HUNTINGTON'S CHOREA WITHOUT CHOREIFORM MOVEMENTS.

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THE case recorded below came under observation because of symptoms which, although well recognised as quite characteristic in America, where this rare disease seems to be relatively more common, appear for all that to have received scant recognition in this country. Moreover, several of the relatives of the patient have been afflicted in a way which might have made recognition difficult had British text-books alone been consulted. Here few references are made to the possibility of mental symptoms preceding motor, or even, on occasion, holding the stage alone.

PERSONAL CASE.

I. T., a married woman, aged 37, was admitted to the Maida Vale Hospital in May, 1929. She was brought up because of inability to look after her home affairs and of indistinctness of speech.

There was in addition a remarkable family history of others similarly affected.

History of present illness.—Patient was quite normal until 18 months previously, when, just before the birth of her youngest child, she seemed at times thick in her speech, and became apathetic and "different." Since then marked changes in her personality took place insidiously and progressively. She gradually lost interest in life, spent long hours "gaping out of the window," and her husband volunteered that she had "changed into a cantankerous old woman." She had become dirty in person, and neglectful of her house and children. She lay long hours in bed of a morning. She had become both critical and intolerant of criticism; at times she was "very contrary" and would fly into a tantrum when rebuked. She had on occasion boasted in rather a grandiose manner, especially of her own home and upbringings, but had never expressed any definite delusion. Her registration of passing events had become poorer, and she had become incapable of sustained application.

Overshadowing all appeared to have been a progressive loss of her normal interests and outlet for energy. She had shown no insight, and had predominantly been very content with herself and (lack of) doings.

In addition to these mental changes, her speech had become progressively more slurred, and this, combined with a growing tendency to ramble in her talk, had on several occasions led to the suggestion that she was drunk. Moreover, she had at times appeared rather clumsy in her gait.

On no occasion had any involuntary movement been seen.

Past history.—Previously she was said to have been bright and care-free, and "full of life and go." She was always considered perfectly normal, though possibly not very clever (her school record was not below the average), and her physical health had always been very good. She had been married about 10 years, and had four children (sons) all healthy. There had been no miscarriages.

State on admission.—Her condition was filthy and verminous. She was rather thin, but of quite good physique. There was no evidence of system disease. Her teeth were, however, very septic. There was some slight tremor about the lips. Her speech was markedly dysarthric and very monotonous. Examination of the central nervous system was otherwise quite negative. The tendon reflexes were possibly rather brisk. There was no trace of involuntary movements, ataxia or incoordination. Gait appeared perfectly normal. Her arteries were not thickened—B.P. 120/90. Blood W.R. was negative, and spinal fluid normal in every way. (Protein .02 per cent.; cells none; Lange $\frac{1}{10}$ and W.R. negative.)

She had a curious lack-lustre eye, and very little play of facial expression. She looked dull and apathetic, not sad. She sat or stood listlessly and unoccupied, answering questions readily, but indifferently, and her attention soon wandered. Her replies were discursive and rambling, and she was very garrulous, but quite coherent. She often gave vent to an apparently causeless unemotional laugh. She was perfectly orientated, not deluded or hallucinated. When pinned down her memory appeared very fair, and she made no worse a show than many hospital patients, although on superficial examination she appeared to have marked memory defect.

She had no insight. She said she supposed she was in hospital for a rest, and that she was quite capable of looking after her affairs. She was quite unconcerned about her filthy condition, and by the questions put to her.

Progress.—A few days after admission the patient was doubly incontinent, whilst up and dressed. Afterwards her habits were clean; indeed she spent quite a lot of her time washing her hands (she was unwilling or unable to explain this). At first she slept in a room by herself, and spent her time gazing out of the window, or trifling with the daily papers, but on being transferred to a ward, perked up a little, and although she usually paid no attention to the other patients, would dance with them on occasion to the gramophone in the day-room.

She continued unchanged until discharged early in August, her emotional poverty always strikingly in evidence.

Since returning home she has reverted to her former ways. She has neglected her household duties so much that her husband—who works in a garage—has had to employ a daily help to look after his home and her. She went to the cinema with considerable enjoyment on most evenings, and did not get up till lunch-time in the day. Seen in November, she said she was perfectly well, but found some difficulty in speaking since her

teeth had been removed (this had been done when in hospital). Her husband told me that perhaps she was rather clumsy sometimes, and would knock into door-posts occasionally, and trip on the stairs. Objective physical examination was still quite negative, except for the dysarthria which was more marked. Her memory, too, appeared to be worse. She was very vague as to when and for how long she had been in hospital.

Family History.—Patient's sister, a sensible North Country individual, and of especial value as a witness since she had a child who had suffered from Sydenham's chorea, provided the following family history. What she told me was supplemented by letters from her father and from Morpeth Mental Hospital. She had known two cases personally—those of her mother and of another sister.

(1) It was found impossible to trace the family further back than the *grandmother* (who married her cousin), and of her few details are available. Family tradition has it she was afflicted "in the same way." She died in Morpeth Asylum in 1863, her case being diagnosed as general paralysis. In view of the subsequent family history, it seems not unreasonable to assume that hers was in reality a case of Huntington's chorea.

(2) Coming next to the *mother* of the patient: The symptoms in this case started early in the fourth decade, and she died at the age of 49, having previously been bedridden for two years. She was never in an institution. At the end she had considerable dysphagia, and became very thin. Here again the first symptoms appear to have been changes in character, very similar to those of her daughter. My informant had to leave school when young, and when her mother was still in the thirties, in order to look after the home. Unlike the patient she, however, showed marked ataxia very early, and "reeled like a drunk." This became progressively worse, and for many years she also made a mess feeding herself. My informant was emphatic that her mother never made any choreiform movements (it will be remembered she had her own child to judge such movements by), or at most very slightly, and in her last years. She too was very garrulous and rambling in her talk, but never senseless. She was markedly dysarthric early, and for many years was intelligible only to her own relatives. She was usually apparently cheerful and contented, and never realised there was anything the matter with her. She became increasingly selfish and irritable, soon lost emotional control, and would fly into passions, "swearing like a trooper." Her memory was affected little, if at all. She was always a great reader, and remembered what she read.

(3) The *sister* of the patient first showed symptoms at the age of 24, and died 10 years later. She also was bedridden and emaciated towards the end. My informant assured me that all she had said of her mother was true in this case too. She also was markedly ataxic, dysarthric and garrulous, though always coherent. The whole course, however, appears to have been on a lower key; she was far less emotional, and more apathetic and depressed. She also never had any involuntary movements.

(4) The *uncle* of the patient the Morpeth authorities state to have been a classical and typical case of the disease. At the age of 24 he experienced some difficulty in walking and readily became tired. Two years later his hands became affected. His fingers were tremulous and he had difficulty in doing up buttons. His handwriting also became shaky. Simultaneously the muscles of his face and neck became involved. Admitted into Morpeth in 1898, he was a well-built man, six foot tall, and weighing over 14 stone. His speech was slurred, and he showed twitching movements of the fingers and toes. His tendon reflexes were very active. He was diagnosed as a case of general paralysis, and the diagnosis maintained for four years, when it was changed to Huntington's chorea. His legs became very unsteady and weak, and his articulation almost unintelligible. With the passage of years he became progressively worse, and the choreic movements were always exaggerated on making any effort. Eventually he became bedridden, and had to sleep on a mattress on the floor, as the movements were liable to fling him from an ordinary bed.

His mental state from the date of admission was one of progressive dementia. He died 11 years after reception, in the early forties.

The information as to the other affected members of the family is less ample.

(5) Patient's *aunt* died at the age of 44, after an illness of about 10 years' duration. She was never in an institution. She is said to have been markedly ataxic and dysarthric, and evidently deteriorated mentally.

(6) One of her three *sons*, now in the twenties, is also showing signs of the disease. My informant, apart from having heard from a relation that he was "beginning to go," knew no details, and was unwilling to give the address.

DISCUSSION.

Elaborate studies on an extended scale have, in recent years, been made in America. The mass of material investigated has been very large. Davenport and Munsey² have analysed 962 known choreics. Enquiry into the families involved the consideration of no fewer than 4,370 individuals. Estella Hughes⁴ has studied 218 cases and their families—1,308 relations in all. The conclusions reached are substantially the same.

They emphasise that the usual definitions such as "the insidious onset and progressiveness, in middle life, of involuntary jerky movements of the muscles in groups, accompanied by mental deterioration" need considerable amplification, for although the motor symptoms usually precede the mental, the reverse may occur; either may be present single; the disease is not invariably progressive, and the onset may be relatively early. Davenport and Munsey go so far as to cite cases in affected families where choreic movements were noticed soon after birth, and continued throughout life, without the supervention of further symptoms. The majority of cases, however, do conform to the general pattern, and have certain features in common, though individual families appear to have their own idiosyncrasies.

The disease in this country is certainly rather rare, although no statistical enquiry appears to be on record. Allbutt¹, even with his exceptional opportunities, stated in a demonstration that he could only call to mind having seen four such cases before, and one he quotes was seen in Italy. In America it is much more common. For example, it accounts for 1.9 per 1,000 of admissions into Michigan State Hospitals⁴. For everyone institutionalised, it is estimated there are four others living at home, more or less socially incapacitated.

The majority of cases are drawn from rural communities, and from the small professional classes, and in general the families are respectable and industrious. The balance of evidence seems to be against any special neuropathic manifestations in tainted families. Hamilton³, in a study of 27 cases, found such traits "conspicuously absent" and Rosanoff⁴ considers them to be so infrequent as to be accounted for by coincidence. With this view Hughes appears to agree.

The heredity is direct, that is, passes through generations without a break, and does not ordinarily appear in the children whose parents have been free

from chorea unless these parents died before the age of incidence ; it is suggested that the apparent exception to direct descent could be explained away, for the simplex carrier of a dominant trait might well show that trait imperfectly developed, or even, rarely, not in a recognisable form. There appears to be no means of foretelling which members of any given family will become affected. On an average half the children will develop the disease. The sexes are affected equally. The usual age of onset is between 30 and 45, but the widest variations have been recorded. It is perhaps of interest that Davenport and Munsey consider that evidence adduced in favour of antedating, as generations pass, is founded upon a statistical fallacy.

The motor choreiform symptoms are well known and very characteristic, and when well developed can hardly be mistaken for those found in any other disease. They need not be gone into here. Ataxia may be a prominent feature. Similarly the mental changes are on the whole distinctive. The deterioration is more pronounced in the affective sphere than in the intellectual. The patients usually show unconcern and inertia, and the statement that a "weakness in judgment and initiative, absent-mindedness, a growing selfishness and irritability, are amongst the earliest symptoms" is usually true. There is a notable lack of true appreciation of the disease. Only five times out of the 218 cases of Hughes is attempted suicide recorded, in spite of the emphasis laid by many authors (such as Stoddart and Craig and Beaton in their text-books) upon depression as a common symptom. Quite a large proportion show paranoid delusional formations ; hallucinatory states and schizophrenic reactions are sometimes seen.

The average duration of the disease is from 10 to 15 years, though very wide variations can occur (1 to 37 years and more).

It will be seen how strikingly the family here recorded exemplify the points given above. It springs from a small yeoman stock, respectable and industrious. No members, apart from those afflicted, have shown any marked mental abnormality. It exhibits direct descent very clearly ; no member not descended from a tainted parent has developed the disease. Roughly half the children in each generation have been affected. Neither sex is exempt. Two only out of the seven have been, so far, institutionalised. The known cases are said not to have been in any way remarkable before they developed symptoms. The average age of onset has been just under 30, with limits of 36 and 24. The average duration is about 10 years.

The patients have commonly shown inertia, unconcern and irritability, and mentally seem to be in no way remarkable. The family appears, however, to have been rather exceptional in that the most striking motor manifestation—choreic movements—has only been conspicuously present in one member. In all dysarthria has been a prominent symptom, and in the case personally observed this has been the sole striking motor disorder, even though marked mental deterioration has taken place. Moreover, it is noteworthy that two members

of the family were actually diagnosed as suffering from general paralysis, and doubtless the same mistake would also have occurred in the case here recorded, were the family history and negative serological finding not available, for the clinical picture was certainly suggestive of that disease.

DIAGNOSIS.

When the motor symptoms predominate, cases clinically indistinguishable have rarely been observed in certain post-encephalitics^{5, 7} and sufferers from neurosyphilis⁷.

In these coming under observation primarily because of behaviour changes and mental deterioration, the diagnosis must be made from general paralysis (as in this family), and that very difficult group, the presenile arteriosclerotic degenerations. Against the latter will, of course, be the absence of definite changes in the vessel walls.

In all, the distinctive heredity will finally clinch the diagnosis.

SUMMARY.

1. A case of Huntington's chorea has been described, and the family from which the patient springs investigated.

2. The case is of interest because of the absence of motor symptoms, with the exception of dysarthria, although marked mental deterioration had taken place. Two members of her family had in the past been diagnosed as cases of general paralysis, and, clinically, this case also simulated that condition.

3. A brief summary of the common features of Huntington's chorea is given, drawn from detailed American investigations.

4. It is pointed out that even the complete absence of motor symptoms need in no way negative the diagnosis of the disease.

5. The distinctive heredity will in doubtful cases settle the diagnosis.

I must thank Dr. Anthony Feiling for permission to publish the case, which was in the Maida Vale Hospital under his care, and Dr. East, Superintendent of Morpeth Mental Hospital, for kindly supplying details of the case which was in that institution.

REFERENCES.

- ¹ ALLBUTT, C., *Brit. Med. Jour.*, 1918, i, 389.
- ² DAVENPORT and MUNSEY, *Amer. Jour. Insan.*, 1916, lxxiii, 195.
- ³ HAMILTON, *Amer. Jour. Insan.*, 1908, lxiv, 403.
- ⁴ HUGHES, E., *Amer. Jour. Psychiat.*, 1924-5, iv, 537.
- ⁵ LHERMITTE and LAMAZE, *Revue neurol.*, 1920, xxvii, 768.
- ⁶ ROSANOFF, quoted by Hughes (ref. 4).
- ⁷ JELLIFFE and WHITE, *Diseases of the Nervous System*, 1929, 669.