

MATTERS ARISING

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

Tyrrell *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 while 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 group⁵ and others⁶ have pointed out that this disorder usually begins with asymmetric limb 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 Tyrrell *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 elsewhere⁷ has died recently and pathological confirmation of CBGD has been obtained. This patient was not included in our series of 15 cases of CBGD⁵ 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 Tyrrell *et al*.

Briefly, this 73 year old male developed difficulties pronouncing selected words at the age of 64. This problem slowly progressed over the next five 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, crosscountry 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 palmonental reflexes. The remainder of the neurological examination at that time was entirely normal. Investigations were unrevealing and PET scanning using both 6[¹⁸F] fluoro-Levodopa 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. He died of an aspiration pneumonia. Pathological assessment revealed the classic changes of CBGD.^{3,4}

Our preliminary PET results using fluoro-deoxyglucose rather than ¹⁵O as used by Tyrrell *et al*¹ 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 Tyrrell *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 correlative studies will be required in a number of similar patients before this can be resolved. Posterior frontal hypometabolism (again using ¹⁵O) as described by Sawle *et al* in CBGD⁵ 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*⁶ cannot be used as a diagnostic marker for this disorder at all stages of development. The failure of fluoro-dopa scans to demonstrate definite pre-clinical evidence of nigral dopaminergic pathology, which to date has been invariably present in CBGD, is extremely disappointing. ¹⁸F-dopa scanning was 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*,⁶ 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 pre-clinical nigral degeneration proposed in idiopathic Parkinson's disease where 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 when reporting patients at a relatively early stage in the course of a progressive "degenerative" disorder. It will be extremely important for Tyrrell and colleagues to provide a follow up report on the course of their patients over subsequent years.

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1 Tyrrell PJ, Kartsounis LD, Frackowiak RSJ, Findley LJ, Rossor MN. Progressive Loss of Speech Output and Orofacial Dyspraxia As-

- sociated with Frontal Lobe Hypometabolism. *J Neurol Neurosurg Psychiatry* 1991; 54:351-7.
- 2 Tyrrell PJ, Warrington EK, Frackowiak RSJ, Rossor MN. Heterogeneity in Progressive Aphasia Due to Frontal Cortical Atrophy. *Brain* 1990;113:1321-6.
- 3 Riley DE, Lang AE, Lewis A, *et al*. Cortical-basal ganglionic degeneration. *Neurology* 1990;40:1203-12.
- 4 Gibb WRG, Luthert PJ, Marsden CD. Corticobasal Degeneration. *Brain* 1989;112:1171-92.
- 5 Rosenfield DB, Bogatka CJ, Viswanth NS, Lang AE, Jankovic J. Speech Apraxia in Cortical-Basal Ganglionic Degeneration. *Ann Neurol* (Abst in Press).
- 6 Sawle GV, Brooks DJ, Marsden CD, Frackowiak RSJ. Corticobasal Degeneration. A Unique Pattern of Regional Cortical Oxygen Hypometabolism and Striatal Fluorodopa Uptake Demonstrated by Positron Emission Tomography. *Brain* 1991;114:541-56.

Rossor and Tyrrell 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 al*¹ 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 Lang. We recognise the importance of follow up of our own cases and have presented preliminary data on the neuropathological features in some of these cases, and to date Pick's disease is the most frequent association.²

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- 1 Lippa CF, Cohen R, Smith TW, Drachman DA. Primary progressive aphasia with focal neuronal achromasia. *Neurology* 1991;4:882-6.
- 2 Kennedy AM, Tyrrell PJ, Warrington EK, *et al*. The clinical metabolic and histopathological correlates of orofacial apraxia. *J Neurol Neurosurg and Psychiatry* (Abst) 1991.

BOOK REVIEWS

All titles reviewed here are available from the BMJ Bookshop, PO Box 295, London WC1H 9TE. Prices include postage in the United Kingdom and for members of the British Forces Overseas, but overseas customers should add £2 per item for postage and packing. Payment can be made by cheque in sterling drawn on a United Kingdom bank, or by credit card (Mastercard, Visa or American Express) stating card number, expiry date, and your full name.

Recent Advances in Psychogeriatrics—2. Edited by TOM ARJE (Pp 224; Price: £29.95). 1992. Edinburgh, Churchill Livingstone. ISBN 0-443-04468-6.

It has been seven years since the first *Recent Advances in Psychogeriatrics*. A new volume is timely as there have been significant scientific advances and the roles of the social

and health services are undergoing major changes.

Professor Arie has selected a panel of distinguished contributors from home and abroad. The range of topics is appropriately wide and there is enough here to interest the academic as well as the jobbing psychogeriatrician and geriatrician. There are sensible reviews of ubiquitous, diffuse Lewy body disease, the potential biochemical tests for Alzheimer's disease and the epidemiology of dementia. Those interested in therapy however will have to wait for the next edition. More clinically orientated were the chapters on depression, mania and paranoid disorders.

A review by Walker on the social services, with particular emphasis on the individual and community care is especially topical: will they match vision with funding? Three chapters on carers emphasize the importance of listening to and caring for the carers in any good service. Dr Jolley and Professor Arie look at psychogeriatric services with reference to the international scene and the current political context: there is plenty here to discuss with one's colleagues and managers. An overview on audit shows that as in so many specialties it is still really only in its infancy, and no doubt this will be a chapter to repeat in a future edition, perhaps with a section on memory clinics? Sensitivity to the needs of the disabled in hospitals is a current issue and the chapter on designing for confused old people deserves to be read by managers and architects.

Required reading for all clinicians interested in the elderly.

JGC COX

Soviet Scientific Reviews/Section F: Physiology and General Biology Reviews Vol 5, parts 1-4. Part 1: Problems of Internal Inhibition, The Dominant and the Conditioned Reflex. Edited by GASSANOV/PAVLYGINA. (Pp 75; Price: \$20.00; £11.00) ISBN 3 7186 5160 2

Part 2: Relay Functions of Hippocampal Monoamines in Acquired and Inborn Forms of Behavior. Physiological Mechanisms of Complex Behavior in Anthropoids. Edited by GASANOV, ET AL/FIRSOV ET AL. ISBN 3 7186 5161 0. (Pp 83; Price: \$21.00; £12.00).

Part 3: A New Ideology of Studies of the Neurophysiological Correlates of Mental Activity. ISBN 3 7186 5177 7. Edited by MEDVEDEV/PAKHOMOV. (Pp 54; Price: \$14.00; £8.00).

Part 4: The Neural Mechanisms of Conscious and Unconscious Perception. By E A KOSTANDOV. (Pp 55; Price \$14.00; £8.00). ISBN 3 7186 5176 9. 1991. New York, Harwood Academic Publishers.

These four small booklets are edited by two members of the USSR Academy of Sciences, as it then was when the papers were commissioned and written. The four sections cover higher nervous activity. Part 1 is dedicated to the problems of internal inhibition and the dominant and conditioned reflex. The first paper gives an interesting history and analysis of the work of Pavlov and brings readers up

to date with suggestions for the neurophysiological basis of internal inhibition (a process that suppresses the elaborated reaction). The second paper of part 1 is a further discourse on conditioned reflexes with original data on goal directed reactions.

The second part of volume 5 discusses relay functions of hippocampal monoamines in acquired and inborn forms of behaviour and the physiological mechanisms of complex behaviour in anthropoids. The papers in this section contain original experiments on the effects on monoamines on hippocampal function. Previous work has suggested that an increase in the 5HT content of the hippocampus weakens memory and learning. Damage to 5HT containing neurones in the raphé nucleus accelerates the development and persistence of the conditioned reflex of active avoidance. The relevance of these observations may take on additional importance bearing in mind the provisional data suggesting some benefit of 5HT receptor subtype active drugs on cognitive function in pathological states.

The third part of this volume on higher nervous activity discusses studies of the neurophysiological correlates of mental activity and deals with methods of studying the brain's system for maintenance of "mental activity". The final part of this volume is on the neural mechanisms of conscious and unconscious perception and contains an interesting section on the functional relationships between the hemispheres and the unconscious.

The English translation of these papers is of a high standard but despite this I found the concepts elaborated difficult to penetrate. Nevertheless, there may be material here which some psychologists, psychiatrists and neurophysiologists may find of interest. To the general neurologist however, I suspect there will be little here to hold his attention. The papers in volume 5 reference material only up to 1986 and therefore these booklets are already significantly out of date. This may of course represent the difficulty of publishing scientific material in the old USSR.

AHV SCHAPIRA

Childhood Epilepsies: Neuropsychological, Psychosocial and Intervention Aspects. (Wiley Series on Studies in Child Psychiatry). Edited by BRUCE P HERMANN and M SEIDENBERG. (Pp 264; Price: £18.95.) Chichester, J Wiley & Sons Ltd. 1989. ISBN 0-471-93562-X.

In recent years neurologists, neuroanatomists and pharmacologists have highlighted exciting new developments in the understanding and treatment of epilepsy. There is a danger that the needs of the individual will be swamped by science. With their careful choice of contributors, Hermann and Seidenberg show that the study of psychological and social aspects of epilepsy can be equally rewarding. The issue of a paperback, after the hardback published in 1989, confirms the appeal of their approach.

Clinicians can find ample information to assist them in improving the quality of care of children and adolescents with epilepsies and

their families. Classification of seizures and epilepsies is followed by details of neurological and psychological approaches. The prognosis of cognitive functions, academic achievement and school performance, information processing, and neuropsychological functioning are dealt with comprehensively, yet succinctly. Several authors present results of their continuing studies.

One of the aims of the editors was to provoke further research, and most of the contributors have highlighted gaps in current knowledge. Strategies for controlling for the many variables involved in cognitive and psychosocial studies are required. Problems related to the infant and pre-school child have received little attention. Family developmental factors are understudied. Longitudinal investigations of cognitive functions are few. Work on compliance has concentrated on adolescents and there is little information about carers. Limitations of current vocational assessments are highlighted. Researchers are provided with a mine of information about potential projects. The real need for more financial resources is emphasised by several contributors.

All those working with children with epilepsy should read and note this book. Inevitably the standard of care will be improved as a result.

S J WALLACE

NOTICE

World Federation of Neurosurgical Societies Awards to Young Neurosurgeons

The World Federation of Neurosurgical Societies will give five awards to young neurosurgeons for the best papers submitted for presentation at the Tenth International Congress of Neurological Surgery in Acapulco, Mexico on 17-22 October, 1993. This will be open to all neurosurgeons born after 31 December, 1957. Each award will consist of an honorarium of US \$1500, a certificate, complete waiver of registration fees and also accommodation for the Congress. The papers will be judged by a committee and must contain original, unpublished work on basic research or clinical studies related to neurosurgery.

Young neurosurgeons should submit 8 copies of the manuscript (not more than 10 double spaced typewritten pages exclusive of figures and tables) to: Dr Albert L. Rhoton, Jr, Chairman WFNS Young Neurosurgeons Committee, Department of Neurological Surgery, University of Florida College of Medicine, PO Box 100265, JHM Health Center, Gainesville, Florida 32610-0265, USA.

The last date for submission is 31 November 1992.

If there is more than one author, the senior author's name and address (including country) must be mentioned.