

A familial cluster of Parkinson's disease identified in Milne Bay Province, Papua New Guinea

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SUMMARY

Parkinson's disease is a chronic debilitating condition, the prevalence of which has not been fully established in Papua New Guinea. We describe a cluster of 9 cases of the disease, restricted to two generations of one family, and the key ideas and beliefs held within the family regarding disease aetiology. Many of the concerns and feelings of guilt expressed by family members were alleviated following supportive listening and culturally appropriate counselling, explanation and advice from trained health professionals assisted by bilingual family facilitators. This is the first time that such a family has been reported in Papua New Guinea and may warrant more detailed assessment. Addressing patient and community perceptions of disease aetiology should be at the heart of health promotion initiatives and counselling.

Introduction

Parkinson's disease (PD) affects approximately 1% of the population over 50 years of age in the developed countries of the world (1) but there is little information on its prevalence in countries of the South.

The aetiology of the disease is complex with multiple genetic and environmental factors likely to be involved (2).

Environmental factors were favoured throughout the 1980s by many authors (3,4) particularly after evidence that intravenous drug addicts exposed to the pethidine analogue 1-methyl-4-phenyl-1,2,3,6-tetrahydropyridine (MPTP) developed a condition almost indistinguishable from classical PD (3,4). Cycad use and exposure to isoquinoline derivatives and other environmental toxins have also been associated with neurodegenerative conditions, including parkinsonism (5-7).

An increasing number of families containing

multiple PD cases have been identified during the last 10 years (1,3,8,9). Recently, genetic markers on chromosome 4q21-q23 were linked to the PD phenotype in one large family sharing a common ancestor who lived in Contursi, Italy, in the 18th century (1). Autosomal dominant inheritance with incomplete penetrance has been identified in other families (4,8).

Parkinsonism has been reported in Papua New Guinea in the past (10), but multiple cases occurring within a single family have not previously been described. The objective of this study was to investigate cases of PD within a known multicaser family living in Gaunani village in the isolated, mountainous interior of Rabaraba District, Milne Bay Province.

Cases of Parkinson's disease were first recognized in this family during the 1980s, but no formal clinical assessment has previously been conducted. In addition we wished to quantify prevailing ideas and beliefs held within the family as to the aetiology of the condition and to provide culturally appropriate,

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accurate health information and family counselling.

Methods

Two non-affected members of the family were identified and interviewed at Agaun Health Subcentre, which is adjacent to Gaunani village. The nature of the disease and objectives of the study were explained to the two facilitators, who returned to the village and arranged for all family members living in the village to meet with investigators on the day of the study. We carried out an unstructured natural group interview followed by semi-structured interviews with individual family members. A family tree was constructed. Neurological assessment, including history taking and neurological examination, was carried out (by AV) on all individuals.

Cases of Parkinson's disease were defined according to standard neurological reference texts (9,11).

Beliefs within the family about the origins of the disease were appraised and discussed. Key health information messages and genetic counselling were provided in ways that were sensitive to prevailing cultural and religious ideas, with facilitators acting as interpreters where necessary.

Results

Cases of Parkinson's disease identified and clinical features

All members of the family living in Gaunani village were seen on the day of the study. Some family members from Bonenau village, a 3-hour walk away, also attended. 6 cases of PD were identified, all of whom were already receiving benzhexol therapy, the only readily available therapy at that time. No additional cases were found among the remaining 22 adults examined and no children suffering from neurological disease were identified.

It is likely that 3 other family members had the disease prior to their deaths during the 1980s because they appear to have experienced typical symptoms of the disease, according to other members of the family (Figure 1). It is difficult, however, to assess the validity of this

historical evidence; recall bias may have been significant given the nature of the study.

Discussion with the family suggested that no other cases existed among family members living away from the area.

The cases identified had features typical of the disease (9,11), with pronounced pill-rolling tremor, rigidity and bradykinesia, despite benzhexol therapy (Table 1). The average age of onset of PD in the family was 29 years (range 21-42 years), with pronounced rigidity in 5 of the 6 cases. Among children in the family, 43 were aged 21 years or over at the time of the study, with 19 aged 29 years or over.

Family beliefs regarding disease aetiology

Communities in this area are strongly supportive of the Anglican Church and its work in education, health and other activities. Christian religious beliefs provide a context in which to frame other ideas, perceptions and beliefs, with health and disease causation sometimes being viewed within this overarching belief system.

A number of key themes were identified:

- The disease is a punishment due to some wrongdoing by a family member in the past, or a curse or form of witchcraft placed on the family by another family or by a sorcerer living in the village or possibly in another village close by;
- Many felt guilty, as if the disease was a punishment from God, the reason for which was unclear;
- The disease is due to some sort of poison in the stream from which drinking water is drawn, or something present in their food or in the soil in which vegetables are grown;
- The family is the only family known with this condition in Papua New Guinea or anywhere else in the world;
- All family members might one day get the disease since the disease lives inside them all.

The family had not received counselling or advice regarding the disease in the past and both the investigators and the family

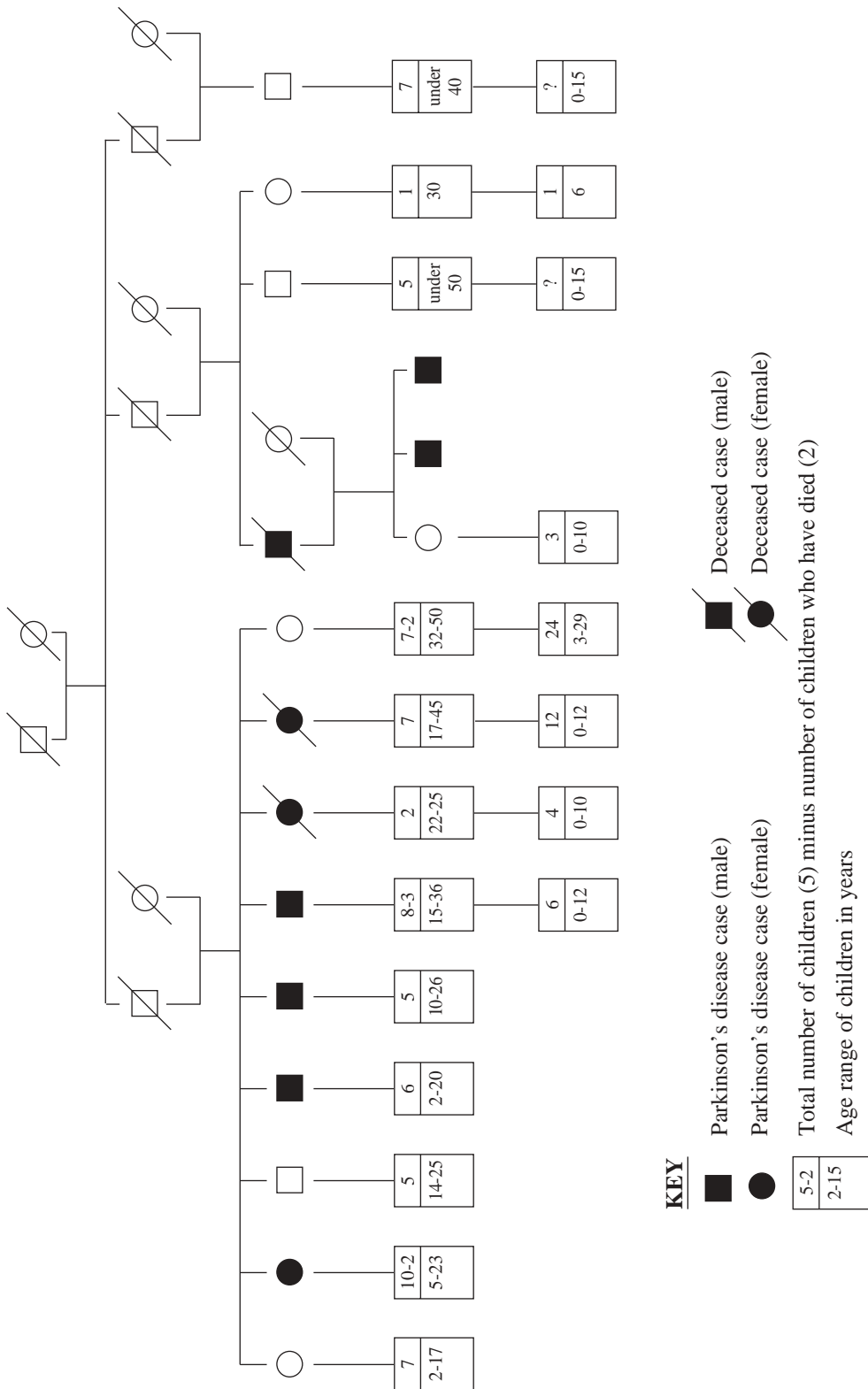


Figure 1. Familial cases of Parkinson's disease, Gaunani village, Rabaraba District, Milne Bay Province.

TABLE 1

SUMMARY OF THE CLINICAL FEATURES OF PARKINSON’S DISEASE CASES IDENTIFIED IN GAUNANI VILLAGE

Patient gender	Age at time of study (years)	Age at onset of symptoms (years)	Year of onset of symptoms	Preceding history of encephalitis	Typical clinical features of PD	Pronounced rigidity (+) or not (-)	Other features* present (+) or absent (-)
M	70	42	1969	No	Present	+	-
M	54	27	1970	No	Present	+	-
M	50	34	1982	No	Present	+	-
F	43	21	1975	No	Present	-	-
M	48	29	1978	No	Present	+	-
M	44	21	1974	No	Present	+	-

* Other features assessed:

- Presence of dementia
- History of urinary incontinence
- Presence of Keyser-Fleischer rings or other features of Wilson’s disease
- Evidence of severe autonomic nervous system dysfunction

facilitators felt it essential to address their concerns.

Health information and counselling were provided in small groups and carried out in a number of family homes. We provided information in ways that were sensitive and respectful of the ideas and concerns raised and that were appropriate, both in terms of prevailing local customs and the background of Christian religious belief. The family facilitators assisted in this process. A larger question and answer session was also held in the village meeting place for the family and the wider community.

Discussion

The relatively young age at onset and pronounced clinical features of rigidity are atypical findings that are in keeping with other multicase families described elsewhere (1,3). If we assume that genetic factors predominate in the aetiology of the condition in this family, the risk of first-degree relatives developing the disease may be around 10-15% (3). This has obvious implications given the number of relatives aged 21 years or over.

The data only partly support this assumption, however. There have been no additional cases in the family since 1982 and

there is no history of affected individuals in the first two generations of the family. The 6 cases confirmed during the study all developed symptoms within a 13-year period. It is possible that they were exposed to a common environmental toxin at some point in the past, the toxin having a variable latency period before producing clinical disease. Slow toxins and long-latency neurological disease have been described in Guam, the western Pacific and elsewhere (6,7). A change in diet prompted by crop failure or famine, or the use of traditional herbal medicines may have provided this common exposure. Certainly, cycads have been used in this area in the past as a famine food and in the treatment of yaws (12).

Many family members were greatly reassured by the health promotional work carried out as part of the study. In particular, many were relieved to hear that they were not alone in suffering from this condition and that the perceived universal risk to first-degree relatives of developing the disease was unfounded. The burden of guilt experienced by many people was lifted following this work.

Patient and community perspectives of disease aetiology need to be at the heart of health promotion initiatives, which should also ensure that adequate attention is given to

cultural and religious influences on illness behaviour (13). In this paper we have illustrated how such beliefs had been overlooked in the past, despite the best intentions of the health workers involved in providing care for this community.

This is the first time that familial aggregation of PD has been reported in Papua New Guinea. This family may warrant more detailed assessment and investigation, particularly in view of recent advances made in the understanding of this disabling disease (1,3,8,9).

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Consent for publication was gained through the family facilitators by formal, verbal agreement. Many family members expressed the desire that their story and experiences be more widely told.

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