MUSCULAR ATROPHY OF THE PERONEAL TYPE AFFECTING MANY MEMBERS OF A FAMILY.

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Two years ago Dr. Tooth described, in a thesis for the M.D. of Cambridge,¹ a new type of progressive muscular atrophy, beginning in the legs and showing a hereditary tendency. It had been independently distinguished, a short while before, by Professor Charcot and M. Marie. For the last year I have had under my care, at the West London Hospital, a remarkable case of the disease.

Charles Wheeler, aged 22, a fine-looking young man, by trade a holder-up for riveters, was sent to me at the West London Hospital by Dr. Seymour Taylor, for electrical treatment. It is now five years since he noticed a weakness in his legs, the right being considerably the earliest affected. It has caused a deformity in his feet, for which he once underwent tenotomy of the right tendo Achillis, and which leads him to wear out the toes of his boots; he suffers much from cramp in the legs; for fifteen months he has noticed weakness and wasting in the left hand, and for nine months in the right.

The feet are much arched, the instep being raised, and the length of the foot proportionately diminished. On the right side the first phalanx of the big toe is bent upwards, the second downwards; the same upward bend is less observable in the first phalanx of the other toes. The heels are slightly drawn upward, by contraction of the tendo Achillis.

There is great wasting of the muscles in the tibial and peroneal regions, and some, which is increasing, in the calf. The muscles in front of the thigh have also begun to waste.

Since the foot cannot be properly bent upwards owing to the weakness of the tibial muscles, the knees have to be raised higher than usual to keep the toes off the ground, and when the foot is lowered the ball of the toes touches the ground first. At the same time, since the thigh muscles are not early affected, the foot points straight forward as in health, and there is no irregular throwing of the feet. I have never seen a case of infantile paralysis affecting both legs where there was not some paralysis of one or both thighs,

¹ See 'BRAIN,' Vol. X. 1887, p. 243.

and this causes the foot to be turned out in a way quite different from the gait I am describing. I should think therefore that in a young person, in whom alcoholic paralysis is not likely, this way of walking might usually be ascribed to this disease. I have seen two or three cases in the streets since I have been on the look-out for it. The toes of the boots are of course worn out quickly, just as in spastic paraplegia.

The look of the hands was just that of the usual form of progressive muscular atrophy, the thenar, hypothenar, and interosseous muscles being wasted. Since he has been under me the forearm muscles, both flexor and extensor, have wasted on both sides. The extensors of the thumb were the first muscles above the wrist to waste.

Fibrillar tremors have been very marked throughout, and have been seen in muscles which did not perceptibly waste till later.

I could not find any anaesthesia to touch or temperature, and except for the cramps he has had no pain.

When he first came, faradic contractility was diminished in the wasted muscles; but there was no R D to galvanism. The muscles contracted more easily to stimulation of the nerve than when they themselves were touched. During this year R D has appeared in several muscles, both of arm and leg.

The knee jerks were normal at first, but are now diminished.

I took a tracing of the tremor, and found it to be at the rate of ten or twelve per second, that of normal muscular contraction.

He brought me a cousin—George Milton, 29, also a big strong man. He was quite healthy until five years ago, when his legs and feet began to grow weak. He has noticed weakness in his hands for two years. He presented the same appearance of the legs, though in a less degree. In the hands the thenar, hypothenar and abductor indicis muscles are slightly wasted. There was tremor of the calf, and no trace of anaesthesia. I have not had the opportunity of testing the muscles electrically.

I could not in either case find any hypertrophy of muscles, and with the exception of syphilis in the first patient both had been perfectly healthy till the onset of the present affection. They now enjoy good health in every other respect, and the viscera are quite sound.

The disease is quite familiar to them, for, as will be seen, many males of the family are similarly affected; but they do not usually seek medical treatment for it. On going into the country to see the relations, I could not find that any of those whom I saw had ever been under a doctor for the complaint.

The family history can be traced through five generations, and is as follows:—

Sear married Facer, and had seven children; six sons and one daughter:

1. William S., affected in legs and arms, died at 70, of apoplexy.
2. Joseph S., affected.
3. David S., slightly affected in legs.
4. Samuel S., affected, died at 65.

5. Robert S., affected.
6. George S., affected, died at 74.
7. Mrs. King, healthy.

1. William S. had nine children; four sons and five daughters. The four sons were all quite healthy; three of them are married. One lives at home; his children are healthy.

Two others are in America, and correspond frequently. They have never said that anything of this sort has occurred in their children.

The five daughters were all healthy and all married:

- 1a. Mrs. Wheeler had two sons; an elder died at 19 of phthisis. He was a shorthand clerk, and had before death become affected in the hands so that he could not write comfortably. He died ten years ago. The state of his legs is doubtful.

The next son is Charles, the patient.

- 1b. Mrs. John Milton has had two sons and four daughters. The eldest son is George, the patient.

The second son has written lately to say that his feet are getting bad. He is about 20 years old.

The daughters are all healthy. Three are married, viz.:

Mrs. John Milton, jun., has one son, aged 6, who is "weak on the foot; not right in the feet," and three daughters under 9, who are healthy.

Mrs. Sibley has two sons under 11, healthy, and one daughter, who has abscesses in the legs.

Mrs. Page, no children.

- 1c. Mrs. Huckle has two sons and four daughters:

William, 38, healthy, is married, has two sons and four daughters. The eldest son is about 13, and both are unaffected.

Charles, 24, began to waste in the legs at 10. The left leg is the worst; it is extremely wasted below the knee, both in the calf and front of the leg. The foot is a hollow claw, the arch being so high and narrow that the tarsus projects on the dorsum like an abnormal swelling; it is also slightly curved inwards. There is some wasting and tremor of both thenar muscles. He says his hands are getting weak, and that he cannot hold things steady. There is no pain or anæsthesia.

The daughters are all healthy. Three are married:

Mrs. Pates has five children. Willy, aged 12, the eldest boy, walks badly, and is considered affected.

Mrs. Cobb has only a baby.

Mrs. Ale has no children.

- 1d. Mrs. Sam Milton has six sons and two daughters:

Fred, 23, wasting of legs, feet arched, talipes equinus slight, and tremors in legs and hands; no pain or anæsthesia. He thinks the disease began two years ago.

Harry, 19, and Albert, 18, healthy.

Edward, 16, legs much wasted, feet much arched, tremors in legs and hands; no pain or anæsthesia. He has walked badly since he was 6.

Arthur, 6, and Herbert, 3, are healthy.

The two daughters, both under 15, are healthy.

1e. Mrs. Carter, now dead, had three sons and two daughters :

The two eldest sons, of whom the elder is only 25, are healthy.

James, aged 20, is affected.

The daughters are quite healthy. One, Mrs. Dalton, has two babies.

None therefore of William Sear's children are affected; but of fifteen daughters' sons, eight are already affected, two are under 20, and two more are under 6 years of age.

2. Joseph Sear had two sons, who were healthy, and one daughter who had no children.

3. David Sear had no children.

4. Samuel Sear had two sons, and five daughters. The sons were both healthy. The daughters were also all healthy; four were married.

4a. Fanny, died of phthisis, had one daughter, who was healthy.

4b. Jane, Mrs. Wade, had one son, now 38, and one daughter, both healthy.

4c. Betsy, had one son, healthy in limbs, but weak in the eyes.

4d. Eliza, Mrs. Kitchender, has children; the eldest son Harry, now 23, is affected.

5. Robert Sear had two sons and two daughters; they were healthy, but nothing has been seen of them lately.

6. George Sear had one son and one daughter.

6a. The son, Reuben, is very badly affected.

6b. The daughter, Mrs. Baker, herself healthy, has a family of sons and daughters. The eldest son, now in America, became affected in the legs.

7. Mrs. King (born Sear) had one son and one daughter.

7a. The son, William, now dead, was badly affected.

I can discover nothing about the Facers, except that Mrs. Sear (born Facer), the mother of this tree is said by her grandchildren to have been crippled in her legs.

There are a few points on which I will add remarks.

1. These cases do not give any corroboration to the opinion that the disease is a neuritis.

2. This, the daughter's son inheritance, is peculiar to this family, not to the disease.

3. This form makes one wonder what inheritance is. That the diseased tissues of a consumptive father should be so represented in his spermatozoon as to cause his child to fall

into consumption is remarkable enough. But that the women of this family, themselves even uncommonly buxom and healthy, should be able to select their children, and transmit to the males alone tissues unlike their own, and endowed with a regular form of weakness which they do not themselves possess, is still more marvellous. It seems as if the daughter of a diseased father carried from the beginning of her life ova of two sexes, the female healthy, the male containing within it the representation of the father's disease.

4. Although in these cases both peroneal and tibial muscles are alike atrophied, so that it cannot now be said for certain which were first affected, yet there is in several a slight lifting of the inner border of the foot, and a slight increase of the concavity to the inner side, which seem to show that the peronei were at one time the most diseased. The talipes equinus is due to the unopposed contraction of the muscles at the back of the leg, at a time when those at the front were alone affected.