REMARKS

ON THE DIFFERENTIAL DIAGNOSIS OF INSULAR SCLEROSIS FROM HYSTERIA.*

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I PROPOSE to offer a few remarks upon the value of certain symptoms in aiding us to diagnose a case of insular sclerosis, or at least to differentiate in a doubtful case signs which point to organic disease of the central nervous system from those of "functional" character. Especially where the patient is a young woman, the early stages of this disease are very apt indeed to be attributed to hysteria. The initiatory symptoms are often so insidious that great difficulty is experienced in forming a correct diagnosis; and, owing to a kind of opprobrium which still lingers amongst us as attaching to the hysterical condition, no little injustice, sometimes of a very cruel kind, is apt to be done to the unfortunate patient. I am frequently asked in an imploring tone by a sufferer from insular sclerosis, "You don't think, do you, that I could walk well if I tried?" evidently referring in these terms to suggestions which had been freely made that the difficulty of progression simply arose from want of will.

I have not time, nor is it necessary, to enter generally upon the etiology, symptoms, course, anatomy, and other features of insular sclerosis. I need only remark that in a fully developed and typical case all or many of the following symptoms may be present: loss of power in one or more extremities with tendency to spasticity in the affected limbs, intention tremor, exaggeration of knee-jerks and wrist-jerks, ankle clonus, imperfect control over the bladder and (less frequently) the bowel, localised muscular wasting, bulbar symptoms, giddiness, double vision, more or less impairment of vision, optic atrophy, nystagmiform movements of the eyes, scanning articulation, apoplectic attacks. The likelihood of a condition characterised by the presence of the greater number of these symptoms being ascribed to hysteria, although I am sorry to say that it is not altogether absent at the present day, diminishes with the advance of knowledge. But in the early stages, when only a very few symptoms are present, the probability of such an erroneous diagnosis being made is still very strong indeed. It has appeared to me, therefore, that some useful purpose might possibly be served by contributing to the study of a subject of great interest an estimate of the value which experience has led me to attach to certain symptoms—either singly or combined—as helping us at least to feel sure that we are dealing with a case of organic disease of the central nervous system, and not a purely functional affection. When the conclusion is reached that certain symptoms point to structural change a diagnosis is likely to be arrived at by a combined process of synthesis and exclusion. Apologising for the necessarily fragmentary character of the comments, let me at once proceed to deal with some of the symptoms.

The most frequent of all modes by which insular sclerosis is ushered in is by a loss of power, often of very sudden origin, being complained of in one of the limbs. This may or may not be accompanied or preceded by a feeling of "numbness" or "tingling." The loss is only partial; the patient lets things drop from the hand, perhaps, or what is still more common, in the course of a walk drags one of the legs. The limb, however, is still employed, though awkwardly. It is not uncommon for this to occur in the sequel of a strong mental emotion, and the patient, if a young woman, is practically sure, then, to have the symptom set down to hysteria. Should it be accompanied by no other sign I do not think it is possible to be quite certain that the conjecture is not a correct one. At the same time it should be remembered that loss of power of hysterical origin is likely to be more complete than that which is an early symptom of insular sclerosis. And it is much more likely to be accompanied by very marked anæsthesia. So that a paresis of a limb which is only associated with "tingling" or "numbness," and in

which anæsthesia is not present, should be looked upon with doubt, and the patient subjected to observation. It is very likely that in the course of some days, or a few weeks, the power is regained. In these circumstances it is too often imagined that the diagnosis of "hysteria," which has in all probability been made, is amply confirmed. I have satisfied myself, from observation of numerous cases over many years, that a very characteristic feature of insular sclerosis is the liability of the patient to a transitory paresis of a limb, which is succeeded, after a longer or shorter interval, by a recurrence in the same or some other limb or limbs. On the other hand, such attacks as these, according to my experience, do not belong to hysteria.

As an example, I extract the following from notes of a case which had been supposed to be hysteria, but which died some years afterwards from insular sclerosis: "Sixteen months previously there had been a weakness of the right leg and the left hand, of which she got the better..... Again, in the autumn, there occurred a weakness of the right leg. But just before she came to me the left leg became the weaker."

In not a few cases an occurrence of squinting (paresis of an external ocular muscle) ushers in the disease, and the attacks of this kind are usually transitory, like those of the extremities. This, again, is a symptom which, according to my experience, is not one of hysteria.

It sometimes happens that blurring of the sight in one eye, which may go on to considerable loss of vision, is the first symptom, or it may occur later. The ophthalmoscope may show no change. Under the idea that the symptom is due to hysteria, the patient is encouraged to expect recovery, and this will very likely happen in due course (like the recovery of power in a limb). It will, however, be liable to be followed some time afterwards by a similar affection in the other eye, or a return of the difficulty in the same one. Here, again, let me say that this course of events, in my experience, constitutes no part of hysteria. I cannot remember ever to have seen a case thus characterised which the event showed to have been of purely functional origin. I have seen many, on the other hand, which the sequel, under my own observation, has shown to be examples of insular sclerosis. Although, as I have said, in many of these cases in which some loss of sight is complained of, no change is to be found by the ophthalmoscope in the fundus oculi, yet in others (still more often) some pallor of the optic disc (partial atrophy) may be observed. In at least 50 per cent. of cases of insular sclerosis I have found some (often very slight indeed) atrophy of one or both optic discs. It need scarcely be said that this appearance (especially if one eye only is concerned) should be reckoned as furnishing decisive proof that the patient is affected with disease of organic and not simply functional character. The discovery of the remarkable tendency to optic atrophy in cases of this disease, which has been arrived at in the last few years, has very greatly lessened the difficulty of diagnosing insular sclerosis. It should be remembered that the amount of atrophy is often very slight indeed, so that the examination by the ophthalmoscope requires to be of the most careful description to avoid erroneous inference. It is an interesting circumstance that a patient whose fundus oculi exhibits no change may have considerably more impairment of visual power than one in whom there is found a distinct though partial atrophy of the disc. I cannot now discuss the probable explanation of this anomaly nor attempt to enter upon a consideration of the visual troubles generally. They have been dealt with by me in previous publications.¹

In turning over the pages of an old notebook the other day, I came across particulars of the case of a lady whom I had seen in March, 1883, with a view of deciding whether she should be submitted to a course of Weir-Mitchell treatment for the hysteria which her case had been diagnosed to be. Without going into details, I may say that I thought it to be an example of insular sclerosis, which the sequel showed it to be. But in my notes I find: "A suspicious history of loss of sight of the left eye, with gradual recovery of vision, and later on loss of sight in the right eye with gradual recovery of vision. In spite of this I believe the case to be one of insular sclerosis." At that time such symptoms were commonly believed to point to hysteria, and my note appears to

* Read at a meeting of the West Surrey District (South-Eastern Branch) of the British Medical Association at Godalming.

show that I then shared the general belief. Subsequent large experience has perfectly convinced me, as I have already mentioned, that such an opinion is quite erroneous.

The excess of knee-jerks, which is marked in insular sclerosis, is not of itself evidence of organic disease, but is common enough in emotional subjects. Loss of knee-jerks, on the other hand, is not caused by hysteria. Where their absence is clearly established (and considerable care is sometimes required for this) the symptom furnishes most valuable evidence of structural disease. In cases of insular sclerosis, however, the knee-jerks are very rarely indeed lost (they are almost invariably exaggerated), except that in certain conditions of severe muscular contracture it may be almost impossible, from mechanical reasons, to elicit them. A curious experience happened to me a week or two ago. A girl of 16 years of age (introduced by my colleague, Mr. Marcus Gunn) presented ophthalmic symptoms which pointed to insular sclerosis, and in the circumstances I felt confident of finding excess of knee-jerks. Instead of this there was very considerable difficulty in eliciting any at all, even with reinforcement. Her mother, who was present, seeing my surprise, suggested that I should examine her. She was a woman of absolutely perfect health, but in her I succeeded in eliciting a very slight response indeed, and that only in one knee. There is here evidently a family peculiarity which thus masks the indication which would otherwise be furnished by the state of the knee-jerks.

Ankle clonus is so very characteristic a symptom of insular sclerosis that the question of its possible existence as a purely functional phenomenon becomes of very great importance. Twenty years ago, in the early days of the discovery of tendon phenomena, I was disposed to think that ankle clonus was a not very uncommon symptom in purely functional disorder. The fact that with the progress of time and increased experience I no longer hold this opinion depends, it is probable, on the circumstance that in the distant past I considered as examples of hysteria many cases which really belonged to the class of insular sclerosis. However that may be, I do not now come across well-marked ankle clonus in cases of purely functional character, although in these a modified form of the phenomenon is not very rare. I think it must be looked upon that well-pronounced and persisting ankle clonus is a symptom of some structural change in the pyramidal tract, the exceptions to this rule being too few to be safely regarded. In the question of diagnosis of insular sclerosis the existence of ankle clonus would be a still more important sign but for the fact that it is in such cases almost invariably associated with other characteristic symptoms, and the combination is sufficient to constitute overwhelming evidence where a lingering doubt might possibly be felt if this symptom stood alone.

Absence of knee-jerk associated with ankle clonus must be taken as clear evidence of organic disease. I have more than once met with this peculiarity in cases of insular sclerosis.

Intention tremor holds a very important place in the diagnosis, but it is not always present. In the hysterical there is sometimes a clumsiness of movement of the hand when directed towards an object (arising probably from a loss of muscular sense), but it is not of a kind to give rise to error. I cannot call to mind any instance in which well-marked intention tremor was observed by me in a case which the sequel showed to be of purely functional character. Nor am I familiar with nystagmiform movements of the eyes in cases of hysteria. (As is well known, these occur in other diseases of organic origin besides insular sclerosis.) The same must be said of the scanning articulation, which, however, is not a very common feature of insular sclerosis.

The bladder troubles in insular sclerosis, which are of very frequent occurrence, usually in the early stages assume the form of delay in the passage of urine. This may possibly depend upon the muscular spasticity which characterises the disease. I cannot pretend to be able to safely differentiate this symptom from that which is apt to occur in hysteria. It rarely or never amounts to the total inability to pass urine which is characteristic of hysteria, where the symptom is possibly the result of vesical anaesthesia. In the course of the disease there is not infrequently also too frequent action of the bladder and inability to retain it. In the advanced stages there may be a veritable incontinence from paralysis of the bladder.

In these remarks I have only touched upon symptoms of insular sclerosis, about which there is the most chance of a difficulty arising in regard to their value in indicating the existence of organic disease. In cases of hysterical paraplegia, the condition most likely to be confounded with insular sclerosis, there is liability to the occurrence of other symptoms to which it is necessary to allude.

Anæsthesia of the lower extremities, if very complete, is one of these, an anæsthesia which is bounded above by their junction with the trunk. This is a symptom of distinctly functional character, and taken by itself points in no way to the existence of organic disease. It is common, though not necessarily present, in hysterical paraplegia.

Hysterical contracture of the lower limb is a rather more difficult matter to deal with. This, like the contracture which is very apt to come on in advanced stages of insular sclerosis, is associated with excess of knee-jerk, except where there is mechanical difficulty in the way of its exhibition. On the other hand, it is not in the functional case accompanied by well-marked and persisting ankle clonus. Moreover the history of the case should be sufficient to guard against error. Hysterical contracture is apt to occur rapidly, indeed often suddenly, in a person previously well. The contracture in insular sclerosis arrives gradually after more or less long-continued or varying loss of power in the extremities. Contracture of the arm is comparatively rare in insular sclerosis, whilst it is perhaps more common than that of the leg in hysteria.

I have already referred to the fact that in hysterical paraplegia there is often profound anæsthesia of the lower extremities, which forms no part of the picture of insular sclerosis. But there is another point on the sensory side of great differential value to which I must allude. In by far the great majority of those cases of hysterical paraplegia in which anæsthesia is either altogether absent or but slightly expressed it will be found that the plantar reflex is absent. I first called attention to this interesting point many years ago.² A large subsequent experience has established the correctness of this statement. It will often be found on inquiry that the patient had been previously peculiarly susceptible to titillation of the footsole. Dr. Julia Brinck, who has been occupied in personally applying massage for many years in England, and whose opportunities for observation of the condition of the plantar reflex in cases of hysterical paraplegia are therefore unusually favourable, has informed me that her experience entirely accords with my observation. Moreover she frequently finds the plantar reflex, which had been quite absent at the commencement of a course of treatment, return on the patient's recovery. It is scarcely necessary to say that the plantar reflex may be absent in connection with destructive disease of the spinal cord, but in such cases the concomitant symptoms should be sufficient to prevent error. But the occurrence of absence of plantar reflex in the circumstances (cases of hysteria) described is proof that it may also be, what absence of the patellar tendon reflex never is, a purely functional symptom. Unfortunately its value as a distinguishing mark between organic and functional disease is diminished by the fact that the plantar reflex is probably not invariably absent in hysterical paraplegia, although exceptions to the rule are extremely rare. On the other hand, the plantar reflex as a general rule will be found present in cases of insular sclerosis and its retention in a patient suffering from loss of power in the lower extremities thus serves to lend evidence of some considerable importance against the case being one of purely functional disorder. But it sometimes happens that a temporary absence of plantar reflex occurs as a functional symptom superadded to structural change. This tendency for functional symptoms to be mixed up with those belonging to organic change constitutes a great difficulty in the diagnosis of insular sclerosis.

THE "TOE PHENOMENON."

There has recently taken place a very important development in the diagnostic value of plantar reflex to which I should like to refer. In February, 1896, J. Babinski brought before the Société de Biologie de Paris some most interesting observations, which he has amplified in succeeding contributions.³ He shows that in normal conditions excitation of the foot sole provokes, amongst other reflex movements, such as

flexion of the foot on the leg, of the leg on the thigh, of the thigh on the pelvis, a flexion of the toes on the metatarsus. In some persons, on exciting the sole the toes remain to all appearance motionless, but they do not ever (in health) execute a movement of extension. This applies especially to the big toe. In certain pathological conditions, however, excitation of the sole provokes extension of the toes, especially of the big toe. This modification of the reflex he calls the "toe phenomenon." Without proceeding to the consideration of various details, I may say that in all the cases in which Babinski has observed this phenomenon its relation to disorder of the pyramidal system has been incontestably established, and he has not once noted its existence in a case in which the pyramidal system was certainly in a state of integrity. According to his experience it is never seen in hysterical paraplegia, of flaccid or spastic character. During the last five or six months I have had the opportunity of testing the observation in a variety of cases, and especially in many examples of insular sclerosis, and am able so far to confirm the exactness of Babinski's description. It appears, therefore, that by means of this very interesting test our power of distinguishing between organic and functional paraplegia is likely to be greatly increased. If the foot-sole, on being tickled, responds by a movement of extension of the big toe (care being taken to be sure that it is a movement of extension of the metatarso-phalangeal joint) we may take it as tolerably certain that there is an affection of the pyramidal tract, and that we have not to do with a case of functional paraplegia. In insular sclerosis it must be a rare event for the pyramidal tract in some part, at least, of its extent, not to be invaded by islets of sclerosis, and the value of the test is therefore strikingly exhibited. Time will not permit me to dwell further on this interesting discovery. The subject has been ably discussed by Dr. James Collier, and I cannot do better than refer you to a contribution from him which appears in the spring number of *Brain*.⁴

NOTES AND REFERENCES.

¹ *Simulation of Hysteria by Organic Diseases of the Nervous System*, 1891; Atrophy of the Optic Nerve as a Symptom of Chronic Disease of the Central Nervous System, *BRITISH MEDICAL JOURNAL*, October 7th, 1893; Insular Sclerosis and Hysteria, *Lancet*, January 2nd, 1897; Retro-ocular Neuritis, *Optic. Soc. Trans.*, 1897. ² "Absence of reflex from the sole of the foot is a very constant symptom in hysterical paraplegia.....Persistence in the application of a stimulus to the sole will, in hysterical cases, frequently but not always succeed in provoking a response after a few minutes."—Clinical Lecture on Diseases of the Nervous System, 1882. ³ See especially *Le Phénomène des Orteils*, *La Presse Médicale*, July 27th, 1898. ⁴ An Investigation upon the Plantar Reflex, with reference to the significance of its variations under pathological conditions, including an inquiry into the etiology of acquired pes cavus.

THE LOCALISATION AND SYMPTOMS OF DISEASE OF THE CEREBELLUM CONSIDERED IN RELATION TO ITS ANATOMICAL CONNECTIONS.*

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IN bringing this communication before the Medico-Chirurgical Society to-night, my main object is to draw attention to the importance of certain nerve tracts, which have been proved by the recent investigations of neurologists to bring the cerebellum into intimate relation with the spinal cord and higher parts of the central nervous axis.

I do not propose to approach the subject from the point of view of the anatomist, but rather with the intention of impressing upon the clinician that a knowledge of their position and relations, when considered in connection with the results of experiment on the cerebellum and its peduncles, may afford valuable data for the explanation of some of the symptoms of cerebellar disease, and may assist him in the difficult task of its localisation.

The symptoms of cerebellar disease may for our purpose to-night be divided into two classes: first, those which are without special significance beyond the fact that they indicate the

existence of an intracranial lesion, but which have no further localising values; and, secondly, those which are regarded as being more definitely characteristic of affections of the cerebellum. Among the former may be placed headache, optic neuritis, hebetude, and vomiting; among the latter the disturbances of equilibrium and co-ordination indicated by the peculiar reeling gait, the ataxia, the severe vertigo, and the deviations and oscillations of the eyeballs.

One important and puzzling feature of cerebellar disease is that these characteristic symptoms, as is well known, are not invariably present in all cerebellar lesions, for some such lesions may run a course which is more or less latent, or may be indicated by symptoms of so vague a nature as, in the present state of our knowledge, to be practically useless for localising purposes; and on the other hand similar symptoms may result from lesions which do not affect the cerebellum directly. One of the first steps towards the explanation of these peculiar variations in the phenomena of cerebellar disease was the recognition of the fact that these differences in the symptoms are not due to differences in the nature of the lesion (occurring as they do equally in tumour, hæmorrhage, softening, and sclerosis), but depend, as Nothnagel was the first to show, on differences in its situation. This author states in his *Topische Diagnostik der Gehirnkrankheiten* that "the conclusion to be drawn from all these observations is that the situation of the lesion is the factor that determines the presence or absence of symptoms; only when the lesion is limited to one hemisphere can it run the course without symptoms" (p. 47). "There are certainly many cases which show that lesions in the hemispheres may also produce symptoms....., but the analysis of these cases shows, in our opinion, without doubt, that the lesion in such cases involved adjacent parts in one way or other." It may be concluded, therefore, that the loss of the substance of one hemisphere does not of necessity produce any symptoms, or at least any symptoms as yet known to us. "Lesions actually limited to one hemisphere cannot be diagnosed" (p. 48). "Disturbances of co-ordination only occur when the lesion indirectly or directly involves the vermis—that is, the middle lobe of the cerebellum" (p. 50).

"Another question (p. 51) is whether disturbances of co-ordination must invariably appear when the vermis is involved. In my earlier publications I answered this in the affirmative on the strength of the observations then at my disposal."

"The later observations show undoubtedly that tumours may be situated in the vermis without causing any symptom of cerebellar ataxia, and therefore lesions of the vermis need not be invariably associated with disturbances of co-ordination."

"In this connection I may mention that these exceptions are always cases of tumour—that is, of slowly-growing lesions, and that in all cases a greater or less part of the vermis was left intact."

Nothnagel is frequently credited with the unqualified statement that it is only tumours in the middle lobe that cause disturbances of equilibrium, and that tumours of the lateral lobes always run a latent course. From what has just been quoted, however, it will be seen that this is not a correct interpretation of his opinion.

I shall now endeavour to explain from the anatomical standpoint why it is that disturbances of equilibrium occur specially in connection with lesions of the middle lobe, and why in certain instances these disturbances may also be present in lesions of the lateral lobe. The anatomical data have been obtained partly by the collation of observations of various investigators on the human fœtus, partly by the study of degenerations following experimental and pathological lesions of the brain and cord.

The cortex of the middle or vermiform lobe of the cerebellum forms the central termination for at least six important tracts derived from the spinal cord, two arising from the cord directly and the remainder from the nuclei of the postero-medial and postero-external columns, namely, the clavate and cuneate nuclei (two arising from these nuclei on the same side and two from those of the opposite side, each represented by a single line in Fig. 1). The uninterrupted fibres consist of the direct cerebellar tract and the ascending antero-lateral tract of Gowers. All these tracts, except the last mentioned,

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