and iritis. She was carefully treated for at least six months by calomel and opium, afterwards by a course of potassium iodide, and was apparently cured. Her hair grew again, and her general health improved in a most striking way.

About the age of 50 she became unsteady in her walking. She

described her sensations as "like walking on a cushion". A few months later she complained of 'darting pains' in both legs; there was also some loss of bladder sensation and occasional incontinence of urine. Three years later she sustained a severe compound fracture of the leg owing to a slight fall in her kitchen; the fracture united with great deformity after considerable treatment. She sustained another fracture, not so severe, two years later, as a result of a second trivial fall. The husband died as the result of an accident. He was not tabetic.

Objective Examination.

NERVOUS SYSTEM.—A typical case of pronounced tabes dorsalis. Patient is profoundly ataxic, and unable to stand without support. Complete loss of pressure pain in the legs. No tendon-jerks obtained in either the upper or lower limbs. Cranial nerves healthy except that the pupils are unequal, irregular, and of the Argyll Robertson type. Mental functions unimpaired, though she is depressed at her constant invalid state. The tongue and lips steady; handwriting good.

Subsequent History (given by her daughter who nursed her during the latter years of her life).—The muscular pains became gradually worse; at times she had crises of pain causing her to scream in agony, and only obtained relief by hypodermic injections of morphia in increasing doses. Between the years 1914 and 1918 a few slight strokes occurred, but there was not much permanent paralysis as a result. In the periods of freedom from her attacks of pain she was mentally very active, and played a good game of whist, on which she was particularly keen. There was complete incontinence of urine during the last four years of her life. She became aphasic after a severe stroke, but recovered speech some few weeks later. Death occurred on Sept. 14, 1918, following a severe apoplectic attack, in her 68th year.

Case 4.—Mrs. W. Examined June 9, 1914, her age then being 57.

History.—This lady had measles and scarlet fever as a child, but was healthy afterwards until her marriage. Her husband had acquired syphilis previously, and suffered from symptoms of constitutional syphilis after marriage. His wife's health became affected, but she had no definite chancre. Both patients were treated over a period of many months by mercury and iodide. The wife suffered from sore throats; her hair was shed, and there was some skin rash, but her health subsequently improved. There were no children of the union, and no miscarriages. The husband is not tabetic.

At the age of 55, two years before she came under observation, she noticed the onset of unsteadiness when she walked. She could not stand in the street, nor turn round sharply. Her sight deteriorated, but she is able to see fairly well yet, even in a poor light. There has been a little difficulty in starting the act of micturition.

Objective Examination.

NERVOUS SYSTEM.—Mentally good. Speech perfect. No fits. Cranial nerves: No palsies; Argyll Robertson pupils; discs pale, but vessels of

good size; tongue steady, no tremor. Motor functions: No paralysis; marked inco-ordination in the movements of the legs to all tests; hypotonia in the leg muscles; knee- and ankle-jerks lost; plantar reflexes flexor in type. Analgesia on deep pressure over the nerves and muscles of the legs. Upper limbs healthy in function, and all arm reflexes present.

Subsequent History.—Re-examined Nov. 2, 1920. The disease has progressed slowly since the patient was first seen six years ago. She cannot go out of the house now because her gait makes her an object of interest to passers-by. The bladder disorder is much worse; there is troublesome incontinence, with the occasional passage of blood and slime. The legs are much more inco-ordinate, and the disease is now affecting the upper limbs, tendon-jerks being obtained with great difficulty in the arms, and there is now some hypotonia in these members. There is no mental impairment, and no trophic change. For personal reasons no Wassermann reaction with the blood-serum or spinal fluid was proposed to the patient.

Case 5.—R. L. X., age 46 years.

History.—This patient had a healthy boyhood until the age of 15, when he was supposed to have acquired gonococcal urethritis, which was promptly treated. He twice acquired a definite gonococcal infection subsequently, at the age of 27, and again about the age of 35. There was no hard sore so far as he knows, and certainly no rash. He had no mercurial treatment internally, though he had black wash applied locally to the penis. He has always been 'nervy', easily excited, and somewhat lacking in self-control. For the last few years he has had some difficulty in walking at times, especially in the dark, and is liable to what he calls attacks of dizziness. In a darkened room he usually likes to be near a table or other ready support in case of need. He has a tight feeling across the upper part of his abdomen, but this he associates with flatulence, because the feeling does not persist when he has brought up some wind from his stomach.

Objective Examination.—The patient is explosive in his expressions, and unduly emphasizes each remark. He is emotional, and mentally very quick in all his reactions. The pupils are slightly smaller than the average size; they give a definite reaction to light, but the movement is sluggish, and restricted in range. The co-ordination, tone, and power of all muscles are excellent. The knee-jerks are obtained without re-enforcement. The upper limbs are healthy in all respects. There is some analgesia of the muscles of the legs, and also on pressure over the peroneal nerve at the neck of the fibula. There are no other signs of established disease in the posterior columns of the cord. Joint and muscle sense are excellent. Generative and bladder functions are healthy.

Case 6.—C. X., age 55. This member of the family has never suffered from venereal disease. He married at the age of 27, and has a healthy family. Apart from an attack of renal colic about three or four years ago, he has been perfectly healthy. He was carefully examined by me, and I can safely say that his nervous system is absolutely healthy. He is the only member of the family who has escaped venereal infection, and in whom one can definitely exclude tabes.

SUMMARY.

Of the six surviving members in a family of eight, born of healthy parents, four have acquired undoubted syphilis from various different sources, with a resulting tabes dorsalis in all. A fifth member of the family acquired gonorrhœa (syphilitic infection being doubtful), and probably has early tabes dorsalis. A sixth escaped venereal infection and remains healthy.

It is suggested, in the light of the above facts, that tabes dorsalis cannot be attributed to a special strain of spirochæte introduced at the initial infection, but is much more likely to be due to the spirochæte acting upon tissues specially sensitized, either by natural family peculiarity or by certain methods of treatment.

The discovery of the spirochæte in the tissues (brain and cord) of general paralytics and tabetics should stimulate further research into the other, still unknown, factors which are concerned in the production of these diseases.