SYPHILIS OF THE SPINAL CORD.¹

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THE *post hoc propter hoc* argument has been used so freely with regard to syphilitic diseases of the central nervous system that it seems eminently desirable to be able to make the diagnosis of cerebral or spinal syphilis without reference to the etiological factor. We should be able to recognise the symptoms of syphilitic disease whether or not the patient concedes, or we can prove previous syphilitic infection. My own experience during the past few years leads me to believe that this can be done with tolerable certainty in the case of the diseases under discussion.

Some time ago in a paper on "Multiple cerebro-spinal syphilis,"² I referred to the morbid lesions and to the important points of differential diagnosis for cases of syphilitic disease involving both the brain and the spinal cord. In this paper I propose to confine myself entirely to the diagnosis of spinal cord syphilis.

A fresh impetus has been given to the writings on this subject by Erb's short article³ which appeared in the *Neurologisches Centralblatt* for March 15, 1892. With the same keen clinical insight which led to admirable results in his studies on muscular atrophies and of Thomsen's disease, Erb has sought to establish "a type" of spinal cord disease which he proposes to call "syphilitic spinal paralysis."

¹ Read before the New York Neurological Society, April, 1893.

² *New York Medical Journal*, 1891.

³ A detailed analysis of Erb's cases will be found in an interesting article by Dr. S. Kuh, of Chicago, published in the *Zeitschrift für Nervenheilkunde*, Bd. iii.

This special type is to be recognised by the following characteristics :

- (1.) The usual symptoms of spastic paraplegia :
The peculiar gait, carriage and movements.
- (2.) Marked exaggeration of the deep reflexes.
- (3.) Muscular contractures slight as compared with the exaggeration of the reflexes.
- (4.) Involvement of the bladder.
- (5.) A slight, yet distinct disturbance of sensation.
- (6.) Gradual onset of the disease.
- (7.) A decided tendency to improvement.

We may concede at once that these points are well taken and that the symptoms stated above leave little doubt of the syphilitic nature of a given case. It is due to Rumpf to say that, on p. 351 of his well known monograph, he alludes to all these diagnostic symptoms in a case which was proved to be due to specific disease of the blood vessels in the spinal cord. He mentions spastic symptoms, paresis followed by paralysis of the sphincter of the bladder with incontinence of urine, exaggerated reflexes and contractures ; later on in the course of the disease the diminution of sensation. The onset, too, was a gradual one. Rumpf, naturally enough, in speaking of his case which ended on the *post-mortem* table, does not refer to a tendency to improvement, and he does not insist, as Erb does, on the relatively slight muscular contractures as compared with the great exaggeration of the reflexes.

In developing his spinal type Erb recognises Rumpf's services in this field. The former may claim the special merit of attaching greater importance than Rumpf did to this difference between the intensity of development of muscular contractures and of increased reflexes.

The questions I have been trying to solve for some time past are—first, whether this Rumpf-Erb type represents the most frequent form of syphilitic spinal cord disease ; and secondly, whether the cases that conform to this type represent *a* syphilitic spinal paralysis, or *the* syphilitic spinal paralysis, as Erb would have it. This is not a mere quibble of words. Much mischief was done, as every one will acknowledge, by Strümpell's procedure in describing the

clinical features, of what he supposed to be polio-encephalitis, and in labelling these cases "*Die cerebrale Kinderlaehmung*," whereas it is now well established that if his cases exist at all they constitute a very small proportion of all the infantile cerebral palsies. I do not fear a similar mistake with regard to Erb's type of spinal cord disease, but there is a distinct tendency even now among more recent writers to imply that every case of syphilitic spinal cord disease should conform in most respects to the type described by Erb.¹

This leads to the further question: Are the symptoms described by Erb so characteristic of spinal syphilis that we should trust to them entirely, or are there other symptoms which point with much greater force to the syphilitic nature of cord disease?

For the present the question is a purely clinical one, and one of great practical importance at that. The question in pathology can be brought up after the clinical question has been settled.

Let me record in condensed form a few cases, taken from private practice, of undoubted syphilitic disease of the spinal cord, from which we shall be able to draw the necessary inferences.

Case 1.—Mr. C., age 50, was referred to me by Dr. Gibney, September, 1891, and has been under my treatment ever since.

There was a distinct history of syphilis acquired two years previously, followed by constitutional symptoms.² He has been married 23 years; wife has had no children. He has been an active brain worker for many years, but moderate in his mode of life. In November, 1890, he had an attack of illness, during which his right eye-lid drooped; his left leg was paralyzed. In the course of a few weeks he recovered sufficiently to get about again. He also had "*eczema ani*," and shortly after this was treated by various physicians for the tertiary symptoms of syphilis.

In the spring of 1891 he had a severe attack of influenza. A few months later in May, 1891, he developed a right hemiplegia, and a paresis of bladder and rectum. The left leg was still

¹ It is evident from Dr. Kuh's article (which has appeared since the above was written) that Professor Erb recognises mixed forms, &c., of syphilitic disease of the spinal cord.

² I am indebted to Dr. J. P. Tuttle for the early history of the case.

weak from the first attack. He recovered in a few weeks and was once more active in his business.

Two weeks before I first saw him he was compelled to go to bed again, his legs having grown weak and his bladder and bowels not acting voluntarily.

The condition I found him in was a rather pitiable one. In addition to complete spastic paralysis of both lower extremities, there was complete paralysis of both upper extremities; there was no increase of the wrist and elbow reflexes, but a decided tendency to atrophy of the interossei particularly in the left hand. The knee jerks were enormously exaggerated; there were *extreme contractures* of the adductors of the thighs, and the very marked contractures of the posterior leg muscles inhibited the ankle clonus. The pupils were normal, speech normal; sensation in both lower and upper extremities was normal at the first examination. The electrical reaction of all nerves and muscles was entirely normal. There was retention of urine and inability to evacuate the bowels. This group of symptoms has been materially modified from time to time.

The involvement of upper and lower extremities with symptoms pointing to a very irregular invasion of the various systems of the spinal cord left no doubt in my mind as to the nature of the trouble, and in spite of very discouraging circumstances, I adhered to, and pushed, the mercurial and iodide treatment¹ to the extreme. During the first week the symptoms increased in severity. The paralysis became absolute and anæsthesiæ were soon associated with the paralysis. The anæsthesia was evident only in both feet and legs up to the middle of the thighs and over the sacrum and lower dorsal spine. There was no anæsthesia in the upper extremities, but very decided atrophy of the interossei. Within two weeks of the beginning of the treatment, while the conditions of the lower extremities remained unchanged, the upper extremities recovered completely and have been entirely normal ever since. Scarcely had these symptoms been recovered from before the intensity of the process in the lower cord seemed to increase. A bed-sore began to form which in spite of the most careful nursing increased in size until it was large enough to put one's fist in, and for fully eight months it was a threatening feature of the case, but finally healed completely under the surgical skill of Dr. Gerster. There has been

¹ Inunctions of six grammes of mercurial ointment daily, and gradually increasing doses of the saturated solution of the Iodide of Sodium, from 10-150 drops, three times daily.

a gradual improvement in the paralysis, so that after much patient treatment he is now able to walk about with the assistance of his crutch and an attendant, with every prospect of regaining the normal use of his legs unless a sudden relapse should set in. All anæsthesiæ have completely disappeared, the contractures are relaxed (the ankle clonus is now apparent), but spasmodic twitchings of the legs are still quite frequent. The patient has also recovered from several attacks of cystitis, but is still suffering from a specific ulcer of the rectum.

Recovery in this very severe form of spinal syphilis I consider fortunate enough, and doubt if it would have been possible if the symptoms had not been due to a specific process.

Please note that the contractures were extreme and that at one period very marked sensory disturbances existed.

Case 2 resembles the first in many particulars and still has run a very different course.

Mrs. X., a lady age 53, having five children, all healthy and married, was first seen in June, 1891. She was in good health until two years previously, and extremely active in spite of her weight of nearly 250 pounds. After the "blizzard," March, 1888, she had a troublesome cough and for a long time did not feel well. In spring of 1889 began to notice a heaviness in the left leg; this would remit at times and then would grow worse again, so that she was accused of having an hysterical affection. The trouble was restricted to the left leg for nearly two years, before the right leg became similarly affected. A few weeks before I saw her she was sent to Atlantic City by a physician who thought that the change of air would have a favourable influence upon her supposed functional paralysis.

The first examination revealed a very different state of things : (a) Extreme and absolute spastic paraplegia; not a muscle of a toe even could be moved. (b) Extreme contractures, so that the knees would be firmly clasped together unless forcibly separated by pillows. (c) Violent spasmodic twitchings which gave rise to excessive pain. (d) There was complete anæsthesia of both legs as high up as a line passing two inches above the symphysis pubis. (e) Loss of faradic response without alteration of galvanic formula in all leg muscles and nerves. (f) No tendency to bed-sores; and (g) absolutely no trouble with bladder or rectum.

The upper extremities were not affected. No history of traumatism or of unusual exposure, and, I beg you to remember, no history of an acute infectious disease. In the absence of these predisposing conditions—and more particularly on account of the

extreme spastic symptoms associated with disturbances of sensation and with changes in electrical reactions (atrophy could not be made out on account of adipose tissue) without, however any interference with vesical and rectal reflexes, and no tendency to formation of bed-sores, I made the diagnosis of a specific myelitis. No other pathological process is as selective as this one. The symptoms pointed to involvement of the lateral columns, of the posterior roots or posterior gray matter, and also of the anterior horns or roots on both sides, and yet the column of Clark and the central gray matter were seemingly not affected.

I have persisted in specific treatment up to the point of tolerance with gratifying results, although the proof of any specific illness was a long time forthcoming. Some nine months after the beginning of treatment the lady's husband consulted me for very persistent headaches. Suspecting the nature of the trouble, I discovered not only that he had acquired specific disease many years since, but that he had been under a distinguished surgeon's care for bone syphilis only a year ago. The further progress of the wife's case has been marked by a gradual restoration of the electrical reactions, by total disappearance of the anæsthesiæ, by the cessation of all contractures and of the spasmodic twitchings, and by the slow and steady diminution of the spastic paralysis. She has regained moderate use of all the muscles of the lower extremities, but is still confined to her chair, as her legs will not yet bear her tremendous weight; but I expect her in a few months to learn to walk, as single efforts to stand have been successful, and she moves legs and feet very readily while sitting in a chair. I have not seen any but specific cases of spinal cord running such a chronic course and yet showing a tendency to improvement after the lapse of one or two years and even longer. I hold that in this case, next to the very remarkable improvement, the entire absence of bladder and trophic symptoms compared with the intensity in development of the other symptoms is the most characteristic diagnostic feature.

I could parallel this case by at least two others, but I do not wish to go into wearying details.

Case 3 is that of a colleague now practising in Hot Springs, Ark. He is 36 years of age; married, no children; formerly in good health; first symptoms of fatigue on slight exercise in 1880; began to look pale and run down. Was in Ottawa, Canada, at the time, and went to sea for a couple of years; did not improve, and he began to have rheumatic pains about joints, and suffered somewhat from dizziness. After his return he developed a very

marked spastic gait which has slowly increased up to the present time; a slight irritability of the bladder, and loss of sexual power were symptoms from which he suffered at various times; but both these functions are now normal. He suffers most, however, from headaches. On further questioning I learned that one year ago after the onset of spastic symptoms he had some difficulty in speaking, associated with a numbness of the left side of the body; once had an attack of aphasia lasting 24 hours, and remembers a temporary weakness of left hand. As a result of my examination, I noted inequality of pupils, no other head or ocular symptoms; grasp excellent in both hands, but very exaggerated wrist and elbow reflexes; spastic gait with considerable rigidity, so that in walking the knees are pressed quite closely together; exaggerated knee jerks; no anæsthesia anywhere, no trophic symptoms, no tremors, no stammering speech, &c. I made the diagnosis of spinal syphilis, and then learned that fifteen years ago he had been inoculated with syphilis on the middle finger of left hand. I urged repeated mercurial and iodide treatment, but this has not relieved the spastic symptoms.

Please note again extreme spastic symptoms with contractures, slight involvement of the bladder, no sensory symptoms, and no atrophic symptoms, proving once more the selective character of the process. The case resembles in most respects those described by Erb as typical of syphilitic spinal paralysis.

Case 4.—This case, differing as it does from all other cases of spinal or cerebro-spinal syphilis I had seen, led me to seek for safe guides in differential diagnosis. I saw the young man, F. I., age 26, in consultation with Dr. Lustgarten, Oct. 14, 1892. He comes of a very markedly neurotic stock with the history of insanity in the family, and has one sister who is deaf and dumb.

Four years ago he had acquired undoubted syphilis. Since last spring has experienced difficulty in walking and in the use of his hands.

There was also the history and evidence of paralysis of the right superior oblique. At the first examination the young man was quite unable to lift himself up in bed; there was distinct atrophic paralysis of both trapezii, of infra and supra spinati, of deltoids and pectorals; a paresis of the entire arms and hands; grasp being almost nil.¹ All forms of sensation, particularly tactile and temperature sense, were normal. This was true of the upper extremities and of the lower extremities and on the trunk to the level of the umbilicus. Elbow reflexes could not be

¹ At this first visit a careful electrical examination could not be made.

elicited. The lower extremities were much emaciated, more so the right than left; great weakness, scarcely able to lift legs an inch from the bed, and yet if put on his legs could stand a little; no ataxia, no absolute paralysis anywhere. Right knee jerk lost, left knee jerk very feeble; all superficial reflexes lost; no disturbances of vesical and rectal reflexes: no bed-sores, but a severe burn in the anæsthetic area on the right side just below the margin of the ribs.

The diagnosis of wide-spread syphilis was clear to me, although the paralysis tended to the atrophic rather than to the spastic type.

The good results of treatment were very evident at my next examination about two months later, Dec. 6. Patient was able to sit up in bed and to walk about though feebly; the pupils now reacted to light, though sluggishly, and the muscles about the shoulder joint had recovered surprisingly from the atrophic condition. Normal sensation had returned, leaving a relatively small area of anæsthesia on the trunk. It is worth noting that at a still later examination the area of anæsthesia had again increased, while all other symptoms had receded.¹ Grasp of hands nearly normal, all muscles with the exception of the extensor groups in both forearms having been restored to their normal volume. Faradic responses diminished but present in all muscles even in the extensors, with the exception of the anterior tibial group of the right side. Both knee jerks can now be elicited by reinforcement, and his mental condition has much improved.

Four months after first examination patient was able to call upon me at my office, improved in every respect, walking without assistance and simply using a walking-stick to get about with. He was still affected with double vision, in consequence of paralysis of right superior oblique.

If we except the slight ocular trouble, this was a spinal case clearly enough. The paralysis was of the atrophic and not of the spastic order. The involvement of both the upper and the lower extremities; the unequal development of the symptoms (the paralysis and anæsthesia being very pronounced, while the bladder was not involved, and the electrical conditions and the reflexes were less distinctly altered, and trophic disturbances were wanting entirely), this peculiar and unequal development of symptoms, as well as the rapid recovery, proved the case to be one of spinal syphilis with slight cerebral involvement.

¹ Drawings illustrating this variation in the areas of anæsthesia were exhibited at the meeting.

In the cases just recorded the diagnosis of spinal cord syphilis can hardly be questioned. I share the fate of other writers, in not being able to offer *post-mortem* evidence of these diagnoses. Recent syphilis of the central nervous system if properly recognised should not lead to *post-mortem* examinations, excepting in rare instances; and it is true, moreover, of all syphilitic diseases that the clinical manifestations are in many respects far more reliable than are the results of histological examinations of the diseased tissues, for, curiously enough, the pathologist in making his diagnosis has to rely very largely upon points obtained by clinical examination.

Now what were the features which led to the diagnosis of syphilis in the cases I have detailed? In one there was wide-spread atrophic paralysis, in three of them there was spastic paraplegia of the most pronounced type. In these the reflexes were excessively exaggerated. In two of them the muscular contractures were slight; in one of them they were extreme. In one case there was permanent involvement of the bladder. In the others no involvement. In all but one of them sensation was disturbed. In two the onset was gradual. In the others it was quite sudden. All of them have shown a decided tendency to improvement.

I have mentioned first the features in which they conformed to Erb's type, but in other and equally important, if not more important, points they differ from that type. In one instance there was a distinct atrophic paralysis, with all the symptoms that pointed to a wide-spread affection of the gray matter of the cord, that would have reminded one of a poliomyelitis, with the exception that, as predicted on the strength of the syphilitic character of the disease, the patient made a very rapid recovery.

In Case 1, in which the diagnosis of syphilitic disease is to my mind beyond dispute, the contractures were extreme: the bladder has remained involved up to the present time, and bed-sores formed as in the most typical case of acute transverse myelitis.

These points of difference did not conceal the true nature of these cases, and I beg of you now to consider a few facts which have impressed themselves upon my mind as more

characteristic of spinal cord syphilis than any other I have been able to fasten upon.

(1) The most striking of all is the unusual distribution of the disease over the greater portion of the cord, involving as it did in several cases the cervical, dorsal and the lumbar enlargements.

(2) The relatively slight *intensity* of the morbid process as compared with the *extensive area* involved, as evidenced by the preservation of some of the functions of the cord with complete loss of others.

(3) A rapid dwindling of some of the symptoms, and the very chronic persistence of others. In several of the cases, for instance, the anæsthesia lasted but a very short time, while the paralysis was recovered from with extreme slowness.

(4) The very frequent history of other symptoms pointing to specific disease in the same or other parts of the central nervous system.

I shall be inclined to suspect specific disease of the cord if the patient presents symptoms of paralysis, whether they be of the spastic or flaccid character, and whether the contractures be slight or not, provided he furnish evidence of a morbid process affecting a very large part of the cord, and yet showing a relatively slight intensity at any given level of the cord; if, for instance, he exhibit the symptoms of extreme paralysis, spastic or atrophic, with partial or slight anæsthesia of the parts paralyzed, with little or no involvement of the bladder, and particularly if we can find traces of specific disease in other parts of the central nervous system.

These cases remind one more or less of those which were formerly diagnosticated as irregular cases of subacute or chronic myelitis; and if we refer for a moment to the nature of the morbid process we can readily understand why this irregularity in the development of the symptoms should be the most characteristic of all of the diagnostic features of these cases. There is not in them as in the cases of acute myelitis a morbid process which is rapidly destructive and which rapidly advances across the entire cross section of the

cord, involving all the symptoms due to loss of function of the various spinal systems. If the syphilitic disease be the result of a specific endarteritis of the vessels of the cords, we know that some and by no means all of these vessels are affected, and that the disease advances slowly from one group of vessels to another. If there be diffuse specific infiltration it also invades but very slowly one part after the other. It has a remarkable tendency, too, to increase for a time and then to recede, whether as a result of treatment or not, and then possibly to increase with renewed force. If the infiltration start from the meninges it invades most frequently the lateral columns first, often at symmetrical points, and advances very slowly from white to gray matter. The intensity of the process is spent often upon the lateral columns, hence the frequency of the spastic symptoms. It may invade the gray matter, giving rise to sensory symptoms, sometimes to atrophic symptoms. It is very apt to recede from the gray matter, restoring sensation, the conditions of nutrition and the normal condition of the bladder; but it seems to halt at the lateral columns, leaving the spastic symptoms intact for a very long time.¹

If we try to harmonize the clinical symptoms with the pathological conditions it would seem much more natural to lay greater stress upon those symptoms which are due to the peculiar retrograde features of the morbid process, than upon those symptoms which are dependent upon the area attacked by the disease.

For all these reasons it seems better to fasten upon *the relatively slight intensity of the disease*, and consequently of all symptoms, *as compared with the extensive area involved*, and upon the *tendency to remissions and relapses* than upon any mere association of symptoms due to involvement of the different parts of the cord at any one given area. I do not mean to dispute the existence and the propriety of establishing Erb's type of spinal disease, but I do believe that if we do not regard other and broader points of diagnosis we shall often fail to recognise spinal cord syphilis.

¹ In a paper entitled "Tabes and Syphilis" (not yet published), read before the American Neurological Association, July, 1893, I have laid particular stress upon the anatomical features of this process.