A case of Dementia with Lewy Bodies

HISTORY

A 74 year old gentleman with a 30 year history of reactive depression presented with a one year history of cognitive decline and speech impairment. His family described a relentlessly progressive expressive dysphasia. Initially his speech had become more effortful and had lost fluency. As time progressed his speech output had markedly declined with stuttering, hesitant and repetitive speech. His cognitive function had also declined with increased difficulty remembering day-to-day activities, forgetting names and faces and losing his way in familiar environments. During this time he was very depressed and he had been admitted 5 months prior to neurological presentation following a suicide attempt.

His wife had noted bradykinesia and a loss of facial expression in the last 6 months. She had also observed some jerky irregular movements of his upper limbs as well as a resting tremor.

On direct questioning the family reported significant day-to-day fluctuation in both speech and memory. Frightening visual hallucinations had occurred in the past with the patient reporting seeing men coming out of the mirrors in the house. These were no longer a prominent feature following treatment with Olanzapine. His extrapyramidal symptoms had clearly predated the use of Olanzapine.

Past medical history was unremarkable as was family history apart from depressive disorder. His current medications were Aspirin, Olanzapine, Mirtazipine and Lactulose.

EXAMINATION

The patient had a marked non-fluent aphasia. Formal cognitive testing showed problems not just with language production, but with attention, frontal executive function, memory, and visuo-spatial processing. There was relative preservation of naming, visual memory and perception.

Physical examination revealed bilateral palmo-mental reflexes and a pout reflex. Cranial nerves were normal. He had hypomimia and mild bradykinesia with a slow gait and decreased arm swing. There was a rest tremor and myoclonus of the upper limbs that was relatively symmetrical. Power and sensation were normal. There was a mild degree of upper limb apraxia.

INVESTIGATIONS

Routine bloods were normal as was a CT head. An EEG showed an excess of theta in keeping with an encephalopathy but no other specific abnormalities. MRI head was unremarkable.

CONCLUSIONS

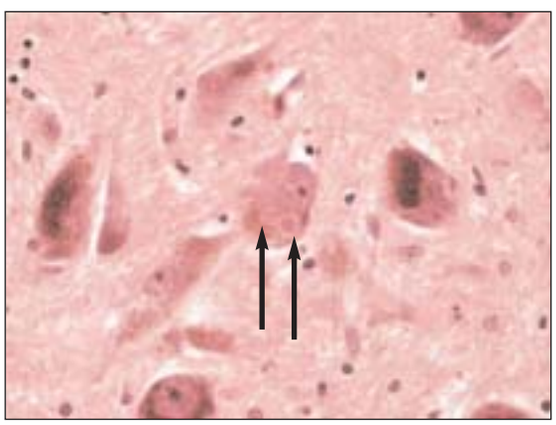
Treatable causes of dementia were excluded. This gentleman's presentation with an extrapyramidal picture, rapid cognitive decline, fluctuation and visual hallucinations were felt to be most compatible with Dementia with Lewy Bodies (DLB) and he was started on Rivastigmine.

Although he had significant dysphasia, his cognitive testing revealed more global deficits than would be associated with a primary progressive aphasia. DLB can present very rapidly and has been confused with more sub acute causes of dementia such as CJD.[1] The typical triad of fluctuating cognitive impairment, visual hallucinations and an extrapyramidal syndrome suggests the diagnosis although a wide variety of cognitive deficits have been reported.[2] Post mortem analysis of these patients shows typical Lewy Bodies (see picture) widespread throughout the cerebral cortex.

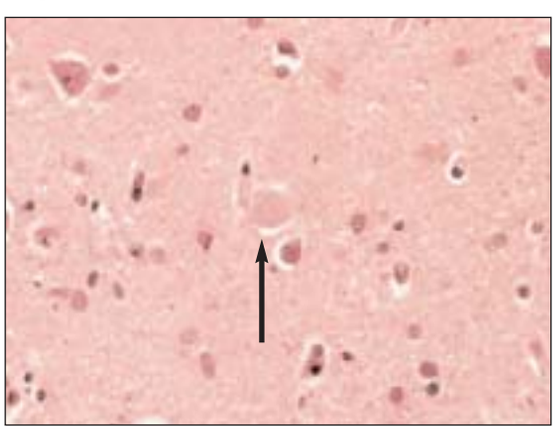
References

1.Tschampa HJ, Neumann M, Zerr I, Henkel K, Schroter A, Schulz-Schaeffer WJ, Steinhoff BJ, Kretzschmar HA, Poser S (2001). *Patients with Alzheimer's disease and dementia with Lewy bodies mistaken for Creutzfeldt-Jakob disease.* J Neurol Neurosurg Psychiatry 71(1):33-9.

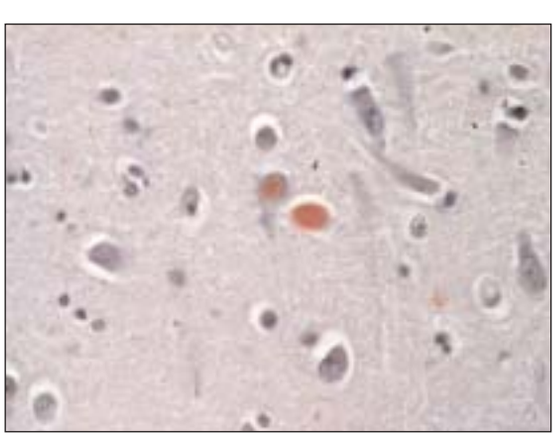
2.McKeith I (2000) *Clinical Lewy body syndromes* Ann N Y Acad Sci;920:1-8.



A: Substantia nigra, H&E, x400: two classical Lewy bodies within a nigral cell.



B: Cingulate cortex, H&E, x400: a cortical Lewy body, which is less strongly eosinophilic, lacking a peri-inclusional 'halo' and lacking a concentric pattern of staining.



C. Cingulate cortex, ubiquitin immunohistochemistry, x400: two Lewy bodies positive for ubiquitin (would also be positive for alpha-synuclein).

Figures supplied by Dr Rhys Davies and Dr John Xuereb, University of Cambridge

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