CLINICAL LECTURES

ON

DISEASES OF THE NERVOUS SYSTEM.

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VII.—ON BULBO-SPINAL ATROPHIC PARALYSIS.

GENTLEMEN,—I beg to direct your attention to a case which at present occupies one of my beds in the hospital, and which presents a clinical picture of disease, to which the term bulbo-spinal atrophic paralysis may appropriately be given. This condition is worthy of consideration, not only on account of its comparative rarity, its occurrence in the female sex, and at an early age, but because it supplies a text for the discussion of many intrinsic features of interest connected with it.

Jane C., aged 28, seamstress, tells us that there is no history of nervous disorder in her family, and that she herself has always been in the most robust health till about a year ago. About this time, and without apparent cause, a slight hesitation in her speech was observed; but this was at first so trifling, that no notice was taken of it. This peculiarity in talking, however, very gradually increased, and, a month or so later, in addition, weakness and awkwardness of her hands and fingers were experienced, and clumsiness in performing accustomed acts. This also very slowly progressed, and, shortly afterwards, the same condition appeared in the lower extremities. All these symptoms, which began very insidiously, have continued to increase up to the present time, the general health of the patient remaining all the while in an excellent condition.

On personal examination (January 3rd, 1884), the patient is seen to be stout and ruddy in appearance, in good spirits, and in robust general health, and all the organs and functions are healthy, except those of the nervous and muscular systems about to be indicated. She is a well educated and highly intelligent person, and there is no trace of any mental deficiency; there is no pain in the head or other cerebral abnormalities, and all the special senses are intact. Although the patient is naturally of a cheerful disposition, the expression of her face is peculiar, having a sad and almost comically dismal appearance. The orifice of the mouth is enlarged, and remains partly open, with its angles drawn downwards. The lower part of the face is flat and expressionless, and its natural furrows are smoothed out. The movements of the mouth are imperfect, and it can only with difficulty be closed; and the patient cannot purse the lips, whistle, spit, or perform those labial acts which she formerly accomplished. The tongue is seen, apparently of ordinary size, lying at the floor of the mouth. All its movements are very sluggish and imperfect, and are accompanied by a slight tremor. It can with difficulty be protruded beyond the teeth, or raised upwards at its extremity. Speech is very indistinct, so as at times to be almost incomprehensible, and is in character exactly like that of one who tries to talk in keeping the tongue fixed to the floor of the mouth, thus making it an impossibility to pronounce the palatal and dental consonants. In addition, owing to the deficient action of the palate and lips, the voice has a nasal character, and the labial consonants are imperfectly uttered. Finally, it will be observed that the voice itself is weak and low, and cannot be raised to a loud tone. The patient next tells us that she has some difficulty in swallowing, especially fluids, which not only run out of her mouth, but sometimes through her nostrils. The saliva has a tendency to accumulate; it becomes viscid, and dribbles from the angles of the mouth, and you see that she has a handkerchief constantly in her hand to wipe the moisture away. On looking at the fauces in this case, there are no visible signs of paralysis of the uvula or palate; and when these are tickled, the reflex actions appear to be active. Neither is there any determinable paresis of the lower jaw, the movements of which seem to be as in health. The movements of the eyeballs, the pupils, the sight, and the ophthalmoscopic appearance of the eyes, are normal. There are no abnormalities of the respiratory or circulatory systems.

In addition to these conditions, we find that there are more extensive morbid peculiarities. The movements of the neck are slow and deliberate, and it has the appearance of being stiff; and when the patient is suddenly addressed, although her eyes at once follow the speaker, her head follows more slowly. There is, however, no real rigidity of the muscles, and the want of activity is obviously due to their paresis. Although very soft and flabby, there is no appearance of

special wasting of the muscles of the neck, trunk, or upper arms; but, as you observe, the patient is stout, and there is a considerable deposition of subcutaneous fat, which may conceal a moderate attenuation of their substance. The muscles, however, of the forearms and hands are distinctly atrophied, especially on the extensor aspect of the former, and in the thenar and hypothenar regions of the latter. The hands are thin, flat, hollowed out, and their natural eminences have disappeared, so that the bones stand prominently displayed. Moreover, there is a marked deformity of the hands and fingers. The wrists are permanently somewhat flexed, the near phalanges are extended, and the two distal phalanges are in a state of fixed flexion. In this way the hands present a clawed appearance, or the "*main en griffe*" of the French; and this, as you know, is mainly owing to paralysis of the interossei muscles, with some retained activity of the flexors and extensors of the forearms. Accompanying these appearances is a corresponding weakness of movement, and this bears a relation to the obvious wasting of the muscular substance. In the trunk and upper arms the various actions can be performed, but slowly, feebly, and with obvious effort and fatigue. The movements of the hand and fingers are especially imperfect, and both flexion and extension of the former are very weak and limited. The actions of the thumb and fingers, in every direction, are greatly curtailed, and the patient cannot sew, arrange her dress, feed herself, or in fact perform, except very awkwardly, any of the usual manipulations of ordinary life.

The lower limbs have the appearance of being well developed, but the large development of subcutaneous fat would conceal any loss of muscular substance, unless it was very considerable. The patient can walk, but does so with great difficulty, and the gait is slow, hesitating, and unsteady, but with no evidence of inco-ordination or ataxia. The steps are short, and the feet are shuffled along the ground. The muscles of the trunk and upper limbs are soft and flaccid, while those of the lower extremities are somewhat stiff and rigid; but this condition does not exist to a great extent.

On applying electrical tests, it is seen that the stimulation of all the nerve-trunks of the body by the *faradic* current is practically followed by normal muscular contractions; the only exception being that the movements of the hands and fingers are less vigorous than natural to a standard strength, and that in the lower extremities, for proportionate effects to be produced, stronger currents are necessary. The same results follow when this stimulus is applied directly to the muscles themselves; only that, in those obviously atrophied, the vigour of the contraction is diminished. Nowhere, however, is there complete loss of reaction; and the contraction is in proportion to the bulk of the muscular substance; and even in the most wasted muscles, if the current be augmented, an action can be developed. The *galvanic* current applied to the various nerve-trunks causes analogous effects to faradism, and without abnormal qualitative change. The muscles, however, when directly stimulated by this agent, for the most part demonstrate alteration in their structure, as evidenced by the occurrence of the partial form of the reaction of degeneration. In the lower portion of the face it requires a current from eight elements, when applied to the nerve-trunk, to produce a minimum muscular contraction, while one from six elements causes a well-marked action on the muscles themselves. The character of the contraction also is altered, it being slower, more prolonged, and tonic than natural. Finally, the polar formula is modified, the anodal closing being as powerful as the cathodal closing contraction ($ACC = KCC$). The same general features exist to a greater or less extent in most of the muscles of the body, and more especially in those of the fore-arms and hands. In the last, a comparatively stronger current is required to produce proportionate results, and the anodal closing is more powerful than the cathodal closing contraction ($ACC > KCC$). Very similar conditions exist in the lower extremities, still more powerful currents being requisite to induce action in the muscles, and notably in the extensors of the feet and toes, where a strength derived from forty elements, indicating 12 M.A. on the galvanometer, produced only feeble contractions ($ACC = KCC$). Investigations proved that the resistance of the body presented no abnormal conditions.

The reflex actions of the skin throughout are seen to be normally developed. The knee-jerk phenomena and the mechanical irritability of the muscles are everywhere considerably exaggerated. Ankle-clonus is elicited on both sides, and fibrillary contractions, both spontaneous and artificially produced, can be demonstrated. All these motor abnormalities may be said to be symmetrical on the two sides, but they are most marked throughout on the left. The sensibility of the skin is everywhere natural. The skin of the arms and hands is somewhat cold and cyanotic, but not to any great extent.

COMMENTARY.—This case comprises a typical combination of sym-

ptoms which are usually differentiated into two distinct groups, and which are described as separate diseases, under the names of bulbar paralysis, and progressive muscular atrophy. To Duchenne, medical science is indebted for his researches on, and a definition of, both these affections; and that observer believed that each of these constituted an independent malady, which was characterised by clinical and pathological features peculiar to itself. He further maintained that, in those rare cases in which the two conditions occurred in the same individual, it was a mere coincidence, the one disease being an accidental complication of the other. This view has been adopted by his followers. In the light of recent advances in neurological science, it will be of advantage to discuss this question.

From a clinical standpoint, it is asserted that progressive muscular atrophy is a disease characterised by atrophy without paralysis; whereas bulbar, or, as it is otherwise termed, glosso-labio-laryngeal paralysis, is paralysis without atrophy. In addition to this, further distinctions have been drawn between these two conditions, such as their etiology, the age at which they occur, with other minor points, all of which, however, are so uncertain and indefinite as to constitute no definition between them. The differential character on which main stress has been laid, is the question of atrophy or paralysis, as already stated; the accuracy of this assertion may, however, be disputed.

In progressive muscular atrophy, along with the wasting there is undoubted weakness, which goes hand in hand with the disappearance of the muscular substance; and, in chronic cases, owing to the secondary degeneration of the entire pyramidal tract and peripheral motor nerves, it is probable that the paresis considerably exceeds that due to actual loss of muscle.

In bulbar paralysis, on the other hand, while granting that there is paresis, it cannot be admitted that there is no muscular degeneration. From anatomical considerations subsequently to be considered, this tissue-alteration may not reach so advanced a stage as it sometimes does in progressive muscular atrophy of the limbs, but that wasting of the fibres actually takes place is evidenced not only by cases on record in which the tongue was visibly atrophied, but by the revelations of the microscope, and the more recent discoveries in electricity as a diagnostic agent. It is also to be observed that, owing to the condition of the mucous membrane, and the general situation of the parts near the face and mouth, added to the comparative smallness of the muscles involved, anything but very gross diminution in their volume is difficult to demonstrate by inspection alone. When, however, as in the case before us, electrical tests are employed during life to tissues which present no visible alterations in size, or when histological investigations are made after death, there is conclusive proof that changes exist in the structure of the muscular substance. The normal responses when the nerve-trunks are irritated, and when the muscles are directly stimulated with the faradic current; the increased contraction when galvanism is applied to the muscular fibre; the long drawn-out and tonic spasm produced; and the altered polar reactions, all give abundant evidence that the muscle has undergone those structural changes characteristic of degeneration. Granting, then, that in bulbar paralysis muscular atrophy may not be visibly apparent, and that it does not reach the same degree as in the so-called progressive muscular atrophy, it is maintained that it cannot be asserted to be altogether absent, and, therefore, does not constitute a distinction between the two diseases. On clinical grounds, then, such a distinction is not tenable.

Duchenne and his followers further believed that their views on this subject were supported by pathological facts. They admitted that bulbar paralysis was due to a central lesion, but they maintained that progressive muscular atrophy was essentially a morbid condition originating in the muscles, and that any nervous alterations complicating the situation were of a secondary nature. The histological anatomy of the spinal cord was, however, not so developed in the days of Duchenne as it is at present; and, owing mainly to the researches of Lockhart Clarke, Hayem, Charcot, Joffroy, Vulpien, and Erb, the myopathic and neuropathic discussion concerning the nature of progressive muscular atrophy may now be said to be practically at an end. In this disease, it has been for long admitted that structural alterations are present in the muscles, peripheral nerves, spinal roots, and anterior cornua of the cord. The question at issue has been, in which of these localities did the primary lesion appear? Numerous cases have now been recorded by the authors mentioned, in which the entire history and progress of the disease, the nature of its histological appearances, the comparison between it and other affections of an allied kind, force us to the irresistible conclusion that the essential degeneration is that of the motor cells of the anterior cornua of the cord, and that the other morbid nervous and muscular alterations are secondary consequences of that condition. More than one instance has been reported where there existed well marked degeneration of the motor

cells, accompanied by muscular wasting, without discoverable change in the spinal roots or peripheral nerves; this not only disproves the myopathic theory, but suggests that these central ganglia preside over the nutrition of the muscle, independently of that of the nerve, or of the voluntary impulses derived from the brain, both of which may remain intact. This general invasion of the multipolar cells would also seem to best explain the imperfect and irregular distribution of the wasting, the integrity of certain fibres of the muscle, the corresponding responses to the electrical stimulus, and the absence of true paralysis or loss of sensibility. Further development of this spinal localisation has been attempted by Remak, who has endeavoured to show that the various groups of muscles which act in physiological harmony are represented in the spinal cord by special arrangements of cells, which can be anatomically distinguished from one another. A gradual sclerosis of these centres would explain the characteristic distribution of the wasting in progressive muscular atrophy. Secondary changes may result from a lesion in any part of the cord; and doubtless the slight rigidity of the muscles of the legs, and the exaggerated tendon-phenomena observed in this case, are due to such alterations extending to the pyramidal tracts.

The morbid central lesion associated with bulbar, or glosso-labio-laryngeal paralysis, is generally admitted to consist of degeneration of the nuclei of the motor nerves of the medulla oblongata. In this disease it is the hypoglossal, the facial, the motor branch of the fifth, the glossopharyngeal, the pneumogastric, and the spinal accessory nerves, which are involved.

Now anatomy teaches us that the cells forming the nuclei of these nerves are embedded in that portion of the grey matter of the medulla which is a direct continuation of the anterior cornua of the spinal cord. These, when traced upwards into the medulla, are seen to be divided by the decussating fibres of the pyramidal tract into two portions, the anterior of which contains the nuclei of the glosso-pharyngeal, spinal accessory, pneumogastric, and motor branch of the fifth; and the posterior, the nuclei of the hypoglossal, facial, and motor oculi nerves. In short, these masses of grey substance are simply a prolongation of the anterior cornua, and their nuclei in the medulla oblongata are analogous to the motor cells in the cord, the former presiding over the nutrition of the muscles of the tongue, lips, and larynx, and the latter over those of the trunk and extremities. Disease, therefore, of the one resembles, in essential nature, that of the other, the symptoms of the lesion only depending upon its locality and extent. The anatomical distribution and the clinical phenomena of the malady are in perfect harmony with each other.

Immediately above the decussation of the pyramids in the bulb is found the nucleus of the hypoglossal only; at a higher level, from before backwards, are the hypoglossal, and the mixed and sensory nerves. Above this point the nuclei of the facial, motor oculi, and acoustic nerves are situated. As destructive disease spreads, it thus involves the centres in special directions, and in a certain order; and, as a matter of fact, it is found that the hypoglossal nerve is, as a rule, the first and most seriously affected, and next in order is the facial. The spinal accessory, pneumogastric, glossopharyngeal, and motor branch of the fifth, show distinct, but less profound changes. The motor oculi, the sensory nerves, along with the entire white substance of the medulla, remain intact.

To attempt thus to found a distinction between bulbar paralysis and progressive muscular atrophy does not appear to be more successful on pathological than it is upon clinical grounds. The two conditions are, in short, identical, and are due to analogous neuropathic changes in the ganglion motor cells, and are alike in anatomy, symptoms, and progress. In neither is there any distinctive histological alteration, the modification of the centres, nerves, and muscles, being the same in both, consisting of a pigmentary granular fatty degeneration, with atrophy of substance. In the majority of cases met with in practice, the disease is so far limited that, on the one hand, symptoms of affection of the spinal cord only are produced, and, on the other, those of the medulla, which, for convenience, may be named respectively progressive muscular atrophy and bulbar paralysis. This difference in terminology may be given rather to retain the ancient nomenclature than to indicate any distinct clinical or pathological characteristics.

Less frequently, but by no means uncommonly, progressive muscular atrophy spreads upwards to the medulla, and induces all the typical phenomena of bulbar paralysis. The disorder when thus complete may then be termed bulbo-spinal atrophic paralysis. Still more rarely, as in the case under consideration, the morbid change begins in the bulb; and subsequently extends downwards, so as to involve the cord in the same physiological tract, and thus produce a similar clinical tableau. Of these, M. Hallopeau has described several instances in his

thesis of 1875. Why the morbid process should at one time spread upwards, and at another downwards, the present state of science does not permit us to say.

There are other interesting features connected with this patient well worthy of attention; a consideration of which must be postponed to a future occasion.

ABSTRACT OF THE HARVEIAN LECTURES

PROGNOSIS IN HEART-DISEASE.

Delivered before the Harveian Society of London, November, 1883.

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LECTURE III.—concluded.—MITRAL STENOSIS.

THIS is a common form of valvular disease, and is generally admitted to be the most serious. Of the one hundred and fifty-one cases taken by Dr. Phillips from the St. Mary's Hospital *post mortem* records, it furnished fifty-three, of which thirty-eight were females, and only fifteen males; this predominance in the female sex being constant. The average age at death among these cases is higher than the frequency of a fatal termination at a very early period of life would have led me to expect, being 33 for males and 38 for females.

A suggestive inquiry is, why mitral stenosis should be so serious, and especially why it should be attended with greater danger to life than mitral incompetence. One reason is, that the effects of obstruction here are not so easily neutralised as are the effects of regurgitation. The high pressure maintained in the pulmonary circulation and in the left auricle, by hypertrophy of the right ventricle, will antagonise incompetence of the valve in two ways—by resisting the reflux during systole, and by more rapid filling of the ventricle during diastole. In stenosis, this high pressure can only be of service by increasing the rapidity of the passage of blood through the narrowed orifice during diastole; and, as diastole only lasts for a certain time, if the contraction be extreme, the ventricle cannot be properly filled before the systole is again due. When such is the case, the compensation is inadequate. On the other hand, since the constriction may reach a degree which, in the absence of experience, would seem quite incompatible with life, and since the subjects of such extensive change must have lived through all the intermediate degrees, it can scarcely be simply that obstruction, as such, is specially dangerous. Probably the explanation of the greater danger of obstruction, as compared with regurgitation, lies in the fact that, when once adhesion between the flaps of the valves has set in, it tends to go on, the friction and strain keeping up chronic inflammation, which glues the valves further together.

There are clinical features about this form of valvular disease which are worthy of note. The patient, up to a very advanced stage, has not the look of heart-disease, is neither pallid, nor dusky, nor anxious-looking, but often has a good bright colour and cheerful expression. Not unfrequently, up to the moment when, from some cause or other, serious symptoms have set in, he or she is unconscious of embarrassment of the circulation, and capable of ordinary work. On the other hand, such patients are sometimes liable to congestion of the lungs and hæmoptysis after exertion. It is, moreover, the valvular affection which, independently of inflammation or ulceration, most frequently gives rise to arterial embolism. This is not the result of detachment of fragments from the valves, but is due to the formation of fibrinous coagula between the muscoli pectinati of the left auricle, or in the appendix, or, less frequently, between the columnæ carneæ of the ventricle, which are shaken out of their beds by exertion or excitement, and carried into the circulation. Another condition more frequently met with as a consequence of mitral stenosis, according to my experience, than in other valvular affections, is great enlargement of the liver, with true pulsation of this organ; and it is not uncommon to find fluid in the peritoneal cavity before there is œdema of the feet and legs, or the œdema will disappear with rest in bed, while ascites remains for a time; whereas cardiac dropsy, as is well known, usually begins in the connective tissue.

Finally, the pulse of mitral stenosis is interesting. It is, according to my experience, almost always regular until the heart is obviously failing, unless the obstruction be complicated by regurgitation, or by valvular affection of the right side of the heart; and, when irregu-

larity comes on, it is usually at first the effect of some of the beats of the heart failing to reach the pulse—no doubt, from inadequate filling of the ventricle. The action of the heart may thus continue to be regular when the pulse is irregular. In some rare instances, there is only one beat of the pulse for every other beat of the heart; and, on listening to the heart, there will be no aortic second sound with the alternate systoles; the rhythm, as expressed by the sounds, being one-two-one, one-two-one. But perhaps the most interesting point about the pulse is that the artery, while rather small, remains full between the beats, and has the character of high tension. This would scarcely have been anticipated, and is not easily explained.

Coming, now, to the indications by which the degree of obstruction may be inferred, we cannot, as has been stated in the first lecture, rely upon the amount of hypertrophy and dilatation present. There is no dilating influence brought to bear upon the left ventricle, and no demand for increased exercise of systolic energy. This ventricle, therefore, as a rule, undergoes little change; but it is conceivable that, in an early stage of contraction, the increased pressure in the auricle might distend the ventricle during its diastole, thus giving rise to a certain degree of dilatation, which will call for some hypertrophy; and, perhaps, it is in this way that an increase in the capacity and thickness of the left ventricle occasionally met with, especially in young subjects, is produced. While, however, the left ventricle undergoes little change, all the other chambers of the heart are affected. The auricle is dilated, often to an extraordinary degree, and the walls may either be hypertrophied, or stretched and thinned. But it is upon the right ventricle that the greatest strain falls, and it becomes greatly hypertrophied with or without dilatation. Unfortunately, there is no such correspondence between the hypertrophy and the obstruction as can be relied upon for the purpose of prognosis. It would appear as if the refusal of the left ventricle to enlarge, set limits to the enlargement of the right ventricle.

We are thus compelled to fall back upon other indications, and these are, according to my experience, afforded by changes in the sounds of the heart. I have, indeed, been led to divide mitral stenosis into three stages, distinguishable chiefly by means of modifications of the sounds. The diagnostic sign of obstruction at the mitral orifice upon which most stress is usually laid, is the well known presystolic murmur of vibratory character, heard over a limited area to the inner side of the apex, and often accompanied by a thrill perceptible to the hand. When present, it is perfectly characteristic, though not absolutely pathognomonic, and probably it exists for a time in all cases; but, in a very large proportion of the occasions on which the diagnosis of this condition of the valve has to be made, it is absent. The following, then, is the course of the modifications which I have traced in numerous instances, verified by *post mortem* examination.

In the first stage, there are presystolic murmur and thrill, with more or less accentuation of the pulmonary second sound, and, perhaps, reduplication of the second sound; its distinguishing mark is the presence of the second sound at, and to the left of, the apex. An apex-murmur, followed by two sounds, can scarcely be taken for anything but presystolic, and accordingly the diagnosis is extremely easy. At this stage, according to my experience, there are no symptoms.

The second stage is marked by the disappearance of the second sound from the apex. At the same time, the first sound, as heard here, has gradually undergone a complete change of character, and has become short, sharp, and loud, so as to be extremely like, in fact, an exaggerated second sound. Now, mistakes in diagnosis are easily made; the presystolic murmur, ending in a short sharp first sound, is taken to be a systolic murmur, followed by a loud second sound, and the disease is supposed to be mitral insufficiency. Thus the most serious of the valvular affections, at a period when symptoms are impending, is taken for the least serious. There will, of course, be accentuation of the pulmonary second sound, with or without reduplication. It would, perhaps, scarcely be worth while attempting any explanation of the modifications specified, were it not that a fact will be borne in mind, for the sake of a reason, which might otherwise be forgotten. First, then, it may be asked, why does the first sound become short and sharp? No very definite answer can be given. It cannot be the direct result of the state of the valve; and seeing that, in hypertrophy, we have a modification of the first sound of a reverse kind, while in dilatation and in palpitation the first sound approximates in character that of mitral stenosis, it would seem that the peculiar modification has its origin in the walls of the ventricle; and it may be conjectured that the imperfect distension of the cavity, and the consequent absence of resistance at the beginning of the systole, is the determining cause. This sharp first sound has been spoken of as the remains of the presystolic murmur which leads up to it; but such an explanation is inadequate.