NOTES OF A CASE
OF
DUCHENNE'S PSEUDO-HYPERTROPHIC
MUSCULAR PARALYSIS,

WITH SPECIAL REFERENCE TO THE TEMPERATURE OF THE
OVERGROWN LIMBS, AND WITH GENERAL REMARKS.

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SINCE Dr. Duchenne first drew attention, in the year 1861, to the occurrence of a form of progressive paralysis, accompanied by hypertrophy of certain muscles, several English observers have brought before the notice of the profession illustrative cases. Among these I may mention Dr. Meryon, Mr. Wm. Adams, Dr. Hillier, Dr. Langdon Down, Mr. Kesteven, and Mr. Butlin. But the pathology of the disease is only in part understood, and the disease itself is of sufficient rarity to justify me in communicating to this Society the record of a case in which one new, or at least seldom noticed, fact has been observed.

A. M—, æt. 7, was admitted for the second time into St. Thomas's Hospital, under Dr. Bristowe, on the 1st of May, 1873.

History.—His mother stated that he was a “beautiful baby” when born; that he began to say words at nine months, and “spoke properly” before he was two years old. He had no fits during infancy or afterwards. [About two years ago, a year after an attack of measles, he began to show signs of weakness in his limbs. To use the mother’s very apt words—“his right foot hanged with him.” He soon began to tumble down while walking, but was at first able to get up again without help. About ten months ago it was noticed that he could no longer pick himself up again after a fall, and about the same time the calves of both legs were seen to be over-grown (See Plate I). No particular extension of weakness since then appears to have been recognised by his parents. His mother says that he learned the alphabet at six years of age, and she considers that he has a good memory.]

He has two brothers living: one, aged ten years, an idiot from early childhood; the other, aged eight months, as yet a “beautiful baby.” One sister is living, aged three years, healthy. One sister and one brother died in infancy, one of whooping-cough, the other of teething. The mother knows of nothing which could have caused his illness. He was in Victoria Ward from November 25th, 1872, to January 12th, 1873, and is said by the sister of the ward to be stronger now than then.

Examination.—The boy stands with his feet placed about nine inches apart, the toes being turned a little out; the feet are large and flat. The knees are bent a little forwards, the buttocks backwards; the calves, very much overgrown as compared with all other muscular parts of the body, project far beyond the line of the thighs in profile. His back forms a deep curve forward from the hip to the shoulders, so that the belly and lower chest are made very prominent, and the shoulders are thrown back. A line falling from the point of the shoulder to the ground would be quite clear of the buttock. The head, rather large in proportion to the body, is bent forwards. The muscles of the back are evidently much wasted.

The right shoulder is contracted and drawn downwards, and its muscles are also wasted. The muscles of the rest of the body, excepting the calves, are poor and thin for a boy of his age (See Plate I).

Measurements :—

Circumference of head from occipital protuberance round frontal protuberances	20 inches.
Right upper arm below insertion of pectoralis major	$5\frac{1}{8}$ „
Left ditto	$5\frac{1}{2}$ „
Right forearm two inches below elbow	$5\frac{1}{4}$ „
Left forearm two inches below elbow	$5\frac{1}{4}$ „
Circumference of chest, nipple line	$20\frac{1}{4}$ „
Circumference of abdomen at umbilicus	$17\frac{3}{4}$ „
Right thigh, middle	$9\frac{1}{4}$ „
Left thigh, middle	$9\frac{1}{2}$ „
Right leg	$8\frac{3}{4}$ „
Left leg	$9\frac{1}{16}$ „ ¹

He walks from his hips, shuffling the feet along the ground without getting them clear, and swaying his body from side to side in the alternate raising and lowering of the hips. There is evident weakness in the movements of all muscles below the

¹ I have since measured the legs and thighs of a number of healthy children, and find that the calf is usually about a fourth less in girth than the thigh, the ratio being in six cases 12 to 8 $\frac{1}{2}$. Comparing slim children of about the same age, I found that the calves in this case were at least half an inch larger than they should be. They appeared to be overgrown therefore—1st, as compared with the rest of the muscles of the body; and 2nd, as compared with their natural bulk. They were much firmer to the touch than the other muscles of the body, and when thrown into contraction became very hard and knotty.

hips. The toes are turned inwards in walking. On the lightest touch, and often without external impulse, he falls down. The left leg is the first part to give way; it is doubled under him, while the right is extended from the knee and flexed from the hip; at the same time the body comes crashing to the ground in a sitting posture. In this his favorite attitude he moves readily along the floor by the aid of his arms. When he wishes to rise he bends his trunk forwards, and places his hands upon the floor in a way that reminds me of a baboon; he then bends the right leg and raises the trunk in an oblique position, but cannot rise to his feet; then, by seizing a chair or similar support with both hands, he is enabled to straighten his legs and stand, but with the trunk now horizontal. The sacro-spinal muscles are too weak to lift the head and shoulders without further assistance. It is necessary therefore, lastly, that he seize some higher fulcrum (such as a bedpost), and climbing, with hand over hand, gradually raise the shoulders through the agency of the dorso-humeral muscles.

As concerns his intellect few indications of deficiency or damage can be detected. His manner is gentle and somewhat lively; he is well-conducted and obedient, but the sister of the ward says that his memory is short and that he stammers at times. His speech is clear and free from stuttering.

All the muscles of the body are sensible to faradic electricity. The muscles of the calf are the most sensitive; the left thigh more than the right; the right pectoralis major, deltoid, and sacro-spinal muscles, less than the rest. The left leg and the right arm are distinctly weaker than their fellows. When the sensibility of the legs and other parts is tested with callipers his answers are so vague and contradictory as to make it quite uncertain whether there be any paralysis or excess of sensibility.

Observations.—The skin of the calves was not different in tint from the skin elsewhere, that is to say there were no signs of the red discoloration spoken of by Duchenne. On

the other hand, when the hand was applied to them they appeared to be warmer than the thighs. I had not been able to find any observations of the existence of a higher temperature in the skin over the hypertrophied muscles, either in Duchenne's works or others, although the possibility of the occurrence had not escaped that acute observer. I now endeavoured to ascertain by the use of a thermometer the relative temperatures of the legs and thighs. For this purpose I made use of a surface thermometer, made by Mr. Hawksley, principally, I believe, at the suggestion of Dr. Sibson. It consists of an ordinary clinical thermometer having the bulb lengthened and twisted into a flat coil. One side of the coil is supported by a little plate of boxwood, to which it is cemented by putty. Over the wooden plate an oval sheet of india-rubber is stretched, having its margin extended by a ring of metal fixed to the stem of the thermometer. When the thermometer is applied with the free face of the coil in contact with the skin, the coil is enclosed by the above arrangement in a little chamber with non-conducting walls, and is defended from atmospheric and other disturbing influences. In taking the temperature the thermometer was always brought to 80° Fahr. as a starting-point, and the further rise of temperature during three minutes' application of the instrument to the surface was noted.

The first observation was made on the 3rd of September, when the thighs and calves were compared. The results were—

Right thigh 83·0	Left thigh 83·6
Right calf 85·6	Left calf 85·7

showing a difference in favour of the calf of 2·1, and 2·6, on the left and right sides respectively.

On September 8th, right thigh 85·3; right calf 87·1; a difference of 1·8.

The observation was repeated at the end of ten minutes, during which the child being under examination was exposed in a temperature of 62° Fahr.

The figures were now—

1st. Right thigh 81·5, after 3 minutes of exposure.

2nd. „ 82·5, after 3¼ „ „

3rd. Right calf 85·4, after 3 „ „

Both parts being cooler, the difference was more marked than before, amounting to nearly four degrees with equal exposure.

September 11th.—Left thigh 87·5; left calf 89·7; 2·2 deg.

September 13th.—Left thigh 86·3 (3¼ minutes).

Left calf 89·2 (3 „). (2·9 deg.

Left shoulder 87·6

Left cheek 92·5, after a fit of crying,
the face being flushed.

Similar observations were made on three other occasions, with the same result. There was an excess of temperature in the calf of from 1·8 to 3·9, as compared with the thigh.

The microscopical condition of the muscles of the calf was next investigated. For this purpose an emporte-piece, or tissue-explorer, devised by Mr. Hawksley, was used, Dr. Duchenne's instrument appearing to be unnecessarily large. Mr. Hawksley has made for me a small cannula, with oblique-pointed and sharp-edged end, through which, when thrust into the tissues, a stylet, terminating in a screw with a broad sharpened thread, might be introduced, and by a twisting movement might be made to entangle small pieces. In practice this worked very well. It gave little pain, caused little or no bleeding, and after bringing away enough tissue to suffice for three or more microscopic slides, it left only a tiny puncture, which healed without any discomfort.

The muscular tissue brought away was paler than natural. Under the microscope the muscular fibres were of full size, well defined, and free from fat or other accompaniments of degeneration. The striæ were fine, clear, and close. Between the fibres an unusual proportion of fibrous tissue was observed, and it appeared also to me that there was an excess of nuclei on the surface of the sarcolemmata. There did not appear to be an excess of fat.

I have spoken of this case as illustrating Dr. Duchenne's pseudo-hypertrophic muscular paralysis, because the symptoms agree with those of the cases published by him. The disease with which this disease might be most easily confounded, would be the progressive muscular atrophy of infancy. But, as Duchenne shows, this disease advances from the face to the trunk, from the upper limbs to the lower; it destroys the muscles in a progressive but irregular way, and, after the destruction of the muscular fibres, produces local and partial deformities. Whereas, in the case under consideration, as in other similar cases, the disease attacks first and principally the lower limbs, producing a gradual and general enfeeblement, and an overgrowth instead of a wasting of certain muscles. The atrophic paralysis of infancy (infantile paralysis) is even more clearly defined by its sudden occurrence—fits or fever often introducing it; by its completeness at first, tending to diminution afterwards, with localisation in a certain number of muscles; this localisation issuing at a later stage in various partial deformities of limbs, owing to disturbance of the balance of muscular tonic force. In this I again quote Duchenne. The considerations which led me to read the note of this case before the Society were connected with the observations of the temperature of the calves. The disease is sufficiently rare to furnish individual observers with but few illustrations in a long time. The pathology of the disease cannot be said as yet to be understood. And therefore any observations tending to throw light upon this point will be useful if they are followed up by those who have at one time or another opportunities of investigation. It is cited by Duchenne as one of the points of difference between this disease and progressive muscular atrophy, that whereas in the latter the papers of Dr. Lockhart Clarke and Dr. Bastian have proved the existence of degeneration and wasting of the anterior column and grey matter of the cord, in examination of the spinal cord of subjects of the pseudo-hypertrophic muscular paralysis made under Dr. Duchenne's own directions, no morbid appearances have been detected. In one of the latest reported, only a year ago, the sections of the spinal

cord are recorded to have been altogether healthy, and the testimony of Cohnheim and Eulenburg is to the same effect. Mr. Kesteven, a most trustworthy authority, reported in the 'Journal of Mental Science' for January, 1871, upon the "Microscopical Anatomy of the Brain and Spinal Cord, in a Case of Duchenne's Paralysis, associated with Imbecility." He found the following morbid conditions and changes of structure:—dilatation of the peri-vascular canals, consequent upon long-standing congestion; and numerous circumscribed spots of granular degeneration of the nerve substance. These were found irregularly scattered in the white substance adjoining the grey matter of the convolutions, while very few were noticed in the grey matter itself. They were sparse in the corpus striatum and thalamus opticus; and in the medulla and cord they were few and far between. They were evidently spaces caused by loss of tissue replaced by the morbid matter. It must be remarked that Mr. Kesteven found similar appearances in a case of idiocy with convulsions, and in other cases, so that it cannot be deemed to be characteristic of the pseudo-hypertrophic paralysis.

Of the condition of the muscles in this disease we have evidence from many observers. From a comparison of various statements it results that the appearance differs in the several stages of the disease. There is an early stage in which the muscular tissue is little altered, but there is excess of intermuscular fibrous tissue; and there are further stages in which the muscular tissue becomes attenuated and degenerated, and in which there is present a growing quantity of fat between the fibres. In advanced stages, described by Cohnheim, there is actually a large quantity of adipose tissue formed between the muscular fibres. This latter form would appear almost a distinct one. Between the irregular deposition of fat in granules and the formation of adipose tissue the difference is of course between degeneration and tissue-formation; the formation of fat-cells from the corpuscles of the connective tissue being a very generally accepted fact. The present case, however, does not throw light upon this point. The history and the histology both demonstrate an

early stage of the disease in which the muscular fibres are still perfect, and the overgrowth of fibre simple and untainted by intrusion of fat.

With this I am inclined to connect the comparatively high temperature of the calves. It would indicate, in one way, what the "hyperplasia," to use Duchenne's word, indicates in another, hyperæmia of the affected muscles. In relation to this I noted very carefully the power of the muscles. Under the influence of the will and under the stimulation of electricity they contracted firmly, and, as I have noted, became knotty; but the power they exerted was but feeble. More than once I have heard it argued that the hypertrophy was a real hypertrophy, and was due to the extra work thrown upon certain muscles by the weakness of others. The cases of general pseudo-hypertrophy described by Duchenne made it certain that at least in one group of cases this would not be the explanation. And after watching for a long time on many occasions the movements of the child, I arrived at a certain conclusion that the calves were, relatively, not stronger than the other muscles, and that they did not do more duty. He made use to an overwhelming extent of the thin but much stronger muscles of the upper limbs. After this I think that the conclusion is fair that first the hyperæmia, and second, the overgrowth are due to internal causes—to causes affecting nervous centres. And I would further suggest that it is possible that the disease may have its origin in the sympathetic nervous system, outside the spinal cord and brain. The whole story is rather one of weakness following faulty nutrition, than of paralysis of muscular nerves, followed by wasting of muscles. There is at least evidence of vasomotor disturbance in connection with the overgrown calves, and I should consider any future post-mortem examination incomplete which did not include a careful investigation of the state of the prævertebral and other ganglia. Such an examination was in one case made by Cohnheim, who reported that he had not found any alteration of the ganglionic system.

In the present case, the possibility of vasomotor distur-

bance being set up by peripheric irritation was not lost sight of. None could be detected, however. The child's viscera gave all the indications of health, and excepting locomotion, the functions were well performed.

In referring the disease mainly to vasomotor disturbance I am but following the indications of Dr. Duchenne, whose views appear to me to be confirmed by the observations in this case.

DESCRIPTION OF PLATE I.

Case of Duchenne's Pseudo-Hypertrophic Muscular Paralysis.

Fig. 1. Front view.

Fig. 2. Back view.

Fig. 3. Side view.

Fig. 1.

Fig. 2.

Fig. 3.