Remarks

ON

MANIACAL CHOREA.*

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CHOREA is one of the commonest of all nervous affections met with in hospital, as well as in private practice, and is very often associated with rheumatism. It presents varying degrees of severity, from the mildest form, consisting of awkwardness of movement and facial grimaces, to incessant, irregular movements and contortions, with such jerking and throwing about of the limbs, that the patient must be confined to bed, and watched to prevent her from injuring herself.

In these latter cases (chorea gravis) some delirium, with delusions, is not uncommon. But there is yet a still more grave form of chorea, in which mania may supervene and so completely overshadow the motor disturbances that the true nature of the disease may be overlooked and the patient sent to an asylum.

So wide is the difference in the symptoms that it seems hard to realize that it is really not another and a new disease, but only an exaggerated variety of the chorea simplex of Sydenham.

The following is an example of maniacal chorea, or chorea insaniens*:

CASE (From notes taken by Mr. Thos. Graham, Resident Pupils).—Annie O., aged 17 years, unmarried, was admitted to Sir Patrick Dun's Hospital on June 14th, 1906, suffering from marked choreiform movements, particularly of the left arm and hand. Six years ago she was treated in another hospital for enlarged cervical glands. She had enjoyed good health ever since up to a fortnight ago, when she was treated for rheumatic pains in the legs; two days before admission these pains left her, and then the choreic movements became manifest. The patient herself said she had a "fright," but could give no explanation of its nature, and the accuracy of her statement was questioned by her sister and mother, with whom she lived. She referred her trouble to her head, and said her mind was being affected. The motor disturbance was not confined to any part of the body—the trunk, head, arms and legs participating, but the left side was most affected; the eyes rolled about, and she continually uttered a sort of sob, and had delay and difficulty in answering questions, which she did in a sudden, jerky fashion, but with intelligence. The deep reflexes were not increased. The pulse was about 80, and of low tension; temperature 98°. A systolic murmur was distinctly audible in the mitral area. The skin over the lumbar vertebrae was congested, but not broken, and the toes were of a dusky blue colour. A mixture of ammon. brom. and liq. arsenicalis was ordered; and chloral 15 grains given at bedtime. She did not sleep, but became so noisy and obstreperous with shouting and screaming that she was moved into a ward by herself, and our house-surgeon considered she was a case of acute mania, and suggested her removal to the Richmond Lunatic Asylum. The choreic movements next day were very violent, and in a day or two after admission, in spite of all care, she barked her elbows, rubbed her heels sore against the bed-clothes, and with the heel of one foot abraded the skin of the opposite instep. Her back was also stripped over the sacrum. She could hardly speak, but at times would become calm for a few minutes, and could be fed with soft, thick food. She was given $\frac{1}{100}$ grain hyoscine hydrobrom. with good results for a few hours, but a second dose was useless either to calm the motor excitement or to procure sleep, while 20 grains chloral gave about three hours' sleep. She was quite maniacal at times, striking out, shouting and screaming, or uttering deep, raucous sounds, refusing food at times, and then taking it readily. She passed her motions and water involuntarily in the bed, as, owing to the extreme violence of the muscular system, the use of a bed-pan was out of the question. The urine obtained on one occasion was found to be acid, specific gravity 1014, and free from albumen and sugar; it contained calcium oxalate crystals. During the short sleeps obtained by chloral she was perfectly motionless, and her breathing quiet. The temperature, which was normal on admission, rose to 100° on the evening of the 18th, the fourth day, and fell again to 97.5° next morning. It remained normal till the evening of the 21st, when it shot up to 103.4°. The pulse, when possible to count it, was usually about 80-90, but on the morning of the 21st it rose to 103. The murmur in the heart was audible all through, and proportionately as the heart beat more violently in the times of great motor excitement it became more distinct.

During the 19th and 20th her condition continued in much

the same state—periods of calm, in which she would say some words intelligently, such as "No, thank you, sir," alternated with periods of wild activity in which she had to be held down in the bed, and in which she flung her legs and arms about in the strangest fashion. Owing to the involuntary biting by her teeth, and the rubbing of them by her hands, her lips were sore and bleeding. She was indeed a pitiable object. The mental disturbance was maniacal in type—at one time with delusions, and again with hallucinations, and lastly one of semi-coma.

In the afternoon of the 21st she became particularly violent, after a rather calmer forenoon, and remained in a continuous state of turbulent activity till about 9.30 p.m., when I went to see her. She could not be restrained by six attendants from knocking herself about, and all the wadding and padding and bandages which had been put around her limbs to prevent her injuring herself were rubbed or torn loose. A strait-waistcoat was then put on and her legs wrapped round in a sheet; although these means prevented her springing out of bed and injuring her limbs, she kept up, nevertheless, an incessant wriggling and movement, while she was bathed in perspiration. She had had chloral, a hypodermic of morphine, hyoscine, but the only remedy that succeeded in calming her was chloroform inhalation. Under it she became quite quiet, and the various appliances to keep her in bed were loosened so that she was not oppressed in any way that night. She slept quietly, and took nourishment fairly well at times. Rectal feeding could not be managed. Next day she looked, however, very ill. The heart was beating excitedly (the mitral systolic murmur being present), the pulse was 140, very compressible, respiration 44, and temperature, which had risen to 103.4° at 7 p.m. the previous evening, was 102.4° at 7 a.m., 101.8° at 10 a.m., and 101.4° at 2 p.m. The motor disturbance was much less, and she was quiet, but in a semi-comatose condition. Loss of flesh had been noticeable for some days, but now that the limbs were quiet they were seen to be extremely emaciated. This was the ninth day of her sojourn in hospital. At 4 p.m. she suddenly became unconscious, her respirations very laboured, and her extremities and face cyanosed; $\frac{1}{100}$ grain of digitalin and $\frac{1}{20}$ grain of strychnine were immediately injected hypodermically, but without avail, and she died in five minutes.

Post-mortem examination was made on June 24th. Dr. O'Sullivan, F.T.C.D., Pathologist to the Hospital and Lecturer on Pathology in Trinity College, Dublin, has kindly supplied me with the following report:

The longitudinal sinus contained a small quantity of mixed clot. The vessels of the cortex of the brain were engorged. A very small amount of fluid was found in the lateral ventricles. The brain presented no abnormal appearances to the naked eye. It was fixed in formalin and pieces were subsequently removed from various parts and examined microscopically. The only portion which presented any abnormal appearance was in the Rolandic area. Here some of the small vessels in the cortex were thrombosed. In others collections of cells with oval nuclei lay heaped up in the perivascular lymphatic spaces, and there were clumps of what appeared to be broken up nuclear material in these spaces. The spinal cord was also examined and presented nothing abnormal. The cerebro-spinal fluid was examined bacteriologically in the media recommended by Wassermann for the cultivation of the coccus found by him in rheumatic cases, but the culture remained sterile and no micro-organisms were found in the meninges on section. The other organs of the body showed no abnormality, with the exception of the heart, which was in a very atrophic condition, and some calcified tuberculous glands in the mesentery and in the root of the lung.

The prominent features presented by this case were: Rheumatic pains in the legs for a fortnight, upon which chorea supervened. A mitral systolic murmur, audible on admission, of varying loudness, and lasting up to the ninth day—the day of her death. The pulse quiet and regular, about 80 each day up to the last three days, when it rose to 110, 130, 140. The temperature normal or sub-normal until the day before death, when it touched 103.4°. The choreic movements were slight for about forty-eight hours, and then became so violent that two special nurses were required to watch her and prevent her from falling out of bed and from striking her limbs against parts of the bed. (The idea of treating her by placing her mattress on the floor, with pillows around, was considered, but discarded in favour of the bed with nurses at either side.)

The psychical phenomena were prominent, out of all proportion, and at first quite overshadowed the motorial. They differed, however, by the absence of incoherent speech and wild garrulity, from the forms one is accustomed to associate with the acute delirium of fever or acute mania.

Another point worth noting was the occasional, though temporary, mental calm, which my visits, once or twice a day up to the last three days of her life, seemed to produce. At these times she would cease shouting, the movements became quieter, she could always understand

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what was said, and sometimes she succeeded in speaking and making intelligent replies, such as "Yes, sir," "No, sir," and "Thank you, sir."

Treatment by medicines totally failed to have any effect on the course of this terrible disease, and the only help it gave was by securing some hours of sleep, and muscular rest during sleep. The drug which seemed most useful for that purpose was chloral. On two days $\frac{1}{2}$ grain hyoscine hydrobrom. (once repeated) was successful in calming the violence of the delirium, but it failed to have any effect the last two days of her life; while morphine hypodermically was practically useless.

The occurrence of a systolic murmur at the mitral area, which was considered on admission to be probably due to rheumatic endocarditis of some two or three weeks' duration, must now, in the light of the autopsy, be judged to have been dynamic, and due to simple and acute ventricular dilatation. It is not to be explained on the grounds generally accepted—namely, anaemia and choreic cardiac excitement.

The foregoing case is an example of one of the rarest diseases to be met with in the practice of medicine. It is one of the most grave of the varieties of chorea, whether regard be paid to the extreme violence of the muscular movements, the psychical disturbance, or its acute course, usually terminating in death. "Truly a terrible disease is maniacal chorea" (Osler).

In evidence that this variety of chorea is very uncommon I would point to the very scant literature on the subject, the paucity of published clinical records, and the very short description it receives in standard works of reference on medicine and on insanity.

Two of the most recent works I have been able to consult are *The Textbook of Psychiatry*, by Bianchi and MacDonald, 1906; and *Clinical Psychiatry*, by Kraepelin and Johnston, 1904. No reference to "maniacal chorea" appears in the index of either, while under the title of "Choreic insanity" a very different condition is described.

Bianchi writes: "A choreic subject is always mentally affected, the character of the child or young adult is changed; he becomes irritable and fearful, his sentiments are altered, his power of attention is defective, and he may suffer from hallucinations."

Kraepelin writes: "Senselessness and bewilderment, with choreic movements, occasional hallucination, a morbidly cheerful mood and great restlessness, with rapid physical sinking, are particularly diagnostic of choreic insanity."

From these excerpts it is evident that these authors never met with a case of maniacal chorea, and it is a matter of regret, if not of negligent omission (to say the least of it), that they should not have made themselves familiar with published cases of this disease in the British Isles.

The same adverse comment is applicable to the majority of books I consulted, be they written by specialists in mental and nervous disease, or textbooks on the practice of medicine. Thus Mercier, Bevan Lewis, Hack Tuke, Ziemssen, *Quain's Dictionary*, Reynolds's *System of Medicine*, Strümpell, and many other textbooks in medicine, give no description of any such marked psychical complication as was portrayed by my case. In some, such as Ross and Allchin, it is said "Chorea in adults is sometimes followed by acute mania." Gowers makes the short comment: "The mental state may not appear till the disease be well developed, or it may precede the chorea. The latter may be slight or quickly cease, while the mental disturbance continues in an intense form. There are often delusions, sometimes wild, violent excitement without the garrulity common to ordinary mania. This excitement may be followed by apathy or dullness." However, Hilton Fagge, Clifford Allbutt, Roberts, and Osler are notable exceptions, and they attest to the reality of the extent and the gravity of maniacal chorea; while Clouston (*Mental Diseases*) gives in detail a typical case of rheumatic chorea with acute maniacal insanity.

A fatal termination of chorea in children is practically unknown, but it may occasionally occur between the ages of 15 and 25.

Dr. Handford¹ in thirteen years (1875-88) had 154 cases of chorea, with 4 deaths—that is, 2.6 per cent. This number tallies very closely with the 2 per cent. of the

collective investigation report of the British Medical Association² (439 cases, with 9 deaths).

The association of pregnancy or parturition with chorea gravis is more than an accidental coincidence, and such cases, more than others, add to the death roll in chorea. But, perhaps, of all the grave varieties maniacal chorea (chorea insaniens) is the gravest as well as the most fatal. Of 23 cases collected from various sources by William Gay, M.D.,³ complete recovery took place in 10, death occurred in 8, 1 was followed by permanent weak-mindedness, and 4 were lost sight of, having been sent to asylums.

The duration of these cases may be very short; death occurred in my case on the ninth day; in some cases it happens as early as the sixth, or even the second day, although generally not until after some weeks. The mode of death seems to be due to the exhaustion resulting from the wild muscular movements, inanition, and want of sleep and rest. Many cases end in unconsciousness and coma, although the intellect may remain clear almost to the last—a fact first noticed by Wilks, and confirmed by subsequent observers.

Dr. Tuckwell⁴ had his attention first called in 1861 to maniacal chorea, in a woman aged 24, by Professor Trouseau. He saw a similar case in Vienna in 1862, in a pregnant woman, aged 20; and he describes a case of a boy, aged 17, under his own care—"probably embolic"—in 1867. All three had furious choreic movements, maniacal delirium set in early, and death ensued within ten days of the onset, after semicoma, with partial or complete cessation of the movements. In each of these cases a mitral systolic murmur had been audible during some part of the illness, and a fringe of beads, or bright warty vegetations, the size of a pin's head, was found at the mitral valve. Dr. Tuckwell analyses 34 fatal cases, recorded up to 1867, to which an autopsy was attached. In 4 the heart was healthy, in 5 no mention was made of its condition, and of the remaining 25, 20 had vegetations on the valves. He attributes the fatal results to the vegetations being set free and becoming embolic in the brain, and probably also in the cord, and thus favours the theory attributed to Kirke and supported by Hughlings Jackson and Sir William Broadbent.

On the other hand, as negating the embolic theory, and as bearing a close resemblance to the case of Annie O., I would refer to two fatal cases of acute chorea, with insanity, reported by Dr. Evan Powell,⁵ Medical Superintendent, Nottingham Borough Asylum, in which there was a complete absence of valvular disease or cerebral embolism. The first was a male, aged 19, who had been cured of a second attack of rheumatism ten days before the chorea developed. In three days' time acute mania set in, with delusions and hallucinations and great violence, and death ensued by coma after fifteen days, during which there were lulls in the tempest, and in these quiet intervals the mind was quite clear to all that had occurred. A mitral murmur was present.

The second case was a female, aged 20, without any history of rheumatism or heart disease. After fourteen days of chorea, insanity and hallucinations of hearing and sight set in, and it took four people to hold her in bed. She was sent to the asylum, and died in a week, but she was quite conscious, and her mind was clear up to the last two days.

The autopsy in each case was made by H. Hanford, M.D.,⁶ and the only abnormal feature present was hyperaemia of the brain. After hardening he found (a) thrombosed vessels in the anterior cornua of the cord; (b) haemorrhages into the lymph sheaths; (c) haemorrhages into the anterior horns, causing destruction of the nerve tissue; (d) haemorrhages into the deep transverse fibres of the pons. He concludes that the "numerous haemorrhages from small vessels as well as capillaries, the thromboses, and general dilatation of vessels, afford evidence of a very unusual degree of vascular engorgement of the nervous centres during life."

These cases, while they by no means conclusively prove that the pathological changes discovered in the nervous centres were the cause of the psychical disturbances, negative the theory of emboli, as there was an absence of mitral valve disease in either.

Still more recently further evidence against the embolic theory has been adduced by Drs. Poynton and Gordon Holmes in their admirable contribution to the pathology

of chorea (*Lancet*, 1906). They found, on microscopical examination of three cases of fatal chorea, "very little evidence of embolism," although great hyperaemia was present, with thrombosed vessels in all parts of the brain.

My case is practically another of the same type, as there was neither mitral valve disease nor cerebral embolism. From what I have stated, the embolic theory as causative of chorea—be it chorea major, chorea minor (Sydenham's chorea), or the maniacal type illustrated by my case—can be no longer tenable. Rather must we recognize the difference in the severity of the choreic manifestations to be due to the presence of a toxin, and to the virulence or mildness of its action on the nerve elements of the cerebral cortex and of the lower nervous centres. In direct proportion as the psychical and the higher centres of ideation and volition or the motor centres are those most invaded, so the symptoms each case may present will be either those of mania and other disordered mental states, or those of inco-ordinate muscular contractions and movements of the body and the limbs.

The intimate association of chorea and rheumatism has been long recognized, and has of late years been more and more insisted on, and the consensus of opinion of many clinical observers is in favour of there being one common underlying cause for each of them. It is a well-recognized fact that the first indication of rheumatic fever may be chorea, and that myocarditis, endocarditis, or pericarditis may equally occur in either.

Sir Dyce Duckworth,⁷ in his paper read before the Medical Congress at Lisbon last June, emphasized the view that chorea should be considered as "cerebral rheumatism." He has for many years consistently maintained the intimate relationship between the two diseases, and clinical experience tends, with increased authority every year, to confirm the truth of that relationship.

In 1892, Clouston,⁸ writing upon the varieties of insanity, under the heading of "Rheumatic Insanity," describes the typical case of rheumatic chorea with acute maniacal insanity to which reference has been made above, and asks the question, "Was the rheumatism the true cause of the mental symptoms of the chorea or of both?" and asserts his view, from a clinical standpoint, that the poison of rheumatism was the true cause of the insanity.

It may be interesting to note, *en passant*, that this view is by no means a new one. It simply reasserts what was propounded in 1850 by M. Germain Sée.⁹ His expression is, "that chorea was due to the rheumatic diathesis acting on the brain and spinal cord." Trousseau,¹⁰ who quotes Sée's view, and acknowledges its probability, did his best to refute it and to prove that chorea was a purely functional derangement, similar to hydrophobia and tetanus. The ground upon which he based his theory was that no gross evidences of inflammation of the brain or its membranes are present in fatal cases of any of these diseases.

It is curious to see that now, in these latter days, the older views—based on clinical observation—have again gained ground, and that the keen and exhaustive researches of scientific minds, combined with highly skilled bacteriological dexterity and acumen, have shown that the pathological conditions of these so-called functional nervous diseases are really due to microbic infection and toxic influences. The first ray of light which illumined this difficult subject—after which so many thoughtful physicians were groping in the dark and feeling after the truth, if possibly they might find it—was the discovery by Wassermann, in 1899, of a diplococcus which he had isolated from the cerebro-spinal fluid and inflammatory exudations of a patient who had died of acute rheumatism and chorea. With this diplococcus he had experimentally inoculated rabbits and produced in them polyarthritis.

Drs. Poynton and Paine, in 1900, confirmed the observations of Wassermann, and found the diplococcus in a case of chorea. In 1905 they reported another case of fatal chorea, in which they demonstrated its presence. Isolating the diplococcus from the cerebro-spinal fluid, they had produced by inoculation arthritis, endocarditis, and pericarditis in a rabbit.

In a valuable paper¹¹ Dr. Beattie accepts the theory of a bacterial origin of rheumatic fever, and deals with the

objections raised against it, while he calls the organism *Micrococcus rheumaticus*.

It is not my intention to follow further this most interesting branch of the subject, but would refer to the lucid and convincing contribution on the pathology of chorea which Dr. F. J. Poynton and Dr. Gordon M. Holmes published in the *Lancet*, October 13th, 1906, and from which I have already freely quoted above. These observers found the diplococcus in the brain and its membranes, and also in the diseased valves of the heart, in 3 fatal cases of chorea and rheumatism, and they assume that the presence of micro-organisms goes far to prove that these inflammatory pathological changes are due to a toxin derived from these micro-organisms, and that chorea is caused by the action of bacterial poisons on the brain.

"The conclusions they come to, in their research on this subject, in the first place support the clinical evidence that chorea is a manifestation of acute rheumatism; and secondly, that the demonstration of diplococci in each of their three cases is a striking confirmation of the assertion that the *Diplococcus rheumaticus* is the infective agent in acute rheumatism."

My case does not exactly add support to the views just stated, in so far as no micro-organism was discovered by Dr. O'Sullivan in his careful bacteriological examination of the brain and spinal cord. I have, however, felt it my duty to record it, as a contribution to the literature and clinical history of this most interesting subject, since it is only by the accumulated records of cases of chorea, in which a full and reliable pathological report is made by capable bacteriologists, that a sure foundation can be laid and a true pathology erected.

REFERENCES.

- ¹ *Brain*, vol. xii, p. 129. ² *BRITISH MEDICAL JOURNAL*, February, 1887. ³ *Brain*, vol. ii, p. 151. ⁴ *Med.-Chir. Review*, vol. xi, 1867, p. 506. ⁵ *Brain*, vol. xii, p. 157. ⁶ *Ibid.*, vol. xii, p. 151. ⁷ *BRITISH MEDICAL JOURNAL*, p. 1454, 1906. ⁸ *Mental Diseases*, p. 485, 3rd edit., 1892. ⁹ *De la Chorée Memb. and l'Acad. de Méd.*, Tome xv, 1850 (quoted by Trousseau). ¹⁰ *Clinique Médicale*, 1868, vol. ii, p. 786. ¹¹ *BRITISH MEDICAL JOURNAL*, December, 1905.

ON THE VALUE OF AN ABNORMAL RISE IN THE AVERAGE LEVEL OF THE DIAPHRAGM AS A GUIDE TO THE VOLUME OF THE BLOOD IN ACTIVE CIRCULATION.*

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THE object of this paper is to bring forward clinical evidence in favour of the following propositions:

1. When, from any cause, the total volume of the blood in circulation is materially diminished, the total bulk of the intrathoracic viscera is correspondingly diminished by the relative emptiness of the thoracic blood vessels, especially those of the lungs. This diminution in the bulk of the intrathoracic contents shows itself by an elevation of the diaphragm, which has to be maintained at a higher average level than the normal in order to adjust the cubic content of the thorax to the altered volume of its contained viscera.

2. That such a rise of the diaphragm may frequently be detected clinically, and, when present, is of value in diagnosis, and affords important indication for treatment.

NORMAL POSITION OF THE DIAPHRAGM.

The position of the diaphragm can be recognized clinically by ascertaining the upper level of the gastric resonance in the left nipple line, or of the upper border of the liver in the right nipple line. In both cases percussion is best done with the patient recumbent, and in the case of the gastric resonance the recumbent posture is essential to accuracy, because in the erect posture the gaseous contents of the stomach, if scanty, may rise up into the arch of the diaphragm, and the dullness of the liquid contents of the stomach be continuous with the dullness of the left lobe of the liver and the heart.

Of the two methods named, the percussion of the gastric resonance is easier and more reliable for the following reasons:

1. There is much variation in the upper level of the liver dullness of healthy individuals, dependent upon

* A paper read before the Birmingham Branch of the British Medical Association.