Advanced Neuroradiology Cases
Advanced Neuroradiology Cases

Challenge Your Knowledge

Edited by

Lázaro Luís Faria do Amaral
Neuroradiologist and Chief of the Neuroradiology Department, Medimagem – Hospital Beneficência Portuguesa and Hospital São Jose, São Paulo, SP, Brazil
Neuroradiologist and Head of the Neuradiology Department, Telemedimagem – Hospital Santa Catarina, São Paulo, SP, Brazil
Neuroradiologist of the Neuroradiology Department, Santa Casa de Misericórdia de São Paulo, São Paulo, SP, Brazil

Asim K. Bag
Assistant Professor, Section of Neuroradiology, The Department of Radiology, School of Medicine, The University of Alabama at Birmingham, AL, USA

Fabrício Guimarães Gonçalves
President of the Radiology Society of Brasilia, and Neuroradiologist and Chief of the Radiology Department, Department of Radiology, Children’s Hospital of Brasilia, Brazil

Prasad B. Hanagandi
Pediatric Neuroradiology Fellow at Hospital for Sick Children, University of Toronto, Ontario, Canada

Associate Editor

Suzanne Byan-Parker
Birmingham, AL, USA
With love to my wife, Denise, inseparable companion of my life, and to my son Danilo, and my daughter, Beatriz, for giving my life immeasurable meaning. To my parents Iolanda and Pedro: thank you very much for your love and support.

“Wisdom is supreme; therefore get wisdom. Though it cost all you have, get understanding.”

Proverbs 4:7

Lázaro Luís Faria do Amaral, MD

To all the residents, fellows, and trainees who are striving to be the best: I hope this book makes you stronger and sharper. To all the physicians who want to familiarize themselves to the imaging appearances of some of the rare diseases of the brain for confident diagnosis: I hope you find this book helpful.

To Ras, my soul mate now and forever:
To Rishi, my loving son:
To my parents: Thank you for your EXCEPTIONAL patience, Love, and STRONG support.

Asim K. Bag, MD

To all my teachers, fellow colleagues, and residents.
A special thank you to my parents for their support.
“Neel” my adorable son.
Very special thanks to my lovely woman; for standing by me and extending emotional support and encouragement during my tough times.

Prasad B. Hanagandi, MD

To God, who gives me strength, to my wife Flávia for her love and patience, who provided support and encouragement and who took care of me and especially of two princesses Rafaela and Marcela, during those months dedicated to the completion of this dream.

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Fabrício Guimarães Gonçalves, MD
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Contributors

Taleb Al Mansoori
Department of Medical Imaging, the Ottawa Hospital,
University of Ottawa

César Augusto P. F. Alves
Santa Casa de Misericórdia de São Paulo

Heitor Castelo Branco R. Alves
Santa Casa de Misericórdia de São Paulo

Anderson Benine Belezia
Hospital Beneficência Portuguesa de São Paulo

Karenn Barros Bezerra
Diagnostic Imaging, Radiology Department, Hospital
Universitário de Brasília, Universidade de Brasília

Christiane Monteiro Siqueira Campos
Hospital Beneficência Portuguesa de São Paulo

Guilherme Cassia
Department of Radiology, Hospital Santa Helena

Santanu Chakraborty
Department of Medical Imaging, the Ottawa Hospital,
University of Ottawa

Jeffrey Chankowsky
Department of Diagnostic Radiology, the Montreal
General Hospital

Philip R. Chapman
The Department of Radiology, School of Medicine,
the University of Alabama at Birmingham

Auro Augusto Junqueira Côrtes
Hospital Beneficência Portuguesa de São Paulo

Antônio José da Rocha
Neuroradiology Department at Fleury Medicina Diagnóstica
and Santa Casa de Misericórdia de São Paulo.

Raquel del Carpio-O’Donovan
Department of Diagnostic Radiology, the Montreal
General Hospital

Ricardo Tavares Daher
Hospital Beneficência Portuguesa de São Paulo

Marc Dilauro
Department of Medical Imaging, the Ottawa Hospital,
University of Ottawa

Kelly Fiorini
Hospital Beneficência Portuguesa de São Paulo

Leonardo Furtado Freitas
Hospital Beneficência Portuguesa de São Paulo

Salo Haratz
Tel Hashomer Medical Center

Harry S. Hardin
The Department of Radiology, School of Medicine,
the University of Alabama at Birmingham

Ricardo Heusi
Clinica São Lucas, Universidade do Vale do Itajai – Univale
and Unimed de Balneário Camboriú

Bruno Shigueo Yonekura Inada
Hospital Beneficência Portuguesa de São Paulo

Sunila Jaggi
Department of CT Scan and MRI, Bombay Hospital and
Medical Research Centre

Jennifer Kamps
Stavanger University Hospital

Stephanie Lam
Department of Radiology, McGill University Health Centre –
Montreal General Hospital
List of contributors

Leslie Lamb
Department of Medical Imaging, the Ottawa Hospital, University of Ottawa

Afonso Celso Pedrotti Liberato
Hospital Beneficência Portuguesa de São Paulo

Ingrid Aguiar Littig
Santa Casa de Misericórdia de São Paulo

Bruno Siqueira Campos Lopes
Hospital Beneficência Portuguesa de São Paulo and Hospital Santa Catarina

Márcio Olavo Gomes Magalhães
Radiology Department, Hospital Universitário de Brasília, Universidade de Brasília

Victor Hugo Rocha Marussi
Hospital Beneficência Portuguesa de São Paulo

Lucídio Portella Nunes Neto
Hospital Beneficência Portuguesa de São Paulo

Renato Hoffmann Nunes
Santa Casa de Misericórdia de São Paulo

Taciana Mara Filomeno Orsini
Hospital Beneficência Portuguesa de São Paulo

Satya Patro
Department of Medical Imaging, the Ottawa Hospital, University of Ottawa

Ivanildo Castro Pereira Junior
Hospital Beneficência Portuguesa de São Paulo

Glenn H. Roberson
The Department of Radiology, School of Medicine, the University of Alabama at Birmingham

Rasmoni Roy
The Department of Radiology, School of Medicine, the University of Alabama at Birmingham

Sonali H. Shah
Department of CT Scan and MRI, Bombay Hospital and Medical Research Centre

Aparna Singhal
The Department of Radiology, School of Medicine, the University of Alabama at Birmingham

Inder Talwar
Department of CT Scan and MRI, Bombay Hospital and Medical Research Centre

Bruno Augusto Telles
CETAC/INC CURITIBA – PR

Benson Tran
The Department of Radiology, School of Medicine, the University of Alabama at Birmingham

Rahul J. Vakharia
Department of CT Scan and MRI, Wockhardt Hospital
Foreword 1

I am delighted to write a foreword for this book, prepared by a group of enthusiastic and committed physicians who have dedicated their careers to the practice of diagnostic neuroradiology. All four authors participated in and successfully passed the two-year Pierre Lasjaunias European Course of Neuroradiology, in which I had the pleasure and honor to be a faculty member and past-Director. Energized by a mutual passion for honing their diagnostic skills, they set about building a collective repository of definitive diagnostic neuroradiology case studies.

The fruit of their thirst for knowledge, this book harmoniously unites a unique collection of atypical neuropathologic case studies in a methodical, readily accessible manner. Framed as "Diagnosis Please" format of the Radiology journal, each case study presents a brief clinical history, relevant laboratory results, and supporting diagnostic images. The reader is then invited to diagnose the case based on the clinical and imaging information provided. This interactive format stimulates the reader to actively ponder over the data and work out the appropriate diagnosis, while simultaneously and systematically considering potential differential diagnoses.

The cases conclude with a thorough discussion of the definitive primary diagnosis in light of the analytical, clinical, and imaging findings, effectively eliminating the less likely differential possibilities.

Thus, in a concise, inviting manner, the book provides readers with an essential basis for day-to-day use to develop and strengthen their diagnostic neuroradiology skills. This volume will serve as a valuable resource for neurology and radiology trainees preparing for advanced certification, a reference for established neuroradiologists, and a guide for neurologists in search of readily available examples of atypical neuropathologies. The authors should be congratulated on their earnest wish to share their desire for excellence in practicing neuroradiology.

Àlex Rovira, MD
Head of the Neuroradiology Section, and Director of the MR Unit. Vall d’Hebron University Hospital, Barcelona, Spain
Director of the X and XI Cycles of the European Course of Neuroradiology
Vice President of the European Society of Neuroradiology
Foreword 2

As editor-in-chief of *American Journal of Neuroradiology* and someone who has been practicing neuroradiology for over 25 years, I am well aware of the paucity of relevant and easily accessible information on rare neurologic cases amongst today’s academic publications. I also understand the need for practicing neuroradiologists to remain abreast of current trends and practices in order to accurately diagnose complex patient pathologies. The electronic age and *information highway* have certainly greatly affected academic and clinical medicine by creating vast amounts of information. Paradoxically, this sheer volume of information can be an impediment for a busy clinician in search of a rare neurologic finding, an unusual presentation, or confounding patient history.

Recognizing this difficulty in searching for information on atypical neurologic cases, the authors of this book have created a compendium of cases featuring rare, adult neurologic brain diseases. This task was a result of a desire to extricate illusive nuggets of pertinent information from the ever-increasing amount found elsewhere. Targeting neuroradiologists and other specialists alike, this text incorporates relevant patient information and laboratory values accompanied by typical imaging findings which enable one to offer a diagnosis. Although I agree that multiple pathologies found herein may rarely be encountered, this does not detract from having this book close by as a handy reference.

This book seeks to incorporate recent advances in neuroradiology but also serves as a resource on seldom-encountered neurologic pathologies. Merging these two aspects of neuroradiology, innovations and advances in imaging modalities/techniques with representative samples of atypical patient cases, into one text should be welcomed by those who practice neuroradiology.

The authors sought to include representative cases from all major categories of rare neurologic pathologies ranging from degenerative and infectious diseases, to metabolic diseases and tumors. This text will be an added value to those seeking to diagnose an atypical case or preparing for examinations. The authors have collated a large number of atypical cases that will prove helpful when offering a differential diagnosis based on unusual diagnostic and imaging findings.

I would like to congratulate the authors for creating a resource that unifies rare cases in one reference that will readily enable us to increase our knowledge of atypical neurologic pathologies seen in the brain.

*Mauricio Castillo, MD, FACR*

*Professor of Radiology, Section Chief Neuroradiology*

*University of North Carolina School of Medicine*

*Editor-in-Chief, American Journal of Neuroradiology*
Preface

Neuroradiology is a fast-paced subspecialty. The expanse of worldwide knowledge and global electronic accessibility to journals, databases, and search engines constitutes a daunting wealth of information. However, this vast oasis of information does not readily yield easily accessible information on atypical neuroradiologic case presentations. If anything, the magnitude of available data may impede acquisition of information with any discernibly diagnostic value when searching for unusual cases and atypical presentations.

Traditional data mining or exhaustive literature searches are out of bounds for most practicing neuroradiologists who often find themselves overwhelmed with voluminous case-loads, unable to spare extended periods of time to look up a particular case. This is further complicated by the fact that rare cases find no place in current mainstream radiology journals that are more focused on achieving and maintaining a high impact factor rather than providing practical information to clinical neuroradiologists. Thus, finding consultative or reference materials becomes extremely arduous for a busy radiologist. The introduction of numerous speciality journals and influx of open access journals have added to the cryptic informational overload for clinicians seeking material on unusual case presentations.

The significance of our text lies within the historic origins of neuroradiology as a subspecialty which began, unquestionably, before visual images of the brain were conceivable – or obtainable. A series of benchmarks built one upon another laid the foundation and therefore generated the imperative need to obtain visual access to the human brain, or as William Mac ewen aptly described it, “...the dark continent” [1]. Subsequent fine-tuning and advancement of radiographic technologies enable and sustain exploration of neurologic diseases with such a rapid pace that practicing neuroradiologists stagger to keep abreast of these tidal waves.

The purpose of our book is to create a readily accessible and reliable repository of rare cases that can then be confidently diagnosed by informed assessment of characteristic imaging and clinical findings. The intent is that this text will challenge a reader’s knowledge base and enlighten him or her about the characteristic imaging findings of central nervous system (CNS) pathology. Thus, the reader will be able to then confidently make the same diagnosis in real time, when seen in a clinical setting. In hope to retain the information, the book is formatted as unknown quiz cases with clinical and imaging information in the front facing page (the recto) and the case discussion in the back facing page (the verso). Although this book is targeted towards the full-time neuroradiology professionals, it will benefit any physician who is involved with the care of a patient with a rare CNS disorder. Whether the physician is a general radiologist practicing in a community hospital, an academic neuroradiologist, or a practicing neurologist – they will find value in this text that serves as a readily available guide.

Reference

Acknowledgments

This book is a celebration of exceptional friendship, sharing, patience, and commitment. The preparation of this casebook was a project of several years and could not have been concluded successfully without the support and collaboration of many people. In addition to the people who grace the pages of this book, there are many others needing noteworthy mention.

First, we want to thank Suzanne Byan-Parker, whose orchestrated efforts made this book possible. She has been heavily involved in the project from early on. She spent hours in coordinating, editing, and compiling the manuscript. We find no appropriate words to thank her. We simply want to say, "Thank you Suzanne for everything you have done with this project."

The editors also want to thank their family members for being so supportive and for encouraging them to finish this project that needed countless hours away from their families.

LA, PH, and FG also want to appreciate AB for his leadership role in this project without which it would not have been possible to finalize this process. We also want to thank all the contributors for their gracious help.

LA wants to thank to his colleagues: Sérgio Santos Lima, Christiane Monteiro Siqueira Campos, Victor Hugo Rocha Marussi, Bruno Siqueira Campos Lopes, Lucas Ávila Lessa, Leonardo Furtado Freitas, and Anderson A. Belezia, Neuroradiologists staff at MEDIMAGEM, Hospital Beneficência Portuguesa de São Paulo, São Paulo, Brazil, for their continuous support during this effort.

FG wishes to thank personally the following people for their contributions to his inspiration and knowledge and other help in creating this book: Raquel del Carpio, MD, Professor and Vice-Chair of Radiology at McGill University and Jeffrey Chankowsky, MD, CM, FRCP(C), Associate Professor; Program Director (Residency Program) McGill University, Montreal.

All the editors also acknowledge the continuous friendly and encouraging support of the editorial staff of Cambridge University Press.
How to Use This Book

The patient cases that follow feature pathologies that may present unique challenges to a practicing neuroradiologist, or radiologist. Drawn from both academic and private practice, the samples encompass a range of pathologies categorized as neurodegenerative diseases, neuroinfectious, metabolic disorders and more that specifically target the central nervous system, central nervous system tumors, and a myriad of miscellaneous cases.

The chapters are organized according to overarching extra-neurologic pathologic involvement. Each case correlates with the specific underlying etiology, so to speak, that best represents the pathology or pathophysiology related to the thematic category of one of eight chapters. This framework allows the reader to associate each case discussed with a correlative pathologic or physiologic category of the underlying etiology.

Spanning two continents, this collection of cases represents relatively rare pathologic entities not seen in everyday radiology practice.

The text was designed to be used as a guide, reference, and study companion that houses atypical patient cases with representative imaging and correlative imaging findings. Each case section represents an aspect of a physician-patient encounter beginning with initial presentation, diagnostic procedures, consolidation, reporting, diagnosis, and clinical decision-making. The cases are divided into sections designed to introduce the reader to the patient, observe pertinent clinical and radiologic findings, and engage in the diagnostic thought process in order to arrive at the most likely diagnosis based on the data presented.

Optimally, the reader should be able to identify the most likely primary diagnosis after close reading of the clinical presentation and examining the accompanying images.

Clinical Presentation. Includes the essential patient findings at presentation in a clinical context in order to prompt the reader to begin patient assessment and determine what optimal diagnostic imaging modalities would best ascertain the underlying diagnosis.

Imaging. Images are presented with figure legends in order to complete the clinical presentation.

Primary Diagnosis. Primary diagnosis based on findings, evidence, clinical decision-making, and outcome after careful consideration of the clinical presentation and imaging studies; the most likely diagnosis should be evident.

Differential Diagnoses. A list of the most pertinent differential diagnoses based on the clinical findings and the images featured with each case. The reader should be able to explain why the featured list of differential diagnoses is excluded based on the clinical and imaging studies presented in each case.

Imaging Findings. Images are presented with figure legends describing the important findings, which include functional and anatomic characteristics, striking features, and notation of usual and/or pathologic landmark findings of each image. After examining the images, in conjunction with the clinical presentation, the reader should be able to develop and articulate a basis for discussing the primary diagnosis and eliminating differential diagnoses.

Discussion. Explains the basis for the primary diagnosis based on the clinical, laboratory, and imaging findings. The discussion offers the reader an opportunity to engage in clinical consultation as though reporting on patient findings during a consult in order to arrive at the most likely diagnosis based on imaging findings and characteristic presentations of each case.

Key Points. A summary of combined clinical and imaging features from each case.

Suggested Reading. A list of source and reference material that will provide the reader with additional information pertinent to each pathologic entity.