local hospice where she regained awareness only briefly before her death in early August, 7 months after symptom onset.

GC is an uncommon, diffusely infiltrating primary cerebral neoplasm and is a diagnosis often made after death. The clinical manifestations may mimic many neurological disorders but parkinsonism is not a common feature. In fact, we could only find one other report in recent literature of parkinsonism as the clinical presentation. In this case the diagnosis was only made after death and there were no significant brain MRI abnormalities.

The complex of subacute neuropsychiatric disturbance coupled with parkinsonism and myoclonus is a recognised presentation of sporadic CJD. T2 weighted basal ganglionic signal change is also a feature seen in sporadic CJD although in a recent review this only approached a sensitivity of 63%. Over 20 alternative diseases were described to have these imaging findings, although GC was not included in this list. Our case highlights the fact that GC can present in a manner mimicking sporadic CJD and should be considered in this context.

M Slee
Department of Clinical Neurology, Radcliffe Infirmary, Oxford, UK

P Pretorius
Department of Radiology, Radcliffe Infirmary, Oxford, UK

O Ansorge
Department of Neuropathology, Radcliffe Infirmary, Oxford, UK

R Stacey
Department of Surgery, Radcliffe Infirmary, Oxford, UK

R Butterworth
Department of Clinical Neurology, Radcliffe Infirmary, Oxford, UK

Correspondence to: Dr Mark Slee, c/o Neurology Department, Austin Health, Heidelberg, Melbourne, VIC 3084, Australia; mark.slee@doctors.org.uk

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Neurodegenerative diseases neurobiology pathogenesis and therapeutics


Neurodegenerative diseases are a major global health burden in the Western world particularly with our ageing population. The World Health Organization (WHO) predict that by 2050 a staggering 30 million people will be affected by Alzheimer’s disease in Europe and the USA alone. Other diseases such as Parkinson’s disease, Huntington’s disease, fronto-temporal dementia, and amyotrophic lateral sclerosis also exert a great human toll on the affected individuals, their families, and society. Research into these disorders is therefore of great importance, and this book summarises the current state of research into neurodegenerative diseases. The book is divided into 10 major sections. The first section covers the basic scientific mechanistic aspects of neurodegeneration in 18 different chapters. The role of free radicals, mitochondrial dysfunction, excitotoxicity, neurotrophic factors, nitric oxide, metal ions, and neuroinflammation in the pathophysiology of apoptotic neuronal death are discussed. There is an excellent discussion on the role of prion protein misfolding - a common underlying theme in neurodegenerative disorders. The remarkable advances in understanding the genetic bases of many of these disorders has been a major driving force in all fields of research, and has enabled both transgenic mouse and genetically engineered cell models. It has also increased our understanding of the pathophysiology of sporadic forms of the diseases, and has allowed the design of targeted therapeutics. These concepts are covered in different sections throughout the whole book. There is also a useful section on neuroimaging and a summary of a number of therapeutic approaches to neurodegeneration. There are detailed reviews of Alzheimer’s disease covering mild cognitive impairment, neuropathology, genetics, and then treatments. Equally, other dementias such as fronto-temporal dementia and prion diseases are well covered. The same themes are placed in context with an excellent section on normal ageing. There are nine chapters on Parkinson’s disease and related movement disorders covering multiple system atrophy, progressive supranuclear palsy, corticobasal degeneration, four chapters on cerebellar degenerations, and six chapters on all the motor neurone disorders. Similarly Huntington’s disease, other poly Q disorders, neuroborreliosis, brainstem diseases, Wilson’s disease, and mitochondrial diseases are covered in the final section.

This is the largest and most comprehensive reference text on neurodegenerative diseases available to date. The editors are leaders in the field, and they have brought together a very powerful list of contributors. Each section is nicely coordinated with a clinical introduction summarising the clinical evaluation of the patient, and then leading into neuropathology, genetics, pathophysiology, and therapeutic options. The combination of good clinical information combined with basic science discussion makes this textbook of value to neurologists and psychiatrists as well as non-clinical neuroscientists, and is therefore an excellent addition to any neurodegenerative disease department library. In my view, there is no better overview of the subject currently available, particularly in one textbook. My only caveat is that the pace of scientific research is such that already some of the chapters are out of date. For example, recently RNA interference-based therapies for gene silencing have shown promise in transgenic mouse models of SCA1, Huntington’s disease, and amyotrophic lateral sclerosis, and this is not included. However that is the nature of the beast with this sort of textbook, and I suspect that the editors are already planning the second edition. My only other minor criticism is that a review of biomarker research would have been useful; this is an area of importance in neurodegenerative diseases. A major goal of current research is to improve early detection of disease and presymptomatic detection of neuronal dysfunction. There is also the need for better tools to assess disease progression particularly for the evaluation of disease-modifying therapies. However, these are minor points in what is clearly an outstanding textbook covering an important area both clinically and in neuroscientific research.

S Tabrizi

BOOK REVIEWS

Parkinson’s disease and nonmotor dysfunction (current clinical neurology)