A Lecture

ON

MYOPATHY AND A DISTAL FORM.

Delivered at the National Hospital for the Paralysed and Epileptic.

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In the last lecture I gave here, I considered the various maladies that depend on defective vitality, on the imperfect vital endurance of the tissues which causes their nutrition to fail, generally early in life, sometimes before birth. I suggested in this decay, which results from defect of life, might be called "abiotrophy," and the word has been welcomed as convenient and useful. I illustrated the condition, as some of you may remember, by the wasting of the hair follicles of the skin which causes premature baldness, by the forms of muscular dystrophy which often occur in families, and have thus their essential vital nature strongly emphasized, and also by the many forms of degeneration in the nervous system that are of the same nature, which vary according to the special structure that fails, the time of life at which the vital failure begins, and the cause which sometimes seems to be its incidental excitant.

I have to return to the same subject today in one of its special manifestations. We are all opportunists, more or less, and I am able to show you two examples of abiotrophy which are so instructive as to compel me to take them as my subject to-day. You know that these primary affections of the muscles are called idiopathic muscular atrophy," "myopathy," and also "muscular dystrophy." The last name (due to Erb) means "difficult nutrition," and has a certain aptness, because there is often an overgrowth of the interstitial tissue which suggests the idea of a struggle for existence, such as we sometimes see between weeds and the proper crop which has been sown on the ground. The metaphor is not quite precise, nevertheless well put, and I am able to show you two instructive cases, in which the interstitial growth attains a special luxuriance. Fat cells are formed in the fibrous tissue that develops, and from this there is an enlargement of some muscles, wherein has come the common name of the disease. It was given by Duchenne, who had been specially struck by this feature, the more so because it had chanced to him to meet with some cases in which the enlargement attained a rare extension and degree, being present in most of the muscles. He used to claim Raphael's child-angels as examples of the disease. Some of you may remember the extraordinary development of their muscles as depicted in the celebrated cartoons. In the early days of this hospital there hung, over the fireplace in the consulting-room, an engraving of Raphael's picture, "The Transfiguration"; in it is shown a child with demoniacal possession; it was placed there as an appropriate representation of an epileptic fit, or the most accurate known in art.

On a visit Duchenne once paid to the hospital, I remember that he pointed out the figure as one which might well pass for an example of the disease on which he had bestowed the name.

But there may be no increase in size of the muscles, or there may be a decrease. The interstitial tissue may merely compensate for the wasting of the muscular fibres or it may fail to do so. Hence, in the same case, we meet with muscles that are weak and large, that are weak and normal in size, and that are weak and small. Interstitial growth, which is luxuriant, or moderate, or slight, is seen in the same case; one or the other may preponderate in different cases. Names are often instructive, but they sometimes obscure. I think that Duchenne's designation hindered the recognition of the fact which has been established by Erb and others that neither the amount of interstitial tissue, nor its character, is an essential element of the disease. The constant element is the change in the muscular fibres themselves; some of them may appear at first only slightly large and small, and present the features of degeneration; ultimately this comes general, irrespective of the increase of the interstitial tissue. It is a true abiotrophy.

As to these differences let me remind you of what I said in the last lecture regarding the growth of interstitial tissue which occurs in the nervous system when there is primary atrophy of the nerve elements. I pointed out the variations it presents, and that it sometimes attains a certain vigour and energy which makes it overstep the strict limits of the degenerated tracts. This feature at one time raised a suspicion that it was the primary element in the process, and a similar suspicion regarding the muscular interstitial atrophy may be set at first by the abundant development of interstitial tissue, especially in the pseudo-hypertrophic form. Let me remind you also of another fact I pointed out. When we consider the two features of this double process, the atrophy of the functional structures, and the overgrowth of the interstitial tissue, they suggest that the two elements have a vital relation, common in the necessary connection of the life of each, but opposite in tendency. When the nutrition of the muscular or nerve fibres begins to fail, that of the interstitial tissue is increased. It does not grow merely to fill up space; it is not truly a secondary process, it is the expression of an excess of vitality simultaneous with the diminution of that in the proper structures. The two tendencies, plus and minus, seem to be synchronous in these cases of maladies, and may have the same conspicuous than the diminution. The interstitial growth may be at first much more obtrusive than the atrophy of the functional elements. I saw how, in the opinion of Erb, it seems the natural response of the interstitial tissue to the influence of a toxic agent (tobacco) in the form of interstitial neuritis, the symptoms of which lead the way, but at a later date, it may have been more conspicuous than the diminution. The interstitial growth may be at first much more obtrusive than the atrophy of the functional elements. 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tissue of muscle which is developed in a wrong place, and in a wrong way. It shows how morbid development results in the condition we find in this disease, and illustrates its truly "abiotrophic" nature.

**PSEUDO-HYPERTROPHIC FORM.**

If cannot, for lack of time, and I hope I need not, describe the various symptoms of pseudohypertrophy, but I may point out some symptoms that are illustrated by the cases I have here. The first is a boy of 7, who is still in the early stage. His case seems alone in this family. I insisted in the last lecture on the fact that all family maladies are prone to occur in isolated form, and how important it is to learn to recognize them by their characters and course, because you have not the evidence of their nature afforded by other cases in the same family.

In this child no muscles are sufficiently increased in size to attract much attention, but the calves are distinctly large and firm. The knee-jerks are slight, but are not lost, as they are in the later stage of the disease. He is just acquiring the characteristic method of getting up from the floor, aiding the extension of the knees by putting his hands on them. This transfers some of the weight of the trunk from the upper end of the femur to its lower end, from the place at which the power exerted by the rectus acts least advantage, to the place at which it acts most advantage. You may ask if this action is due to the weakness of the extensor of the knee, why is it characteristic of this disease, since such muscles are often weak from other causes? This is the characteristic one. The action is not absolutely characteristic; it is met with in other maladies, but in such it is very rare. This particular method of aiding the defective movement seems to be acquired only when the weakness comes on slowly during the development of muscular ability.

As a matter of fact it is so rare under other conditions that its diagnostic importance is very great.

The other two other symptoms, the significance of which I wish, the first to point out. One is that the infraspinati are enlarged quite as conspicuously as the calf muscles. When the muscle is put in action, as you can see, it appears as though the fibres were coarser than is common. The other fact is the atrophy of the latissimus dorsi and lower part of the pectoral major. These sometimes seem absent, and often, as here, are reduced to small dimensions. Their loss is often so great, even at an early stage, as to suggest a congenital defect.

The disease is far advanced in the second boy I show you. His weakness became conspicuous at the age of 7, and he is now 14. He lost the power of standing four years ago, about a month after an attack of scarlet fever. The acute disease seemed to induce a rapid increase in the symptoms; you should note this, because the symptoms of abiotrophic disease often appear as if set in action after some acute disease, scarlet fever or influenza; and the malady is ascribed to this influence, although it really existed before. It is easy to understand that an afflication commencing gradually may have been unnoticed, and that a depressing influence, which really only accelerates, may seem to be the cause. In this boy the calves are not absolutely large, but they are so in comparison with the other muscles, which are excessively wasted, and are weak even out of proportion to the wasting. He has hardly any power of flexing the ankle or extending the knee or hip, but these movements are also prevented by the great contraction which has supervened in the opponent.

Permanent shortening occurs very readily in this disease from contraction in the interstitial tissue. When the power of standing has been lost, such shortening quickly fixes the limbs in the position naturally assumed in the recumbent posture. In this child the contraction of the calves is great, and that of the flexors of the knees and hips is such as to prevent extension beyond a right angle. The calf muscles contract earliest, and most, because of the amount of interstitial tissue in them. The tibialis equinus that results often prevents the patient from standing and walking long after the weakness of the other muscles would do so. Once the muscles lose the nutritional influence of use, they become weaker rapidly, and this weakness, once established, is irretrievable. We shall presently see the great disadvantage this has in the present subject.

We can still perceive some enlargement of the infraspinati, although less than in the former case; the characteristic atrophy of the latissimus dorsi and lower part of pectoralis is present in an extreme degree. But of all the muscles of this boy, that which is enlarged in greatest degree, and that which determines the character of the morbid process in the different muscles are quite unknown to us. The muscles of the face have here fair power for the most part, but the closure of the eyelids is feeble, and the left orbicularis is much weaker than the right. This is important; the difference between the two sides emphasizes the morbid character of the weakness. I may add that the electrical excitability of the muscles is lessened in both of these patients in proportion to the wasting, equally to each current; this is the characteristic condition in these diseases. In this patient the knee-jerks can no longer be obtained.

**FACIO-SCAPULAR FORM.**

The weakness of the orbicularis is of interest as another of the many links which connect the various forms of muscular dystrophy. It is especially great in a variety in which the face is affected in an extreme degree, and likewise are those who care the patient is. I must leave it to him to demonstrate the special features, but I would ask you to note the extreme weakness of the orbiculars palpebrarum. It is so great that the patient cannot completely close the eyelids. Common as is the palpebral weakness, in few diseases of this class does it attain the degree seen in this form of myopathy. Yet a characteristic case of myasthenia has lately come under my notice in which the eyelids cannot be brought together by any effort—another link between this myasthenia and myopathy.

One case as is the weakness of many of the muscles in this facio-scapular form, great as it is the ultimate degree of their atrophy, however long its duration, the malady seems to be purely muscular; the nervous system, even the motor nerves, are unchanged. The fact rests on many observations, on no one more striking than that which has been recently published by Spiller of a case described in the early seventies by Duchenne himself, in which life only terminated at 38. We might wonder that this did not involve structural change in the course of so many years; but we must remember that the inability of the muscular tissue to respond to the motor impulse does not mean that the motor impulses themselves are less than normal. This question, however, would entice me far from my present subject.

**DISTAL MYOPATHY.**

The last case I have to show is one to which I would specially direct your attention because it presents unusual features. Indeed, I have not seen a similar case of this. We know that one has been recorded, but it is always improbable that any given morbid state has not been seen before, and has not been described somewhere in the vast expanse of medical literature.

This boy is 18 years of age, and is the eldest of three; the others are healthy. No similar case can be heard of in his family. The symptoms first attracted notice when he was 14 years old; then it presented this: He could not lift his toes against the ground in walking. At a later date his hands...
were found to be weak. This feebleness of hands and feet has slowly, steadily increased, until now it is great. He is quite unable to flex the ankles, although he can just extend the toes slightly, and can move each foot slightly in any direction. Tibialis anticus and the peroneus. He can extend the ankle-joint by the calf muscles with some force. The movements of the knees and hip are performed with good power; the knee-jerks are present, but the left is less than the right. The thigh muscles are of normal size; the anterior tibial muscles are distinctly smaller on the left side; the calves, on the other hand, are rather large, and are firm. They are sufficiently large to suggest a resemblance to the condition in pseudo-hypertrophic paralysis; they may have been larger in the past, for his mother spontaneously told me that one of the children who are now only 2½ years old, as he should. Extension is also feeble; he can get the fingers, with the wrist, into line with the forearm, but cannot fully extend them when the wrist is over-extended. A this a convenient index of the deficiency of power in these cases: I had recently occasion to call attention to it in myasthenia, which is also perhaps a muscular malady. The muscles of the forearms and hands are absent; there is no present, no wasting, of the hand and finger-muscles, which we see in progressive spinal atrophy. Above the forearm the muscles have fair power and present no wasting: only a feeble atrophy of the middle part of the trapezius can be observed. In the affected muscles excitability is lowered in proportion to their feebleness, and equally to paralytic and volitism. To this there is one curious exception; although the muscles in front of the ear, and notably smaller on the left side, the peroneus and tibialis anticus react more readily than on the right side. But this is not all. His neck muscles are normal, with one noticeable exception—the sternomastoids. The platysma here is also feeble, as it should. The condition of his face is noteworthy. The movement of the lips seems unimpaired. His tongue, however, presents a curious aspect of wasting on each side, yet it moves properly, and there is no impairment of the palate. His smile, especially on the right side, presents too little movement outside the corner of the mouth in proportion to the elevation of the eye-lobes, which are pushed down, and the most important weakness is in the muscles of the left side of the face. He cannot raise the eyebrows at all; the frontales are powerless. Closure of the eyes by the orbiculares is also weaker than normal; their contraction is easily overcome, and more easily on the left side than on the right. Conclusive evidence that it is of considerable significance, and so are the weakness of the frontales, and the atrophy of the sternomastoids. All these features stamp this case as a primary myopathy, and yet it differs from all recognized forms in the purely distal distribution of the affection in the limbs, and the normal state of the muscles near the trunk. In other forms the preponderant weakness and wasting are in the muscles moving the proximal parts of the shoulder girdle and the hip, of the elbow and the knee. In the upper limb this is very conspicuous. In the leg the affection of the calf muscles is usually attended with weakness of the fl exors of the ankle joint, even in the early stage the loss of power in the thigh muscles and those of the hip is greater, and as the disease advances the preponderance of their atrophy becomes more and more manifest. Yet in this lat these muscles have got almost to a standstill in the hip joint, and there is no depression of the bones, but not from contraction of the calves but simple failure of the fl exors. The feet can be moved inwards and outwards, and the movement outwards by the peronei is a distinction from the fl exor atrophy of the primary type. The opposition of the great toe is believed not to be a true myopathy. Peculiar is the dis-

**Pathology and Treatment.**

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inherent in the embryonal tissue from which the muscular structures of the body arise—a defect variable in distribution, in character, and in tendency, but essentially the same in all. In a few cases trivial changes have been found in the nervous system; in most it has been absolutely normal, both nerves and central structures, even by the latest methods of research, and in cases of most prolonged duration and profound degree. This facility to detect the muscular changes is in any way the result of the trifling changes that have been found in a few cases, or the more considerable atrophy of the spinal nerve cells found in one case. When we remember the fact, which I pointed out in the last lecture, that abiotrophy is met with in the nervous system, in various structures, and of various course, it is strange that it is not more often met with as an acutage case. It is not surprising, moreover, that slight secondary changes should be met with in the spinal cord when there has been prolonged secondary spinal curvature, with all its effects on the circulation. Such trifling changes, met with in a few cases, irregular in seat and distribution, can have no real significance. Yet they have been thought to suggest that some day it may be discovered that the muscular condition depends, after all, on a morbid state of the spinal cord. This idea is relic of the old fancy, for it was nothing more, that there are in the spinal cord special trophic centres for the muscles apart from the anterior cornua. If this be a truth that should have died of inanition long ago; purely hypothetical, every definite fact is opposed to it.

The vitality of the muscles presents a strange duality. Perhap3 this characteristic maintenance as a focus of vital energy is a double relation may be seen, I think, elsewhere, although less clearly. The nutrition of the muscle depends on that of the nerves through which its function is carried forth. If the nerves slowly degenerate, so does the muscle; if rapidly, from descending irritation, the muscles undergo speedy complete degeneration. Yet the muscle has a life which we may call organic, belonging to it as a structural entity, in consequence of which it may undergo morbid changes, apart from the nervous system, and may fail to live on, though the nerves preserve an unimpaired vitality. This vitality, which is not more strongly called muscular abiotrophy, is not nutrition from defective vitality—and for brevity we call it myopathy.

The treatment of this condition is a narrow subject, yet not unimportant. Its importance is positive and negative: to know what we cannot do and what we can. Life itself is beyond the influence of medicinal agents. Some degenerative changes seem to be within control at least in some degree, but it is otherwise with the organic vitality tends to seem inherent. We can discern no clear evidence that they can be thus influenced; we can perceive no positive effect from the administration of medicines. Moreover, the mere cause of any degree of aui5ratory deterioration when such agents are omitted. Yet even vital energy is not altogether beyond indirect influence. There is one agency, but only one, which has a sure effect on the vital nutrition of the muscles, and that is their voluntary use, their stimulation through the nerves with which their nutritional integrity is so mysteriously bound up. That is the lesson taught by all true muscular growth, by the disciples of Sandow, as by every athlete. That application of the use to these diseases is all-important; it is seen alike whether it is enforced or disregarded.

Muscular exercise, adapted in degree to the muscular state, is the one agent which distinctly stunts the ebbing tide of life, and hinders the failure of muscular strength and muscular nutrition. Neither electricity nor massage can exert more than a trifling fraction of the influence of voluntary use. Without it no definite effect can be observed from their employment; with it they seem sometimes to do a little good, and upward massage to promote the circulation in the muscles and the renewal of the blood, which promise into rods or columns. Those who promise with assurance that which cannot be, always find too ready credence, as is often only found too late.

Notes and references.