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PSEUDO-HYPERTROPHIC PARALYSIS

(OF DUCHENNE)

OCCURRING IN AN ADULT.

BY

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CASE OF PSEUDO-HYPERTROPHIC PARALYSIS.

THE following case is extremely interesting, as being a well-marked example of a very rare disease,—a disease which has only been known to exist since 1861, when the attention of the profession was directed to it by Dr Duchenne (de Boulogne). But the case is still further interesting, in fact unique, as showing the disease occurring in an adult, whereas it has hitherto been believed to be exclusively a disease of infancy and childhood. Dr Duchenne, to whose elaborate and able researches, published in the form of a monograph on the subject,[1] we are indebted for all our information on the disease, says (p. 114, *op. cit.*):—"During more than twenty years that my researches have specially been made on affections of the muscles, and that, I may say, on a very extensive scale, I have never observed in the adult anything analogous to the pseudo-hypertrophic paralysis of infancy. In short, from what has already been said, there follows the important fact, that pseudo-hypertrophic paralysis is a disease of childhood."

The case is as follows:—J. C. M., æt. 26, was admitted into the Aberdeen County Prison on 2d June 1869, under a sentence of nine months' imprisonment. He had been at first a gardener by occupation, but for the last four years he had been a book-hawker. In this latter occupation he had not had any excessive amount of walking, and had been quite healthy up till about a month before being sent to prison. About this time (end of April) he began to feel "stiffish" in walking, had pains "like rheumatic pains" in the outside of the thighs, and found himself often involuntarily rubbing them. About this time he had a gonorrhœa, and on admission to prison he had eczema of the inside of both thighs, which was soon cured. On admission, he complained of pains in the lower part of the back and thighs, which might, however, have been caused by the eczema. He continued to feel his walking difficult, and his legs were getting weak; but he did not observe any muscular enlarge-

[1] De la Paralysie Musculaire Pseudo-Hypertrophique, ou Paralysie Myo-Sclérosique. Par le Dr Duchenne (de Boulogne). Paris, 1868.

ment till about the beginning of July, a month after being sent to prison. At this time he told me that he thought his thighs were much larger than natural. He did not sleep well, looked heavy and stupid, but ate his food well, and his bowels and other excretions were normal. I examined his thighs at this time, and found them to be unmistakably larger in girth than natural. To the feel, the muscles felt firm, almost hard. Three weeks later, the calves of the legs had followed in the same enlargement, and were very firm and hard to the feel, the unnatural size of the thighs and calves being at once visible on inspection. The oblique abdominal muscles were also visibly enlarged. About the beginning of September, he complained of his arms feeling weak, and that he was unable to do his task. I found the biceps of both arms larger than natural, and rather firm, especially on flexion. The forearm was not enlarged, but rather the reverse, and the hands were thin and skinny. He had difficulty in closing his hands, and the grasp of both hands was extremely weak. When holding anything heavy in his hand, he felt considerable difficulty in preventing its fall. He complained now of no pain in his arms, thighs, or legs, but of general weakness, inability to use his hands, with stiffness and difficulty in walking, which he did with a slow, waddling, side-to-side gait, and with the legs far apart. There was no pain or tenderness in any part of the spine, no anæsthesia, no increased difficulty in walking with the eyes shut, no headaches or other "head symptoms." The skin was of normal colour. I did not take the measurements of the limbs till 27th October 1869, when the size of each was as follows:—

Girth of arm at middle of biceps, when extended,	10 inches.
„ „ when flexed, .	11½ „
The same in both limbs.	
Girth of thighs opposite the trochanter major, .	23 „
„ at middle, ...	22 „
„ at lower third, ...	19 „
The same in both.	
Girth of calf of right leg, ...	15½ „
„ „ left leg, ...	15 „

12*th January* 1870.—State much the same, except that the enlargement of the oblique abdominal muscles is more marked. The muscles of the back do not seem to be affected. The glutei had probably been affected at this time also with hypertrophy, although I did not observe it till a month later. There is no albumen in his urine, which has a specific gravity of 1·020, and is acid. No difficulty whatever in micturition, and action of the bowels regular and natural. He complains now of weakness of sight, especially of right eye. As there was no convenience for ophthalmoscopic examination in prison, this was deferred till his release, which took place on 9th February 1870. He then came

to my house, where I examined his state again. The measurements of the limbs were found to be the same as on 27th October 1869, except that the girth of the calves was the same in both limbs, viz., 15½ inches. The glutei muscles were now visibly larger than usual, but the muscles of the back seemed unaffected, and there was not the deep anterior curvature in the lumbar region which is usually observed, and which is produced by the involvement of the muscles of the back in the paralysis. The state of the eyes, as reported by Dr Alexander Ogston, the Ophthalmic Surgeon to the Hospital, who kindly examined them, was as follows:—

Right eye.—S. = $\frac{1}{18}$. Retinal vessels small; pigment external to optic disc. Otherwise healthy.

Left eye.—S. = $\frac{1}{12}$. Vessels of retina small. Papillary edge of choroid showing pigment maceration. Colour vision perfect.

The grasp of his hands was at this time, if anything, weaker than before, and the movements of the fingers in flexion and extension, especially the former, difficult. After sitting down on a chair, he had considerable difficulty in rising, and for a few minutes after felt very stiff in walking. His walk was markedly of the side-to-side waddling character. He could only walk very slowly,—running, he felt, was out of the question. He had much difficulty in pulling off and on his boots.

I should state that his weight on admission to prison was 157 lbs., and at his discharge 162 lbs., showing an increase of 5 lbs. He was going home to the country, as he felt unable for any work; and in case I should not again meet with him, I think it better to record the case at this stage.

Remarks.—This disease is so rare, recognised as a definite disease so lately, and so little generally known, that in the way of making a few remarks on the foregoing well-marked example of it, the profession may perhaps think that I am not uselessly occupying the pages of this Journal by giving a general sketch of the disease in question; and in doing so I shall simply give a resumé of Dr Duchenne's interesting memoir.

That there existed such a disease as pseudo-hypertrophic paralysis was not known till so recently as 1861, when that very able observer, Dr Duchenne, met with a case of it, and recorded it. Since then, he has in his memoir collated thirteen cases, partly from his own practice, and partly from those of other physicians. Other cases have from time to time been recorded in various journals. All the cases recorded by Dr Duchenne commenced in early infancy or in childhood, the most advanced age of *commencement* which he has met with being ten years.

The general phenomena of the complaint are as follows:—There is—

(1.) A stage of general feebleness and weakness of the lower limbs.

(2.) There is a peculiar gait of side-to-side waddling, with unnatural separation of the limbs in walking.

(3.) A deep anterior curvature in the lumbo-sacral region of the spine, when in the upright position.

(4.) A state approaching that of talipes equinus, with clawed nails.

(5.) The muscles of these limbs enlarge to a very marked degree; are apparently hypertrophied.

(6.) There comes a period when the symptoms remain stationary.

(7.) After this, the paralysis becomes general over the body, and increases progressively in degree, the patient ending by being unable to move himself off the couch, and generally dying by some intercurrent disease, which his debilitated condition renders him unable to resist.

Let us look more particularly at each of these symptoms.

Those attacked by the disease have generally been in perfect health, and have been fine, healthy-looking children. No pains or fever precede the morbid state, and in only two of the recorded cases did an attack of convulsions occur shortly before the symptoms of the malady appeared. The first symptom in order of time is the weakness of the lower limbs. If the child is under the age when walking would be looked for, the enfeeblement may be entirely overlooked until this period arrives, when it is found that it cannot stand, and does not, as usual, instinctively move about its limbs, but cries, when put down, to be taken into its nurse's arms. This state is apt to be thought little of by mothers when they find that the little limbs, instead of growing thin, are large and firm. If the weakness commences at a later period, the child gets easily fatigued in standing or walking, falls frequently, runs with difficulty, or not at all, and finally will not walk, but desires to be carried.

(2.) Next we have the separation of the lower limbs, and the lateral balancing, or side-to-side movement in walking.

The separation of the limbs is an instinctive means of keeping up the equilibrium, and strengthening the base of the body in motion. This may occur in other diseases when the lower limbs are feeble. The lateral balancing or side-to-side waddling movement in walking, is caused by the weakness of the muscles of the back and buttocks. This also is observed in other diseases, but it is never to the same extent, and instead of soon disappearing, in the disease we are now speaking of, it only becomes aggravated.

(3.) There is the spinal curve when standing upright and walking. This is one of the most constant symptoms of pseudo-hypertrophic paralysis, but is only present during walking, or in the erect position. During the first period of the disease, there is only a slight bending backwards of the trunk, which by degrees increases till a deep lumbo-sacral curvature is formed in the upright posture. This curve in the lumbo-sacral region of the vertebral column is caused by the paralyzed condition of the extensor muscles of the spine, which generally exists in this complaint. Whenever these extensor spinal muscles are paralyzed, then the curve is produced.

What proves that this is the real cause of the curve is, that Dr Duchenne found that in all cases where it existed, if, in making the patient stand upright, he tried to prevent the spinal bend, the trunk fell forwards, and the patient could not rectify his position without the assistance of his hands upon the legs and thighs. It is then a natural result of the effort to keep upright; the patient bending backwards, and supporting the body by means of the abdominal muscles, which act as flexors of the spinal column. When a patient having this curve stands upright, a plumb-line touching the spines of the vertebræ at the most prominent point falls far behind the sacrum. Dr Duchenne calls the curve thus produced "paralytic lordosis of the lumbo-spinal muscles," to distinguish it from that curve produced by weakness of the abdominal muscles in other diseases; in which case, though a deep lumbo-sacral curve may be formed, a plumb-line applied as above falls over the centre of the sacrum.

(4.) Equinism, or talipes equinus. This, Dr Duchenne says, is one of the most constant phenomena in the disease in question. It does not appear in the early stages of the malady, but, beginning very slightly, it increases gradually till the heel with difficulty touches the ground when in the standing posture. Then afterwards it takes the form of talipes equino-varus. The plantar arch is extended, the first phalanges are placed in a position of unnatural extension with respect to the metatarsal bones, while the two last phalanges are unaltered. The toes thus take the appearance of claws. The cause of this equinism, Dr Duchenne has satisfied himself, arises from the excessive predominance of the extensors of the foot over the flexors in this disease. He found that flexion of the foot was performed with difficulty, and feebly, whereas the reverse is obtained in extension.

(5.) The apparent muscular hypertrophy. This is perhaps the most striking feature in the whole disease,—the existence of enlargement, firmness, and even hardness of the voluntary muscles, along with a state of extreme weakness. This apparent hypertrophy always follows in point of time the state of well-marked feebleness, which is observed as the first symptom. Any of the voluntary muscles may be thus attacked with apparent hypertrophy. It generally begins in the lower limbs, in the gastrocnemii, in the thighs, and the buttocks; then the lumbo-spinal muscles may follow; the oblique abdominal muscles also, and the biceps of both arms, as in my case; the deltoid, and even the muscles of the face, especially the temporals. But while we find some muscles hypertrophied, we may find others emaciated. Thus the muscles of the upper extremities may be smaller than natural, while those of the lower are hypertrophied. In my case, the biceps of each arm was enlarged, while the forearm was rather thinner, and the hands quite skinny. In other cases, the pectorals and the sterno-mastoids have been found atrophied. Thus, it appears that although

nearly all the voluntary muscles may be paralyzed, it does not follow that they should all be hypertrophied; or, in other words, the degree of paralysis does not proceed in the direct ratio of the hypertrophy. The description of the anatomical state of the muscles causing this apparent hypertrophy, I shall reserve till we finish the consideration of the general symptoms.

(6.) Then comes a period when the progress of the disease comes to a standstill for a varying period, generally several years, during which time the hopes of the friends are apt to rise. But—

(7.) After this stationary period, the disease starts afresh. The weakness of the lower limbs increases till the patients are unable to stand upright, and are confined to their bed or couch; the upper limbs become likewise enfeebled, and often become meagre and atrophied, so that the patients are in a nearly helpless condition, during which they are cut off by some intercurrent disease, such as phthisis. There are no cerebral symptoms which are uniformly present. In some cases there has been slowness of speech, with blunting and dulness of the intellectual powers, even idiocy, while in others the mind has been perfectly intact.

The electro-muscular contractility is at first quite normal, and it is only in the advanced stage of the disease that this is diminished or lost. The temperature of the enfeebled and hypertrophied muscles is generally lower than normal, and the skin has sometimes a rosy or bluish tinge, but neither of these phenomena are constant. It is also to be observed that, in general, no pain is complained of, and there is neither hyperæsthesia nor anæsthesia. The functions of the bladder and rectum remain normal, and the general health is only affected at an advanced stage of the disease.

We come now to inquire what are the anatomical alterations which are found to exist in pseudo-hypertrophic paralysis. As to the state of the nervous centres, the results are purely negative. Only one autopsy has hitherto been obtained, and in it no lesion of the nervous centres was detected. Then, as to the condition of the muscles: this has been ascertained in the dead body, and in several other cases in the living body also. This latter has been accomplished by a peculiar instrument, used by Dr Duchenne for extracting a piece of muscular tissue from the living body, and called by him "emporte-pièce histologique."

In the first place, the apparent hypertrophy of the muscles has been found to be the result, not of increase in number or size of the muscular fibres, but of an hypertrophy of the connective tissue, which normally exists. This is what occurs in the second stage of the disease, namely, when the muscles begin to enlarge. Besides this hypertrophy of the ordinary connective tissue, there is also a formation of numerous fibres, with fine undulations. A little further on, in the course of the disease, disseminated among the connective and fibrous tissues, there are found fatty particles of different sizes, and sometimes drops of oil, in clusters.

Next, as to the state of the muscular fibres themselves, they are found (1.) discoloured, presenting a pale appearance, and resembling the subcutaneous cellulo-adipose tissue. (2.) The normal transverse striation becomes extremely fine, and in some parts hardly to be seen, till (3.) the transverse striation completely disappears. (4.) The transverse diameter of the muscular fibres becomes very much diminished; there is, however, no parallelism between the diminution in the transverse diameter and the fineness of the striæ, as it has been found that in fibres whose transverse diameter has been normal the striæ have become very fine, and have even entirely disappeared. (5.) There is a marked diminution in the number of muscular fibres in proportion to the quantity of hypertrophied cellulo-adipose tissue. Griesinger found that in one case the intermediate tissue occupied six or eight times larger space than the muscular fasciculi. If the muscular fibres had not really degenerated, the size of the muscles involved ought to be larger than it is, judging from the enormous hypertrophy of the intermediate connective and fibroid tissues.

But there is good reason to believe that the muscular fibres do really degenerate and disappear, and that probably these undulated fibres, which exist among the connective or areolar tissue, are in reality what remains of empty sarcolemmata. The fatty globules, however, are not part of the muscular fibres in a state of fatty degeneration, but are entirely outside of them. The anatomical state of the muscles shows completely that it is an entirely different disease from that known as progressive fatty atrophy of the muscles. The symptoms of the disease are so marked, and so different from those of any other, that the diagnosis is hardly to be mistaken.

What, then, is the ultimate cause of this muscular disease?

It has been already mentioned that no lesion of the brain has been found; and although in some cases there was obtuseness of the intellectual faculties, even going the length of idiocy, yet in others there was no symptom of this sort, the mind being quite intact.

Besides, as Dr Duchenne remarks, a cerebral lesion would not be sufficient to account for the morbid condition of the muscles. He also observes that, reasoning by analogy, we should not expect to find the seat of the disease in the spinal cord; and at the autopsy above referred to, the spinal cord was found intact. Dr Duchenne is inclined to place the seat of the disease in the vaso-motor nerves, and states that he cannot conceive of the production of such an increase of connective tissue, unless the vaso-motor nerves are involved and paralyzed. The facts that in some cases the skin has been of a rosy or bluish colour, and that the temperature of the affected parts is generally lowered, would seem to go in favour of this view. But no lesion of the ganglionic system has as yet been discovered. M. Bergeron believed that the paralysis of motor power was caused by the compression exercised on the muscular

fibres by the hypertrophied connective tissue; but, unfortunately for this view, the paralysis does not always proceed side by side with the hypertrophy, but many muscles may be paralyzed without having their volume at all increased.

In fact, the pathology of this disease is extremely obscure, and must for the present be left unsettled. As to the age at which this disease may commence, none of the cases recorded by Dr Duchenne commenced later than 10 years of age. The case which I have related commenced at 26, and thus forms a rather important contribution to the history of the malady.

In many of these cases there has been no trace of an hereditary taint, while in a few it has attacked two or more members of the same family.

As to the prognosis, Dr Duchenne states, that if observed and treated in the *first* stage—that is, in the period of weakness, before the pseudo-hypertrophy has commenced—it may be cured. Indeed, he records two cases thus arrested or cured. But if it has gone beyond this stage, and the muscles are already enlarged, it is beyond the reach of curative measures. At least, none of the cases thus advanced have at all improved, but have gone on slowly but surely to the end.

The only treatment which has been found to be of the least use, in even the first stage of the disease, is local Faradization of the affected muscles; and it was by persevering in this treatment that two cases were cured by Duchenne. Along with this, however, he used iodide of potassium, or certain medicines supposed to be of use in the diseases of the nervous centres, such as nitrate of silver or the preparations of phosphorus. But used alone, these internal remedies, as well as strychnine and ergot, with baths, frictions and blisters, cauterizations in the lumbar regions, and hydropathy, were all found useless.

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