Cortical basal ganglionic degeneration presenting with "progressive loss of speech output and orofacial dyspraxia"

Tyrell et al recently described 3 patients with progressive loss of speech output combined with pronounced orofacial apraxia.1 Cases 1 and 2 had been symptomatic for only three years, and case 3 had had symptoms for six years. In addition to the speech disturbances, case 3 also demonstrated dyspraxia of the limbs as did their case 4 in another recent publication,2 a patient who had been symptomatic for only two years and six months. CT scanning revealed asymmetrical cortical atrophy and PET scans demonstrated profound reduction in frontal lobe metabolism. The authors discussed this clinical entity as it relates to other "focal cortical degenerations" which have a somewhat heterogeneous underlying pathology.

One important clinicopathological entity that the authors overlooked in their discussions, which I believe may account for one or more of their cases, is cortical-basal ganglionic degeneration (CBGD).3,4 Although our group and others have pointed out that this disorder usually begins with asymmetrical limbic, cortical and/or basal ganglionic dysfunction, I have now seen patients who present with progressive loss of speech output and orofacial apraxia identical to the cases described by Tyrell et al.1 The delay between the onset of speech symptoms and more typical clinical signs has been as long as five to six years. One such patient, to be reported in greater detail elsewhere, has died recently and pathological confirmation of CBGD has been obtained. This patient was not included in our series of 15 cases of CBGD3 because at that time his signs and symptoms, which had been present for five years and six months, were restricted to those described in the cases of Tyrell et al.

Briefly, this 73 year old male developed difficulties pronouncing selected words at the age of 64. This problem slowly progressed over years to the point that it was difficult even for his wife to understand him. Despite his speech problems he had no other complaints. Eating was unimpaired and he maintained an active exercise programme comprised of swimming, cross-country skiing and walking. At the age of 69, when I first saw him, he demonstrated markedly impaired speech and almost all words were unintelligible. The mouth was held open much of the time. There was severe apraxia for all movements of the lower face and tongue. He had occasional but inconsistent difficulty performing or maintaining eyelid closure and eccentric gaze. There was an inability to suppress blinking in response to glabellar tap (Meyerson's sign), an easily elicitable jaw jerk, snout and palatalmy reflexes. The remainder of the neurological examination at that time was entirely normal. Investigations were unrevealing and PET scanning using both 6[18F] fluoro-Levadopa and [18F] fluoro-deoxyglucose was essentially normal.

Over the next four and a half years he became anarthric and other bulbar functions became involved, eventually necessitating feeding gastrostomy. He developed stimulus sensitive myoclonus of the right side of the face only and a severe akinetic-rigid syndrome with pronounced limb apraxias resulting in him becoming bedbound and unable to care for himself or even to turn his head or aspirate when pneumonia. Pathological assessment revealed the classic changes of CBGD.1,4

Our preliminary PET results using fluorodeoxyglucose, rather than 11C as used by Tyrell et al1 indicate that bifrontal hypometabolism is not a universal feature of patients presenting with progressive loss of speech output and orofacial dyspraxia. The changes described by Tyrell et al were most marked in the inferior and lateral portions of the frontal lobes with some extension into the parietal and temporal cortices. It will be interesting to determine if the early presence of these changes predict the subsequent course, and even underlying pathological disturbances, if more than one disease state can result in the same clinical picture. However, clinicopathological correlates will be required of similar patients before this can be resolved. Posterior frontal hypometabolism (again using 11C) as described by Sawle et al in CBGD4 was also absent in my patient at a time that his symptoms were limited to cranial structures. This indicates that the pattern of disturbances found by Sawle et al cannot be used as a diagnostic marker for this disorder at all stages of development. The failure of fluorodeoxyglucose scans to demonstrate definite clinical evidence of nigral dopaminergic pathology, which to date has been invariably present in CBGD, is extremely disappointing. 18F-dopa scans were done at a time when we were not able to quantify 18F uptake. For this reason it is impossible to exclude a mild but significant generalised and symmetrical reduction in striatal accumulation. However, the prominent reductions found by Sawle et al, which were strikingly asymmetrical, were not seen. Further quantified analyses will be required in CBGD patients with isolated speech disturbances before this issue is resolved. Our results indicate that 18F-dopa PET scanning may not be a reliable marker of this disorder in its earliest stage. Unfortunately, no other diagnostic or predictive tests are available in CBGD and brain biopsy usually fails to reveal the classic pathological changes. The lack of striatal dopaminergic changes in this patient may suggest that nigral degeneration may be a later developing and rapidly progressive feature in some CBGD patients. This contrasts with the slowly progressive, prolonged presynaptic nigral degeneration proposed in idiopathic Parkinson's disease when 18F-dopa scanning might serve as a more reliable marker of very early disease.

The clinicopathological experience described here emphasises the need for caution and restraint in reporting patients at a relatively early stage in the course of a progressive "degenerative" disorder. It will be extremely important for Tyrell and colleagues to provide a follow-up report on the course of their patients over subsequent years.

MATTERS ARISING

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Rossor and Tyrell reply;

We note with interest Dr Lang's comments and details of his patient with cortical basal degeneration who also presented with progressive loss of speech output and orofacial apraxia. Lippa et al4 have also reported a case of primary progressive dysphasia with cortical basal degeneration and this should certainly now be considered as a neuropathological substrate of focal degenerations as suggested by Dr Rossor. We recognise the importance of follow up of our own cases and have presented preliminary data on the neuropsychological features in some of these cases, and to date Pick's disease is the most frequent association.

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