A clinical lecture on bulbar paralysis and its counterfeits / by G. A. Gibson.

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A CLINICAL LECTURE
ON
BULBAR PARALYSIS AND ITS COUNTERFEITS.

Delivered in the Royal Infirmary, Edinburgh.

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IN the realm of Nature the most whimsical resemblances often occur between different plants as well as different animals absolutely unconnected with each other save by contiguity. Many examples of such mimicry are to be found in the works of our naturalists. The similarity that exists to all outward seeming between the humming-bird moth and the humming bird has been described in the delightful work of Bates, while the likeness between the gentle oriole and the savage friar bird may be found in the no less charming work of Wallace. The unconscious efforts of the mimics in such remarkable instances may well entitle them to the phrase "masterly imitators" employed in another connection by Isaac D'Israeli. They are so far from rare that numberless instances might be culled from the works of natural historians past and present, and they have been very thoroughly studied by Poulton. Among those aspects of the ever-changing web of life which come under our clinical notice, and which may well be termed the natural history of disease, there are also many curious resemblances. The simulation of diseases, which consist in structural alterations, by others in which the symptoms, so far as our means of investigation as yet permit us to determine, are those of purely functional disturbance, is a well-known and fertile source of difficulty in diagnosis. The converse of this is not so well recognised, yet it is certain that many diseases of a real organic kind may present to the superficial observer a wonderful counterfeit of certain purely dynamic affections, and Buzzard conferred a real benefit upon medical science when he delivered his Presidential Address before the Neurological Society on the Simulation of Hysteria by Organic Disease of the Nervous System.

We are fortunate to-day in being able to consider two most interesting cases of nervous disease which afford at once considerable superficial resemblance and wide intrinsic divergence; they are therefore excellent illustrations of the subject to which reference has been made. Let me without further preface show you the patients and demonstrate their symptoms. The first case is that of a married woman, aged 57, sent to me from Shetland on account of difficulty in speaking and in swallowing. There appear to be no hereditary tendencies to disease, but there are certain difficulties in obtaining a valid history on account of the tragic results that have accrued from the dangers to which her family has always been exposed. Her father was lost at sea, and her mother died in extreme old age. Two of her brothers were, like their father, drowned. The two remaining enjoyed perfect health, and she never had a sister. Her husband shared the fate of her father and her deceased brothers. She herself has had seven children, six of whom are perfectly well; one died when a few weeks old. Her surroundings are satisfactory in all respects. Her house has two rooms, and it is dry and warm. Her occupation and that of her daughters is to look after the house and a small farm while the sons are at sea. As regards food and drink there seems to have been no room for improvement, and it may more particularly be added that she has not been in the habit of taking much tea. There have been no previous diseases of importance excepting measles as a child, but there has been a tendency to catarrh throughout most of her life. The present illness began exactly one year ago, and its first symptoms were noticed by her friends, who began to have some difficulty in understanding what she said. When her attention was directed to this point, she was conscious, not merely of some indistinctness in her utterance, but of some change in the tone of her voice. Shortly afterwards there was some trouble in swallowing both solids and fluids, and one of the catarrhal attacks to which she is subject having made its appearance about the time referred to, she found that her cough had undergone considerable change. These different symptoms became gradually more marked, and as no improvement appeared to follow the treatment of her doctor, he recommended her to come to the infirmary.

The patient (Fig. 1) is very thin. Her expression may be described as one of apathetic melancholy. The cheeks are sunken, the mouth is large, with thin relaxed lips which have a tendency to fall in when the face is at rest. There is external strabismus, but this is of an alternating character. On asking the patient to open the mouth, the tongue is seen to be small and tremulous. It manifests a lack of symmetry, its right half being long and narrow, while the left is short and broad. As it lies in the floor of the mouth it shows beautiful fibrillary movements. There is great difficulty in protruding it, and on testing the strength by holding the tongue between the thumbs and index fingers it is found that both sides are feeble. The palate is relaxed, and moves very slightly with the respiratory movements. Swallowing is a matter of considerable difficulty. The patient is able to take a little solid food, which is swallowed slowly and in small quantities. Fluids very frequently find their way into the nasal passages. The voice is of low tone and hoarse character; it is also absolutely monotonous. Another important point is that when the patient coughs

the sound produced, as you hear for yourselves, is not that of an ordinary cough; it is very similar to the cough of the herbivora, and has therefore been termed "bovine" by my friend and colleague, Dr. Wyllie. The explanation of the modified voice and altered cough is easily found on examining the larynx. On phonation the vocal cords are seen to have some diminished movements of abduction, adduction not being obviously interfered with. When the patient coughs the false cords do not close over the space which they ought to bridge in this act, hence the resemblance of the patient's cough to that of the ox, in which animal there are no false cords. Her utterance is extremely indistinct and guttural; she is unable to pronounce palatals, dentals, and labials in any degree.

On investigating the sensory nervous system it is found that there is absolutely no impairment of ordinary sensibility in any part of the body. All ordinary stimuli, from the slightest to the most severe, elicit immediate and correct response. In the same way the muscular sense is intact. The acuity of sight and the fields of vision are almost perfect, the fundus is healthy, the discs well coloured, and the vessels of ordinary size. There is not the slightest trace of neuritis or other fundal alteration. As regards the movements of the eyeball, in the investigation of which I am indebted for assistance to my friend and colleague Dr. Sym, there is divergence of either eye according to whichever happens to be the fixing one at the time. The movements are quite free. There is no weakness of any muscle, and the divergence is what is to be seen in patients who have never had, or have early lost, the power of binocular fixation. Hearing, taste, and smell are acute. The muscular sense is absolutely unimpaired in any way.

Turning to the motor functions, the organic reflexes as regards the lower portion of the spinal cord are unaffected. Deglutition, as has been mentioned, is interfered with, and on tickling the fauces the soft palate does not give the usual response. It is, as has been described, relaxed, and moves very slightly with respiration. The breathing is of a somewhat shallow character, and increased frequency, being usually about 30 per minute. The pulse has a rate of between 80 and 90, but it is perfectly regular and shows no nervous disturbance. We may therefore conclude that, while the reflex act of deglutition is diminished and that concerned in the respiration is probably modified to a slight extent, there is no disturbance of the reflex mechanism concerned in the circulation. The cutaneous reflexes are all present, and seem to be within the limits of health. The tendon responses are also unimpaired. The knee-jerks are active, possibly even slightly exaggerated, but there is no increase in the responses as regards the upper extremity, and there is no clonus in any part. Voluntary movements are diminished, not merely in the regions mostly affected, but throughout the whole muscular system. On inquiring closely into the condition of the muscles of the face it is found that the buccinators are weak, while whistling is an impossibility on account of inability to use the lips. The tongue movements have already been sufficiently described. As regards the movements of the extremities, it is found that all the muscles are feeble. The grasp of the right hand registers 30 with the dynamometer, while the left only reaches 18. The same diminution of muscular energy is found in the lower

extremities, although there appears to be no inequality between the strength of the two limbs. The muscles generally are flabby, and this condition is particularly observable in the neck and arms. The interossei are somewhat atrophied, especially those of the left hand. There is a considerable amount of fibrillation in the wasted muscles of the hands. Tested by means of electricity there are considerable differences in the reactions of the different accessible muscles. You are able to observe for yourselves that in response to the faradic current there is diminished response in the orbicularis oris and in the intrinsic muscles of the hands, with a ready contraction to galvanism, which appears about equally to the closing anode and cathode. The cerebral functions are quite unimpaired. The patient answers slowly when she is spoken to, but this is entirely due to her difficulty in speaking, and not to any deficiency in her understanding. Her attention and memory are good.

In this case the diagnosis cannot for a moment be matter of doubt. The wasting of the muscles, the fibrillary movements which they exhibit, and the altered electric reactions that you have seen afford ample evidence that the lower neurons are affected, while the distribution of the symptoms point unmistakably to an affection of the great nuclei in the pons and medulla or their efferent tracts. The patient, in a word, furnishes an excellent illustration of the disease first described by Duménil and Duchenne, commonly known as chronic bulbar paralysis, but termed also, in the more recent nosology of the nervous system, "polio-encephalitis inferior chronica." This is a somewhat cumbrous designation, but it is one that serves to distinguish it from the analogous lesions of the nuclei in the region of the Sylvian aqueduct, first noticed by Jolly and Hutchinson, and now very generally called "polio-encephalitis superior chronica." The patient, however, cannot be regarded as an instance of uncomplicated bulbar paralysis; she manifests symptoms showing, as is so common in such cases, that the morbid processes have involved other nerve centres. It is highly improbable that there is any structural change in the oculo-motor nuclei, since the external muscles of the eye have perfectly free movements, and there is no change in those of the iris and ciliary muscle. In short, there is no ophthalmoplegia. But it is very obvious that there is some progressive muscular atrophy. There is not only wasting of the muscles of the hands, but they also show fibrillary movements and altered electric reactions. The patient, then, has evidently some chronic poliomyelitis as well as chronic inferior polio-encephalitis.

The evidence clearly leads to the conclusion that degeneration has occurred in the hypoglossal nucleus and, probably to a less extent, in the combined nuclei of the glosso-pharyngeal and vagus nerves with the accessory nucleus of the latter, as well as in the facial nucleus. We know very well how variable is the distribution of the morbid changes found after death in this disease; one of the most striking proofs of this fact is to be found in a very interesting case described by Howard Tooth and Aldren Turner. In this instructive example of the disease the motor nucleus of the trigeminal nerve and the nuclei of the facial and hypoglossal nerves had undergone atrophic processes, while the glosso-pharyngeal, pneumogastric, and accessory nuclei were healthy.

Fig. 1.—Glosso-labio-pharyngo-laryngeal paralysis of bulbar origin.

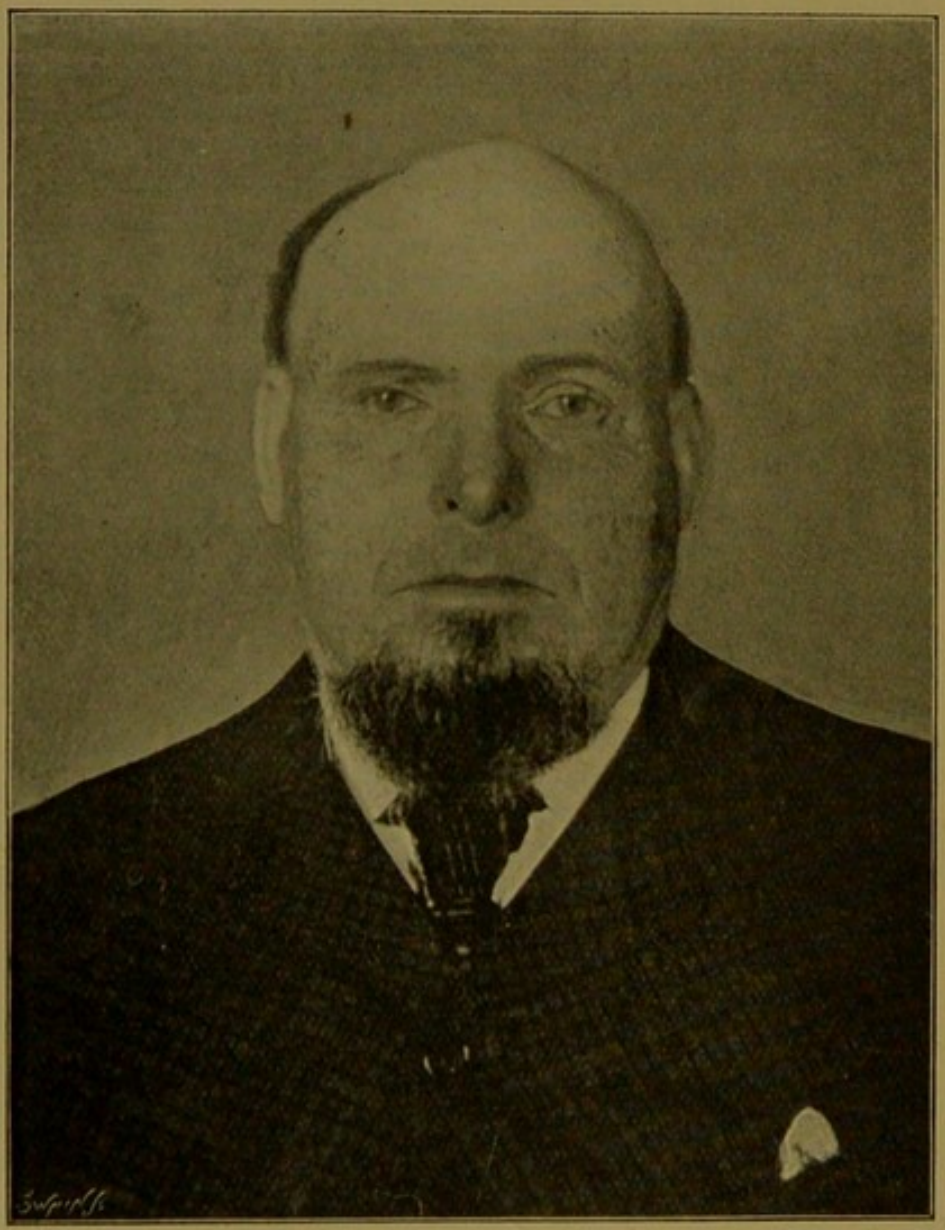

Fig. 2.—Glosso labio-pharyngo-laryngeal paralysis of cortical origin.

The second patient whose case we have to consider to-day is a man, aged 59, belonging to Moray, but occupied for many years in Edinburgh as a skinner. He complains of defects in his speech, stiffness in his back, and weakness of his legs. His father died of diabetes after a short illness, and his mother of heart disease, which was likewise of brief duration. His brothers and sisters are alive and well, as are also all the members of his own family. His general surroundings are in every respect perfectly comfortable, and his habits have throughout his life been eminently worthy of praise. His occupation, however, has not been such as to conduce to health, as his work exposed him to all kinds of weather, in which he had frequently to stand for hours with water up to his knees. He has, further, had extremely heavy weights to carry up and downstairs. With the exception of some attacks of bronchitis and a broken leg, he cannot remember having had any previous disease. The present illness began, about five years ago, with an attack of bronchial catarrh, for which he came to the medical waiting room and saw me. My attention was at once attracted by the monotonous character of his voice and the clumsiness of his utterance. Even at that early period of his disease there was some sluggishness in the movements of the lips and tongue, and on examination of the larynx it was obvious that there was some interference with the approximation of the vocal cords. The patient is quite certain that up to that time neither he himself nor his friends had noticed any alteration in his speech beyond a hoarse monotony which they attributed to cold. Shortly after this, however, they observed some thickness in his speech, which was more marked at night when he was tired, or if he tried to read aloud for any length of time. His fellow-workmen began about the same time to have some difficulty in understanding him, and they used to laugh at him on account of his thick speech, which they said might lay his conduct open to suspicion. At the same time, or very shortly afterwards, he began to find a difficulty in swallowing, which rendered him liable to choke, and to allow food to pass up into the nose. A couple of years afterwards some weakness in the legs was first observed. It gradually increased, yet the patient was able for a time to continue his work. When, however, a short time later the arms began to lose their power, he found it necessary to give up his work. Since that time the weakness in the extremities has progressively but slowly increased, and the change has been much more rapid in the right arm and leg than in the left. During the last two years speech and swallowing have, if not improved, at least become no worse, although the patient is still liable to choke if he attempts to swallow quickly. The treatment which has been adopted, and which certainly seems to have been useful in retarding the progress of the disease, consisted in the hypodermic injection of strychnine, with massage of the muscles accessible to this method of treatment.

The patient is a healthy-looking man of average height and more than average bulk. His attitude on standing is slightly stooping, with the feet rather widely separated. He walks very slowly and with some difficulty, often dragging the right foot to some extent. Although he can walk without assistance he prefers to use a stick. His gait is somewhat unsteady, and he keeps his eyes fixed on the ground unless this is perfectly smooth. When asked to turn round, he does

so slowly, as if afraid of falling. He says he often feels as if he would fall, and that he has done so several times on attempting to turn quickly or on being stopped suddenly by anyone crossing his path. He knows that very little would knock him down, and he cannot walk on a windy day. When asked to rise from the sitting posture, he does so slowly and with difficulty. This he says is due to the stiffness in the back and legs. If he stoops he is very apt to fall forwards; otherwise he has noticed no tendency to fall in any definite directions.

The patient's face in repose is rather melancholy, as you may see (Fig. 2), but he is easily roused. He has himself noticed that a very trifling circumstance amuses him, and that when he begins to laugh he cannot readily control himself. There is no change in the natural lineaments of the face when at rest save a slight degree of apathy. The eyes can be shut and opened perfectly well though somewhat slowly. When the patient is asked to open his mouth he does so widely but gradually. He is able to approximate the lips and to purse them up, but not sufficiently well to whistle. When the mouth is open the tongue is seen to have no trace of atrophy; it can be protruded, slowly, indeed, but fully and without any loss of power. The soft palate moves perfectly with deep respiratory efforts. There has been some difficulty in swallowing, but this has apparently improved during the last two years. Lately there has been little tendency towards the passage of anything up into the back of the nose, but without care there is a liability to choke. The voice is low in tone and husky in character, but it is not absolutely monotonous, for, as you are able to make out for yourselves, he can sing two or three low notes of the register.

The patient complains of shooting pains in the legs, more particularly on the right side, where they extend from the heel to the thigh, but are worst at the knee. Sensibility to tactile, thermal, and painful stimuli are practically normal both as regards locality and acuity. The muscular sense is quite unimpaired. His sight, he tells us, is good for his age; the pupil reflex is active, all the movements of the eyeball are perfect, and convergence is good. The hearing is slightly diminished in both ears. Taste and smell are normal. The muscular sense is free from impairment. The organic reflexes are almost intact, except as regards deglutition, which is still somewhat imperfect. The cutaneous reflexes are very easily elicited. All the muscle and tendon responses are excessive. The supinator and wrist-jerks are much increased, especially on the right side. The knee-jerks are greatly exaggerated, especially on the right side, and ankle clonus is present in both extremities, but is more distinct on the right side. Voluntary movements of the muscles of the upper part of the face are but slightly impaired, although rather slow, but there is great sluggishness of the muscular movements of the lips and tongue. The patient can move his arms perfectly well, and the co-ordination of their movements is unimpaired. The strength of the left hand, however, is much greater than that of the right. The dynamometer registers 46 on the right and 86 on the left side. As you can see for yourselves, the patient is able to lift a chair to the level of his shoulder with the left hand, while he can only with difficulty raise it from the floor with his right. He complains of weakness of the legs, and this is more marked on

the right side. There is perfect freedom of movement in both legs, but on testing their strength, by making him flex the knee and then push against the resisting hand, the right leg is obviously weaker than the left. As already mentioned, the gait is rather unsteady as well as very slow, with a slight tendency to drag the right leg. He says that he feels the ground under his feet to be quite natural. He is easily tired, and the sensation of fatigue comes on more easily in the right leg than in the left. There is no alteration in the size of the extremities of the different sides of the body, but the muscles on the left side show more firmness and better tone than on the right. No rigidity is present in any part. There is no wasting of any muscle in the body, and there is not a vestige of fibrillation to be observed. The electric reactions, as you see on stimulation with faradism and galvanism, are quite unaltered. The speech is thick and slurring, but quite intelligible. When we ask him to repeat our common test sentences, the syllables are apt to run into each other. Labials appear to give the greatest difficulty amongst the consonants, but the dentals and palatals are also modified. One most interesting point about the patient is that his cough is usually, although not invariably, of the bovine character. This is explained by examination of the larynx, which shows not only some slowness in the movements of the vocal cords, but diminution in the approximation of the false cords on coughing.

In attempting to form a mental picture of the processes at work in this rather unusual case, where must we place the lesions? It is perfectly clear that they are to be sought in the upper neuronic realm. The absence of any wasting or fibrillation, and the existence of normal electric reactions in the muscles, are sufficient to prove that the lower neurons are not primarily implicated. The increased cutaneous reflexes and the exaggeration of the tendon responses furnish ample evidence of some interference with the control of the lower by the higher neurons. The disease in this case is what is commonly termed pseudo-bulbar paralysis of organic origin. With such widespread symptoms may there not be multiple lesions? In many cases of this kind, which are to be found in medical literature, lesions have been discovered in the lenticular region, but in such instances there have been definite hemiplegic attacks, often separated from each other by long intervals of time. Here we have no such history; the onset has been eminently gradual. It seems to me that we must in this case assume a chronic degenerative process affecting the motor cortex of both sides almost symmetrically, and producing lesions of descending degeneration; we may term it cortical glosso-labio-pharyngo-laryngeal paralysis. Such a widespread but uniform change is much more probable than a series of scattered and incoherent lesions, for we must ever remember that the words of Newton are singularly applicable to the processes of disease: "*Natura enim simplex est, et rerum causis superfluis non luxuriat.*"

When we compare and contrast these two cases, we find how great apparently is the superficial likeness—how profound really the underlying difference. In both there are difficulties in the movements of the lips and tongue, in the act of swallowing, as well as changes in the character of the voice and the manner of coughing. There is, however, in the first case atrophy of muscles, together with fibrillary movements and

degenerative electric reactions in those so affected. In the second case there is no wasting, no fibrillation, and no reaction of degeneration. In these points lie the essential and intrinsic differences. There are, moreover, other clinical features more accidental in their presence, yet linked to the central facts in a manner most interesting—on the one hand, symptoms of the spread of the degenerative processes to other nuclei; on the other, the invasion of the descending motor tracts.

Before concluding, let me say in few words that there is another malady which sometimes mimics true chronic bulbar paralysis of organic nature. It is the affection known by the name of asthenic bulbar paralysis, first recognised by Jolly and Strümpell, whether following, as it sometimes does, an acute disease, or arising, as is more common, without any definite antecedents; the group of symptoms produced is not so definitely connected with the bulbar mechanism; there are frequently disturbances of the oculo-motor and masticatory functions showing some changes in the third, fourth, fifth, and sixth pairs, and there are also almost as commonly alterations in the functions of the extremities. In this very interesting form of disease the symptoms are of a neurasthenic kind, and the feebleness of the affected muscles is always more marked towards evening. There is in asthenic bulbar paralysis no wasting of muscles, no fibrillary movements, and no electric changes, unless such an easily induced muscular exhaustion by electric stimuli as was seen in the case described by Jolly. The outlook in cases of this kind is serious, and a considerable proportion have died from respiratory failure. In several instances no changes were found on *post-mortem* examination, but a case described by Widal and Marinesco revealed some disintegration of the chromophilic cells. It seems probable that such cases are of toxic origin.

Bulbar paralysis of organic nature and progressive course may therefore be imitated by affections which may be truly organic, although with lesions situated elsewhere—cortical, as in the case you have seen, or basilar, as in some cases of chronic meningitis, or neuritic, occurring in multiple neuritis, or, so far as our present lights allow us to determine, functional.

It was no part of my intention to discuss the treatment of such affections to-day, and the only further remarks to be made concern etiology.

The two cases which have been brought before you to-day belong to the great group of chronic degenerative processes. In both of them the causal factors are obscure, unless we accept the fact of diabetes in the paternal parentage of the second case as proof of a neuropathic inheritance of which in our own time we hear so much. They are also insidious and gradual. We may well apply to the etiology of such diseases those words of Seneca which so aptly served as the motto for Playfair's memorable *Illustrations of the Huttonian Theory of the Earth*—"*Nunc naturalem causam quærimus et assiduam, non raram et fortuitam.*"