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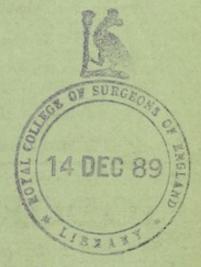
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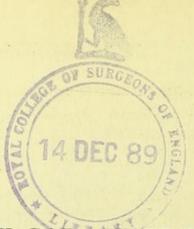
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CASE OF REMARKABLE HYPER-EXCITABILITY OF ALL THE MUSCLES AND TENDONS OF THE BODY TO DIRECT MECHANICAL PERCUSSION, WITH CLONUS OF VARIOUS REGIONS, INCLUDING THE LOWER JAW. RECOVERY.

BY A. HUGHES BENNETT, M.D., F.R.C.P.

A CASE of Clonus of the Lower Jaw, published in a recent number of 'Brain,' by Dr. Beevor, induces me to put on record the following case which has come under my observation, and which was kindly sent me by Dr. Francis Hawkins. The etiology and pathology of the condition is obscure. The patient will doubtless be considered as having suffered from hysteria, as this convenient neurosis has to answer for a multitude of mysterious phenomena, and to provide a refuge for our ignorance. This would be, however, at best, only an imperfect explanation of the condition about to be described. Physically, intellectually, and morally, this woman presented none of the protean or emotional characters which are usually associated with that disorder, and, with the exception of the symptoms from which she suffered, was in all respects of sound bodily and mental health.

Ellen C——, aged 26; single, housewife. The chief complaint of the patient is weakness of her legs, and the occurrence of frequent attacks of spasms in her extremities. The family history is unimportant. The patient herself has always been in good health till three years ago, when she had a discharge from the right ear, accompanied with deafness, which has continued since. Two years ago, without apparent cause, she was somewhat suddenly attacked with headache, vomiting, and general malaise. This illness lasted about a week. Ever since, she has been subject to seizures of trembling and spasm of the limbs, which occur spontaneously and without loss of consciousness. Her powers of walking, also, have become impaired, and her legs are weaker than formerly,

otherwise her health has been good.

On examination, the general health is robust, and all the organs and functions normal in every particular, except the

following exceptions regarding the nervous system. The intelligence of the patient is perfect; she is of a calm and stable temperament; there is no evidence of special nervous. ness, or signs of a hysterical character. In short, the patient has an unusually composed and well-balanced disposition. The special senses are all normal except the old-standing deafness of the right ear. Vision, and the appearance of the fundus oculi normal. There is no paralysis, tremor or nystagmus of the eyeballs. There is nothing abnormal to be detected in the condition of the face or limbs, the movements of which are normal, although the patient states that her extremities feel weak, and that she is unable to walk above a few yards without fatigue, and bringing on the attacks about to be described. There is no rigidity of the muscles or incoördination of their movements. Several times a day the patient is seized with a sort of cramp, sometimes in the left arm, sometimes in the right leg, and sometimes in both together. These occur either spontaneously, or as the result of a sudden movement of, or blow on the limb in question. This consists of a tonic spasm of all the muscles of the limbs which renders them rigid and distorted, and which continues for a few minutes, after which it quickly relaxes. In the upper extremity the arm is firmly adducted to the side. the forearm is flexed, the palm of the hand extends upwards over the shoulder with the fingers clenched. The lower extremity is in a state of rigid extension, the knee adducted, the foot in a condition of equino-varus. These are the most common forms of distortion, but they vary somewhat on different occasions. Immediately after these attacks the parts implicated feel weak, but soon regain their normal condition, and in the intervals the patient feels quite well in every respect. Besides these, the chief forms of spontaneous spasm, the patient experiences other more special and occasional phenomena of a like nature. For example, every now and then there is a sudden spasm of the throat, including the larynx and pharynx, which for a time impedes respiration and causes temporary aphonia. Again, occasionally there is a sudden contraction of both orbiculari palpebrarum muscles, so that both eyelids are tightly closed for half a minute or so. The cutaneous reflexes can everywhere be elicited, but are not exaggerated.

The most noticeable feature of this case is the remarkable increase of muscular contraction which follows percussion of the tendons, the muscles themselves, and the bones. From above downwards the most prominent phenomena were as follows. A tap on the bone over any portion of the

frontal or temporal regions caused at once tonic contraction of the orbiculari palpebrarum muscles, immediately followed by a clonus, in which the eyelids were opened and shut with great rapidity. This lasted for perhaps half a minute, when it somewhat suddenly stopped, to be again produced by another tap on the bone in the neighbourhood. This phenomenon was preceded by fixation of the eveballs in an upward direction. On the mouth being opened, one finger of the observer was placed on the teeth of the lower jaw, and when this was tapped, a well-marked jaw-jerk was obtained. the pressure was kept up, a rhythmical clonus ensued, which continued as long as the tension was maintained. clonus was large in degree and slow in rhythm, being about 6 or 8 to the second. At other times if the lower jaw itself was tapped with the percussion hammer, a very fine, rapid, rhythmical tremor of the jaw was produced, roughly speaking about 15 or 20 to the second, which continued for a few seconds and ceased by itself. Occasionally the artificially produced jaw-clonus was followed by temporary spasm of the larvnx and pharynx, in which there was inability to breathe for some seconds, and suffusion of the face, a feeling of oppression and lump in the throat, and inability to This aphonia lasts for some minutes afterwards. speak. Percussion of all the tendons throughout the body causes the most violent manifestations, in the shape of an immediate jerk of the muscle operated on, which is at once succeeded by clonic trepidation of the entire limb. Frequently the local convulsion spreads thoughout the entire body. For example, a slight tap on the patellar tendon produces a violent extension of the leg, at once succeeded by clonic shaking, of a large type, of the whole lower extremity, after a second or two of which the back stiffens, the head is bent backwards, and a condition of opisthotonus ensues, with rigidity and trepidation of the head, neck, and all the This lasts for 10 or 12 seconds, and then extremities. suddenly relaxes. The same ensues when any tendon is percussed, especially those of the left arm and right leg, and also when a blow is given to any of the prominent bones, or a jar to any part of the body is received. This spasm is not accompanied by actual pain, but the sensation is unpleasant and arrests the respiration. There is no loss of consciousness during these paroxysms, the patient being perfectly acute intellectually, and able to describe her sensations. Ankle-clonus cannot be elicited; but when vigorous attempts are made to do so, there is a tendency for universal spasm to be produced. Response of muscles to mechanical percustion lively, but not excessively exaggerated. Sensibility is everywhere natural. There are no trophic changes, and the electrical reactions of the nerves and muscles are normal.

The patient was kept for a few days without treatment while the foregoing observations were being made. She was then directed to lie in bed, and ordered chloral hydrate, gr. xx.; pot. bromid. gr. xxx., every six hours. Fourteen days afterwards all the symptoms were notably diminished in degree, but still preserved the same characters as before. About this time she was seized with a mild attack of pleurisy of the left side. The draught prescribed above was discontinued, and poultices were applied to the chest. In one week she was convalescent from the pleurisy, and in two had quite recovered. It was then found that all her nervous symptoms had disappeared. On getting out of bed she felt weak, but after a few days expressed herself quite well. There was no trace of spontaneous spasm. The gait was normal, and the patient moved about the ward all day without fatigue. No clonus could be elicited either of eyelids, jaw, throat, upper or lower extremities. Everywhere percussion of the tendons produced natural responses, except at the knees, where they were somewhat lively, but not excessively so. The patient, when seen a month after discharge from the hospital, was found to have retained her improved condition.

Commentary.—This woman, as far as could be ascertained, was healthy in every respect except in the particulars just described, namely an abnormal readiness of all the muscles of her body to contract on the application of local percussion to their substance, and especially to their tendons; and also for this contraction to be succeeded by a rhythmical clonus, sometimes located in the individual muscle, and sometimes spreading to the entire limb or throughout the whole body.

The nature of the morbid process which led to the production of these symptoms is very obscure. That it was not the result of serious gross organic disease is proved by the fact that, although of two years' standing, the affection completely recovered in a few weeks. In the absence of a positive anatomical demonstration we are reduced to speculation, and may theoretically assume the position to be most plausibly explained by a condition of extreme functional over-activity of the ganglion cells of the anterior cornua of the cord and medulla, causing hyper-tonicity of the entire muscular system and its consequent tendency to tonic and clonic spasm. Even granting this to be the proximate cause of the physical phenomena, we have in this case no means of knowing whether it was primary

or secondary to changes elsewhere. There was certainly no evidence of the latter.

The spontaneous tonic spasms, the increased tendon reflexes. and the rhythmical clonic trepidations have all probably the same physiological signification, and their occurrence in different parts of the body is not of special pathological import, but depends only on the distribution and extent of the morbid process to which they owe their origin. The jaw clonus, for example, is not characteristic of any particular disease, but only implies involvement of the medulla or those central regions the irritability of which leads to hyper-tonicity of the muscles which move the inferior maxilla. The symptom is not so rare as seems to be supposed, and doubtless, when looked for, it will be found more commonly. I have repeatedly demonstrated it in cases of disseminated sclerosis, spastic paralysis, and in so-called hysteria. Others have also observed and recorded this symptom.1 Although in itself of no special significance, jaw clonus, being one of the highest available deep reflexes that can be obtained, is on that account of great clinical importance. The same applies to what Dr. de Watteville calls "the jaw-jerk," a condition which has already been described by Dr. Lewis of Philadelphia, under the term "Chin-reflex.2

In this case the jaw-jerk was well marked, as also was the clonus, when pressure was kept up by the finger on the teeth of the extended inferior maxilla. In addition, however, to this ordinary clonus of six or eight contractions to the second, a tap on the masseter or on the lower jaw, not necessarily in an extended condition, caused another form of tremor, consisting of a fine, rapid, rhythmical clonus about twice as fast as that produced by continuous extension, and this lasted for ten or twelve seconds after the percussion blow, and then ceased of its own accord. Similar phenomena followed a tap of many of the other muscles of the body, and also from a jar of the bones in their neighbourhood. This was notably observed in the orbiculari palpebrarum and oris, in the other muscles of the face, in the muscles of the neck and larynx, and in some of the small and superficial muscles of the trunk and limbs, although in these latter they were complicated with the larger clonus and tonic spasm. This fine rapid trepidation, the result

¹ Ballet, "Note sur l'état de la réflectivité spinale dans la fièvre typhoïde; "Prog. Méd.,' 1881.

Delom-Sorbé, 'De la trépidation épileptoïde provoquée.' Bordeaux, 1885. Weir-Mitchell and Lewis, "Physiological Studies of Knee-jerk," &c.; 'Medical Times,' New York, Feb. 1886.

² 'The Chin-reflex. A new Clinical Observation.' By Morris J. Lewis, M.D., Medical and Surgical Reporter. Philadelphia; May, 1885.

of a direct blow or general jar, was very similar to what is seen during a rigor; the chattering of the teeth being almost identical in both. The condition necessary for its production is probably an exaggeration of that necessary to produce ordinary clonus; the greater rapidity of the tremor being possibly due to its velocity not being impeded by passive flexion of the muscles. The two phenomena may, on the other hand, depend on entirely different mechanisms. Further observations on the subject are necessary to illuminate the question; and these may throw light on many obscure points in connection with rigors, shiverings, tremblings, and such-like conditions.

The attacks of tonic spasm with distortion of the limbs in this case are also of interest, and not easy of explanation. some respects they resemble a condition described and illustrated by myself in a former number of 'Brain.' Those seizures of tonic spasm were usually spontaneous, arising without apparent exciting cause at irregular intervals. On the other hand, it is to be noticed that the attacks of clonic trepidation already described rarely did so, but almost always had to be artificially provoked. It is possible to conceive that the same pathological basis may be the origin of these two conditions. Assuming a continuous irritation of the central nerve-elements to be the cause, the accumulation of motor energy exploding at intervals might give rise to the spontaneous attacks. Under the same conditions a mechanical stimulus would originate similar phenomena, and passive stretching of the muscles give rise to clonic spasms. But why the spontaneous seizures should always be of a tonic nature, and those provoked be clonic, furnishes matter for speculation.

The treatment of this case was based on purely hypothetical considerations, and turned out to be successful. Primary functional irritability of the grey matter of the cord having been assumed, hydrate of choral, and bromide of potassium in large doses were administered. These drugs are known in health to depress the functional activity of these centres. Although the affection had lasted for two years, and the patient had previously tried the effects of rest and other methods without benefit, the result of these remedies was immediate, and in a fortnight all the symptoms were notably diminished. An accidental attack of pleurisy seemed to

complete "the cure."

^{1 &#}x27;BRAIN,' January, 1885.

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