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Buzzard, Thomas. University College, London. Library Services

# **Publication/Creation**

[London]: [William Clowes and Sons], [1878?]

#### **Persistent URL**

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ON A PROLONGED FIRST STAGE OF TABES DORSALIS: AMAUROSIS, LIGHTNING PAINS, RECURRENT HERPES; NO ATAXIA; ABSENCE OF PATELLAR TENDON REFLEX.

BY THOMAS BUZZARD, M.D.,

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In a paper published last January ('Berlin. Klin. Wochensch.') Professor C. Westphal has again drawn attention to a symptom of tabes dorsalis, which he has now had the opportunity of observing in a large number of cases, and which may be described as follows:-In health, if a man sits with one leg crossed upon the other, and the ligamentum patellæ be then smartly struck just below the knee cap, there occurs a contraction of the extensor muscles in front of the thigh, and there is consequently a sudden kick upwards of the leg. Dr. Westphal pointed out in 1875 that this, which he calls by the somewhat awkward title of the "knee-" or "leg-phenomenon," was absent in typical cases of tabes dorsalis. In his late communication he gives reasons for thinking that the test may prove very useful in the early stage of tabes, before there is any ataxia. He says that he has seen cases of pains without ataxia or sensory troubles, but with coincidently commencing optic atrophy, and absence of "knee-phenomenon." One very striking example is related. A woman, thirty-six years of age, suffered from pains, "like knives," especially in the left lower extremity, frequent and severe. She had also hyperalgesia of the skin, so that the least contact was painful. Pains came much more rarely in the right leg, and were not to be compared in severity with those in the left. Repeated examination showed that the knee-phenomenon was absolutely wanting in the left leg, whilst it was present, but not strongly marked, in the right. The patient's gait was not ataxic. No sensory troubles of the left leg appeared objectively, nor was there loss of muscular sense, as tested by closing the eyes. In spite of all this the case was certainly one of tabes, for there was, besides the pains, white atrophy of the optic nerves, inequality of pupils, along with very frequent and scanty urination.

In all the cases of confirmed tabes in which I have used this test since I became acquainted with Dr. Westphal's suggestion, I have found, as he has done, that the knee-phenomenon was absent. The case which I proceed to relate is one which goes to confirm the diagnostic value of the symptom described. It is one of the nature of which there can, I think, be no question; and it is published, not only on account of the support which it lends to Dr. Westphal's valuable suggestion, but also from the extreme interest which attaches to some of its other symptoms. Amongst these are the great prolongation (already, if we include the two years of herpes unaccompanied by pains, seventeen years) of the first stage of tabes—the remarkable recurrence of herpes every few weeks during fourteen years, as well as certain points connected with the slight amount of vision which still remains in the amaurotic eyes.

It was in April 1864, just fourteen years ago, that I first examined A. B., who was then 36 years of age. His complaint was of bad nights, and troubles in his organs of digestion. Incidentally he mentioned to me the state of his sight, which had become greatly impaired. With the right eye he could only discern light from darkness; with his left he could recognise faces, but all outlines were blurred and confused. He had consulted several ophthalmologists on account of his eyes, and they had described his disease, he told me, as atrophy of the optic nerves. For this he had undergone a variety of treatment, including hypodermic injection of strychnia and the application of the constant current, without any effect.

Family history.—His father, who was gouty, died at 61 years of age. His mother, who was then still living, had always

enjoyed excellent health. (She has since died, at 84 years of age.) No history of tubercle or neurosis in the family.

Previous history.—In infancy, so he had been told, he suffered from "inflammation of the bowels" and bronchitis, besides an attack of whooping-cough. All his life he had been subject to take cold easily, and to suffer from bronchial attacks; and there had always been from his childhood a tendency to looseness of the bowels. When twelve years old he had measles badly, from which he recovered after a long illness. After this, for twenty years he enjoyed excellent health. His habits were temperate in all respects; and the mode of his life, a great part of which was spent in the open air, conduced to healthfulness. He had never had syphilis, but had once or twice suffered from gonorrheea.

In the year 1860 he had an attack of "muscular rheumatism," which lasted about a fortnight, but only kept him in bed a day or two. There was no fever; his joints did not swell; and he was able to do his work except on one or two days. As the pains in his limbs ceased, the attack was immediately followed by jaundice, from which he suffered for three weeks, and during which he lost two stone in weight. He is quite clear that since this attack he has never been "the same man." He never got up his health or strength again. A very notable sequela had been insomnia. He would retire to bed between eleven and twelve o'clock, lie anxiously restless till three or four, and then drop off to sleep for four or five hours. Sometimes, on the other hand, he would sleep on first going to bed, but in that case would wake at three or four o'clock, and sleep no more. Altogether, he had rarely more than four or five hours' sleep at night.

In 1861 he had for the first time an attack of herpes. It was after undergoing great fatigue that he noticed what he took to be a boil on his right buttock, but which really consisted of a crop of vesicles, which burst and formed scabs. During the next two years he had, on four or five occasions, a recurrence of the eruptions in the same situation. There was rarely more than a single group. These attacks were not accompanied by any pain, beyond a little burning at the site of the vesicles. The eruption did not always follow the same

course. There would be a group which appeared to be going on to maturity, when it suddenly became, as he describes it, stagnant and abortive. The vesicles would not burst, but would wither up and disappear in a few days, without any scab being formed. At another time, after apparently aborting, or at least taking no action for twelve or twenty-four hours, they would then, as it were, re-develop and go through the ordinary course.

It was not till 1863, two years after the attacks of herpes had commenced, that they began to be associated with pains. These were at first confined to the right lower extremity; and attacks had been since occurring, when I saw him, at intervals of a month or so. Their character will be particularly described later on.

The account which he gave of the onset of his ocular disturbance is curious. It seemed that in October 1862, after a severe attack of diarrhoea, he suddenly found an imperfection of sight in his right eye. In reading he observed that he failed to see the end of the word at which he was looking; next he found that whilst reading the top line of a page he could not see the second or third line without lowering the eye, so that ere long he had to put his finger on a line, dropping it as he read, and so spell out the words and lines. This condition, as regards his right eye, stretched over a year. In the autumn following he took a holiday, and amused himself by sketching from nature. He found it necessary to paint by the aid of his left eye, shutting out the right by means of a piece of cardboard suspended from his hat. His health, though then improved, was not up to the old standard, sleep being uncertain and digestion imperfect.

In October 1863, exactly one year (almost to a day) after the failure of the right eye, similar symptoms attacked the left. He had been out sketching, the sight of the left eye being perfect, and went home and dined. In the night he was attacked with diarrhœa, which recurred in the morning. Next day he found that he could not see the ends of words with his left eye, and, as with the right eye, so the sight of the left gradually deteriorated, and was diminishing at the time when he first came to me. Examination showed the heart, lungs, and liver to present no sign of disease. The urine, which was slightly acid, deposited much urate of ammonia; it contained neither albumen nor sugar, and showed no excess of phosphates.

A. B. remained under my observation at this period (1864) for three or four months. During this time the symptom of which, next to the insomnia, he most complained was the very frequent occurrence of nocturnal emissions. These would happen sometimes four or five nights together, recurring after an interval of a few nights, and repeated altogether from ten to fifteen times in a month. He had always since puberty been troubled a good deal in this respect, but the discomfort had rather increased than diminished. The discharges were involuntary, and were not followed by any feeling of exhaustion.

Meantime the pains which had begun in 1863 continued to recur at short intervals. In 1867 my attention was particularly directed (as I find by my notes) to the pains, and also to the herpetic symptoms from which he suffered, and which he informed me were a repetition of what had occurred on various occasions during the preceding six years. One such attack occurred in December. He woke up in the night with terrible pain in the right sciatic nerve, which he described as "knifelike spasm." It persisted, occurring at intervals of a few minutes only, all the following day. This kind of pain, he said, had been liable to attack one or other of his lower extremities—and especially the right—in the thigh, knee, leg, or ankle. It would generally last a week, and was always associated more or less with an eruption of herpes. The pain usually came simultaneously with, or preceded, the herpes by a few hours, but on the other hand it sometimes, although less frequently, followed the appearance of the eruption. The pain might be in various parts; the herpes always came in one place. It always occupied the upper part of the right buttock. Even when, e.g., the pain attacked the left lower extremity, the herpes came in the right buttock. He calculated that during the preceding four years he must have had at least fifty or sixty of such attacks of herpes. A "good healthy crop," as he described it, would take from ten to fourteen days ere it

subsided. The pains generally continued throughout, and ceased about the time that the scabs were scaling off. On a very rough average he would have such attacks once in four or six weeks. It appeared that between twenty-four and thirty years of age he used to be affected very frequently indeed with herpes præputialis, which occupied sometimes the prepuce, at others the base of the penis and scrotum.

For fifteen years this patient has suffered every few weeks (with a certain exception) from attacks of pains which he describes as "forked lightning." The seizures were closer together and more violent in character from the end of 1872 to the middle of 1874. During this period of about eighteen months he would have a bad recurrence every two or three weeks.

I have notes of one such attack which I may transcribe.

"1873, Feb. 14.—Pains which were formerly rare come now very frequently, and with tremendous force. The stomach seems, to him, to be the directly exciting cause. The pains always affect now the left leg; formerly the right was alone subject. He shows a point two or three inches above the left knee, which has been an especially frequent seat of pain. Over a space not bigger than a crown-piece it is as though some one was giving him repeated violent digs with a knife. The present bout came on yesterday at 5 P.M., and has never ceased for more than two minutes at a time. He knows from experience (and the event proved that he was right) that this bout will go on for four or five days, gradually getting less and less acute, and with longer intervals, until he ceases to suffer. And he will be pretty sure to have herpes. Sometimes the herpes appears first. He will have a patch of heat and itching on the buttock, and then he gets a "fine crop" of herpes. Sometimes the vesicles are abortive, and do not come to the breaking stage. Then the pain does not come so rapidly. The herpes is always confined to the right gluteal region, and never passes to the left of the spinal column. I can see now some scars of recent attacks . . . .

"The part where the pain first begins, always remains as it were the camp or head-quarters of it—pains radiating from it

as a sort of focus. So to-day he has had occasionally pains in the left shin and calf, but the left heel, where the pain commenced, has continued to be its principal seat."

From the middle of 1874 till August 1876, he enjoyed almost complete immunity from these attacks. There would be, it is true, at rare intervals a little herpes, accompanied by a faint reminder of his pains, which he graphically describes as "sheet lightning," in comparison with their ordinary forked-lightning character.

Present State.—As regards the relation of herpes to pain, he says that perhaps once in six times the eruption would occur without any pain whatever, or, at the most, with a very little "sheet lightning." Very rarely indeed has he had pains without any herpes, although the eruption has been sometimes so insignificant that it would have escaped notice but for his long experience of it. As a rule, there would only be one crop of herpes, but occasionally a second crop would appear near to the first. In the years of only moderate severity the intervals of attacks of pain would be from six weeks to two months or so. Since they recurred severely in August 1876, they have continued to trouble him at about this rate.

I have lately (June 10) taken the opportunity of examining carefully an eruption of the kind. About two inches above, and to the left of the sulcus between the nates, very near therefore to the sacro-iliac synchondrosis of the left side, I found a single group of seven or eight vesicles of about the size of a large pin's head-some larger, some smaller. They were collected (almost all touching each other) upon a base the size of a threepenny piece, the skin of which was slightly pink in colour, and when pinched up felt hard, as compared with the adjacent integument. The vesicles contained transparent and colourless fluid. This group had appeared two days previously. At the same time I observed one or two brown scabs (the remains of vesicles) on the upper and inner part of the left thigh, where the scrotum rests upon it. On the following day the base was less hard, and paler in colour, the vesicles flattened and shrivelling.

It is only since 1873 that herpes has appeared to the left of the spine. Before that time it always came upon the right buttock. Since then, however, it has been, if anything, a little more frequent on the left buttock than on the right, and most frequent of all, perhaps, about the middle of the sacrum and the sulcus of the nates. Herpes has never shown itself on both sides at once.

It is to be noted that his pains do not appear, at least commonly, to originate in the buttocks, which, however, are the constant seat of the herpetic eruption. Occasionally, it is true, the pain will seize him in either groin, and then shoot round over the hip to the buttock; but these are much less frequent sites of pain than others. He cannot say which is the most frequent site.

The patient has a firm conviction that all his attacks of pain and herpes are associated more or less with intestinal disturbance. If he feels that he has eaten too much at dinner, he is apprehensive of an attack, and is pretty sure to have a "quiver," or flash or two of "sheet lightning." Then if there be, as is sometimes the case, a violent discharge of flatus downwards, he knows he is safe. But if this does not happen, the expected attack comes on. Even if a bad attack of pain has started, should there be an escape of flatulence he will often get relieved of pain in ten minutes. During bad attacks there are constant recurrences of a pressing desire to pass flatus by the bowel, but the passage does not take place. He does not associate the attacks with the use of any particular kind of food, nor does weather seem to influence them.

Fatigue appears to induce attacks, and, as he thinks, in the following manner. Overtired, he sits down without appetite, which comes, however, as he eats, and then, perhaps, he exceeds a little in the quantity of food taken. Under these circumstances an attack is pretty sure to follow. If, however, he is hungry, and after eating a moderate meal is quiet for two or three hours, he either escapes altogether or there may possibly be a flash of "sheet lightning," which does not go on to the "forked" variety. The last attack began in the night, after taking a small glass of stout at bedtime. At times, when his pains are very bad, there is a

sense of weight in the lower bowel, and a feeling as though something wanted to come away and could not.

Vision: Right eye.—At the present time he can see two window-lights as two, but there is no evidence of sashes. Very thick black hands on a white-faced watch are quite indistinguishable.

Left eye.—He can see the angle formed by the black hands of his watch, and thus get an approximation to the hour. He cannot in the slightest degree recognise any one by sight, nor can he walk about without a companion to lead him.

There is a point of interest as regards colour. The colour blue, he tells me, is not seen as such by him, but as bright light. A piece of bright blue ribbon looks like a bit of burning magnesium wire. If he is walking under a bright sun upon yellow gravel, the shadow which he casts (and which under such conditions is of course very blue in tone) is actually felt by him as something lighter than the sunlit gravel upon which he treads. And so also the shadows of trees, at a little distance, came out quite light from, as he supposes, the influence of the blue in their composition. In photography, he reminds me, the colour blue comes out white.

The sight of his eyes has continually, but very slowly, deteriorated since they were first affected. The pupils are of moderate size, and equal. There is no manifest affection of the external muscles of the eyes.

Direct Ophthalmoscopic examination. Right eye.—The optic disc presents a marked change in colour and character. Instead of the normal warm white or pinkish tint with fine striation, it is of a cold, slightly bluish-grey tone, which suggests the idea of its being painted in opaque oil colour. Its surface is flat, dense, and uniform, no signs of any central depression or lamina cribrosa being perceptible, and the sclerotic ring appears to be merged into the disc, the outline of which comes out sharpagainst the rich red of the choroid. There is no marked diminution in the size of the retinal vessels, but they appear as though applied to the disc, and subdividing near its upper and lower border leave the large bulk of the disc free from any trace of a bloodvessel. Both the branches of the artery and the veins are

bordered by a distinct white line on each side for a distance of nearly two disc-lengths from the optic papilla. This white line would measure about one-third of the diameter of the vessel.

Left eye.—The appearances are practically the same, except that there are a few more minute vessels on the disc than are to be found in the right eye.

Organs of locomotion.—A. B. walks without the slightest tendency to stagger, and this even if (as he will sometimes attempt upon a quiet and straight country road) he dispenses with the arm of a companion. His gait is somewhat peculiar, as he rises high upon his toes at each step; but I have no reason to suppose that this "equine action," as it might be called, perhaps, is not natural to him. His walking power is a good deal diminished; he cannot now manage three or four miles at a stretch without distress.

Tendon reflex.—If he sits with his legs crossed, and the ligamentum patellæ be smartly struck, there is no contraction of the quadriceps extensor muscle, consequently the leg is not jerked up. If the foot be suddenly and forcibly brought into a state of dorsal flexion, and the tendo Achillis struck, there are no rhythmic movements of the foot. This applies to both lower extremities.

Cutaneous sensibility is absolutely normal in all respects throughout his body. Nor is there any affection of the muscular sense. He knows exactly where his legs are, and can direct his feet readily. So also he can make his two forefingers touch at their points with as great precision as any one in health.

Pains in the trunk.—Within the last twelvementh he had on three occasions experienced what he calls "sheet lightning pains," inclined now and then to develop into a passing "forked lightning," in the right side of his chest. A week ago he suffered for twelve hours, at intervals of only two or three minutes, from attacks of forked lightning pains, which invaded the right side of the chest from about the second to the sixth rib, and appeared to go through to the scapula, corresponding apparently with the intercostal nerves. They were not accompanied by any herpes.

General condition.—A. B. is of healthy aspect and fair complexion, his brown hair being but slightly tinged with grey. His disposition is of the happy kind which is, curiously enough, found so often in patients suffering from tabes. There is marked arcus senilis in each eye. His arteries show no signs of thickening. Bladder normal. Bowels regular, and less disposed to diarrhea than formerly. Tongue clean. He does not suffer from any constrictive feeling in the waist. His digestion has improved, and he has increased in weight during the last four years.

Examination shows the nates fleshy and firm, the skin covering them, and the sacral region generally evincing no sign of anæsthesia. There are a few slight scars of recent attacks of herpes. One may see, perhaps, traces of three or four, but all except the most recent are very faint, and in strong contrast with the marked cicatrix which follows an eruption of ordinary shingles. Of the attacks which took place at periods antecedent to a few months ago, not a vestige remains.

The character of the optic atrophy, and of the pains, leaves the diagnosis of the nature of this case in no doubt. It is evidently one of sclerosis of the posterior columns, although that particular symptom which has caused the name of progressive locomotor ataxia to be given to this class of disorder is as yet absent. On the other hand, it is interesting to note that the test which we owe to Westphal—the absence of reflex contraction of the quadriceps extensor muscle on striking the ligamentum patellæ—here also lends support, if such were needed, to the diagnosis of tabes dorsalis.

The absence of inco-ordination of movement, whilst other symptoms of the disease—notably the flying pains—are well marked, is, I need scarcely say, not at all uncommon in the first stage of tabes; but it is exceedingly rare, so far as I am aware, for the immunity to extend over so long a period as has happened in this instance. Fifteen years have elapsed since the commencement of the characteristic pains, and as yet no sign of muscular ataxia has presented itself. So long a duration of pains, with exemption from locomotor troubles, is

not, however, altogether unexampled. Charcot and Bouchard have published ('Comptes Rendus des Séances de la Société de Biologie,' Année 1866) particulars of a female patient who died of some accidental disorder after having suffered during fifteen years from lightning pains. Up to the time of her death she walked without embarrassment or projection of the legs; she did not strike the ground with the heels, and closure of the eyes did not affect her security in standing or walking. At the autopsy, the only appreciable lesions (except some doubtful traces of meningitis) were found in the posterior columns, and consisted in a multiplication of the nuclei of the neuroglia, with thickening of the meshes of the reticulum, but without concomitant alteration of the nerve-tubes.

For seventeen years A. B. has suffered from attacks of herpes, and according to his own computation the eruption has come out upon him not less than 180 times. Whilst the pains have been principally referred to the periphery of the sciatic nerves, the accompanying herpes has always, it will be noted, occurred on the skin, which is innervated either by the small sciatic or by the posterior branches of those sacral nerves the anterior divisions of which go to form the sciatic plexus. Some of the pains, it is true, have been referred to districts innervated by the anterior crural nerve; but I think I am right in saying that where these have occurred they have not been alone. On the same occasion the district of the sciatic nerves has also participated in the paroxysms. What appears, therefore, at first sight as a great anomaly, viz. the confinement of the herpes to a very small district, the pains meantime ranging over various parts of the lower extremities, is found to be no longer so much so when we remember that the nerves which supply the skin at the part where the herpes was wont to appear, have a common origin with those which are distributed throughout the length of the lower extremities.

The case of Dr. Charcot's which I have quoted goes far, by exclusion, to show that we may look for an explanation of the lightning pains of tabes to the irritative alteration of the posterior columns of the spinal cord. The researches of Bärensprung especially have shown that inflammation of the

spinal ganglia will produce herpes in the district of the corresponding nerve, and it seems not unfair to conclude that an irritative process attacking the nerve-tubes within the cord would be attended with a like result. The concomitance therefore of herpes, in my patient, with paroxysms of pain, would appear capable of explanation by supposing that such a condition of the cord obtains in his case as was found in Dr. Charcot's patient. But it does not appear so easy to explain the occurrence of herpes alone for two years before the pains began. So far as it goes, this circumstance would tend to support the theory of a special system of trophic nerves, which it may be imagined were alone influenced until the growth in the neuroglia attained a certain development. Nor does it seem unreasonable to suppose that the herpes præputialis from which this patient began to suffer when he was twenty-four years old may have been really the first outcome of that diseased condition which has since taken so marked a form. Here, however, we are met by the difficulty of accounting for the fact that trophic disturbances of the skin (like herpes and some other eruptions) are, after all, of very exceptional occurrence in tabes. It happens that I have a case of the kind at present under my care in the hospital, but I do not know that I have had one before. The patient is a man affected with typical locomotor ataxia, who has suffered from highly characteristic pains for the last seven years, the longest interval being a few days. On both legs below the knee, and on the anterior aspect, there is a crop of bullæ. At first the skin comes up, so the man describes it, in a small blister, just as though he had been scalded. This breaks, and discloses (as I could see) red flesh, which was very tender, and which he said might go into an ulcer if irritated, but if not irritated would scab and dry up. One of these places had a scab on it. and this had begun, I was informed, a week previously. whole duration of a bleb would be about fourteen days. subcutaneous cellular tissue would appear to be inflamed, as the leg is somewhat swollen, and the skin looks as glossy as a chilblain. The patient began to have these spots five or six years ago. They only lasted a few weeks, and then disappeared till three years ago, when he "knocked both his legs

(owing to his greatly disordered gait), and then they broke out again." Then nothing more was seen until five months ago, when they appeared again after blows upon the legs, and have been coming out pretty constantly ever since. The blebs smart, burn, and sometimes itch. During this time he has pains like electric shocks in both legs, centring in the shins "as if in the marrow of the bones." He thinks that he has had as sharp and continuous pains without as with a coincident eruption.

There can be no doubt about the association of gastric disturbance with lightning pains in the case of the patient A. B. I have watched the commencement and progress of an attack on several occasions, and can confirm his account. But whether, as he supposes, the intestinal irritation set up by undigested food provokes the pains, or whether the indigestion and flatulence are not themselves caused by the same condition, whatever it be, which causes a periodical recurrence of pains, I am not enabled to decide, but should incline to the latter view. As is well known, a tendency to frequent action of the bladder as well as of the bowels is common enough in tabes, to say nothing of the occurrence of seminal emissions, which is so often a result of the malady. In the connection between the abdominal plexuses of the sympathetic and the lumbar and sacral nerves, one is disposed to find an explanation of the coincidence of the affection of abdomino-pelvic organs with that of the lower extremities of the body.



