CASE
OF
MOLLITIES OSSIIUM,
PRECEDED BY
DEGENERATION OF THE MUSCLES.

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MARY G— was admitted into St. Mary's Hospital, under my care, March 26th, 1852. She was twenty-six years of age, unmarried, and had never been able to follow any avocation, on account of weak health. She was about four feet ten inches high, and six stone seven pounds in weight, not emaciated, and of symmetrical figure. A waxy, yellow complexion, with bright scarlet colour in her cheeks, gave her the aspect of a delicate person. No history of hereditary disease of any kind could be elicited from herself, or from enquiries made at her native village. The ankles were œdematous, and she walked slowly, as if from languor, on a flat surface; when she attempted to ascend steps or to raise herself from a stooping posture, defective power in the muscles of the haunch and thigh became very evident. The flesh of the whole body was exceedingly soft and flabby, the calf hanging down in the baggy way that it does in emaciated persons. She stated that she had first become an invalid seven years previously, her illness commencing by weakness and pain across the loins, especially at the end of the short ribs on the left side. The same symptoms had continued, with occasional variations, up to the date of admission. She

usually felt worst in early spring, and got better during summer and autumn. The bowels were somewhat costive; the catamenia generally regular, but occasionally postponed for a fortnight beyond the customary time. The urine, examined on admission and frequently while she was in the hospital, was variable in quantity, and variable in specific gravity, in proportion to its quantity, from 1·020 to 1·028. It was sometimes neutral, rapidly turning alkaline; but generally acid, depositing a considerable sediment of lithate of ammonia, soluble in heat. In all cases, boiling immediately threw down a cloud of phosphates, soluble in mineral acids, and showed the absence of albumen.

With the exception of a stitch under the short ribs of the left side, caused by turning in bed, and a sense of great debility in the back on standing, there was no pain experienced at any time, either with or without pressure.

The patient remained in the hospital, taking steel, for five weeks, without any change in the symptoms except the disappearance of œdema and improvement of appetite; after which she left at her own desire. She returned in three weeks, and again remained under observation for ten days, when she was advised to go to the seaside, and left with the expressed intention of doing so. During her residences at St. Mary's, the bones of the back and limbs had been carefully examined several times, without any deviation from the natural state being discovered, except that the ribs on the right side, viewed from behind, were not quite symmetrical, being more prominent than the left. On her going away, I told her of the difficulty experienced in arriving at a diagnosis, and desired her to let us know when any medical man could discover the nature of her complaint.

From St. Mary's she went to St. George's, and was under the care of Mr. Cutler for about six weeks; after which she left at her own desire. The symptoms continued equally obscure at St. George's, and no diagnosis of the case was formed till one night spontaneous fracture of the left femur occurred. She was then removed to her home, at Islip, Oxfordshire, and placed under the care of Mr. Blick. She

informed him of the wish I had expressed to learn the result of the case, and through his kindness I am enabled to detail the remainder.

It appears that when he saw her on the 11th of August, 1852, a fortnight after leaving St. George's, fracture of both femora had taken place in the upper third of the bones. Obtuse angles were formed at the seat of fracture, by the thighs being drawn upwards and outwards, and twisted on themselves, so that the external border of each foot lay on the bed, and the soles approximated to one another. The only parts of the lower extremities capable of voluntary motion were the toes. The shin-bones felt soft on pressure, producing a sensation to the finger described by Mr. Blick as like that of a fibro-cartilaginous tumour. She breathed, ate, drank, slept, excreted fæces, urine, catamenia, as well as usual; taking meat, wine, and beer, but refusing medicine. She had no pain, except when the tumefied parts about the fractured ends of the bones were touched, and those were excessively sensitive. No important change seems to have occurred—except that the body kept shortening, and the leg-bones getting softer, so that the foot could be raised three or four inches from the bed without altering the position of the knee—till April, 1853, when the right arm became painful to the touch and paralytic.

In May, the same misfortune happened to the left upper extremity also.

In June, the pelvic arch gave way, the mons veneris being drawn upwards and the anus thrust forwards, the alæ of the ossa ilii falling inwards.

In July, the ribs on the right side gave way, and she began to suffer much from dyspnœa and cough, with quick pulse, fever, and restlessness.

In August, the bones of both arms were found quite soft.

In September, the ribs on the left side fell in, and she was now much distressed by increased dyspnœa and palpitation of the heart. The contractions and dilatations of that organ were distinctly visible through the fleshy parietes. The lower jaw and bones of the skull also felt soft on pressure.

Towards the end of October the distortion of the lower parts of the trunk was so great, that the fæces could not be naturally expelled, and had to be removed by mechanical means.

She at last died of dyspnœa, on the 6th of November.

Several times during the illness, Mr. Blick sent me some of the urine secreted by Mary G—. It presented always very similar physical characters to that passed in St. Mary's. Whilst acid, the microscope showed a great quantity of lithate of ammonia, vesical epithelium, and a few crystals of oxalate of lime. After it became alkaline, there were to be seen in it a number of yellow spherules, some of them furnished with thorn-like processes, which Dr. Hassall informs me he considers to be uric acid in combination with an earthy base. There was also a large quantity of vesical epithelium, numerous crystals of triple phosphate, and a few stellæ, pronounced by Dr. Hassall to be phosphate of lime. A quantitative analysis of this urine, by Dr. Beale, gave the following result :

<i>Urine of Mary G—.</i>		<i>Healthy Urine (Berzelius).</i>	
Water	971·00		933·00
Solid Matter	28·10		67·00
<i>In 100 Parts of Solid Matter.</i>			
Urea	17·7		44·7
Extractives	36·3		39·0
Lithic Acid	0·6		1·4
Earthy Phosphates	3·3		1·4
Fixed Alkaline Salts	41·9		25·9

It will be seen, therefore, that the solid matters of the urine were diminished below the natural standard by more than half, and that this diminution was due to deficiency of animal matter; that the alkaline and earthy salts were nearly doubled in amount, the principal comparative augmentation taking place in the earthy phosphates.

After death, the body was found to have shrunk from four

feet ten to three feet one inch and a half, and could be rolled up upon itself, to use Mr. Blick's forcible expression, "like an ill-stuffed bolster." No examination of the viscera was made, but portions of the tibia, sartorius, and rectus femoris muscle were removed, and a sharp instrument passed into many parts of the bony skeleton, which was found universally soft and unresisting.

The section of tibia sent to me was of the colour of muscle, soft and friable throughout, presenting to the knife scarcely more resistance than brain, and retaining its shape solely by the aid of the tough periosteum. No remains of bone could be felt except on cutting the periosteum, where a slight grittiness was perceived on making a section with sharp scissors for microscopical examination. Under the microscope, the whole of the bone, to within half a line of its external surface, was seen to consist of large fat-vesicles, containing, some white, others a reddish oil, and thus accounting for the colour of the texture, with the intervals between them filled up with spherules of various magnitude, mostly about as large as blood globules, of a dull red tinge. They were formed apparently of aggregated grains, and some had an indistinct nucleus. No fibrous structure could be detected in this situation.

The part next to the periosteum, which felt gritty when under the scissors, exhibited, when examined under a quarter-inch lens, small islands of opaque bone, which, however, did not retain a perfectly healthy appearance; the bone corpuscles being indistinct, and the caniculi not to be discerned. These islands were surrounded by some structure more transparent, and that again bounded by a reddish fibrous structure, in which were oil globules of various sizes, and a few oval fat-vesicles towards the inside. The addition of hydrochloric acid caused a slight disengagement of gas.

The portion of rectus muscle was, to the naked eye, of the natural colour, but of too homogeneous an appearance. Under the microscope, it presented no traces at all of fibrous structure, or even linear arrangement. It had become a mere con-

geries of fat-vesicles, the interspaces between which were filled up with globular granular corpuscles of various sizes, estimated by Dr. Seiveking as from $\frac{1\text{ to }2}{5000}$ ths of an inch in diameter. Many of the larger had a granular nucleus.

The circumstances which have induced me to lay this case before the Society are the following :

1st. The portrait which is afforded of an early stage of the disease—a stage at which it is rarely the subject of observation.

2d. The impression produced by it upon my mind that the degeneration of the bones was preceded by that of the muscles—that the degeneration of the two tissues was dependent, in this instance, on the same crasis, and the probability, therefore, that such is its history in other cases also.

3d. The opportunity of placing on record a careful quantitative analysis of the urine in this disease.

4th. The fact of the degeneration being least advanced in the external circumference of the bone.

5th. The formation of perfect fat vesicles in both bone and muscle.

As respects the symptoms which precede the known softening of the bones, attention may be called to the absence of those rheumatic pains which are usually stated to be precursors, nay by some supposed to be the actual causes of the malady. When the disease is fairly established, these pains may conjecturally be referred to the pull which healthy muscle exerts on the periosteum, deprived of its usual firm base of resistance. But, as in the case before us, the muscle appears to have yielded first to the morbid influence, it did not strongly contract, did not drag on the periosteum, and so no pains of consequence were experienced. We see, also, that previous to the softening being demonstrated to exist, the same appearances were noted in the urine which were observed when the disease was fully established, pointing to a great probability that the chemical constitution was not dissimilar. We see also that the degeneration was purely automatic, not arising as a consequence of any other morbid state, nor, as far as could be ascertained, of an hereditary taint.

That the degeneration of the muscles preceded that of the bones is of course a matter of opinion. A person observing the circumstances only after death, would naturally suggest that it was due to their necessary inertness for so many months. But the condition of the patient during her stay at St. Mary's discountenances this idea. The mechanism of the bones was complete so far as their ordinary uses demand, yet so peculiar was the partial paralysis, or rather torpidity of fibrous contraction, that, in spite of the rarity of the disease, I was induced to select fatty degeneration of the muscles as the only explanation I could give.

As respects the quantitative analysis of the urine, it is confirmatory of the suspicion usually expressed, though not hitherto proved, that a great loss of lime takes place through this channel.

The observation that the degeneration was least advanced in the external surface of the bone, shows that its course is from within outwards; and that, therefore, till the shell of osseous structure bends or breaks, the bone is as useful as ever for the purposes of muscular motion. The mere thinness of the plate of bone remaining intact can make no difference mechanically to the action of the muscles implanted in it, so long as it is strong enough to bear the strain. It will be seen that this last argument has an important bearing on the first observations made concerning the early diagnosis of the disease.