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Lewy Body Dementia

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Dementia with Lewy bodies (DLB) is a common form of dementia. The characteristic features of DLB are progressive dementia particularly affecting attention, visuo-spatial and executive ability, fluctuating cognition, spontaneous parkinsonian symptoms, persistent vivid visual hallucinations, hypersensitivity to neuroleptic medication and REM sleep behavioural disorder. Patients with DLB frequently have mixed pathology. The presence of Alzheimer's disease (AD) pathology modifies the clinical features of DLB, making it harder to distinguish DLB from AD clinically during life, with AD being the main differential diagnosis. Clinical diagnostic criteria for DLB applied at presentation can fail to identify up to 50% of cases. An accurate diagnosis is important for carers, so that they are aware of the symptomatology of the illness, the course and the prognosis, and for the professionals, so that they manage appropriately motor, cognitive, psychiatric, sleep and autonomic symptoms, and avoid neuroleptic medication which frequently leads to increased confusion, alterations in consciousness and worsening of parkinsonian symptoms. In addition, failure to diagnose DLB affects AD treatment trials, making it more difficult to develop and test drugs that specifically target the different underlying pathology of AD or DLB.

At present there are several imaging techniques that can improve the identification of DLB during life. Whole brain atrophy, rate of atrophy over time, and white matter lesions on MRI are not helpful in differential diagnosis. Hippocampal and medial temporal lobe atrophy can detect difference between AD and DLB at a group level but have limited sensitivity and therefore utility for individual patients[1].

More helpful are metabolic studies of occipital lobe hypometabolism using FDG PET. A number of single centre studies, some including autopsy diagnosis, have shown that FDG PET provides good discrimination between AD and DLB, with sensitivity between 86-92% and specificity 80-92%[2]. Studies with HMPAO SPECT looking at occipital hypoperfusion with relative preservation of medial temporal perfusion show less consistent results[3].

Patients with DLB have pronounced cardiovascular autonomic dysfunction and this is well demonstrated with MIBG cardiac scintigraphy. This investigation has excellent sensitivity of 95-100% and specificity of 87-100%[4]. The main drawback is that abnormal scans are difficult to interpret in the elderly as diseases common in this age group, such as diabetes, myocardial infarction, ischaemic heart disease and cardiomyopathy, can all lead to abnormal scans increasing the risk of a false positive diagnosis.

Patients with DLB, compared to AD, have a pronounced pre-synaptic dopaminergic deficit in the striatum. At present the most studied technique for assessing dopaminergic pathways is ^{123}I-FP-CIT SPECT, although other techniques such as DTBZ PET have also yielded promising results. Following a number of single centre studies of FP-CIT SPECT[5] which used both semi-quantitative and visual analysis there is now good evidence from an autopsy study and a European multicenter study that FP-CIT SPECT has high sensitivity and specificity for distinguishing probable DLB from non-DLB dementia[6,7]. The autopsy study[7] is on-going and additional results continue to support the published data. Follow-up data from the European Trial indicate that FP-CIT SPECT is also helpful in clinically less clear cases of possible DLB.

References

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