EDITORIAL

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Lewy body dementia: myth or mystery?

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Introduction

Psychiatry is a medical discipline long on disorders and short on explanations.[1] The current debate concerning Lewy Body Dementia is surely enough to confirm this verdict. Although reports only number 300 cases, its rise to prominence has been rapid, provoking heated exchange in the literature and leading more recently to a demand for a review of existing clinical (and histological) classification systems.[2]

Lewy body dementia (LBD) (for the sake of nosological neutrality since up to 20 different rubrics currently exist) has been variously hailed as a new form of dementia with a distinct clinicopathological profile, so called diffuse Lewy body disease (DLBD) or senile dementia of the Lewy body type (SDLT), a variant of Alzheimer's disease – the Lewy body variant of Alzheimer's disease LBV or as part of a spectrum of Lewy body disorders including Parkinson's disease.

What is the clinician to make of this nosological quagmire and has any advance really been made since the early reports of 1961?

It seems clear that LBD is not a new illness and that it represents improved neuropathological detection and a higher index of suspicion which have highlighted its presence. The research base, although small, has identified several important considerations which suggest that LBD should not be ignored.

Firstly, prevalence rates in several studies[3,4] confirm that it is not uncommon.

Secondly, it seems related to two major neurodegenerative conditions, Alzheimer's and Parkinson's diseases.

Thirdly, in the face of mounting clinical and genetic evidence for the heterogeneity of Alzheimer's disease (perhaps accounted for, in part by LBD) and the contamination of existing research data, we require an urgent revision of the current classification system.

Lewy bodies

Lewy bodies (LBs) are eosinophilic intraneuronal inclusion bodies which are the pathological hallmark of a number of neurodegenerative conditions including Parkinson's disease. LBs consist of abnormal accumulations containing the protein ubiquitin and phosphorylated and non-phosphorylated neurofilaments, important components of the neuronal cytoskeleton. Neurofilaments play an important role in axonal transport and determination of axonal calibre. Transgenic animal studies demonstrate that abnormal neurofilament accumulation can lead to neuronal dysfunction and degeneration .

Over the years there has been increased recognition from postmortem studies that LBs also occur in the cerebral cortex in a substantial number of patients with dementia. The numerous rubrics mentioned above have arisen because of the differences between the clinical syndromes which have been described to date in association with cortical Lewy bodies and the diagnostic significance attached to them. Review of the literature suggests differences in the sampling frames from which patients have been drawn may be responsible.[5]

Despite the apparent nosological confusion, a consensus is arising that most research groups are probably referring to a similar condition characterised at the neuropathological level by subcortical and cortical LBs formation, with or without a variable amount of Alzheimer-type pathology, predominantly senile plaques, with only a minority of patients having sufficient neurofibrillary tangles to meet quantitative neuropathological criteria for AD.