Case Studies in Dementia

Volume 2

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Common and Uncommon Presentations

Volume 2

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Contents

Contents

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Preface

Typical dementia cases characterized by a slowly progressive amnestic syndrome in patients older than 65 years-old constitutes a highly prevalent condition in clinical practice. However, clinicians frequently encounter a significant number of atypical dementia cases featuring either rapid progression, or early onset (younger than 65 years old), or dominance of non-amnestic symptoms. As dementia constitutes a rapidly growing topic in neurology, case studies in dementia intends to illustrate advances in diagnosis of typical and atypical cases.

The first volume of Case Studies in Dementia focused on the classical presentations of typical and atypical dementias. Apart from typical Alzheimer's disease, we included cases of Alzheimer's disease associated with congophilic amyloid angiopathy, posterior cortical atrophy, behavioral presentation of Alzheimer's disease, and mixed pathology cases between Alzheimer's and vascular dementia. We also included various non-Alzheimer dementia cases.

On the spectrum of frontotemporal dementia, volume one describes presentations of the behavioral variant, semantic dementia, progressive non-fluent aphasia, and dementia with motor neuron disease. We also included a synucleinopathy case with Lewy body dementia and a poststroke and vascular dementia case following surgery with a Klüver–Bucy syndrome. There are cases of secondary dementias due to toxic encephalopathy and substance abuse. Regarding transmissible causes of dementia, volume one has chapters describing the clinical features of dementia cases secondary to neurosyphilis, HIV, Heidenhain variant of Creutzfeldt–Jakob, as well as Gerstmann–Sträussler–Scheinker disease. Finally, volume one closes with a section on dementias associated with genetic disorders. There are cases of adult-onset polyglucosan body disease, a disorder characterized by a deficiency of the glycogen-branching enzyme; another case of dementia due to genetic mitochondriopathy and Huntington's disease, which is a polyglutamine repeat disease.

This second volume highlights in its clinical cases the conceptual, genetic, and biomarker advances adopted by the recent operational definitions of dementing diseases. To this end, the first clinical case introduces the biomarker conceptual framework for investigating atypical dementia cases.

Subsequently, Chapter 2 provides insights regarding care planning in typical and atypical cases. On the spectrum of typical presentations, we included a asymptomatic and a typical case of Alzheimer's disease both diagnosed by second generation imaging agents. As atypical cases, we discuss an early-onset Alzheimer's disease and a case of cognitive decline associated with neurofibrillary tangle-predominant dementia. As for focal cortical syndromes, this volume includes cases of frontal variant due to Alzheimer's disease. There are cases of behavioral variant of frontotemporal degeneration due to C09orf75, MAPT17, progranulin, CHR3, and TARDBP mutations. The present volume also includes a case of posterior cortical atrophy and an intriguing case with the association between posterior cortical atrophy and logopenia. Regarding language presentations, there are cases of agrammatic, semantic, and logopenic presentations of primary progressive aphasia. There is also an interesting case illustrating alexia without agraphia associated with Pick's disease. A number of chapters illustrate dementia syndromes with motor manifestation such as progressive supranuclear palsy, Lewy body dementia, multiple system atrophy, normal pressure hydrocephalus as well as Wilson's and Parkinson's disease. We include two cases of dementia following cerebrovascular diseases. Regarding prion diseases, the reader will have the opportunity to contrast a Creutzfeldt–Jakob disease and fatal familial insomnia. We added a case of paraneoplastic syndrome and a case of Hashimoto's encephalopathy for illustrating autoimmune causes

of dementia. The differential diagnosis of psychiatric conditions and dementia is exemplified in a case describing a patient with bipolar disorder. The second volume closes with an Appendix with the updated diagnostic criteria of the typical and atypical cases. We hope that this appendix would streamline the learning process for students and residents.

We would like to express our gratitude to the patients, their respective caregivers and family members who have contributed to this case book. We would like to thank the authors for their diligence, time, and patience to accomplish all tasks required and we would also like to thank Monica Shin, MSc, for her valuable voluntary assistance provided in this volume. On behalf of all authors, we would like to thank all our families, as well as the organizations for supporting our work in dementia research and clinical care.

We expect that the cases presented in this book would serve as an inspiration for the next generation of researchers and health professionals to advance the care of dementia patients.