Huntington's disease in Tanzania

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SUMMARY Huntington's disease was studied in a Bantu community in northern Tanzania. Although there is evidence to suggest that the disease has been present here for over one hundred years, this is the first report of the condition in Tanzania. A survey of published reports indicates that the disease is infrequently reported in persons of Negro ancestry.

Huntington's disease is transmitted as a Mendelian autosomal dominant trait and typically presents as chronic choreoathetosis starting in early to middle adult life, accompanied by progressive dementia. The disease causes great distress to the patient and family. Although it is a relatively rare disease, it is important that its presence in a community is recognised, for its transmission can be prevented by skilled genetic counselling.

Family studies

The family pedigree is depicted in the figure and this indicates that an autosomal dominant trait is present. All family members are Chagga, a Bantu group resident on the lower slopes and adjacent countryside of Mount Kilimanjaro. As a result of the wide dispersal of the family throughout Tanzania, and the reluctance of members to be assessed, only two patients could be examined personally. However, subject IV·15, the son of one of these subjects, was a well-educated man who could speak fluent English. He had a wide and detailed knowledge of the family, and proved to be an accurate and reliable observer.

Case reports

Subject III·6 was a male aged 63 years. Grimacing and involuntary movements were first noted in 1959. He continued to work in his general store until 1974 when increasing memory loss and difficulty with calculation became prominent. He later ceased to deal directly with customers but continues to assist in running the store. On assessment he was noted to have grimacing with pouting and smacking of the lips. Vertical conjugate gaze and rapid eye movements were impaired. Mimicral apraxia was demonstrable. He had moderate dysarthria and speech was interrupted by intermittent grunting. Coarse

generalised choreoathetosis was present and a repetitive pattern of pronation and supination of the left forearm with extension and flexion of the left forefinger was observed. The gait was ataxic with a dancing quality. There was no evidence of cerebellar or pyramidal tract dysfunction. Moderate dementia was apparent. Memory and intellectual functions were impaired and he was suspicious and unco-operative. Standards of attire, personal care, and cleanliness were poor and inappropriate for a once-successful businessman.

Subject III·10 was a male aged 58 years. Grimacing and choreiform movements started in 1964 when he was employed as a headmaster. His subsequent deterioration with increasing ataxia and mental impairment resulted in early retirement. On examination, he had a vapid expression with sporadic grimacing, pouting, and wry movements of the mouth. Ocular dyspraxia was evident. Speech was markedly dysarthric. Coarse choreoathetotic movements of limbs and digits were present and his gait was ataxic. Assistance was required to prevent falls. There was no abnormality of cerebellar or pyramidal tract function. Dementia was gross. His Swahili was sparse and barely comprehensible. He was apathetic and inert but was co-operative when examined and obeyed simple commands. Several weeks after discharge from hospital he died and was interred in his village. News of this event arrived one week later and necropsy could not be performed.

Subject II·1 was a male aged over 80 years. He was reported to have had sparse choreiform movements and grimacing for many years. He was notorious for his irascibility and refused to be interviewed by a doctor or to provide information about his family and ancestry.

Subject III·1 was a male aged about 60 years. He was reported to have had grimacing, involuntary movements, dysarthria, and ataxia since middle age. Memory and intellectual function were impaired.

FIGURE Family pedigree.

Subject III·2 was a male aged about 58 years. Grimacing and choreiform movements had developed in his forties, and later deterioration of memory and intellect were noted. He was now unable to walk because of severe ataxia and his speech was incomprehensible.

Subject IV·3 was a male aged 35 years. He was noted to have had grimacing and some choreiform movements for several years. He was otherwise normal and performing his duties as a district education officer. It is believed that he and his wife recently separated.

Subject IV·7 was a male aged 32. He had been observed to have had occasional grimacing and involuntary movements, but was believed to be otherwise well.

Subject IV·34 was a male aged 25. Grimacing and occasional choreiform movements had been pre-

sent for 2 years, but he was believed to be otherwise normal.

Family cases

Subject III·4 developed chronic choreoathetosis with grimacing, dysarthria, and ataxia in his early forties. He later became demented and died in 1969 aged 55 years. *Subject III·8* developed a similar illness when he was aged about 40 and he too became demented. He died in 1974 aged 56 years. *Subject II·6* was believed to have developed an illness similar to that exhibited by subjects III·6 and III·10 and to have died in late middle life. *Subject II·5* had a large family by his first wife and no case of Huntington's disease has appeared in her progeny. No information could be obtained regarding subjects I·1 and I·2.

Discussion

The mean age of onset of Huntington's disease in large series varies from 35.5 to 43.2 years.^{1,2} Consideration of the available evidence in regard to eight adults in this study suggests a mean age of onset of 36 years. In three adults, the mean duration of disease was 15 years. Two living choreics have been suffering from the illness for 18 and 20 years, respectively. The average age at death of the three adults was 56 years. These figures are similar to results reported in other series in which the mean duration of clinical illness varied from 10.5 to 16 years,^{2,3} and the average age at death was 55 years.⁴ In view of his advanced age and relatively mild clinical presentation, subject II.1 appears to be an example of the non-progressive disease seen in some elderly patients.⁴ There were no reports of cases suggestive of juvenile disease in the present study. The prevalence rate for this community is approximately 7 per 100 000.

The diagnosis of Huntington's disease in this community is supported by the presentation of chronic choreoathetosis accompanied by progressive dementia, starting in early to middle adult life, together with evidence of autosomal dominant inheritance. These three features together help to differentiate the disease from the many other conditions in which choreiform movements may be observed. These include Sydenham's chorea in association with rheumatic fever, and hepatolenticular degeneration (Wilson's disease). There was no evidence to suggest these conditions in this study, and in the two patients personally examined, liver function was normal and Kayser-Fleischer rings in the cornea were not observed. Tardive dyskinesia which may complicate treatment with various drugs, including phenothiazines and butyrophenones, may be considered. The only patient known to have been exposed to one of these drugs was subject III.10 who had been treated with intermittent courses of haloperidol in recent years. However, his history and clinical presentation were typical of Huntington's disease and he had marked dysarthria and ataxia which are not features of tardive dyskinesia. In presenile dementia, although familial cases have been reported, a hereditary pattern is absent and choreoathetosis is only occasionally observed in advanced disease.

Huntington's disease has been reported from most ethnic groups including Negroes although it is reputedly rare in this race.⁵ There have been reports of the disease in Negroes in the USA.^{6,7} In Africa a case was reported in a Kikuyu male in Kenya in 1936.⁸ Two further cases were reported in Kenya by Harries.⁹ In Uganda there was an isolated report of a

case in an African in 1956.¹⁰ In Zimbabwe, three cases were described in a Shona family although a positive family history could not be demonstrated.¹¹ In South Africa the disease was observed in a Zulu family.¹² It was not encountered by one clinician in Natal.¹³ However, the prevalence of Huntington's disease is moderately high among Afrikaners having been introduced from Holland in the 17th century.¹² There have been reports of the disease in families of mixed ancestry in South Africa.^{5,14} In Ghana it is reputedly rare.¹⁵ Three cases have been reported from Nigeria.¹⁶ In Trinidad, the disease has been recognised in persons of apparently unmixed African ancestry.¹⁷

The present report appears to be the first study of Huntington's disease in Tanzania. A previous survey of neurological disease in Tanzania indicated that the disease had not been recorded in this country.¹⁸

It has been shown that the abnormal gene rarely develops by mutation and is almost invariably introduced into a family.¹⁹ Subjects I.1 and I.2 were presumably born in the 1870s. This was before the establishment of European missions and settlements in the interior, which only began after the Congress of Berlin in 1884 when the area came under German sovereignty. Before this date, the Chagga territory was only rarely visited by Europeans. However, some of the principal safari routes from the coast, 250 kilometres away, led through this region, and by 1825 the Chaggas were provisioning caravans and trading in slaves and ivory. Caravans were usually led by Arabs and the porters were mainly recruited at coastal towns where many of the inhabitants had varied ancestry as a result of contact and intermarriage with foreign seafarers and traders. The East African coast had been regularly visited by shipping from early historical times and in recent centuries the Portuguese, Turks, Dutch, and Omani Arabs had in turn attempted to dominate coastal trade. In 1820, England was the strongest naval power on the East African coast. There is no tradition in the family of intermarriage with foreigners, and all family members appear to be typically Bantu in appearance. However, it is possible that at some remote time, the anomalous gene was introduced at the coast, and later transferred to the interior, perhaps by a member of a safari.

Within this Bantu community, attitudes to Huntington's disease are similar to those of affected families elsewhere. Members of the family clearly identify the condition as a unique and dreaded disease. Close relatives are fearful that they may contract the condition. There is a widespread belief that it is caused by witchcraft and it is usual for the family to consult traditional healers for treatment and charms. There have been no reports of suicide within

the family. It is hoped that a long term programme to identify new cases and to offer genetic counselling to the individual families will be instituted.

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