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In 1851 I had the honour to communicate to the Royal Medical and Chirurgical Society the details of a case which, after careful microscopic investigation, I ventured to call *Granular Degeneration of the Voluntary Muscles*. The French have denominated it *Atrophie Musculaire progressive*. Since that time two other cases have fallen under my observation, in each of which I scrutinised, with great care, the different tissues of the body; but I would refer to one in particular, which was for a long time under the care of my friend Mr. Skey, and the *post-mortem* examination of which Mr. Savory kindly superintended. In it there existed the same breaking up of the sarcolemma of the elementary primitive muscular fibres, and a segregation of the granules of which the sarcous matter is composed, as in the case described in my former communication; therefore I still adhere to the term granular degeneration, for it is not a simple atrophy or attenuation of the muscular fibre, but an absolute destruction or disruption of that tissue, and if any other name were given it, I think that which has been

suggested by Shultz would be the best—namely, *Necrobiosis of the Voluntary Muscles.*

H. D. C. Delamotte, Esq., a surgeon at Swanage in Dorsetshire, has a family of fifteen sons and daughters. Of the first six, by one mother, there are three sons and three daughters, all strong and healthy. The second portion of the family consists of five sons and four daughters, and the eldest daughter and son, aged respectively 24 and 22, are now the subjects of the disease in question; all the others are perfectly healthy.

Mr. Charles Delamotte, who has kindly consented to be here this evening, was showing symptoms of weakness in the legs when I first saw him in June 1855. His sister had then been affected three years, and walked with considerable difficulty.

The first symptom which manifested itself in both Mr. and Miss Delamotte was a sense of weakness referred to the loins, so as to cause a waddling gait; next, a difficulty in bending the thighs on the body, as in the act of getting up stairs; then a gradual diminution of the power of raising the legs towards the thighs; and, eventually, the feet and toes participated in the paralysing influence.

The period of time during which these successive stages of the disease run their course varies in different individuals. In Mr. Delamotte's case it has been very much prolonged by his perseverance in exercise: but in all the course is progressive and unyielding.

After the legs have been rendered thus useless the upper extremities begin to exhibit the same centrifugal morbid action. I hear that Miss Delamotte, whom I have not seen since 1855, has lost all voluntary motion of her arms and hands; her brother, however, has still a firm grasp of the hands, and is able to bend the forearms on the arms, but he cannot raise his arms to his head. He now amuses himself by rowing, and thus continues the exercise of his upper extremities.

From neither parent is it possible to trace anything like hereditary predisposition, and in both the patients the in-

firmity has appeared and progressed without any other assignable cause. Both cut their teeth easily, and each one passed through the period of childhood without having had fits of any kind, nor was either considered to be nervous. The young lady began to menstruate at the early age of fourteen, and menstruated regularly; she never had leucorrhœa; and when I saw her she had a remarkably healthy appearance, a ruddy complexion, well-developed limbs, and good intellectual power. Her tongue was clean, her appetite good, her bowels regular, but her pulse was somewhat weak and quick (108).

Her brother also gives an excellent account of his general health. He has never indulged in pernicious habits, he has never exposed himself to influences likely to become the source of disease, nor has he ever been the subject of any other cause of weakness. He has always slept well at nights, his appetite is good, his digestion good also, his bowels are regular, and his sexual feelings, he supposes, are like those of other men. His pulse, like his sister's, is somewhat weak and quick. Notwithstanding the inactive condition of his legs he never suffers from cold, and the skin retains its healthy warmth and sensibility.

The peculiarity of the disease in these cases is that it is *not* apt to be accompanied with symptoms of nervous disturbance; and from the researches I have made, I am induced to believe in an idiopathic disease of the muscles (dependent probably on defective nutrition) akin to that to which the bones are liable, from the same cause.

In the Thirty-seventh Vol. of the Society's Transactions, Dr. T. K. Chambers has recorded the case of a female, æt. 26, in whom mollities ossium and granular degeneration of the muscles co-existed. This patient had gradual increasing weakness of the muscles without any pain being experienced at any time. In my treatise on paralysis I have also referred to a case in which both muscles and bones were affected, and apparently in consequence of defective nutrition, but without pain; and the singular absence of every indication of nervous disturbance has obtruded itself on my

mind during the progress of most of the cases of granular degeneration of the voluntary muscles which I have seen.

Mr. Delamotte assures me that throughout the whole course of his infirmity he has had neither ache nor pain. Neither has he at any time experienced the quivering or starting of the diseased muscles which M. Cruveilhier described in the case of Prosper Lecomte, in which the anterior roots of the spinal nerves were affected.

Most pathologists, however, are disposed to attribute to some lesion of the nervous centres these extraordinary cases, and I have often felt a desire to discover a central nervous cause for what we have all been taught to regard as indispensable for such effects. Yet when a paralysis extends in a centrifugal direction, respectless of the course and distribution of nerves; when there is an entire absence of any one symptom of nervous disturbance; when, moreover, such men as Quekett and Savory have failed to detect in such cases the slightest change from the healthy condition, either in the peripheral nerves or in the nervous centres; surely it is not discordant with a sound physiology to suppose that there may be a destruction of muscle, independent of nervous origin; and that the symptoms which I have described may distinguish it from those cases in which manifest nervous symptoms appear during life, and in which lesions of the nervous centres are found after death.

If, then, the disease in question consists in an alteration in the composition of the muscular tissue, are we encouraged by the most recent researches in biology to hope that, by any remedial agent, a formative change may be effected, that the cell constituents of the affected muscles may be incited to healthy action, and that new sarcous substance may be reproduced in the primitive muscular fasciculi?

Thus far the disease has pursued, as I have already stated, a progressive, unyielding course; but Mr. Delamotte affirms unhesitatingly that during the last year his symptoms have, under the influence of Fowler's solution, made very much less progress than they have done during the course of any preceding year.

The accompanying illustrations are taken from the muscles of the Hon. Wm. P—, who suffered from an affection identical with that of the subjects of the present paper, and whose case is described in the thirty-fifth volume of the 'Transactions' of this Society, p. 77.

DESCRIPTION OF PLATE II.

FIG. 1. Rectus abdominis. ($\frac{1}{8}$-lens; low eyepiece.)

2. Spinalis dorsi. (ditto.)

3. Pectoralis major, clavicular portion. (ditto.)

4. Longissimus dorsi. (ditto.)

5. Pectoralis major, sternal portion. (ditto.)

Fig. 1.

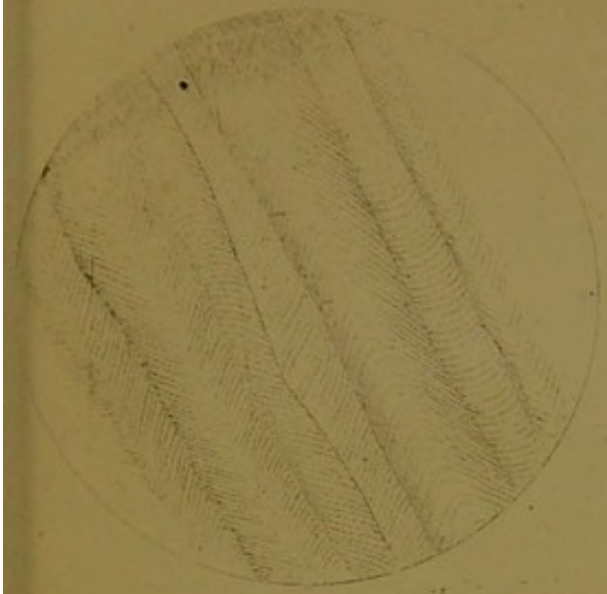

Fig. 2.

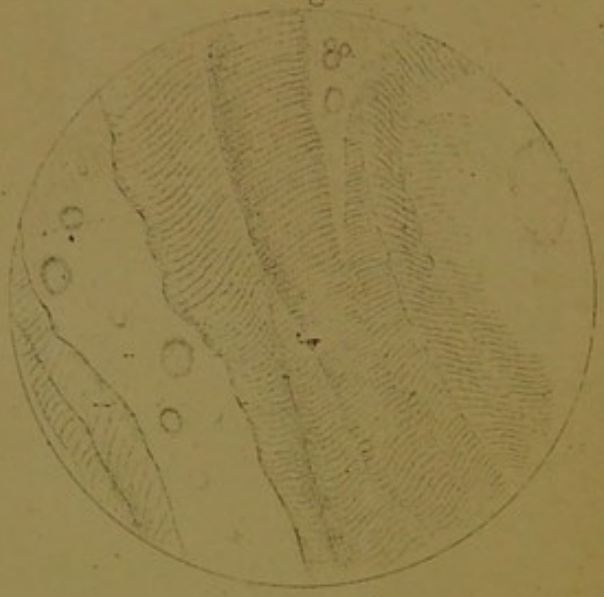

Fig. 5.

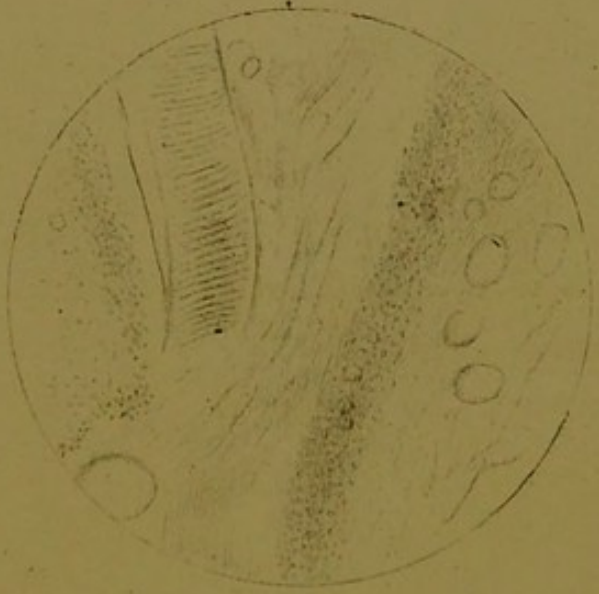

Fig. 3

Fig. 4.