A Lecture ON MYOPATHY AND SYRINGOMYELIA.

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PARALYSED AND EPILEPTIC.

BY SIR W. R. GOWERS, M.D., F.R.S.,
PHYSICIAN TO THE HOSPITAL.

GENTLEMEN,—You may wonder why I have linked together two diseases so diverse in nature as the subject of this lecture. It is because they were connected by a link, the effectiveness of which you will at once admit, and also its actual utility—the link of error. I will show you a case which is certainly one of syringomyelia, and was thought at first to be also the subject of muscular dystrophy. If you study your errors properly and do not yield to the natural temptation to push them hastily out of sight with your foot, you may learn much from them. But you will probably find it easier to give special attention to errors in diagnosis that are not your own.

You will not learn quite so much from them, but the habit, once acquired, will not be easily discarded. That which concerns one's self is naturally fixed more firmly in the mind and the memory.

You know that syringomyelia is a malady that presents many of the features which are produced by various diseases of the spinal cord. But of these two are the more common—loss of sensation of pain and often to temperature without loss to touch; and, secondly, muscular wasting. The case I will show you presents both these symptoms. The sensory loss is a symptom which has to be sought for. This patient was ignorant of its existence. It is very easy for any person to be unaware of that which is only shown by the absence of discomfort. These cases illustrate the fact that pain is a precious danger signal, by the injuries its loss permits. The muscular wasting is that which brought the patient into the hospital, and it happens to present peculiar characters which bear a resemblance to those presented by a man with true myopathy now in the ward, whom I will show you first.

The two maladies ought not to be confused, although they are both diseases of development. The one is defective structural formation in the spinal cord, whereby cavities are left in the process of its development or portions of embryonic neuroglial tissue fail to achieve their change into nerve elements, but remain as tracts of low consistence, which break down into cavities. These enlarge by distension and thus cause symptoms, or the residual tissue may increase by a slow process of growth. Myopathy, or muscular dystrophy, is a defect of muscular growth, not dependent on the nervous system, but inherent in the muscles, by which the fibres fail, sometimes early in life, sometimes later. The interstitial tissue grasps the nutritional influence and increases, but not enough to compensate for the defect in the fibres, unless fat forms among

them, when the bulk of the feeble muscles may be much increased. This is the case in the early variety, the pseudo-hypertrophic form, which you probably know best; in this the muscles vary in size, the calves being usually largest, the extensors of the knees often increase in size in the lower part, the infraspinati are large, the lower parts of the pectoralis and the latissimus dorsi are small, and often they seem almost absent. The face is free. This form affects chiefly males, females seldom; sometimes all the males of a family suffer, and all the females escape, but transmit the disease to their sons. The malady increases in degree and extent as its subjects grow up; it impairs the power of breathing until some pulmonary disease develops; or it may be some slight catarrh, which would be a trifle to a normal individual, ends the feeble life soon after adult age has been attained.

But there are other forms of myopathy which do not present the increased size of muscles. On the contrary, all or most lessen in size, though often not to the same degree as in spinal atrophy. A more abundant growth of the interstitial tissue takes place, but it does not become the seat of fat formation, partly perhaps because the change usually takes place after the period of growth is over. One of these forms, which is

Fig. 1.—T. R., man with facio-scapular myopathy, showing abnormal elevation of the scapulae.

Fig. 2.—The same, from behind.

particularly well marked, is termed the "facio-scapular" form, from the groups of muscles which are constantly involved. It is also called after Landouzy and Déjerine, its first describers. It is to this form that the case belongs which I am now going to show you (Figs. 1, 2, and 3). The malady sometimes begins in early life, but more frequently not until growth is nearly over, and it may go on increasing and becoming distinct in fresh parts even for thirty years. If you observe the face and test the closure of the eyelids by opposing it, the least force is sufficient to keep them apart. In a moment I will point out other indications of this form. This man is 44 years old. His son is here,

who is likewise suffering, for this form of myopathy develops so much later than the pseudo-hypertrophic variety, that the subjects survive to an age at which they can transmit the disease. In the pseudo-hypertrophy, life usually terminates too soon for this, and the ability to pass it on to another generation is unknown. This facio-scapular form is eminently transmissible, as is shown by the family history of this patient. Of ten children in his family he is the only survivor. Three died in very early life, and of the six others all

are known to have suffered—two females who died at 19 and 26, and four males who lived only to 21, 22, 35, and 37. Their mother also suffered, and from her they received it, and so did her own mother. On his father's side there is no indication of the malady.

In the neck the sterno-mastoids are small, but act well. The upper part of the trapezius, which extends between the occipital bone and the shoulder, seems gone on each side; the rest of these muscles is small but present. You know that this upper part is usually the last to be affected in spinal atrophy, the *ultimum moriens* Duchenne called it; but it suffers often alone in myopathy. The deltoids are good; the triceps small and weak, and each biceps is changed in shape, small in the upper and lower parts, but

Fig. 3.—The same, showing contraction of the biceps.

full in the middle, standing out as a lump when put into firm contraction (Fig. 3). The forearm and hand muscles seem normal. The pectorals are irregularly affected; only small parts remain of the lower half. The serrati are gone, and the angles of the scapulae project, from the tilt of the bones; the erector spinae is small. The supraspinatus seems normal, and has not the prominence of the infraspinatus. This appears to be distinctly enlarged, and like most other muscles is the same on each side.

Wasting in the abdomen seems chiefly in the lower part of the oblique muscles on each side. The rectus is strong, but when he attempts to sit up the abdominal wall on each side bulges out, as you see, in a remarkable shape. In the legs, the muscles are generally small, but none are much affected except the anterior tibials and extensors of the toes, which are much wasted and have little power, so that in each leg there is foot-drop; but the peronei are good. The knee-jerks and ankle-jerks are present. He can stand on either leg and walk, but tends to catch his toes against any elevation. Sensation is everywhere normal.

It is not easy to learn the history of his symptoms, although he is not an unintelligent man. The impairment

in the arms became worse about seven years ago, and his walking has suffered more recently. But he has been able to work at his trade until just now; it consisted in stuffing the cases of mattresses with hay, an occupation we should not think laborious, although he avers that it is very tiring, and doubtless it is to him. For seven or nine years he has been conscious of weakness and pain in the left shoulder and upper arm, and later of pain in the lumbar region. Attacks of influenza, twice with pneumonia, have seemed to add to his loss of power. The weakness in the feet has been conspicuous during the last year or two. The pains led to a former diagnosis of neuritis, but were probably caused by the strain on the fibrous tissues which the muscles usually support in part. It is probable that the gradual muscular failure dates from early adult life, but for long it was unnoticed,

Fig. 4.—The son, with facio-scapular muscular myopathy.

because the muscles that were affected could be supplemented by others, at any rate to a large extent. Moreover, the parts that are not wasted are rather large, and are perhaps the seat of some true hypertrophy. The weakness of the face was unknown to the patient until he came here. It involves no conscious disability.

The great variations in the age at which this facio-scapular form begins and also the rapidity of its progress are sufficiently shown by the ages at which the affected brothers and sisters died, which I have mentioned. Other recorded cases illustrate the same fact, and in most of the families some of those who suffer have married and transmitted the disease, and this has happened, as in the family before you, with both sexes.

Our patient has a daughter and a son, but the daughter is only 11, and is said to present no sign of the disease. The sister who died at 26 was married, and left a daughter, but her state is not known. I show you our patient's son (Fig. 4). The orbiculares palpebrarum are conspicuously weak; they cannot overcome the gentlest hindrance to the closure of the eyes. The orbicularis oris is also feeble. The sterno-mastoids are good. The trapezii are greatly wasted. You can even see the projection of the cervico-dorsal spines. The serrati also have little power; the scapulae project when the hands are put

forward. The pectorales are not markedly atrophied, but they seem weak. On the other hand, the infraspinati are abnormally large, as they are commonly in the pseudo-hypertrophic form. There is no distinct wasting of the leg muscles, but the strength of the flexors of the ankle is small. All reflex action, superficial and deep, is of normal type.

I now show you the other case, a woman who has certainly syringomyelia and was at first thought to be the subject of myopathy; but more careful observation only justifies the first. She is 31 years of age and married early, having two healthy children. Nothing can be learnt that suggests any family malady, tubercle or myopathy, and she has never had any indication of syphilis; yet her past history presents one peculiar feature, which is of considerable importance in reference to our present question. In early life she possessed unusual muscular power, such as is rare in women. She was fond of its exercise, especially in swimming and gymnastics. When about 16 she gave public exhibitions in each of these. She could walk thirty miles without fatigue, until her strength failed, five years ago. Her performances necessarily entailed constant practice, and the muscular exertion, long continued, led to widespread hypertrophy of the

Fig. 5.—Analgesia—vertical lines, broken where incomplete. Thermanaesthesia—oblique lines.

muscles, traces of which are still obvious, especially in the arms and calves.

That she is the subject of syringomyelia is evident from the extensive loss of cutaneous sensibility to pain, without loss, or with very limited and partial defect, of sensibility to touch. I show you the distribution of the loss to pain in this diagram (Fig. 5). Its degree is indicated by continuous lines where it is complete, and by broken lines where it is imperfect in degree. It is partial over the right shoulder and the upper half of the arm and on the right front of the thorax; on the middle of the abdomen it is complete, in an area narrow at the ensiform cartilage, broader below, ceasing above the pubes, where it is interrupted by a zone of normal sensibility, but it is renewed again by complete analgesia at the top of each thigh. It becomes partial at the middle of the thigh, and ceases a little above the knees. This loss passes to the back of the hips and outer part of the thighs, and it extends incompletely across the gluteal region, except in the distribution of the lowest sacral fibres, where sensibility is normal on each side of the anus. It passes, in a pyramidal form, up the lower half of the back. Temperature-sense is impaired only in the small regions marked by oblique lines; touch only in slight degree on the right thigh, marked by transverse lines. You will find those directions of lines generally advantageous for depicting the loss of the different forms of sensation—perpendicular for pain, transverse for touch,

oblique for temperature. It is distinctly convenient always to use the same system.

If you compare this diagram with one for the cutaneous distribution of the spinal segments, as shown by that of the nerve roots (which I may assume you know), you will see that there is very little correspondence. The chief correspondence is in the escape of the lowest sacral in the region on each side of the anus (these cavities usually cease at the lower extremity of the cord). Elsewhere little correspondence can be traced. The analgesia only occupies parts of the thoracic and lumbar segments. Yet the region affected is not irregular. Over the back it extends through a diminishing medial area, pyramidal in form, which ends upwards to a point about the ninth dorsal spine. In front a corresponding area is involved, which is continuous with the loss on the right side of the thorax. The loss on the abdomen is interrupted by the zone of normal sensibility, in the position of the twelfth dorsal segment, but larger, and the loss begins again in the higher part of the thigh in each leg and continues, to cease above the knee. Except on the thorax, there is considerable bilateral symmetry in the loss, although there is seldom correspondence with the spinal segments.

The explanation is that the cavities which chiefly interrupt the sensibility to pain are usually situated in front of the posterior horn, in the middle of the half-cord, and probably damage the path for pain, which seems to cross in the posterior commissure. They are often roughly symmetrical. The impressions conducted by the nerve roots seem to be soon rearranged in the cord for their upward conduction, and their interruption has thus an effect different in distribution from that produced by interruption of the nerve roots themselves. It differs also in nature, because touch escapes. Sometimes, however, a cavity occupies the posterior cornu, and may be so situated as to interrupt the root nerves after they have entered the cord. Then all forms of sensibility may be impaired, and the loss may correspond in distribution with one or more of the spinal segments. But the variations in the position of the cavities is so great that no generalization from the loss of sensation to the position of its cause seems yet to be possible.

Next to analgesia, muscular atrophy is the most common effect of syringomyelia. The cavities often occupy the region behind the anterior cornua, as I have just said. Their gradual enlargement may cause an invasion of the anterior horn with destruction of the nerve cells; the related anterior nerve fibres then slowly perish, and the muscles to which they are related. Thus a slow progressive muscular atrophy occurs very frequently in some parts, but the effect is not always slow. A cavity may increase rapidly; the erosion of the wall may expose an artery to influences which induce thrombosis, and sudden necrotic softening in the vicinity, or rupture. Then there is quick, even sudden, loss of power and acute degenerative changes in the electric irritability of the related muscles.

A sudden process evidently must have occurred in a cavity on the right side of the cord in this woman. She was not aware of any impairment of motor energy until it came, suddenly and gravely, five years ago. She was walking out with her child, and she fell, without cause. She could not rise again, but when helped up she did not fall, but managed, with care, to walk a short distance home. It is not easy to ascertain what had happened, but she could never again flex the right hip. The power of flexing the left hip is said to have failed slowly during the next year. Probably wasting came on at the same time in the higher part of the muscles in front of the thigh. Now both are weak and similar. The lower halves of the extensors of the knee are large and strong. When they are contracted they present a striking contrast to the upper parts, projecting just as does the middle part of the biceps in the man we first examined.

The extended legs cannot be raised from the couch on which she lies. We cannot see the flexors of the hips, the psoas and iliacus, or we should doubtless find them much wasted. I think that it is the condition of the thigh muscles, the extensors of the knees, with wasting in the upper parts and not in the lower, together with the large size of the calves, which suggested the presence of muscular dystrophy.

But another symptom seemed to confirm the opinion. If she sits on the floor, or kneels, she is unable to rise with-

out help. In the attempt she makes to get up, to achieve the upright posture, she adopts the same expedient that is so constantly seen in pseudo-hypertrophic paralysis, and hardly ever in any other condition. She puts her hands on the knees or on the lowest parts of the thighs. This is to aid the extension of the knees. The weight of the trunk rests on the hip-joints, at the upper end of the lever formed by the femur. The fulcrum is at the knee and the power, the extensor muscles, acts on the femur, between the weight and the fulcrum, the position of least advantage. By the transfer of much of the weight, by the hand, to a position near the fulcrum, the power is beyond it, and then acts to much greater advantage. The greater ease with which then the knees are extended, and the weight of the body raised, is the reason for the adoption of this device in pseudo-hypertrophic dystrophy. Some patients supplement this also by making the hand climb up the thigh and so aid the extension of the hip, but the patient has not enough defective power in the extensors of the hip to need this. I have often wondered that patients with other forms of loss of power in the extensors of the knee do not adopt the device. Probably it is only discovered by reason of the gradual onset of the extensor weakness in the muscular disease during the period of growth, when the expedient comes, as it were, naturally to the versatile muscles of the growing child. I have often looked for it in other cases, but I think I have never seen it until this case. It obviously seemed to support the idea that she was also the subject of muscular dystrophy, but it is not a symptom, as you see, on which any absolute weight can be placed.

You can now better understand why we decide that she is not suffering from muscular dystrophy, and yet we can perceive that the mistake was easy. The two diseases are not mutually exclusive, nor have they any mutual pre-disposition. If they occur together it would be simply as the consequence of what, in our ignorance, we call chance. Indeed, syringomyelia has been found, I believe, to co-exist with myopathy in 1 case out of more than 300 cases of the spinal affection which have been published, but the facts I have mentioned show how carefully such cases must be scrutinized, since in both diseases the muscles suffer.

In this woman the facts I have mentioned leave no doubt that syringomyelia exists, and also that a sudden lesion must have occurred in a cavity on the right side, extending its forward erosion into the anterior cornu, probably by thrombotic softening, with destruction of nerve cells, paralysing the flexors of the right hip and the upper part of the extensors of the knee. The corresponding grey matter on the left side of the cord, according to the patient's account, did not then undergo damage, but this followed, in the course of the next year, by a process of more gradual character. Where the destructive process by which the nerve cells perished was rapid or sudden, so far as a group of cells is concerned, the electrical condition in the muscle supplied would be the "reaction of degeneration." You remember what this is; faradic excitability, which depends only on the nerve fibres in the muscle, fails rapidly, but the muscular tissue responds in increased degree to the slowly-interrupted voltaic current, and the kathodal closure contraction ceases to be the first, and is replaced by the anodal or positive. The time which has elapsed since the destruction, and the fact that this is not complete, prevents any evidence from the faradic response. The normal fibres that remain are sufficient to give a contraction to this. But on carefully examining the muscle in the upper part of the thigh with the voltaic poles, we find that the first contraction, on slowly increasing the strength of the current, is at the anode, not at the kathode. This shows that in the muscle are many fibres of which the nerves have undergone degeneration, although the fibres of the muscle have not wholly perished, and they still present this feature. It shows that there has been an acute process in the nerve centre. It is a feature never found in myopathy, and compels us to ascribe the state of the muscles to that of the spinal cord.

Thus it is clear that the condition of the muscles in this patient is not the same as in the man whom we first examined. Although they are similar in partial enlargement, the variation has not the same nature or the same distribution. The smaller part presents indications that it

depends on the state of the spinal cord, disease of which is proved by the presence of the change in sensibility of the skin, of which the patient was unaware, and which corresponds in greatest intensity to the region in which the lesion must lie which causes the wasting of the muscles. The remarkable size of those that are not wasted is explained by the patient's history in the rather distant past. At the time when the growth of all structures is most active her muscular system underwent a long course of systematic exercise, very unusual in degree, of which the remains are still distinct.

It seems strange that a girl with this congenital defect in the spinal cord should have been disposed to undertake such remarkable feats of muscular power without any family inducement to cultivate agility and energy. But her defect in the spinal cord is always chiefly in the posterior and medial parts, in the sensory regions of the cord. The change in the motor region is the result of a later extension, and it rarely occurs in early life. We may, perhaps, conceive that the motor processes, which so dimly affect consciousness, have not the normal afferent check from the sensory processes which should influence them. Of this the subject is quite unaware. Restraint may be effective before sensibility is subjectively lessened; we are often determined by influences which do not enter our consciousness. Some years ago a rather distinguished student was working here who was a really great cyclist. He had won many of the best cycle races in the country. Soon afterwards he came to me about some symptoms he felt in his hands, and on examining him I found such extensive loss of sensibility to pain as could only be explained by syringomyelia. I had to advise him to give up all severe exertion, including the races to which he was devoted. We must remember that the central motor nervous system represents the muscles, and all prolonged muscular effort has an influence on the spinal cord similar to that which it has on the muscles, and this disturbance of the circulation, and accumulation of the products of fatigue, resulting from severe exertion, must be of a nature to increase any morbid processes, the extent and progressive tendency of which we cannot safely estimate. I do not know what has become of him. But the history of this woman seems to show that severe exertion is unwise for such a sufferer. Hence it is very important to be familiar with the early symptoms of the malady, and to discover its existence, so that the life of the patient may be wisely regulated. The fact that a sufferer can generally feel well a touch, although pain is perceived dimly or not at all, explains the common ignorance of the malady, in spite of the frequency with which its presence is indicated by spontaneous pain. From this the patient whose case we have been considering was entirely free. It is often also accompanied by a sense of coldness, without any objective indication of the temperature which is complained of. These subjective sensations seem due to the irritation of the conducting fibres within the cord which are in process of destruction, and the pain may cease when sensibility is lost.

But tactile sensation is often impaired. It is very little affected in this patient, only in the front of the right thigh. Sensibility to temperature is lessened in the regions indicated in the diagram by oblique lines. Often the loss is almost as extensive as that to pain. The sensitiveness to pressure is often also lost, and that of the deeper structures may be impaired as well as that of the skin, although often not in corresponding degree. The cavities may extend higher up, into the medulla and pons (syringo-bulbi this is termed), and impair some of the cranial nerves, especially the sensory part of the fifth. The dissociated analgesia of this affection may remind you of the common loss in tabes, in which the perception of pain is so commonly lost and that of touch so rarely. When the analgesia involves the deeper structures an arthropathy may occur in syringomyelia, very similar to that of tabes. But this is most common in syringomyelia in the arms and in tabes in the legs. Other collateral consequences of sensory defect are also found which I cannot now even mention. I have brought the case before you briefly to illustrate the diagnostic difficulty, and to impress on your mind the fact that we must always beware of the significance of a superficial similarity, and remember that the resemblance may be due to very different causes, and may almost cease, on careful scrutiny.

Remarks

ON

ANTISPASMODICS AND THE CURE OF SPASMS.

BY EUSTACE SMITH, M.D., F.R.C.P.,

SENIOR PHYSICIAN, EAST LONDON HOSPITAL FOR CHILDREN;
CONSULTING PHYSICIAN, VICTORIA PARK HOSPITAL FOR DISEASES OF THE CHEST.

For the relief of spasm and the many distressing sensations which may be grouped under that head many kinds of treatment may be adopted. In the case of general convulsions and some forms of local spasmodic contraction we may act upon the nervous system generally, and make use of means which reduce its sensitiveness to morbid impressions. For this purpose we turn to the narcotics—opium, chloral, belladonna, the bromides, etc. But these remedies should never be used indiscriminately or without due consideration of the nature of the attack and the presence of a removable cause. They may form the main treatment of the case, or may be employed with the object of reducing general nervous excitement while special means are being used for the relief of local distress.

In certain forms of local spasm we may make use of remedies which are known to have a special sedative action upon the disturbed part, such as grindelia in spasm of the respiratory tract; belladonna, chlorotone, and codeina in intestinal cramps; valerian and asafoetida in the mysterious functional nervous derangements which are massed together under the head of "hysteria."

Again, we may seek to produce a tonic effect upon the system as a whole by the use of strychnine and nuxvomica, ergot of rye, sulphate and valerianate of zinc, quinine, arsenic, the stronger preparations of iron, and drugs of similar action. These, however, are of little value during an actual paroxysm, but exert their influence gradually by reducing the general nervous irritability, so that the system becomes less intolerant of morbid stimuli, and ceases to respond with such petulant activity to disturbing influences. Their action in this respect is deliberate, and rarely of more than passing value, unless persevered with for weeks or even months at a stretch.

The above may be said to include the special classes of drugs which are generally understood by the term "antispasmodic"; but they do not embrace the whole subject of the cure of spasm. The best and only satisfactory kind of antispasmodic treatment is that which strikes at the root of the evil and suppresses the irritant to which the disturbance is due. In many persons, especially in children and young women, the source of the ill is often a deranged condition of the digestive organs, and when this is remedied the spasm ceases. General and partial convulsions, tetany, and many kinds of tonic and clonic spasm, both general and local, may be induced directly by peripheral and especially by gastro-intestinal irritation. Children, with their immature and impressionable nervous systems, are curiously amenable to these influences, and in some such subjects any form of irritation seated at any part of the body may be capable by reflex mechanism of setting up automatic and irresistible contraction in voluntary or involuntary muscles. The tendency to such morbid response to stimuli is greatly enhanced by a lowered state of the general health. In the case of tetany, for example, the disorder rarely attacks a healthy sturdy subject, but is met with almost invariably in children whose nutrition is temporarily at fault, either from natural delicacy or domestic mismanagement. The spasmodic contractions of this complaint seem to be the consequence, more or less direct, of toxic absorption from the alimentary canal. A poison is generated which shows a special preference for the motor fibres and ganglion cells of the anterior horn of the spinal cord; and, in cases of long standing, atrophy and sclerosis have been noted in these cells, combined with some thinning of fibres. More than this, however, is required to explain the mechanism of tetany, for autointoxication is common in children of all ages, while tetany is only an occasional consequence of gastric dilatation and decomposition of food. It seems