GENERAL PARESIS COMPLICATING HUNTINGTON'S CHOREA

By

H. MERSKEY, M.A., B.M., B.Ch., D.P.M.

Assistant Psychiatrist, Cherry Knowle Hospital, Ryhope, Nr. Sunderland

THAT chorea may sometimes occur as a result of cerebral syphilis has been accepted for some years (Kinnier Wilson 1940) and it has often previously been reported in association with G.P.I. The literature on the topic is dealt with by Lowrey and Smith (1918), Stone and Falstein (1938) and Weickhardt (1945). In all the reports however I can find only three previous instances where clinical and serological evidence of G.P.I. in a patient was associated with a family history and clinical syndrome of Huntington's chorea. These cases were reported by Pagliano and Avierinos (1922), Urechia and Rusdea (1922) and Stone and Falstein (1938) and the case here described appears to be only the fourth similar example to be reported. It has a practical as well as an academic interest since the differential diagnosis of the early stages of Huntington's chorea from those of G.P.I. is still sometimes necessary.

CASE HISTORY

C.E. Housewife. First admitted to hospital in 1942, aged 34 years. Her father died in a mental hospital aged 38 years. He was said to have the physical signs of G.P.I. and no particular evidence of chorea was recorded in his case. Her brother, J. McA., is now a patient in Cherry Knowle Hospital. He has shown a personality change since 1939 and is now demented and subject to choreic motor disorder and involuntary movements. He shows no signs of syphilis and his cerebrospinal fluid is normal as well as his blood Wassermann reaction and Kahn test.

The patient herself had been "not well" for four years prior to her admission to hospital in 1942. She had been a poor scholar and always illiterate but cared for her four children until she developed headaches and inability to walk, diagnosed at a general hospital as neurotic. At the time of first admission she was obese and infected with scabies. The pupillary reflexes were normal and there was no abnormality recorded in the central nervous system except "pseudo-clonus" at the right ankle and a mild symmetrical increase in the tendon-reflexes in the legs. The record of the Wassermann reaction in the blood has been lost but it was negative in the cerebrospinal fluid. The Lange colloidal gold curve read 2221110000. She was then regarded as basically feeble-minded with hysterical and schizoid features. She was dull, quarrelsome and not very clean but worked and occupied herself when given clear instructions.

She was re-admitted to hospital in 1943 in a similar state, being then depressed and tremulous as well, and she remained in hospital from that time. In 1947 she was still able to occupy herself in the laundry but she had some ataxia and fell unduly often. Her pupils by then were irregular and unequal and did not react to light, her speech was slurred, tendon reflexes were increased in the legs and there was bilateral ankle clonus with flexor plantar responses. The Wassermann reaction in the cerebrospinal fluid was strongly positive and the Lange curve read 5554321000. She then received 5 intramuscular injections of a bismuth preparation and a course of 5 megaunits of penicillin. In the two months subsequently she showed little change but an inoculation with malarial blood was then successful in producing rigors and she experienced twelve bouts of pyrexia exceeding 103°F. The rigors were terminated effectively with quinine and she appeared more settled. Although jerky in her movements her ataxia was improved but she remained untidy and dishevelled and liable to snatch food from other patients.

In March 1949, six months after the last observations and nine months after ceasing malarial treatment, she appeared worse. She was confused and staggering and had much coarse tremor of her arms. In 1950 she was demented and in need of constant nursing supervision and showed jerky spastic movements. In 1951 weakness of the facial muscles, particlarly levator palpebrae, was recorded together with "trombone

tongue", ataxic gait and flexor plantar responses. In March 1952 the dementia and physical deterioration were still progressing and she was inaccessible to conversation; she was also very restless and the presence of choreiform movements was recorded in the case-notes.

In May 1952 she received 1,800,000 units of crystalline penicillin and a further 2,000,000 units of "Distaquaine" penicillin for the treatment of a cutaneous infection but her movements continued to be severe until her death after increasing enfeeblement in December of that year. The death was certified at the time as due to General Paralysis.

Comment

A re-appraisal of this case was only made some time after her death and following on the diagnosis of Huntington's chorea in her brother. Even allowing for her poor original intelligence, however, it is clear that there was clinical evidence of mental and physical changes developing at a time when the Wassermann reaction in the spinal fluid was negative and this evidence together with the onset of her first symptoms at the age of thirty and the long course of her illness supports the additional diagnosis of Huntington's chorea in her case. Further when the signs of syphilis were established she received treatment which seems to have affected the progress of the tertiary infection and even appeared to produce some temporary improvement. Subsequently, however, an increasing dementia again became manifest together with progressively more numerous jerky movements until finally the choreic signs were the predominant ones, and this chorea continued to become worse despite coincidental treatment with a second course of penicillin. It is likely therefore that Huntington's chorea was the cause of her presenting symptoms and of her ultimate physical disabilities and we may reasonably suppose that some of the changes of G.P.I. were superimposed upon the Huntingtonian syndrome. Such a conclusion emphasizes the importance of the serological examinations for syphilis in chronic chorea even where a confirmed family history of Huntington's chorea is to be found.

Acknowledgments

My thanks are due to several colleagues and particularly to Dr. G. Fitzpatrick, Medical Superintendent, Cherry Knowle Hospital, for helpful comments and to Miss A. Wilson for secretarial assistance.

References

Kinnier Wilson, S. A., *Neurology*, 1940, **1**, 483.
Lowrey, L. G., and Smith, C. E., *Amer. J. Syph.*, 1918, **2**, 453.
Pagliano, V., and Avierinos, F., *Marseille méd.*, 1922, **59**, 78.
Stone, T. T., and Falstein, E. J., *J. Nerv. Ment. Dis.*, 1938, **87**, 450.
Urechia, C. J., and Rusdea, N., *Rev. Neurologique*, 1922, **29**, 513.
Weickhardt, G. D., *Urolog. cutan. Rev.*, 1945, **49**, 6.