Clinical Lecture

ON A CASE OF

PROGRESSIVE LOCOMOTOR ATAXY, WITH
ANOMALOUS JOINT AFFECTION.

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LET me draw your attention to the man who now enters the room walking with the help of a stick. There is a marked peculiarity in his gait, and when you look at his legs for an explanation of it you observe at once that the greater part of the right lower extremity is enormously enlarged. Examining him, however, more closely, you see that this enlargement fails to account for his peculiar walk, for the left leg, which is not notably altered in size, presents an abnormality in movement which corresponds to that in the right. If we analyse his march we find the following peculiarities equally well marked in the two legs:—In the first stage of progression the foot, which normally should be carried forwards nearly parallel to and at a distance of an inch or so from the ground, is raised some inches higher than this, and in a strongly dorsal-flexed position. When it has reached the measure of the step, the heel is put down noisily, and with a sort of jerk, the toes being then gradually brought down from their upward flexed posture, and laying hold of the ground preparatory to a repetition of the process. All this time the patient's feet are turned out like those of the dancing masters of our youth, and his eyes are fixed upon the ground in front of him. When he wishes to turn he stops short for a second or two, steadies himself, and brings himself round with great caution. He is not able to move in a small circle. He walks best on an even surface, and goes down stairs easier than up, taking care to plant the entire sole of the foot upon the step. If his feet be placed close together when his eyes are shut, he sways to and fro, and would fall but for help, and he cannot take more than two steps without the aid of his sight. His toilette is embarrassed by this difficulty, for he tells us that when he is soaping his face, and consequently shutting his eyes, he is obliged to lean against the wall, or he would fall. With all this, if we try to bend his legs at the knee against his will, or to resist their voluntary extension, we find little or no failure of muscular power.

His condition, then, it is scarcely necessary to say, is one of ataxy, and not of paraplegia. Moreover, the difficulty of co-ordination is not confined to the lower extremities. The patient is a carpenter, and he finds it impossible to use a hammer, for in striking at a nail he constantly misses his aim, and goes to one side or the other of the object; and he cannot saw a piece of wood in a straight line.

So much for the disorders of motility. As regards those of sensibility, they are of two kinds—diminution of various kinds of sensibility, and pain. He complains of a feeling of numbness in both feet, extending some inches above the ankles, and also in his hands, principally in the left. The touch of a finger is not felt at all on either sole; in the same situation, however, the contact of ice is immediately recognised, and its coldness appreciated; heat, on the other hand, is more slowly, but still correctly recognised. Electro-cutaneous sensibility is much diminished in the hands, still more so in the legs, and is quite absent in the soles of the feet. The muscular sense is manifestly impaired, for not only can he not tell when in bed in which direction his legs are lying, but he cannot feel the contraction of the muscles of his thighs when they respond to an induced current. This response is normal in the right thigh, somewhat deficient in the left, and very imperfect (probably for a reason which I will mention presently) in both legs below the knees.

The other disorder of sensibility consists in the liability to "flying pains," and these he has had since 1869, the longest interval of exemption having lasted about three months. His last attack of pains visited him on Sunday,

Monday, and part of Tuesday. A pain would last perhaps five seconds, and resemble some sharp instrument suddenly pushed into the lower part of his shin-bone. It would recur every five minutes or so during the daytime, and almost entirely deprive him of sleep during the nights. And this has been the general character of his pains; which, however, were worse formerly than they have been of late. Since the commencement of his illness he has always had a feeling as of a tight band around his waist, and of distension in his stomach.

Although the symptoms described are those common to progressive locomotor ataxy, they do not of themselves suffice to mark the case positively as one of that class, as pictured by Duchenne. To complete the catalogue, there should be some evidence of functional disorder of one or other of the cranial nerves. As is well known, diplopia from paralysis of one or other of the nerves supplied to the external muscles of the eyeball, and amblyopia from progressive atrophy of the optic disc, are the most common forms which these disorders assume. Now, this man has a well-marked squint; and if we had not inquired particularly about this feature of his case we might easily have jumped to the conclusion that the symptom, as it appears in him, is just that which is wanting to complete the *ensemble* of the requisite conditions. But it seems on inquiry that his strabismus dates from early childhood, and there is little doubt that it is one of those ordinary squints arising from hypermetropia which are so often ascribed (as he, indeed, ascribes it) to a kind of retribution for imitating a school-fellow with the like affliction. We must, therefore, exclude the strabismus from our calculation. Examination with the ophthalmoscope discloses no change in the optic discs; and the man himself complains of no material weakening of his sight. We do find, however, one cranial nerve which shows symptoms of disorder. For the last few months our patient has been growing deaf in the left ear, and now he cannot on that side hear the ticking of a watch however closely it is applied. Duchenne, Remak, and Topinard have each recorded instances in which the auditory nerve was affected; the latter mentioning seven cases in which he has noted this condition. The feature is a slight one, and not common; but in this case so characteristic are the disturbances of motility and sensibility that it suffices to complete the picture. I ought to add, too, that his pupils are minutely contracted. For the rest, we note that at various times he has had great delay in emptying his bladder, and occasional incontinence of urine; and that for a long time past he has been impotent. In the earlier part of his illness he suffered from gastric disturbance of a peculiar kind. He would require to relieve his bowels five or six times in the twenty-four hours, the motions being small in quantity and solid, and he would besides go more than once daily to stool without result. This irritability of the bowels, although not usually included amongst the phenomena of progressive locomotor ataxy, is a symptom which I have occasionally observed in other cases, but never, I think, occurring so early in the history as it did in this man.

It seems that our patient first began to stagger in his gait about June, 1868. The "flying pains" commenced early in 1869, and in March of that year he began his attendance at this hospital. Under treatment with arsenic he rapidly improved so much that he was able to return to his occupation, which he had been obliged to quit, and he continued to work more or less for his living until December last. Since that time he has not been able to follow his employment, but has attended here regularly.

It was in June, 1873, that he called our attention to his right leg, which he said was enlarged. On examining it, we found the leg swollen and cedematous below the knee. The circumstance after this escaped our attention until the following December, when he again complained very much of this limb. On stripping him, the knee-joint and the thigh for some distance above it were now found to be enormously enlarged, evidently with fluid, and this condition has persisted ever since, with but very slight variations of size.

At the present time (July, 1874) the following is the condition of the limb:—The right thigh begins to swell a short distance below the groin, so that at a point eleven inches above the upper border of the patella it measures 17 inches in circumference, as against 16 inches in the left thigh. Descending, the enlargement is more and more marked until

the knee-joint is reached, and here the measuring tape applied over the patella gives a circumference of 19 inches on the right side and only 13½ inches on the left. About two inches below the lower margin of the patella the enlargement almost suddenly ceases. The swelling is hard and elastic, the skin of almost polished smoothness, and traversed by large veins. Extension of the leg is perfectly performed, but flexion is somewhat limited. Since this photograph was taken in March last, although the size of the limb has continued the same, you will notice that a curious alteration in its appearance has taken place. The leg now forms an angle of about 45 deg. with the thigh, the apex being inwards—a deformity which is due to subluxation of the joint from the strained and weakened internal lateral ligament gradually refusing its office. That the swelling is not œdematous is evident for two reasons:—1st. There is no pitting on pressure. 2nd. When the rheophores carrying an induced current of electricity are placed upon the quadriceps extensor muscle just above the patella, where the enlargement is very great, there is immediate and powerful muscular contraction. The electric excitability at this spot is, indeed, much more marked than in the corresponding part of the left thigh. The fluid therefore lies under and not superficial to the muscle, as would be the case in œdema. Lower down the leg is certainly somewhat œdematous; and there, as you see, the conduction of the current is interrupted by the presence of fluid in the subcutaneous connective tissue, and the muscles consequently fail to act to the electric stimulus. The increased excitability of the quadriceps extensor in the affected limb is doubtless owing to the thinning and tension of the strained skin favouring conduction to the muscular tissue immediately beneath it. When the leg is rapidly extended, the hand laid upon the knee-joint is conscious of a peculiar scrunching thrill. Now it is important to remark that all this accumulation of fluid has taken place without any of the symptoms of ordinary joint inflammation. During the process of enlargement the patient had no pain or heat in the joint; he was conscious only of a gradual increase in its size. At the present time, if he kneels or walks much, he gets a little aching pain extending down the leg; but so long as he does not bend the knee much he has no pain whatever, and the only inconvenience which he suffers is from the weight of the swollen limb causing him to be readily fatigued. There is no swelling of the ankles or feet. As I pointed out before, the action of the right leg in walking is precisely similar to that of the left, but the movement of the joint is limited in the direction of flexion.

Such is the very anomalous joint affection which this patient presents. It is, you will remark, a condition which asserts itself so very prominently that, were this man to present himself for the first time for medical advice, it is more than probable that the whole attention of the examiner would be fixed upon the state of the right limb, and the much more serious general disorder with which the patient is affected would be passed over unnoticed.

It so often happens in clinical medicine that we fail to see what we do not specially look for that I am constrained to believe that such a condition as this has probably existed (though not, perhaps, to so marked an extent) in others of the somewhat numerous instances of progressive locomotor ataxy which have come under my observation; but it is a fact that I have never before noticed this affection, and my colleague, Dr. Hughlings Jackson, to whom I showed this patient, tells me that it is likewise the first case of the kind which he has seen. This experience, or lack of experience, contrasts remarkably with that of Dr. Charcot, who describes no less than five examples of the kind out of fifty cases of tabes in the Salpêtrière. It is to Dr. Charcot,* I think, that we owe in the first instance the

recognition of this peculiar arthropathy, which has been well described also by Dr. Ball. In this country, so far as I am aware, the only recorded case is one described by Dr. Clifford Allbutt.*

Dr. Charcot looks upon the affection as one of the multiple forms of spinal arthropathy, by which term he would designate a group of disorders which appear to be directly dependent upon certain lesions of the spinal cord. In his experience the arthropathy in question is always an early phenomenon, occurring between the prodromous period and that of inco-ordination. If it should, however, be late in appearing, it is then always, he says, in connexion with one of the superior extremities, and he attributes this to the circumstance of the sclerosis, which is the cause of progressive locomotor ataxy, frequently invading the upper part of the cord only after the lower part has been for a long time affected. The present case, however, is an exception to this rule, for the symptom in question did not appear until marked inco-ordination of the lower extremities had existed for many years; and it is in the knee-joint—not a joint of the upper extremities—that it has shown itself. Dr. Charcot describes the condition as an extreme tumefaction of the entire limb, composed of (1) a considerable hydrarthrosis; (2) a diffused swelling, for the most part of hard consistence, and in which the ordinary symptoms of œdema are not usually apparent. He remarks that this arthropathy is unaccompanied generally by fever or pain. This description precisely applies to the case before us. You will note besides that, although the knee-joint is enormously distended with fluid, it does not present the appearance of ordinary chronic synovitis, in which the distended capsule of joint projects with marked distinctness in three places—viz., above the patella and on each side of the ligamentum patellæ. There is no such mapping out of the knee-joint in this case; and this is evidently because, in addition to the fluid in the articular cavity itself, there is effusion in its neighbourhood. This effusion, as it does not occasion the appearances of subcutaneous œdema to present themselves, is probably beneath the muscles. The affection, then, is a peculiar one, and its pathology is by no means clear. Experience of these cases shows that, with a striking similarity in their onset, their progress varies. In some, at the end of a few weeks or months, the swelling disappears, and the joint apparently returns to its former condition. This is styled by Dr. Ball the *forme benigne*. In others, on the contrary, grave disorders remain—erosions of the osseous surfaces, creaking movements, various luxations, or even total destruction of the joint (*forme maligne*). As regards frequency, it is first the knee, then the shoulder, the elbow, hip, and wrist which are apt to be affected; but the small articulations may also suffer. Several joints may be coincidentally involved.

The affection, which in certain respects is suggestive of dry chronic arthritis, differs from that disease, as Dr. Charcot points out, in several particulars: in the large quantity of fluid by which it is characterised, and the fact of the effusion extending beyond the limits of the joint; in the luxation which is common in the ataxic class; in the fact that the knee is most commonly affected, not the hip, and that the joint affection of the tabetic patient may retrograde or even recover, which is never the case in dry chronic arthritis; in the suddenness of the appearance, and rapidity of progress of the disease. On the other hand, it is true that in cases of old standing, when the articular surfaces, deprived of cartilage, have continued to rub against each other, the characters of dry arthritis are observed; eburnation and deformity of the articular surfaces, with osseous growths on the extremities of the bones.

The existence of articular affections dependent upon preceding lesion of the nervous system is now generally recognised. Such affections have been observed in connexion with lesions of the peripheral nerves, as well as in limbs which are the subject of hemiplegia from hæmorrhage or softening of the brain, in Pott's disease, in acute myelitis, in certain cases of tumours occupying the grey substance of the cord. The fact that the joint affection is exceptional in progressive locomotor ataxy suffices to show that it is not due to the sclerosis of the posterior columns which constitutes the important pathological change in that disease, but to

* Leçons sur les Maladies du Système Nerveux. Paris, 1873. 2me Série.

* St. George's Hospital Reports, vol. iv.

lesion of what part of the substance of the cord its causation must be referred there is at present no evidence to show.

I think it very likely that, as we shall in future specially examine the joints of our ataxic patients, we shall meet with cases of the same character as that which I have described.

REPORT ON THE ISSUE OF A SPIRIT RATION DURING THE MARCH TO COOMASSIE.

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(Communicated by the DIRECTOR-GENERAL OF THE ARMY
MEDICAL DEPARTMENT.)

(Concluded from page 240.)

EVIDENCE FROM THE 2ND BATT. RIFLE BRIGADE.

Letter from Surgeon-Major WILES.

Regarding Paragraph 1:—The 2nd Rifle Brigade seem never to have had many teetotalers in their ranks. Prior to leaving Ireland for the West Coast there were 34; these diminished on going on board to one sergeant and four men, who continued in their principles during the campaign. Only one of these five suffered from sickness whilst in the country; another had an attack of fever on board ship. They all did their marching well, and looked as well as, if not better than, the other men.

Regarding No. 2:—On commencing the march up country the order was that no spirit ration should be given unless on express recommendation of the regimental surgeon. Whilst on the march I did not see any indication for giving it, and only gave it about three times up to the arrival at Prahsu. At Barraco, where we were halted for some time (ten days), the men suffered from diarrhoea and febricula. I thought the issue of a ration here might perhaps do good, so gave it once or twice; but finding that the result was unsatisfactory, there being more cases the next day, I stopped its issue. (This sickness I considered to be owing to the disturbance of surface soil.) On arriving at Prahsu its issue was taken out of the hands of the regimental surgeon, by recommendation, I believe, of the principal medical officer, and it was afterwards given nearly every day, or at all events whenever it could be obtained, which was nearly every day. It was generally given, by my recommendation, in the evening; but one morning, as it had not arrived the previous evening, it was given at 5 A.M. I considered at the time, and on thinking it over am now more strongly of opinion, that its issue during the expedition, instead of being beneficial, only made the men more susceptible to the malarious poison and to affections of the bowels. Of course, this is only an individual opinion, but it is an unprejudiced one, although I am far from being a follower of teetotal principles. My opinion regarding its use in West Africa was so strong that, personally, I did not take any stimulant of any kind during the whole period I was in West Africa, and during that time I never felt better, and had no sickness of any kind, and marched the whole way up and down, besides doing some amount of hard work. My experience (derived from active service in the Crimea, India, and China) is very much against its use on active service, as I think its use renders men especially liable to dysentery and camp diarrhoea, and I think it was shown to have that tendency especially in such a climate as the West Coast of Africa. Regarding the men's feelings, I did not find that they missed its not being issued except when they came amongst other corps who were getting it. They cared more for the tea and lime-juice, with which they filled their bottles for the day's march.

Although so strongly against the issue of spirits, I am very strongly in favour of supplying the men with their pint of malt liquor daily when practicable, as I have seen that of great good, and especially so on board transports.

(Signed) J. WILES, Surgeon-Major.

Winchester, May 29th, 1874.

EVIDENCE FROM THE 23RD WELSH FUSILIERS.

Letter from Dr. ALLEN NESBIT FOX.

I regret I can furnish very poor information on the subject of your communication. I only joined the 23rd after their arrival at the Prah. They were then about 120 strong, and, as well as I remember, none were teetotalers; all took their ration of rum in the evening. This was first issued to them at a station called Yancoomassie-Fantee, about 25 miles distant from the Coast. They were in the habit of receiving it daily after sunset up to the time I joined them. From the Prah to Coomassie it was not regularly given, not being obtainable at some of the stations from the difficulty of transport. Hot tea or cocoa was the substitute. The men were always better pleased and more contented with the spirit than with any other beverage; but I cannot say I remarked any impairment of health or difference in their marching from its non-issue. Very few fell out on the march up country; those who did, from over-fatigue and debility, were immediately put in a hammock and had a small quantity of brandy-and-water. And this was given at the different halting places with marked benefit; it appeared to revive and set them up again. I consider the issue of a spirit ration in the evening, after the heat of the day was over, was rather beneficial to the men than otherwise. Should the issue of spirits be deemed advisable, I think a gill per man amply sufficient, and the evening the best time for giving it.

*Letter from Sir ANTHONY HOME, K.C.B., late Principal
Medical Officer on the Gold Coast.*

I wish it were in my power to give you any reliable data on the subject you write about. I have no deductions from carefully-made observations to offer, and I hesitate at taking up your time with my individual opinions. I do not think that in the short period of the Gold Coast expedition observations could be made for a sufficient length of time on the same men, in sufficient numbers and under the same conditions of exposure, to decide the most important matter referred to. I doubt, even, whether exactness is attainable. In saying this, do not think I am deprecating inquiry. Whatever answers the painstaking and intelligent medical officers named give you, they will, I am sure, be most trustworthy. But the question is, Can bricks be made without straw? Can a conclusive judgment be formed without exact observations made *ad hoc*?

In the ration I recommended for the Gold Coast I included 1.64th of a gallon of rum (2½ fluid ounces) daily, when the men were in the field—i. e., actually campaigning.

The experience of the Red River expedition, that men could do very hard work without drink, was quite conclusive so far as that and strictly similar expeditions are concerned; and I had myself, in Canada, ample experience to the same effect. (The "lumberers" go into the woods and live there all winter; they sleep in holes dug in the snow, lying on spruce branches covered with buffalo robes; they work very hard; and it is an inexorable rule that all drinks found in the camp are destroyed.) But then the conditions in the Red River expedition were so different, so opposite to those on the Gold Coast. In the first there was an excellent climate; good rations of succulent food, easily digested by healthy men doing hard work and sleeping soundly; no sickness present. On the Gold Coast there was tough, underfed beef; insipid, badly-cooked food, difficult to digest, and after a time loathed by men who were mostly more or less ailing.

I had noticed in the Crimea, in India, and in New Zealand, that the men on campaigns had an absolute craving for varied diet; that even regular toppers used to spend money in buying jams, sardines, pickles, cheese, &c., at the canteens. In fact, the men spent their money generally on food of this kind when it could be bought; they also used to give willingly 3s. 6d. a bottle for beer. Now I believe if a very varied diet were always issued, the men could get on without alcoholic stimulants in all climates whatever, tropical as well as temperate; but it is visionary to hope that this can always be the case in war. Sutlers do a little in this way; Government can do nothing directly. Men cannot keep in health on poor, insipid, badly-cooked rations. Under these circumstances, I believe that, *after their day's work*, rum is desirable (beer and wine are impossible of attainment). There is a moment in which we may so keep